



## EXPOSED POSTERS

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## PE 001. LUNG CYSTS IN HIPERSENSITIVITY PNEUMONITIS: 3 CLINICAL CASES

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**Introduction:** Hypersensitivity pneumonitis (HP) is a complex and polymorphic inflammatory syndrome originated by immune sensitization to inhaled antigens in genetically predisposed individuals. This syndrome is classically subdivided in acute, subacute and chronic. Chest radiograph is generally considered a limited tool. The HRCT, considered the best exam to inform on the (differential) diagnosis also with prognostic value, frequently presents findings of ground glass opacities, reticulation, centrilobular nodules, mosaic attenuation pattern with air-trapping and, in advanced stages, features of irreversible fibrosis. The presence of cysts in HP is not a common feature being reported in only 13% of patients.

**Case reports:** Case 1: Woman, 72 years, nonsmoker with a long-term continuous exposure to pigeons and chickens. She presented with a moderate effort dyspnea and basal non-“velcro” lung crackles. Her workup presented negative autoimmune study and no potential for drug toxicity. Specific IgG for pigeons and chicken showed high titers. BAL presented high total cell count with 53% lymphocytes with some eosinophils, mastocytes and plasmocytes. HRCT rendered patchy ground-glass opacities, low-attenuation centrilobular nodules and disperse cysts, along with some “mosaic” attenuation. A diagnosis of subacute HP (“Bird fancier’s lung”) was sealed at the ILD-MDT (Interstitial Lung Disease-Multidisciplinary Team) meeting (high level of confidence). Case 2: Woman, 64 years, nonsmoker presented with a grade 2 mMRC dyspnea with persistent dry cough in the previous two years, along with some episodic nocturnal wheezing and basal lung crackles. Clear episodes of symptom worsening after contact with hay were claimed. BAL showed a high total cell count with a high lymphocyte count (81%). Immunological workup showed presence of low positive rheumatoid factor, polyclonal hypergammaglobulinaemia, and high IgG titers to *Saccharopolyspora rectivirgula* and *Aspergillus spp.* HRCT presented a mosaic attenuation pattern with some diffuse ground glass opacities, some lobular areas of air-trapping and cystic areas mainly in

the upper lobes. A final diagnosis of Farmer’s Lung HP variant was sealed at the ILD-MDT meeting with a high confidence level. Case 3: Woman, 80 years with a previous history of unspecified lung disease. Past exposures to canaries, turtledoves and chickens, ceased 15 years ago. Clinically with a chronic grade 3 mMRC dyspnea, productive cough and “velcro” lung crackles. High titers of specific IgG only for chicken droppings. Negative autoimmune testing. BAL presented a normal total cellularity, with elevated neutrophils (30%) and some eosinophils. On HRCT there was a reticular pattern with traction bronchiectasis, mosaic attenuation pattern ( $\geq 3$  lobes), some peribronchovascular patchy fibrosis along with isolated cysts. ILD-MDT discussion concluded by a diagnosis of a fibrotic chronic HP (moderate confidence level).

**Discussion:** Isolated cystic abnormalities in HP are unusual but reported in a small percentage of patients. These cysts, different from honeycomb change, resemble the cysts from lymphoid interstitial pneumonia and are presumably caused by partial bronchiolar obstruction by the peribronchiolar lymphocytic infiltrate present in HP patients. These 3 clinical cases exemplify the finding of cystic lesions in cases of subacute/predominantly inflammatory HP and chronic fibrotic HP.

**Key words:** Cysts. Hipersensitivity pneumonitis.

## PE 002. PLEURAL EFFUSION: THE KEY IS IN THE HISTORY

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**Case report:** We present an 84 year old non smoking woman, without a known pulmonary pathology, employed in a cork manufacture whose husband was a naval locksmith. She was referred from the pulmonary diagnostic center to our Hospital where she was admitted for investigation of shortness of breath with progressive escalation and tiredness with right pleural effusion (PE) that required drainage (1,5 L) one month before. A previous chest computed tomography (CT) showed a large PE on the right side in a free cavity with passive pulmonary atelectasis, pleural thickening and bilat-

eral calcification plaques. When admitted, she was submitted to a thoracentesis (1,1L). The biochemical analysis of aspirated pleural fluid (PF) was consistent with an exudate and it was negative for bacterial and cytological findings. It had an adenosine deaminase of 7.80 U/L. The blood analysis showed a normal NTproBNP and renal function within normal range. There were no evidences of cancer in the ecographic study of the pelvic cavity. She was discharged and sent home while awaiting a thoracoscopy procedure. But one month later she was readmitted for symptomatic recurrence of PE. She was submitted to thoracentesis with blind biopsy of the pleura with negative citohistology results and thoracoscopy also with a biopsy which showed diffuse thickening and whitish plaques in both pleuras, without identification of granulomas or neoplastic tissue. A simple talcage was performed. For two years now she remained without new recurrence of PE. A PE can be a diagnostic challenge since 20% of PE etiology remains unknown despite of widespread investigation using chest CT and interventional pulmonology procedures. In recurrent PE the most probable cause is malignancy, however some benign etiologies must not be dismissed, since their diagnosis, although mostly of exclusion, require the integration of epidemiological factors, often missing in anamnesis.

**Discussion:** Taking into consideration the known epidemiology aspects related to asbestos exposure and corroborated by the imagiological and historical findings, with exclusion of other causes of PE and along-lasting follow-up without recurrence or evidence of pleuropulmonary cancer, we consider that the most probable diagnostic would be a Benign Asbestos Pleural Effusion (BAPE) due to exposition to asbestos in the past. It is important to notice that BAPE may only be diagnostic after 20-30 years of first initial contact.

**Key words:** *Pleural effusion. Shortness of breath. Asbestos.*

### PE 003. SEVERE NON-SPECIFIC INTERSTITIAL PNEUMONIA IN DERMATOMYOSITIS: A CASE REPORT

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**Introduction:** Dermatomyositis (DM) is characterized for phenotypical diversity - the clearly distinct clinical subsets can be associated with a specific pattern of autoantibodies which can predict severity. Lung involvement, namely interstitial lung disease (ILD), is very common. Evaluation by interstitial experts is advisable in order to early referral to lung transplantation centers.

**Case report:** The authors describe a clinical case of DM with biomarkers that predict rapidly progressive ILD with multidisciplinary evaluation that allowed an accurate treatment. A 50-year-old woman with subacute onset of a clinical condition consistent with DM with multi-systemic involvement. She presented with myopathy, skin changes, Raynaud phenomenon, arthralgias and progressive dyspnea - hypoxemic respiratory failure was noticed and high-resolution computed tomography showed diffuse ground-glass opacity. Bronchoalveolar lavage was consistent with alveolitis. Myositis and neoplasms were excluded. Positivity to MDA-5, SRP and Ro-52 autoantibodies helped to establish the diagnosis. Clinical response to all organ involvement was noticed with immunosuppression (high dose of steroids and intravenous cyclophosphamide) with the exception of lung - persistent hypoxemia determined referral to specific ILD outpatient clinic, intensive physiotherapy and prolonged immunosuppression. At this moment, lung transplantation is being considered.

**Discussion:** This case shows that multisystemic diseases like DM can have serious ILD and involvement of all specialties is crucial to early diagnosis and treatment. The pattern of autoantibodies can predict the severity of the disease (MDA-5 is very specific to progressive and severe ILD). We point out the importance of knowing

the clinical spectrum of ILD in connective tissue diseases and its implications on diagnosis and prognosis. Early referral to lung transplantation centers is crucial.

**Key words:** *Dermatomyositis. Interstitial lung disease. Autoantibodies. Lung transplantation.*

### PE 004. FROM THE INTESTINE TO THE LUNG - AN IATROGENIC CASE

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**Introduction:** Mesalazine is the top-line drug for the treatment of mild to moderate ulcerative colitis. Its pulmonary toxicity is rare, as are the pulmonary manifestations of inflammatory bowel diseases (IBD) and can begin days to years after starting the therapeutic.

**Case report:** A 74-year-old male, retired construction worker, non-smoker, diagnosed with diabetes mellitus and ulcerative colitis on metformin and oral mesalazine for several years, without known history of pulmonary pathology. The patient presented to the emergency department (ED) with a 3-week history of non-productive cough, dyspnea and wheezing. He was treated with amoxicillin/clavulanic acid and azithromycin and posteriorly with levofloxacin, without clinical improvement. He later returned to the ED due to the persistence of symptoms. On admission the patient was feverless, eupneic at room air, with diminished vesicular sounds and cracklings at the right base. Arterial blood gas analysis revealed partial respiratory failure. Blood tests showed: 11,300/ $\mu$ L leukocytes; 9,350/ $\mu$ L neutrophils; 160/ $\mu$ L eosinophils and 15.9 mg/dL C-reactive protein. Negative *Pneumococcal* and *Legionella* urinary antigen tests. Chest X-ray showed, at the lower half of the lung fields, bilateral condensations, more evident on the right side. Chest CT showed extensive consolidation in the right lower lobe with air bronchogram and consolidation foci in the left lower lobe. The patient was hospitalized and started piperacillin/tazobactam and ciprofloxacin, without clinical or imaging improvement. He underwent bronchofibroscopy (BFC) with bronchoalveolar lavage (BAL) and transbronchial lung biopsies. Histology revealed inflammatory infiltrate with predominance of polymorphonuclear cells and lymphocytes, without cancer cells. Microbiological cultures (blood, BAL and sputum) were negative. HIV serology, immunology testing and ACE were all negative. Systemic corticosteroids were initiated with partial improvement and he was discharged under a tapering course of 20 mg prednisolone for fifteen days. One month later, he returned to the ED, with a two week history of productive cough, dyspnea, marked anorexia and fever. The blood tests, showed maintained high inflammatory parameters, with an increased eosinophilia of 900/ $\mu$ L. Chest imaging showed bilateral peribronchovascular opacities, some with air bronchogram and with upper lobe predominance. BFC showed no changes and BAL had 24% eosinophils; 3% neutrophils; 16% monocytes; 54% lymphocytes. Transbronchial lung biopsies revealed an inflammatory infiltrate with a predominance of polymorphonuclear cells, lymphocytes and some eosinophils. Mesalazine-induced pulmonary hypersensitivity was suspected, the drug was discontinued and steroid therapy (1 mg/kg/day) was started, with clinical, laboratory and total radiological improvement after two months.

**Discussion:** In patients with IBD and pulmonary manifestations, the diagnostic hypothesis of infection, extra-intestinal manifestation of IBD or drug-induced toxicity should always be considered. In the medical literature, one can find about forty cases of mesalazine pulmonary-induced toxicity, being eosinophilic pneumonia the most common reported form. In the present case, clinical, laboratory and

radiological improvement after mesalazine withdrawal supported the diagnosis of drug-induced eosinophilic pneumonia.

**Key words:** *Mesalazine. Eosinophilia. Eosinophilic pneumonia.*

#### PE 005. THE TWO "GREAT IMITATORS" - REPORT OF TWO CASES

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**Introduction:** Tuberculosis and sarcoidosis are both chronic, multi-systemic and granulomatous diseases, with clinical and radiological similarities, which sometimes represents a difficulty in the differential diagnosis between these two entities, especially in areas where there is a high prevalence of tuberculosis.

**Case report:** Case 1: 54-year-old man from Guinea-Bissau, non-smoker, construction worker, with diagnosis of pulmonary tuberculosis 20 years ago. He presented with symptoms of fatigue, dyspnea of exertion, non productive cough, afternoon fever and weight loss of 4 kg for the last 4 months, but without respiratory insufficiency. From the complementary study: leucopenia, increased SR, increased ACE, negative serologies, negative BAAR screening in the sputum; diffuse micronodular pattern on the chest radiography and on the thoracic CT multiple small peribroncovascular nodules with miliary distribution, with bilateral central predominance, consolidation areas with air bronchogram and bilateral hilar conglomerates; cytology, and mycobacterial, PPJ, BAAR, TAA and mycological tests were negative in BAL. Given the high clinical and radiological suspicion of reactivation of tuberculosis, therapy with first-line anti-bacillary drugs was begun. Due to the symptomatic maintenance and the imaging changes, a Pulmonology appointment was sent. LFT with moderate restrictive ventilatory alteration (FEV 1 85%, FEV 1/FVC 82.61%) without compromise of gas exchange. He repeated bronchofibroscopy, whose cultural examinations remained negative, but a predominance of lymphocytes in BAL (73%) and CD4/CD8 ratio 1.69 was found. BB and TBPB revealed non-necrotizing granulomatous inflammatory disease compatible with the diagnosis of sarcoidosis. Corticotherapy was begun, with clinical and radiological improvement. Case 2: 31-year-old man from Angola, non-smoker, construction worker, diagnosed with pulmonary tuberculosis in the previous year. He had symptoms of tiredness, dyspnea of exertion, productive cough, pleuritic pain, febrile sensation and non-voluntary weight loss of 10 kg for the last 12 months, with severe respiratory insufficiency. From the complementary study: leucopenia, increased SR, increased ANA, ENA and IgG; negative serologies; negative BAAR in sputum; disperse micronodular pattern on the chest radiograph; CT-TAP with traction bronchiectasis in the upper lobes, pleural thickening and bilateral micronodularities of random distribution without mediastinal adenopathies and hepatosplenomegaly with millimetric hypodense nodules without abdominal-pelvic adenopathies; LFT with very severe respiratory restriction (FEV 1 25%, FEV 1/FVC 85.52%) with severe compromise of gas exchange (DLCO 19.1%). Bronchofibroscopy BAL had predominance of PMN; cytology, mycobacterial, BAAR, PPJ and mycological tests were negative; flow cytometry showed predominance of lymphocytes (25%) and CD4/CD8 ratio 2.54; BB without granulomas but TBPB suggesting granulomas and inflammatory infiltrate. A TTLB was performed but the result was also inconclusive and complicated by iatrogenic pneumothorax. Lung biopsy of the middle lobe and pleural biopsy by videothoracoscopy showed multiple non-necrotizing epithelioid granulomas with multinucleated giant cells of interstitial, pleural and parietal pleura. Diagnosis of stage IV sarcoidosis (with pulmonary, hepatic and splenic involvement) was

concluded and systemic corticotherapy was begun with resolution of respiratory insufficiency, clinical, radiologic and functional improvement (FEV 1 42%, FEV 1/FVC 84.4%, DLCO 33%) improvement. **Discussion:** The aim of this study is to demonstrate the importance of biopsy for histological confirmation, given the similarities between the clinical presentation and the radiological pattern in cases of miliary tuberculosis and sarcoidosis.

**Key words:** *Miliary tuberculosis. Sarcoidosis.*

#### PE 006. LANGERHANS CELL HISTIOCYTOSIS - AN ATYPICAL PRESENTATION

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**Introduction:** Histiocytic disorders are a group of diseases that occur when there is an accumulation and infiltration of different organs by cells with characteristics similar to bone marrow-derived Langerhans cells. Pulmonary Langerhans cell histiocytosis is a rare disease, occurring in 15% of the cases as part of a multisystem disorder.

**Case report:** A 19-year-old female, medical auxiliary, smoker (10 pack-year), presented in 2007 at Ortophedics outpatient department, with an one-year history of left knee mechanical pain. After complementary investigation with knee radiograph and CT, an osteolytic lesion suggestive of cystic formation, in the external portion of left tibial epiphysis was found. Lesion resection was performed with pathology revealing bone fibrohistiocytic lesion with giant cell tumor characteristics. In 2009 the patient relapsed and was submitted again to surgery. In 2013 she presented at Neurosurgery department with a left supraciliar, frontal swelling with a recent 3-year growth. Brain CT showed osteoblastic lesion. After resection, histopathological examination revealed an osteoma. The patient was maintained in follow-up by Ortophedics, with a new left knee lesion relapse in 2015 (submitted to surgery with total knee prosthesis placement). Follow-up thoracic-abdominal-pelvic CT scans had no relevant findings at first but in March 2017 CT scan showed bilateral pulmonary micronodular pattern. In reevaluation CT from October 2017 multiple irregularly-shaped cysts and multiple nodular opacities with mid and upper zone predilection were found, suggesting Langerhans cell histiocytosis. Diagnosis was confirmed by surgical lung biopsy in December 2017. The patient maintains follow-up in pulmonary interstitial lung disease department. She remains asymptomatic and no further treatment was needed. **Discussion:** The authors point out the atypical presentation of the disease, especially within this age group. Multidisciplinary approach is essential as histiocytosis may present as a multisystem disease. Detection of pulmonary features in an initial phase is also rare and of particular interest in this case.

**Key words:** *Histiocytosis. Langerhans. Interstitium.*

#### PE 007. COCAINE LUNG

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**Introduction:** Pulmonary exposure to inhaled particles is a known cause of respiratory pathology and should be part of the differential diagnosis of acute respiratory failure.

**Case report:** The authors present the case of a 45-year-old male patient, graphic designer, smoker (30 pack-year), with history of heroin use in the past and currently sporadic consumption of inhaled cocaine. Primary right pneumothorax at age 25 and pulmonary tuberculosis in childhood. Admitted to the Emergency Department by cough and pro-

gressive worsening dyspnea within the last 24 hours, marked bronchospasm, no fever. Negative inflammatory parameters, severe respiratory failure and radiological pattern of multilobar interstitial pneumonia. No improvement to the initial therapeutic measures. Computed tomography of the thorax with areas of ground glass opacities of the pulmonary parenchyma. The patient reported inhaled cocaine use in the 24 hours that preceded the symptoms. Imaging findings compatible with the diagnosis of pneumonitis associated with acute cocaine use - cocaine lung/"crack lung". Instituted systemic corticosteroid therapy with gasometric normalization and regression of the imaging alterations initially described. Pulmonary manifestations associated with cocaine use are heterogeneous and acute or chronic manifestations may occur. The radiological pattern most frequently associated with cocaine use is ground glass opacities, but the findings may be multiple and are non-specific. There is no pathognomonic finding, and the clinical suspicion associated with the cocaine use is necessary. However, the diagnosis is unusual, since the respiratory symptoms associated with cocaine use are not always valued and do not motivate the use of medical services. Corticosteroid therapy may be considered, although there are no studies to support it. The avoidance of drug addiction habits is the main treatment.

**Discussion:** The authors share the clinical case fundamentally for their didactic aspect.

**Key words:** *Pneumonitis associated with cocaine. Acute respiratory failure.*

#### PE 008. ALPHA-1-ANTITRYPSIN DEFICIENCY. RETROSPECTIVE STUDY OF PATIENTS UNDERGOING REPLACEMENT THERAPY AT CENTRO HOSPITALAR DE SETÚBAL

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**Introduction:** Chronic obstructive pulmonary disease due to alpha-1 antitrypsin (AAT) deficiency is a rare disease. It is estimated that more than 3 million people worldwide have combinations of alleles associated with severe disability. AAT replacement therapy aims to maintain AAT levels above the protective threshold, delaying the progression of the disease.

**Objectives:** Authors present the experience of five patients undergoing intravenous replacement therapy, followed in Pulmonology Department of Centro Hospitalar de Setúbal.

**Methods:** Retrospective data were collected from all patients undergoing AAT replacement therapy for at least 4 months, including five patients in this study. The demographic characteristics, family history, habits and lifestyle and phenotype of these patients were analyzed. Levels of AAT, data from plethysmography, study of capillary alveolar diffusion gradient and arterial gasometry were analyzed before and after the introduction of substitution therapy.

**Results:** The population consists of 2 male and 3 female patients, 4 ex-smokers and 1 non-smoker. The current mean age of patients is 59 years, with mean FEV1 of  $1.1 \pm 0.4$  L ( $42.8 \pm 11.1\%$ ), undergoing therapy for  $51.2 \pm 45.5$  months ago. In all cases, an increase in serum AAT assay was observed after the introduction of replacement therapy, however, in only one patient an increase to a value above 80 mg/dL was observed. In 4 patients clinical, gasometric, FEV1, FEV1/FVC and DLCO stability were observed after the introduction of substitution therapy. In one of the 5 patients included in the study, there was a slightly exacerbation of gasometry, FEV1, FEV1/FVC ratio and DLCO, that can be justified for the longest evolution of the disease in this patient, being the one undergoing replacement therapy for much time.

**Conclusions:** Based on this small sample, we can conclude that our experience is positive, with both, clinical and functional benefit for

patients. As it is described in other observational studies, a positive effect on lung function was also observed in our small sample. The reduction in mortality has not yet been observed, as it has already been described in some observational studies.

**Key words:** *Alpha-1 antitrypsin. Chronic obstructive pulmonary disease. Replacement therapy.*

#### PE 009. NEW MUTATION OF A1-ANTITRYPSIN DEFICIENCY (Q0SANTANA) FOUND IN PATIENTS IN THE AUTONOMOUS REGION OF MADEIRA

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$\alpha$ 1-antitrypsin deficiency is a co-dominant autosomal genetic disorder that essentially predisposes to lung and liver disease. It is caused by mutations in SERPINA 1 gene, which modify the configuration of  $\alpha$ 1-antitrypsin molecule and compromise its release. The phenotyping of  $\alpha$ 1-Antitrypsin is performed in plasma samples by isoelectric focusing separation in pH gradient and in presence of control samples of a known phenotype; the protein is visualized through general staining of proteins. In every study the screening of rare alleles is performed by PCR-multiplex combined with RFLP, which enables evaluating the homozygosity for the S allele and for the Z allele and detecting heterozygosity for the Mmalton/Mpalermo and Q0ourém alleles. The identification of other rare alleles is performed by DNA sequencing of the entire transcribed region of the  $\alpha$ 1-antitrypsin gene (SERPINA1) whenever reasoned. To date, around 100 different alleles have been identified and classified according to serum levels and molecular function of  $\alpha$ 1-antitrypsin. The M1, M2 and M3 alleles are associated with normal protein secretion (100%) thus not being associated with any clinical pathology. The Z allele is the most frequently identified allele, associated with decreased secretion of  $\alpha$ 1-antitrypsin. It is also possible to identify null alleles, such as Q0, or dysfunctional alleles in which there is a normal serum level of  $\alpha$ 1-antitrypsin but with reduced function. Alpha1-antitrypsin deficiency caused by null alleles is associated with the total absence of the protein and is usually manifested by more severe forms of lung disease. Two cases of  $\alpha$ 1-AD with very low values of  $\alpha$ 1-antitrypsin and whose genetic study was not coincident are described here. A new allele called Q0Santana (Q0S) was then identified which corresponds to a rare variant caused by the deletion of an adenine at codon 278 leading to the formation of a preterm termination codon and the production of a truncated protein. Two cases of  $\alpha$ 1-antitrypsin deficiency were discovered in Madeira Island with identification of Q0S (ZQ0S with levels of  $\alpha$ 1-antitrypsin of 13 mg/dL and M2Q0S levels with  $\alpha$ 1-antitrypsin of 66.8 mg/dL). This variant is not described in the literature. It is an allele which leads to severe deficiency of  $\alpha$ 1-antitrypsin and when in heterozygosity with alleles of severe deficiency it may be associated with the risk of lung disease in adults. In conclusion, research on rare mutations is essential whenever the  $\alpha$ 1-antitrypsin values found do not coincide with the results of the genetic study.

**Key words:**  *$\alpha$ 1-antitrypsin deficiency. A1-antitrypsin. Mutation. New mutation. Q0Santana.*

#### PE 010. 6-YEAR OVERVIEW OF A SMOKING CESSATION PROGRAM IN A CENTRAL HOSPITAL

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**Objectives:** To analyse the characteristics of the population admitted in a smoking cessation program.

**Methods:** We conducted a cross-sectional, retrospective study in which data was collected from clinical records of our smoking cessation consult, between 2011 and 2016. Data was collected about demographic characteristics, pack-year consumption, nicotine dependence (Fagerström Test), motivation to quit (Richmond Test). Statistical analyses were conducted to analyze the data.

**Results:** 669 individuals enrolled the study: 334 males and 335 females. Mean age was  $50.9 \pm 12.2$  years. As it concerns the patients' education level, it was evaluated in 616 patients: 10 patients were analfabet, 277 had basic education, 150 attended high school and 186 attended college. Pack year consumption was divided into three groups, 178 patients consumed less than 30 pack year consumption, 312 between 30 and 59 and 167 had consumption equal or superior to 60. 638 participants had their motivation to quit evaluated according to the Richmond Test that qualifies motivation in to three levels (low -  $n = 313$ , intermediate -  $n = 273$ , high -  $n = 52$ ). Of the total of participants, 611 had their nicotine dependence level evaluated by the Fagerström Test - qualifying their dependence level as low ( $n = 98$ ), intermediate ( $n = 290$ ) or high ( $n = 263$ ).

**Conclusions:** The authors consider that it is important to make a reflection about the population treated in order to know their characteristics making it possible to adjust the approach to that specific population. In a specialized smoking cessation program we have a small number of patients actually motivated to quit, making it highly important to act on their motivation.

**Key words:** Smoking. Motivation.

#### PE 011. PULMONARY INFLAMMATORY PSEUDOTUMOR - HOW RARE IS IT?

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**Introduction:** Inflammatory myofibroblastic tumor (IMT) of the lung, also known as inflammatory pseudotumor are rare pulmonary lesions of unknown etiology. Similar lesions can also occur in other tissues as thyroid, liver, spleen or lymph nodes. Here we present two cases of IMT diagnosed during the past year on a Portuguese university hospital.

**Case report:** A.T., 52-year-old man, non-smoker, presented with cough and mucoid sputum production for half a year aggravated with low fever and weight loss of 2 kg in one month. Complementary study with chest CT showed a right hilar mass. Bronchofibros-copy and transbronchial biopsy of the lesion were performed, with no evidence of malignant cells. PET scan confirmed hypercaptation focus in the described mass. Pursued study with transthoracic needle biopsy that also excluded neoplastic disease. Due to high suspicion of lung cancer, the case was discussed with a thoracic surgery team and Video-assisted thoracoscopic surgery (VATS) was performed for complementary study. During the procedure there was no evidence of neoplasia. Excision of hilar ganglion was performed with extemporaneous examination without signs of malignancy. Lung biopsy documented chronic inflammatory infiltrate with no specific characteristics. Maintained clinical and imaging surveillance, with repeated bronchofibros-copy that showed infiltration of the upper lobar spur mucosa and transbronchial biopsy was performed, with an anatomopathological result compatible with IMT. Reassessed by thoracic surgery, patient underwent right upper lobectomy with definitive pathological anatomy of the surgical specimen showing that the lesion was indeed a pseudoinflammatory tumor. R.A., 63-year-old man with a history of upper left lobectomy due to predominantly lepidic lung adenocarcinoma. During follow-up, thoracic CT was performed and documented a mass in the left costal pleura compatible with pleural metastasis. PET scan confirmed hypercaptation focus in the previously described lesion. Transthoracic needle biopsy was performed and excluded metastasis

of prior neoplasia documenting evidence of fibroblastic/myofibroblast proliferation of probable reactive character.

**Discussion:** IMT are rare pulmonary lesions comprising less than 1 percent of all surgically resected lung anomalies. It is unclear whether these represent a primary inflammatory process or a low-grade malignancy with a significant inflammatory response. Clinical features are diverse, ranging from asymptomatic to nonspecific symptoms as cough, dyspnea, thoracalgia or hemoptysis. The diagnosis requires histologic examination of tissue in order to make a correct differentiation from other malignant or infectious lesions. Transbronchial biopsies are rarely successful, because endobronchial IMT account for less than 5 percent of cases. Usually diagnosis is obtained with surgical resection, which is preferred since complete resection is almost always curative. IMT have a broadly variable history, ranging from a benign course to invasive masses, rarely even with distant metastases. Even after complete surgical resection, long-term follow-up is recommended because there are reports of recurrence for as long as 11 years after resection.

**Key words:** Inflammatory pseudotumor. Myofibroblast proliferation.

#### PE 012. USE OF MITOXANTRONE AS PLEURODESIS AGENT IN PLEURAL EFFUSIONS, SECONDARY TO LUNG CARCINOMA

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**Introduction:** In patients over 50 years of age about 40% of pleural effusions are caused by malignant disease. The most common is lung cancer responsible for 37% of the cases. The treatment is chemical pleurodesis with a sclerosing agent, it is usually palliative and the aim is to improve the quality of life of the patient. Talc is the most common sclerosing agent because it is effective in preventing recurrences success rate of 70 to 100%. However, some associated secondary events are described, namely: dyspnoea, fever, chest pain, atelectasis, pneumonia, arrhythmias, empyema and respiratory failure. The pharmacy of our institution cannot guarantee the sterilization of the talc, so we feel the need to turn to another agent. An alternative sclerosing agent is mitoxantrone.

**Objectives:** This retrospective study aims to describe the use of mitoxantrone as a sclerosing agent in chemical pleurodesis in a group of patients with lung cancer, and pleural effusion.

**Methods:** Patients with pleural effusion secondary to lung neoplasm submitted to chemical pleurodesis with mitoxantrone between 2007 and 2016 were included. A chest tube was inserted and when drainage was less than 200 ml/24h, mitoxantrone was instilled, the dosage, was calculated by the body surface of the patient. The drain was dislodged after 6 hours. We performed follow-up with chest X-rays at 24 hours and every 3 weeks. The success of pleurodesis was defined by the relapse or not of the effusion, and the complications were recorded.

**Results:** We included 24 patients with neoplastic pleural effusion secondary to lung cancer. The overall success rate was 37%. Concerning the complications, there were 3 pneumothorax. No patient was alive at the time of the study.

**Conclusions:** Lung cancer is the leading cause of malignant pulmonary effusion. Approximately 15% of patients with lung cancer have pleural effusion at the time of diagnosis. Pleurodesis is a procedure performed in the palliative context of lung carcinoma. This study was conducted in a group of patients with pleural effusion secondary to lung cancer, and mitoxantrone, as a sclerosing agent, was selected because of the unavailability of talc, and we felt the need to use another agent. The results demonstrate, as in previous studies, that mitoxantrone is a safe agent, without major side effects

or complications for the performance of chemical pleurisy, however, in our study its effectiveness is inferior to talc. The success rate of mitoxantrone in this study is much lower than that found in other similar studies. This may be due to the fact that the tube clamp time is only 6 hours, versus 48 hours in other studies. However, the safe profile of mitoxantrone and the fact that it can be used when the talc is not available is emphasized.

**Key words:** *Pleural effusion. Lung cancer. Mitoxantrone. Pleurodesis.*

#### PE 013. POSITIVE EGFR LUNG ADENOCARCINOMA AND PROGRESSION AFTER TKI- CLINICAL REPORT

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**Introduction:** Adenocarcinoma is currently, the most common histological type of lung cancer. Epidermal growth factor receptor (EGFR) mutation is present in approximately 10% of lung adenocarcinomas. In more than 70% of all cases, these patients tend to respond positively to treatment with tyrosine kinase inhibitors (TKIs). However, TKIs resistance occurs inevitably between 10-14 months after initiating the treatment. Fortunately, in most cases, the progress is caused by the emergence of the resistance mutation, T790m, to which there is targeted therapy, 3<sup>rd</sup> generation TKI. Similarly to the first line therapy, in most cases, the response is very positive during about 10 to 24 months. The rapid progression of T790m positive disease occurs in a small percentage of patients and represents an important clinical challenge.

**Case report:** Woman, 46 years old, secretariat, non-smoker, was admitted in the emergency service of Centro Hospitalar de Leiria in October 2016 for a intense pain in the lower 1/3 of the right hemithorax associated with diminished breath sounds on the right and increased inflammatory parameters. Radiologically had a right base opacification, having been diagnosed with right base pneumoniae and treated accordingly. The patient returned to the emergency service for pain persistence, where it has shown radiological deterioration with pleural effusion and sent to Pneumology for study. Thoracic CT was performed revealing a right hilar mass of irregular contours and perihilar nodules, in addition to the pleural effusion. The PET CT showed hypercaptation of the described masses as well as in the bone. A videobronchofibroscopy was performed afterwards, which biopsies results showed a primitive lung carcinoma, EGFR positive. The staging revealed being a T4N2M1c- stage IVB. In April 2017, therapy with first generation TKI (gefitinib, and afterwards erlotinib because of liver toxicity) was initiated, with excellent clinical response and partial imagological response, being stable until a total of 13 months. By then, a rapid progression was observed, namely pleural with chest wall invasion. The patient was submitted to thoracoscopy for rebiopsy which revealed the presence of T790m mutation, as for it was initiated a 3<sup>rd</sup> generation TKI - osimertinib. Despite targeted therapy, the predictable positive response was not obtained, showing clinical (difficult controlling thoracic pain) and imagological signs compatible with disease progression with need for urgent radiotherapy for pain control. Chemotherapy (platin duplet) was initiated while waiting for results tests trying to identify resistance mechanisms to osimertinib. With the measures applied, there was a slight clinical as well as imagological improvement, that remains until today.

**Discussion:** The rapid progression of T790m positive disease occurs in a small percentage of patients and represents an important clinical challenge, as the case presented. The most common resistance mechanism is C797S mutation and the only one with a therapeutic option with stronger evidence. Unfortunately, in clinical practice, in cases of aggressive disease a prompt approach is needed. With the current therapy, it appears to be a clinical and imagological improvement as the patient waits the results for a better therapy orientation.

**Key words:** *Positive EGFR Adenocarcinoma. T790M. Progression after TKI.*

#### PE 014. ATYPICAL ADENOMATOUS HYPERPLASIA OF THE LUNG, KNOW WHAT?

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**Introduction:** atypical adenomatous hyperplasia of the lung is a premalignant lesion, uncommon condition in the general population. Computed tomography (CT) findings are persistent ground glass opacities. The differential diagnosis should include other lesions with potential malignancy.

**Case report:** a 55 years old female, observed in a pulmonology consultation due to a thoracic CT scan (04/2016) who demonstrated a nodule suggestive of cavitation in the superior lobe of the left lung associated with several ground glass opacities. With constitutional syndrome with 1 year of evolution, she performed several exams at her general practitioner including the cited CT scan. No recent travel history. No night sweats. No relevant personal or family history. She repeated a thoracic CT scan (03/08/2017) with an increase of the lesions: "Dispersed in the pulmonary parenchyma several nodular ground glass opacities, especially in the upper lobes. In the superior lobe of the left lung, we identify a nodule with about 9 mm, blurred boundaries, with apparent air bronchogram - infectious nature? Neoplastic nature? (...)" "Analytically (03/08/2017): normal complete blood count, coagulation, ionogram, renal and hepatic function. Negative autoimmune study. Sedimentation rate and 13mm/h and angiotensin-converting enzyme 69U/L. Respiratory functional study and arterial gasimetry were normal. Videobronchofibroscopy (08/08/2017): Performed B.A.L. in subsegmentar of right b3 with good recovery and bronchial aspirate, without intercurrents: cellularity 82.0/mm<sup>3</sup> macrophages 81.0% Total lymphocytes 5.0%. Culture: negative. Cytology of bronchial aspirate 08/08/2017: negative sample for malignant cells. Therefore it was the patient underwent a surgical biopsy on 18/08/2017, whose result of the pathological anatomy was: "Atypical adenomatous hyperplasia - inferior lobe of the left lung. Tertiary lymphoid tissue - upper and lower lobe of the left lung." Given the premalignant nature of the lesions, the patient remains in follow-up consultation with imaging surveillance of the remaining ground glass nodules.

**Discussion:** Atypical adenomatous lung hyperplasia of the lung is considered to be a precursor lesion of lung adenocarcinoma, which is imagingly revealed as depolarized glass nodules, it is crucial to identify and follow these patients adequately. In the future more scientific knowledge can improve the diagnosis, follow-up and treatment of these patients.

**Key words:** *Atypical adenomatous hyperplasia of the lung. Ground glass opacities.*

#### PE 015. TOXICODERMA SECONDARY TO PEMETREXED: ABOUT A CLINICAL CASE

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**Introduction:** The skin represents one of the major organs affected by the side effects of chemotherapy. Pemetrexed is a multitargeted antifolate drug approved as a single agent or in combination with cisplatin for first-line medical treatment of advanced or metastatic nonsquamous non-small cell lung cancer and is commonly associated with certain adverse reactions as myelosuppression, various digestive tract dysfunctions and cutaneous reactions. Most of the skin reactions occur shortly after pemetrexed administration.

**Case report:** A non-smoking 63-year-old female, non-smoking, was referred to the hospital for cervical/supraclavicular adenopathy and a thyroid nodule. On physical examination, a palpable non-painful swelling was noticed in the left supraclavicular region. Biopsy of the thyroid unveiled a benign nodule. Biopsy of the supraclavicular node revealed adenocarcinoma metastases compatible with a primary pulmonary origin (TTF1 positive, EGFR e ALK negative). Thorax CT showed mediastinal and axillary adenopathy, a 20 mm spiculated nodule in the right upper lobe of the lung and a pericentimetric nodule in the right lower lobe of the lung. PET scan showed a heterogeneous hypercaptation area of 18F-FDG in the left laterocervical region and glycolytic hypermetabolism in mediastinal and axillary adenopathies. The thyroid and the nodules identified in the right lung did not capture 18F-FDG. Transthoracic lung biopsy did not identify any involvement by a neoplastic process. Nerveless, lung cancer (TxN3M1) was considered the most likely diagnosis and the patient started chemotherapy with pemetrexed and carboplatin, having completed 4 cycles with a partial response. After that, pemetrexed was maintained in monotherapy. After 11<sup>th</sup> cycle, violaceous nodules and pruritic skin lesions appeared on the buttocks with later progression to the posterior thighs. Was administered treatment only with a topical corticosteroid. Chemotherapy was suspended with significant improvement of the lesions. Autoimmune and infectious diseases were excluded and a skin biopsy was performed which was compatible with toxicoderma. Given the temporal relationship and exclusion other causes, the most likely diagnosis was toxicoderma secondary to pemetrexed.

**Discussion:** In the case described, toxicoderma presentation was late, contrarily to what is described in the literature, where it typically appears early after drug administration. When dermatologic lesions arise in patients being treated with chemotherapy agent, they may represent a side effect of therapy, but other etiologies need to be considered. So, specific tests such as biopsy may help to guide the diagnosis.

**Key words:** Toxicoderma. Pemetrexed.

#### PE 016. A CASE REPORT OF BRONCHIAL SCHWANNOMA - AN EXTREMELY RARE DISEASE

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**Introduction:** Schwannoma is a benign tumor that arises from Schwann cells that form the neural sheath, usually solitary, found more frequently in the mediastinum and peritoneum and very rarely seen in the bronchic tree. We report the case of a patient with a right lung nodule with characteristics of malignancy, detected in Computed tomography (CT). In the aetiological investigation of this lesion, the anatomopathological examination revealed squamous cell cancer. Bronchofibroscopy detected a nodular lesion in the left main bronchus which revealed a benign schwannoma of the bronchial wall according to the anatomopathological diagnosis.

**Case report:** RR, male, Caucasian, 82, retired taxi driver. Former smoker. Personal history of COPD, hypertension and dyslipidemia. He was asymptomatic up to one year prior to admission, at which point he complained of moderate asthenia, coughing with mucus and wheezy breathing. He denied weight loss, dysphonia, thoracalgia or other symptoms. He performed CT that showed a lesion with spiculated contours of the right upper lobe (RUL) with 25 × 12 mm. Later, while undergoing a bronchofibroscopy, a nodular lesion was detected in the terminal portion of the left main bronchi, and bronchial biopsies were performed at this level. Bronchial biopsy documented a tumor of the peripheral nerve sheath with benign characteristics. PET-CT showed hypercaptation of FDG-18F and a high degree of suspicion of malignancy in the pulmonary nodule in the

anterior segment of RUL with metabolic activity (SUVmax = 9 - > 11). Subsequently, he underwent a transthoracic fine-needle aspiration (TNA) of the RUL node and the cytology obtained was compatible with squamous cell cancer. The patient was referred to oncology and started chemotherapy.

**Discussion:** We report a case of a patient with a benign bronchial Schwannoma whose diagnosis was a finding in a bronchofibroscopy, requested in the context of an investigation of a lesion with spiculated contours of the RUL in a CT. The histology of the lesion in the RUL obtained by TNA was squamous cell cancer but bronchial biopsy documented a schwannoma.

**Key words:** Schwannomas. Endobronchial. Rare disease.

#### PE 017. A RARE CLINICAL PRESENTATION OF SMALL CELL LUNG CANCER

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**Introduction:** Lung cancer can be associated with paraneoplastic syndromes, which can impair several organ functions and manifest as neurological, haematological or endocrinological syndromes, according to the histological type. We report a case of a rare endocrinological paraneoplastic syndrome linked to small cell lung cancer, with prognostic impact that contributed to the adverse clinical outcome.

**Case report:** An 85-year-old man with known history of prostate cancer under hormone therapy, chronic renal disease, arterial hypertension, obesity and a 60 pack-year smoking history. He presented to the emergency department with symptoms of malaise and left thoracic pain. On admission he was eupneic with BP 185/55 mmHg and bilateral lower leg edema. Blood tests revealed leucocytosis ( $15.97 \times 10^9/\mu\text{L}$ ), glycemia 178 mg/dL and hypokalemia (2.5 mEq/L). Chest X-ray showed a right inferior lobe heterogenous lesion. Thoracic CT revealed a nodular lesion in the right inferior lobe, with homolateral and subcarinal lymphadenopathy, narrowing of the intermediate bronchus, middle lobar bronchus and inferior lobar bronchus, as well as a hypodense nodule in the left adrenal gland. The patient maintained refractory hypertension, as well as hyperglycemia and persistent hypokalemia despite adequate supplementation, so further endocrinological tests were made which revealed an increased adrenocorticotrophic hormone (ACTH): 149.0 pg/mL (0-46), serum cortisol: 57.80  $\mu\text{g}/\text{dL}$  (2.90-17.30) and urinary cortisol: > 2,509  $\mu\text{g}/24\text{h}$  (4.3-176). Thus the diagnosis of paraneoplastic Cushing syndrome was made. Cranial CT scan did not reveal pituitary gland involvement. For better assessment of the pulmonary lesion a bronchoscopy was made, which showed an infiltrative lesion of the bronchial tree, and the biopsy revealed the histological diagnosis of small cell lung cancer. He maintained significant leg edema, with diuretic therapy complicated by hypokalemia and worsening renal dysfunction. After multidisciplinary discussion a strategy of palliative radiotherapy was decided, as well as evaluation by the palliative care team. However, the patient had rapid clinical worsening with onset of respiratory failure, and he deceased before starting radiotherapy, four weeks after admission.

**Discussion:** Paraneoplastic Cushing syndrome is a rare entity that results from ectopic ACTH secretion, and can present in patients with previously unknown lung cancer. The two most common histological types are small cell lung cancer and carcinoid tumors. Most patients with this syndrome present at an advanced stage of disease and the prognosis is influenced both by tumor histology and by the severity of hypercortisolism.

**Key words:** Cushing syndrome. Paraneoplastic syndrome. Small cell lung cancer.

### PE 018. POSTERIOR REVERSIBLE ENCEPHALOPATHY SYNDROME AS A PRECURSOR OF LUNG CANCER

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**Introduction:** There are several reports of posterior reversible encephalopathy syndrome (PRES) since it was first reported by Hinchey in 1996. Radiologically, it is characterized by a magnetic resonance imaging of bilateral edema involving the white matter, reaching the posterior vascular territories (parietal lobes and occipital). Clinical findings include headache, changes in consciousness, epilepsy, and nausea. Symptoms and radiological changes are completely reversible when the underlying cause is corrected in a timely manner.

**Case report:** This case refers to an 82-year-old man, with a need for support in his daily life activities, a former smoker of 65 UMA, with a history of controlled hypertension, dyslipidemia, and a 32 mm pulmonary nodule in the left upper lobe in study at the Instituto Português de Oncologia. He was admitted to the neurology department for right hemiparesis, cortical blindness, myoclonias in the right hemisphere, right oculocephalic deviation and blood pressure of 180/100 mmHg. A cranioencephalic computed tomography scan revealed left bilateral and frontal occipital hypodensity of indefinite limits, not suggestive of vascular lesion and a cranioencephalic MRI, which showed suggestive lesions of PRES. During hospitalization a progressive clinical deterioration of the patient was verified. Neurological causes were excluded for clinical deterioration, blood pressure remained controlled from the first day of hospitalization, arterial vasospasm was not identified in a serial transcranial Doppler evaluation, a progressive resolution of the epileptic activity with the institution of antiepileptic therapy was shown, and an infection from the central system was excluded by lumbar puncture. Due to the persistence of clinical deterioration and hematological bronchial secretions, a CT Thorax was requested, which identified a neoplastic lesion of the upper lobe of the left lung, with bilateral and bone lung metastasis. It was transferred to the pneumology department for study. Due to the rapid worsening of the clinical condition, without conditions to conduct a bronchofibroscopy to confirm the diagnosis, it was decided to start palliative treatment and the patient ended up dying on the 10th day of hospitalization.

**Discussion:** PRES is triggered by several factors, including hypertension, renal failure, drugs and autoimmune diseases, but no precise mechanism has been identified. Plausible hypotheses include reduced brain self-regulation and impairment of the blood-brain barrier due to vascular endothelial cell damage, which may result in edematous change. In this case, none of the usual triggering factors have been identified, raising the suspicion of our theory that PRES was probably of paraneoplastic etiology. The mechanisms involved in the paraneoplastic PRES are probably immunomediated and are in the same spectrum of disorders that include paraneoplastic leukoencephalopathy. In the largest retrospective series on PRES, malignancy was present in 32% of the patients, and the paraneoplastic etiology should be considered in cases of PRES with undetermined etiology.

**Key words:** *Posterior reversible encephalopathy. Lung cancer. Paraneoplastic syndrome.*

### PE 019. LOCALLY ADVANCED DISEASE - THE CHALLENGE OF THE THERAPEUTIC APPROACH

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**Introduction:** Despite all the scientific advances in the area, lung cancer is the cancer with highest mortality in Portugal. Being fre-

quently diagnosed in advanced stages, it still entails a reserved prognosis, especially in patients that do not have surgical conditions or indication.

**Case report:** 58-year-old male, retired, former smoker of 50 pack unit years (quit 15 years earlier). Diagnosed with epidermoid lung carcinoma - by bronchial biopsy in a bronchofibroscopy performed in June 2015 - central/hilar inferior nodule of the right lung, invading the hilar structures and part of the mediastinum, with destruction of the 7<sup>th</sup> costal arch with 6 × 7 cm. The rest of the staging exams were negative, thus being classified as a IIIA stage - T4N1M0. We were then sent from another institution to the Pneumological Oncology consult of our hospital to proceed with investigation and treatment. We performed another bronchofibroscopy that showed complete obstruction of the right main bronchus by an irregular mass. He was then submitted to a rigid bronchoscopy: laser photo-coagulation and resection of the mass were performed. It was also observed that the right superior and intermediate bronchi were highly reduced in caliber. In the right lower lobe all the segmental bronchi were permeable, with exception of the apical bronchus that was infiltrated. A bronchial prosthesis was placed in the intermediate bronchus. In a multidisciplinary reunion it was considered that surgery was not an option. In August 2015 the patient was started on first line chemotherapy with carboplatin and vinorelbine - 6 cycles (until January 2016) without complications. Reevaluation with PET-CT showed disappearance of the hypermetabolic lesion of the right lung (with correspondent disappearance of the lesion in the CT component). It presented a low hypermetabolism in the 7<sup>th</sup> costal arch region - compatible with osseous callus. No other foci of hypermetabolism. We opted to remove the bronchial prosthesis, observing mucosal redness in the right basal pyramid and in the apical bronchus which was biopsied - both biopsies came negative. Successive reevaluations since March 2016 have not showed active disease - only maintaining traction bronchiectasis in a postero-basal topography. The patient remains asymptomatic and under periodic surveillance.

**Discussion:** The authors highlight this case as a reflex of an advanced oncologic disease which, with endoscopic treatment and chemotherapy, presented a very favourable clinical evolution, highly uncommon in similar situations. We emphasize the need of a multidisciplinary approach, sometimes with need of using several diagnostic and therapeutic techniques, in order to improve the prognosis and quality of life of these patients.

**Key words:** *Lung cancer. Chemotherapy. Bronchial endoscopy.*

### PE 020. AN ATYPICAL CASE OF NEUROPATHIC PAIN

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**Introduction:** The present case starts with a history of neuropathic pain, a symptom that has developed from a malignant pulmonary etiology. Here, an appropriate anamnesis, objective examination and monitoring of clinical evolution have proved indispensable for an efficient articulation with secondary care.

**Case report:** A 44-year-old female, smoker of 20 pack-years (PY), uses the Emergency Room (ER) for neuropathic pain localized to the right axillary region with irradiation to the shoulder, precordial region and homolateral shoulder blade, which aggravates with thoracic flexion and elevation of the shoulder. It is established the diagnosis of shingles, although the patient does not present skin lesions, being medicated without symptomatic improvement. Three months later, she complains to her Family Physician (FP) of increasingly intense pain with no response to analgesia with opioids and pregabalin in progressively increased doses, at that time with no other associated symptomatology. Once again the therapy is adjusted and she is referred to Neurology and Pain consultations.

Meanwhile she is observed in Neurology consultation, being discharged from it, so that three months later she complains to the FP of persistent symptoms, with the suspicion of a different etiology arising. New exams are requested for investigation, such as mammography, cervical and dorsal spine X-rays and blood tests, which do not present major alterations. It is requested a cervical and thoracic spine CT scan, which reveals an opacification of soft tissue density at the level of the posterolateral segment of the right thoracic cavity, requesting further investigation. A CT scan of the chest is then requested, which reveals an apical pleural thickening to the right upper lobe of indeterminate nature, several dispersed micronodules by the pulmonary parenchyma with a mean size of 2 mm in the right upper lobe (RUL), right lower lobe (RLL) and left lower lobe (LLL) and a 5 mm dominant nodule in the middle lobe (ML), as well as a globous, heterogeneous and substernal thyroid, with 20 mm nodules in the left lobe (LL) and isthmus. At this time, she initiates respiratory complaints with dry cough and weight loss of 8 Kg. Meanwhile, she is referred to the Cardiothoracic Surgery and Pneumology consultations and a pulmonary biopsy is performed, which reveals adenocarcinoma of non-determinable histogenesis (stage T3N0M0) and a PET-CT scan showing an important right apical pleural thickening with intense uptake. In a multidisciplinary team meeting it is decided to perform chest MRI and initiate induction therapy.

**Discussion:** The clinical case here described is a warning for an uncommon presentation of pulmonary adenocarcinoma, which may be easily confused with other etiologies and lead to a delay in the diagnosis, compromising the treatment and eventually the prognosis. Furthermore, it is revealed here the importance of FP in resource management in the health care system.

**Key words:** Adenocarcinoma. Neuropathic pain.

#### PE 021. FEBRILE NEUTROPENIA AND LUNG CANCER - THE REALITY OF A TERTIARY HOSPITAL

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**Introduction:** Febrile neutropenia remains a frequent complication in oncology patients undergoing chemotherapy, compromising disease management.

**Objectives:** To evaluate drug-induced febrile neutropenia in lung cancer patients requiring hospitalization.

**Methods:** Retrospective study of cases from January 2013 to December 2016 in CHLC. Variables were compared using chi-square test, independent T-test, Mann Whitney or ANOVA; statistical significance was admitted for  $p < 0.05$ .

**Results:** 15 cases of febrile neutropenia were identified, of which 86.7% ( $n = 13$ ) were male, with mean age 70.1 years. Within there, 10 cases were of adenocarcinoma, 3 of small cell carcinoma, 1 of large cell carcinoma and 1 of mesothelioma. 46.7% ( $n = 7$ ) were stage IV. Hospitalization had a median of 11 days. The source of infection was respiratory in 60% ( $n = 9$ ) of cases, urinary in 3 cases, gastrointestinal, central nervous system and bacteremia in 1 case. In total, there were 5 microorganism isolations: 3 gram negative agents, 1 gram positive agent and 1 fungi. 7 cases had chemotherapy regimens with intermediate risk for febrile neutropenia (10-20%). According to MASCC Risk Index Score, 40% ( $n = 6$ ) presented with high risk and 20% ( $n = 3$ ) with low risk of complications. 14 cases received broad-spectrum empirical antibiotics of which 2 were de-escalated to directed antibiotics. 1 case was considered palliative. GM-CSF was used for treatment in 1 case. Mortality rate was 26.7% ( $n = 4$ ), with 3 cases from septic shock and 1 from disease progression.

**Conclusions:** Despite the small number of cases, statistically significant associations were found between mortality and the MASCC Risk Index Score ( $p = 0.01$ ), disease stage ( $p = 0.03$ ) and respiratory infection ( $p = 0.036$ ). No statistical differences were found between mortality and age, neutropenia value or duration of hospitalization.

**Key words:** Lung cancer. Chemotherapy. Febrile neutropenia. Mortality.

#### PE 022. NON-SMALL CELL LUNG CARCINOMA IIIA - FIVE YEARS OF REAL LIFE

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**Introduction:** Stage IIIA of non-small cell lung carcinoma occurs in a heterogeneous group of patients for whom the best therapeutic strategy is a multidisciplinary approach with chemotherapy, radiotherapy and surgery.

**Objectives:** To characterize stage IIIA patients with non-small cell lung cancer treated in the last 5 years

**Methods:** Retrospective study of the population of Stage IIIA patients with non-small cell lung carcinoma treated at the Serviço de Pneumologia B do Centro Hospitalar e Universitário de Coimbra between 2013 and 2017. Clinical and demographic data were analyzed using SPSS® software. The size of the sample and the asymmetric percentage of cases censored between the groups did not allow the safe application of Kaplan-Meier curves.

**Results:** Sample comprised of 41 individuals, 75.6% male, mean age 64.5 years. Most patients (70.7%) had a performance status of 1, 48.8% were smokers and 31.7% were ex-smokers. As for histology, 56.1% of the patients had adenocarcinoma, 34.4% squamous carcinoma, 7.3% adenosquamous and 2.4% large cell carcinoma. From the therapeutic point of view, the majority of patients (43.9%) underwent chemotherapy and radiotherapy (9 patients concomitant and 9 sequential), 31.7% surgery and adjuvant chemotherapy, 14.6% neoadjuvant chemotherapy, surgery and adjuvant chemotherapy, 2.4% chemotherapy, 2.4% surgery, chemotherapy and radiotherapy (concomitant), 2.4% chemotherapy, radiotherapy and surgery and 2.4% best supportive care. The chemotherapy most frequently used as the first line was cisplatin with vinorelbine (36.6%), followed by carboplatin with vinorelbine (29.3%) and carboplatin with paclitaxel (19.5%). Of the total, toxicity grade 3 (2 patients) and 4 (1 patient) was reported in 7.3% of patients with the first line of therapy. Patients had median progression-free survival with the first line therapy of 8-months with the most frequent response being the partial response. Death occurred in 43.9% of the patients, with a median overall survival of 17 months. There was no statistically significant difference in mortality according to sex, age, smoking habits, performance status, histology or therapeutic regimen. There was also no statistically significant difference in overall survival as a function of histology or therapeutic regimen.

**Conclusions:** For this sample, there was no statistically significant difference in mortality due to the different clinical variables under study, nor of the therapeutic scheme. Regarding the overall survival, there was also no statistically significant difference in relation to the different histologies nor the therapeutic regimen used. The heterogeneity of this sample is in line with what is known in stage IIIA patients, so the absence of statistically significant differences may be due to the choice of personalized treatment that was made in each case. A larger sample would allow us to draw more reliable conclusions about the different therapeutic strategies.

**Key words:** Non-small cell lung carcinoma. Stage IIIA. Therapy.

**PE 023. ANAPHYLAXIS IN CHEMOTHERAPY**

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Every drug used in chemotherapy has the potential to develop a hypersensitivity reaction, including type one hypersensitivity reactions. At the same time, infusion reactions in chemotherapy are common and do not require immediate discontinuation. The presentation of the latter can share clinical features of hypersensitivity reactions. Recognising reactions that are secondary to mast cell and basophil activation and development of specific IgE is paramount to identify and manage anaphylactic reactions and its potential desensitization. The authors present a case report of life threatening anaphylaxis to cisplatin in a patient with stage four adenocarcinoma of the lung undergoing the fourth treatment cycle. The immediate management and follow up of this case highlight the challenges presented in diagnosis and treatment of anaphylaxis in chemotherapy.

**Key words:** Anaphylaxis. Chemotherapy. Shock.

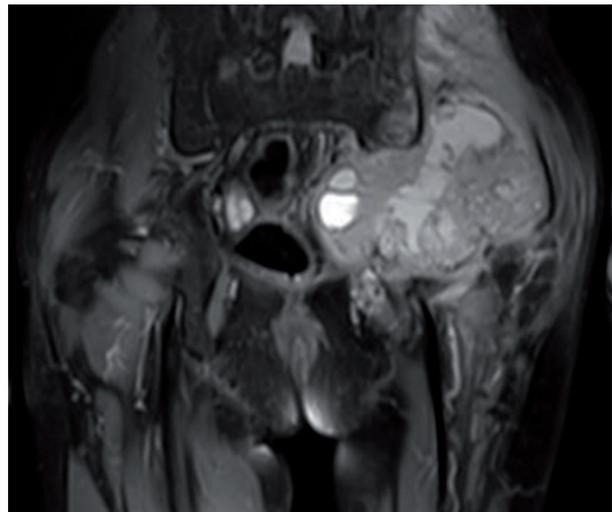
**PE 024. SEARCHING FOR THE PRIMARY LESION**C. Figueiredo<sup>1,2</sup>, B. Mendes<sup>1,2</sup>, S. Amaral<sup>1</sup>, S. Pinheiro<sup>1</sup>, T. Pack<sup>2</sup>, J. Rodrigues<sup>2</sup>, I. Gonçalves<sup>1</sup>, T. Garcia<sup>2</sup>, A.S. Guerreiro<sup>2</sup>, J. Cardoso<sup>1</sup>

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**Introduction:** Soft tissue sarcoma is a rare disease with a severe prognosis. Lung cancer is one of the most common neoplasm and represents the leading cause of cancer mortality worldwide. Paraneoplastic syndromes are rare and occur by a nonmetastatic systemic effect triggered by a malignant disease, for example digital clubbing.

**Case report:** We present a case of a 59-year-old man, smoker (UMA 92), without any previous regular medical care. He was admitted with a tender mass in his left hip with progressive worsening, weight loss for 3 months and complaints of chronic cough with sporadic hemoptoic sputum. On physical examination, the patient was eupneic, there was evident weight loss (BMI 20), a left hip tender mass and exuberant digital clubbing with onychodystrophy of both hands and feet. Laboratory tests without relevant alterations. In the chest radiography, we found bilateral disperse stellate and ill-defined radiopacities. A thoracic and left hip computed tomography were requested and showed multiple pulmonary lesions in various lobes, multiple mediastinal lymphadenopathies and a centred lesion at the left acetabulum with soft tissue component. A hip magnetic resonance was performed and evidenced a bulky mass with an extensive necrotic component, involvement of the iliac bone and adjacent muscles and deviation of adjacent structures. A soft tissue percutaneous biopsy of the left hip revealed adenocarcinoma compatible with pulmonary origin (CK7+, TTF-1+, CK20-; PDL-1 negative). An abdominal-pelvic computed tomography showed mediastinal and retroperitoneal lymphadenopathies, and a cranial magnetic resonance revealed a left temporal lesion, compatible with metastasis. Therefore, the diagnosis was lung adenocarcinoma, stage IV, with metastatic spreading to the lymph nodes, brain and the soft tissues of the hip. Palliative radiotherapy to the brain and soft tissue metastasis was proposed, awaiting the result of EGFR mutation for an eventual targeted therapy.

**Conclusions:** In a smoker with digital clubbing, despite the presentation of an exuberant extra-pulmonary lesion that might indicate primary lesion, one cannot exclude the diagnosis of a primary lung cancer. We reinforce the importance of the clinical history and physical examination in the diagnostic march, as well as the histologic diagnosis in the distinction of the primary lesion and decisions thereafter.



**Key words:** Sarcoma. Lung cancer. Adenocarcinoma. Digital clubbing. Paraneoplastic syndrome.

**PE 025. CONCOMITANT CHEMORADIOTHERAPY IN NONRESECTABLE STAGE III NON-SMALL CELL LUNG CANCER PATIENTS. A CENTER'S EXPERIENCE**P. Americano, S. Lucas, C. Tranvacinha, J. Dionísio, T. Almodôvar  
*IPOLFG.*

**Introduction:** The recommended therapy for non-small cell lung cancer (NSCLC) stage III is concomitant chemoradiotherapy, although the most advantageous chemotherapy regimen, in this context, is not defined. Differing results compare the commonly used

combinations of cisplatin associated with etoposide or a vinca alkaloid, with carboplatin and paclitaxel. In our service, patients are given carboplatin/paclitaxel (CT) or cisplatin/vinorelbine (Pv) associated with radiation therapy. The objective of this study is to compare the efficacy and toxicity of the two therapies, trying to stratify the results according to patients' comorbidities.

**Methods:** All cases of stage III NSCLC, submitted to concomitant chemoradiotherapy in our center, between 01/01/2007 and 12/31/2017, were analyzed. Patients undergoing surgery, isolated chemotherapy or sequential chemotherapy and radiotherapy were excluded. The data were obtained from the thoracic tumors data base and the clinical processes of the patients, and analyzed in the SPSS digital statistical platform, version 24.

**Results:** Of the 252 patients in stage III, 94 patients were included, 70 males, with a mean age of 62 years (between 39 and 78). 48 were stage IIIA and 49 stage IIIB. The histologies determined were: 41 adenocarcinomas, 39 pavement-cell carcinomas, 13 low differentiated NSCLC and 2 adenosquamous. 19 patients had performance status (PS) ECOG 0, 75 PS 1, and 1 patient had PS 2. 57 had no weight loss, 29 had a loss of less than 10% of their body weight and 10 had more than 10%. 58 patients had associated comorbidities, most frequently hypertension (31), chronic obstructive pulmonary disease (24), solid neoplasia (13), and type 2 diabetes mellitus (12). 50 patients were treated with Pv and 44 with CTx. The total doses of radiotherapy ranged from 60 to 70 Gy, usually starting at the 2nd cycle of chemotherapy, but 10 did not complete the dose predicted due to adverse events. A median overall survival of 25.5 months and median progression free time of 12.6 months was found. Twenty-six patients developed chemotherapy toxicity (28%) and 49 (52%) radiotherapy toxicity. The group of patients treated with Pv and CTx were compared in terms of age, PS, weight loss, and comorbidities by the Charlson index with and without age adjustment. There was a slight tendency to use the CTx regimen in older patients, however without statistical significance. The Pv group demonstrated greater overall survival, also without statistical significance. The toxicities were overlapping in both groups.

**Conclusions:** Our results are very similar to those in the literature. Although not statistically significant, there appears to be a tendency to put older patients on a CTx regimen, not being able to prove the benefit of one regimen over the other in terms of both efficacy and toxicity. We highlight the favorable results, with reasonable disease-free and overall survival times, concluding that both schemes are valid options, when well selected according to the patient's profile.

**Key words:** Chemotherapy. Radiotherapy. Concomitant. NSCLC. Lung. Cancer.

#### PE 026. TARGET THERAPY - A PARADIGM CHANGE IN THE TREATMENT OF THE PULMONARY CARCINOMA OF NON SMALL CELLS IN ADVANCED STAGE

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**Introduction:** The development of target therapy significantly altered the prognosis of patients with advanced stage non-small cell lung cancer.

**Case report:** The authors present the case of a 32-year-old man, a former smoker, with a smoking history of 15 pack-units admitted in the Pulmonology Department for fever, cough, chest pain and haemoptysis in February/2016. He presented with hypoxemic respiratory failure in room air and chest CT scan revealed pneumomediastinum, right para-tracheal opacity with tracheal compression (adenopathic conglomerate?), two solid lesions in the right superior lobe, consolidation of right inferior lobe and a hepatic nodule. He was admitted in the Intensive Care Unit and he underwent bronchofibroscopy, which showed neoformative lesion between the trachea

and the right main bronchus, with significant lumen reduction. He was then submitted to rigid bronchoscopy with argon plasma therapy and debulking. The right main bronchus was recanalized and complete occlusion of right inferior lobar bronchus was observed. On the bronchoscopic re-evaluation, a tracheal prosthesis was placed because of partial occlusion of the right main bronchus. The patient was discharged afterwards. The biopsies were consistent with lung adenocarcinoma and the patient started chemotherapy with cisplatin and permethrexed, which was changed to crizotinib due to the presence of ALK rearrangement. Clinical and radiological improvement was evident, tracheal prosthesis was removed and there was complete resolution of both pulmonary and hepatic lesions. The patient shows no sign of recurrence at 21 months of therapy and he is able to work and do his daily activities without limitations.

**Discussion:** This case illustrates significantly the impact of target therapy in advanced stage lung adenocarcinoma, not only in terms of efficacy (given the disease-free survival), as well as quality of life, since this kind of treatment is much better tolerated than conventional chemotherapy.

**Key words:** Crizotinib. Advanced stage non small cell lung carcinoma.

#### PE 027. A RARE CASE OF SYNCHRONOUS TUMOR OF THE LUNG AND MAXILLARY GLAND

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**Introduction:** Lung cancer is the tumor with the highest mortality in Portugal. Patients diagnosed with neoplasia are at increased risk of developing a second tumor. Multiple primary malignant neoplasms are rare and are classified as synchronous if the diagnostic interval is less than 6 months and metachronous if the interval is longer than 6 months.

**Case report:** The authors present the case of a male patient, an active smoker, followed in general surgery consultation by painful submaxillary nodule with greater growth for about 4 months. Reference to weight loss of 30 kg in the last 6 months and marked anorexia. The ultrasound evaluation revealed a solid nodule of the left submaxillary gland and the cytology of the nodule was compatible with basaloid cell neoplasia with cytologic atypia. In preoperative anesthesia evaluation, a chest X-ray revealed a large right hilar mass. In this context, he was referred to a pneumology consultation for etiological research. Due to cachexia and social reason, the patient was hospitalized for staging of probable neoplasia. From the study carried out, the presence of macronodular lesion in the right upper lobe with mediastinal extension and the presence of multiple nodules bilaterally, with mediastinal ganglia of less than 10 mm in the paratracheal and pre-carinal locations, were observed. Abdominal-pelvic CT revealed macronodular lesion of the right adrenal gland and CT-cranioencephalic excluded the presence of secondary lesions. Bronchofibroscopy showed occlusion of the upper right bronchus due to edema, infiltration and extrinsic compression. Trans-bronchial biopsies revealed features consistent with neuroendocrine carcinoma of large cells. The survival of patients with neoplasia has been increasing in recent years, leading to an increase in the incidence of second neoplasms, synchronous or metachronous. This fact correlates with genetic factors of the host, environmental factors, as the exposure to risk factors such as smoking, as well as the oncotic therapy of the first neoplasia and the interaction between all these factors.

**Discussion:** This case alerts us to the importance of the follow-up of patients with primary malignant neoplasia in which, although uncommon, the appearance of a new lesion may correspond to a new primary neoplasm.

**Key words:** Tumor. Lung.

## PE 028. SYNCHRONOUS PULMONARY AND GASTRIC LYMPHOMA

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**Introduction:** MALT (Mucosa-associated lymphoid tissue) lymphomas are extranodal low-grade B-cell lymphomas. Although rare, they are the third most frequent type of lymphoma constituting 5-7% of diagnosed non-Hodgkin's lymphomas. Considering their origin, it is understood that they may arise in virtually any organ being the stomach the most commonly affected site. Although rare, pulmonary MALT lymphoma is the most frequently diagnosed lung lymphoma.

**Case report:** The authors present the case of a 58 year-old male patient, former smoker, with no known history of tuberculosis or autoimmune disease. Patient presented to the Emergency Department with a 3-day course of unproductive cough accompanied by rare episodes of hemoptysis. No fever or other symptoms. The chest X-ray showed an opacity in the lower 1/3 of the left lung field, the laboratory evaluation a  $12.42 \times 10^9/L$  leukocytosis with  $9.82 \times 10^9/L$  neutrophils and elevation of the C-reactive protein to 33.8 mg/dL. He was admitted for diagnostic investigation and started treatment for community-acquired pneumonia with amoxicillin/clavulanic acid and azithromycin. Clinical evolution was good, with resolution of the initial complaints and normalization of the laboratory parameters. Computed Tomography (CT) performed at the pulmonary level showed a heterogeneous consolidation of the left inferior lobe with millimetric nodular hypocaptive images. At the gastric level, there was irregular thickening of the distal stomach wall, with densification and several adenomegalies in the surrounding fat and along the small curvature (the largest centimetric). Based on the radiological changes described, he underwent a bronchoscopy, which revealed no endoscopic lesions. Transbronchial biopsies were performed that showed diffuse proliferation of small lymphoid cells that in the immunohistochemical study were positive for CD20, CD43 and bcl-2. Compatible with MALT lymphoma. He also performed a esophagogastroduodenoscopy that revealed at the level of the gastric body a vegetative protrudent lesion with superficial serpiginous ulcerations. Biopsies were performed which showed infiltration by large and small lymphoid cells positive for CD20, CD43 and bcl-2. Also compatible with MALT lymphoma. After the diagnosis the patient was discharged and referred to the hematologist where he is currently followed-up and has already completed two cycles of chemotherapy with rituximab, vincristine, cyclophosphamide and doxorubicin.

**Discussion:** MALT lymphomas usually have an indolent course and a good prognosis with good response to chemotherapy, progression and transformation into high-grade B-cell lymphoma is rare. When there is multiorgan involvement, as in the case described, it is usually two independent primary tumors and not dissemination. This case emphasizes the importance of an organized and objective diagnostic approach and the correct staging and follow-up of cancer patients.

**Key words:** MALT lymphoma. Hemoptysis. Lung. Stomach.

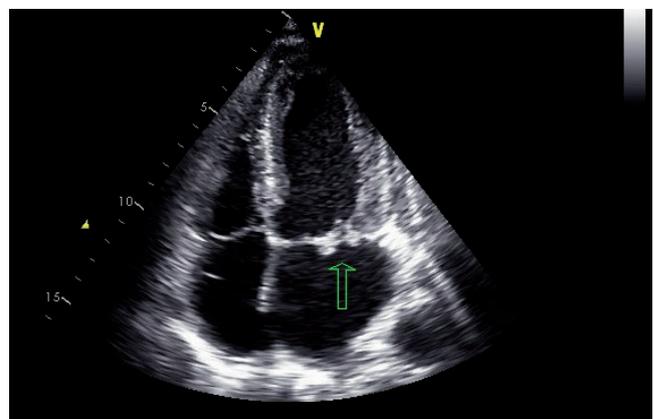
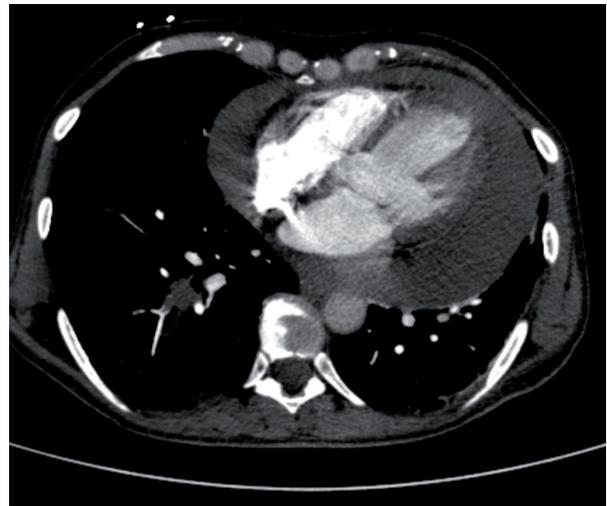
## PE 029. THROMBOPHILIA AS A MANIFESTATION OF LUNG ADENOCARCINOMA - A CASE REPORT

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**Introduction:** Primarily described by Armand Trousseau in 1865, the association of thromboembolic phenomena and malignancy is well documented and can occur in approximately 15% of cases during the clinical course of the disease. These are especially common in mucin-producing adenocarcinoma, including the lung. The cause of

hypercoagulability is multifactorial and includes procoagulant factors associated with cancer cells, as well as an inflammatory response of the host. It can present as Trousseau's syndrome (an extensive, migratory and recurrent venous thrombosis even with anticoagulant treatment), arterial and more often venous thrombosis, disseminated intravascular coagulation or nonbacterial thrombotic endocarditis (marantic endocarditis). Unlike thrombosis from other causes, cancer-associated venous thrombosis is often migratory, may involve superficial and deep veins, and generally affect unusual areas such as the face, arms and chest. Thrombotic endocarditis is a rare event detected in approximately 1-1,5% of cancer autopsies. It is characterized by the presence of thrombi in aortic or mitral valves in the absence of bacterial infection, associated with multiple embolic events, with acute heart failure rarely occurring.



**Case report:** We present a case report of a female patient, 56 years old, smoker (38 UMA), with no other relevant personal history, that was initially admitted to investigate a two month history characterized by weight loss (7 kg), fever, night sweats, cervical and supraclavicular adenopathies, and bilateral acrocyanosis of the 2<sup>nd</sup> and 3<sup>rd</sup> finger. Due to pericardial effusion with hemodynamic instability she was in the intensive care unit, where a diagnostic and therapeutic pericardiocentesis was performed. The complementary exams permitted lung adenocarcinoma diagnosis, obtained from the pericardial fluid and cervical lymph node biopsy, in stage IV with pericardial, pleural, adrenal and extensive supra and infradiaphragmatic lymph node metastasis. It should also be mentioned the paraneoplastic syndrome characterized by venous thromboembolism with evidence of bilateral subsegmental pulmonary thromboembolism and thrombosis of the left brachiocephalic trunk; thrombotic

endocarditis of the mitral valve with moderate mitral insufficiency complicated by multiple arterial embolic phenomena with spleen and bilateral kidney infarction and occlusion of the right common iliac artery; and digital acrocyanosis. After initiation of therapeutic anticoagulation and given the clinical stability, the patient was transferred to the Department of Pulmonology. In spite of the established therapy, there was a progressive clinical worsening with recurrence of thromboembolic events with multiple ischemic cerebral infarctions and subarachnoid hemorrhage, with death occurring at the 31<sup>st</sup> day of hospitalization. She was still awaiting the genetic study of the lung tumor.

**Discussion:** This is a unique case of thrombophilia in a patient with lung adenocarcinoma characterized by extensive venous and arterial thromboembolism, nonbacterial thrombotic endocarditis associated with heart failure, and ischemic and hemorrhagic stroke. We intend to emphasize the multiplicity of possible presentations for this disease, remembering that this is a rare initial manifestation of lung adenocarcinoma associated with poor prognosis.

**Key words:** *Thrombophilia. Venous and arterial thrombosis. Nonbacterial thrombotic endocarditis. Paraneoplastic syndrome. Lung adenocarcinoma.*

### PE 030. DIAGNOSTIC CHALLENGING OF PULMONARY LYMPHOMA: CASE REPORT

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**Introduction:** Primary pulmonary lymphoma (PPL) is very rare, responsible for almost 0.5 at 1% primary pulmonary lung cancer. PPL is defined as clonal lymphoid proliferation affecting one or both lungs in a patient with no detectable extrapulmonary involvement at diagnosis.

**Case report:** We present a case of female patient 70 years age, Caucasian, domestic, as a nonsmoker with no prior history of lung disease, who referred to Pulmonology physician for left thoracalgia, asthenia and anorexia about one year of evolution. Chest CT scans revealed a mass about 10 cm in the LUL (left upper lobe). Bronchoscopy was performed and revealed direct signs of cancer on LUL. Immunohistochemical was negative for AE1/AE3, CK7, CK20, CD56 and Synaptophysin; being only positive for LCA. Patient was admitted later to the Pulmonology Department with dysarthria and prostration. Brain CT scan showed no any alterations but Chest CT scan revealed worsening of pleural effusion. During hospitalization the computed tomography (CT)-guided percutaneous biopsy of mass was performed and revealed only reactivation and inflammation of mesothelial cell. Due to the lack histological diagnosis the Endobronchial Ultrasound (EBUS) was performed. Immunohistochemical analysis showed Lymphocytes B (CD19 + 9) with monotypic expression of Kappa (heavy) chains and aberrant phenotype, compatible with Non-Hodgkin's lymphoma B CD10 + (diffuse large B cell lymphoma - DLBCL). Further examinations of fluorodeoxyglucose (FDG) positron emission tomography (PET)/CT showed left lung neof ormation with homolateral mediastinal involvement. Following the final diagnosis patient was transferred to Hematology department and standard treatment with CHOP (rituximab, cyclophosphamide, doxorubicin, vincristine, and prednisone) was performed. After chemotherapy the patient developed clinical deterioration accompanied by respiratory failure. Patient was admitted to the ICU (Intensive Care Unit) and died three weeks after the standard treatment.

**Discussion:** Non-Hodgkin's lymphoma B CD10 (diffuse large B cell lymphoma - DLBCL) is rare, more frequent in patients receiving post-transplant immunosuppressive therapy, human immunodeficiency virus (HIV) and Gougerot-Sjögren syndrome. Pulmonary diffuse large B cell lymphoma in the immunocompetent patient is

exceptional, represent a diagnostic challenge, since it has several forms of presentation, being confused many times with lung carcinoma. This case clearly illustrates the difficulty in obtaining LPP diagnosis.

**Key words:** *Pulmonary Lymphoma. EBUS. NHL.*

### PE 031. NASAL OXYGEN HIGH FLOW THERAPY - CLINICAL CASE

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**Introduction:** Oxygen therapy by nasal cannula or facial mask is a part of the standard therapy in cases of chronic hypoxemic respiratory failure (RF). However, in cases where high oxygen levels are required, it may have some limitations. Patient discomfort from upper airway drying, difficulty in secretion elimination, difficulty in supplying such high levels of oxygen at home to these patients with advanced disease are examples of such limitations. Nasal Oxygen High Flow (NOHF) is a recent respiratory therapy and is mostly used in a hospital environment in cases of acute hypoxemic failure. It provides high oxygen flow up to 60 L/min through a system composed by a nasal cannula, humidifier and heated circuit. This therapy provides better oxygenation (with lower levels of oxygen), humidification and lung clearance lowering respiratory effort and improving ventilatory efficacy.

**Case report:** Male, 75 years old, Body Mass Index of 27.41 kg/m<sup>2</sup>, retired merchant, ex-smoker for 10 years (50UMA). COPD GOLD D, severe emphysema, type 1 RF on LTOT since 2015. Patient with a good general condition with respiratory symptoms for about 20 years: complaints of dyspnea and fatigue to small efforts. From 2015 to July 2017 there was a marked worsening of the clinical status with multiple and long hospitalizations due to the evolution of the underlying pathology (exclusion of acute factors and therapy optimization). This patient needed very high oxygen levels making the discharge home impossible. NOHF was initiated at a much lower oxygen flow level, with immediate improvement of respiratory pattern, comfort and ability to eliminate secretions.

**Discussion:** NOHF allowed a patient with severe chronic respiratory failure, in a such advanced fase of disease, to be discharged home. This therapy performed continuously at the patients' home contributed to a marked improvement in quality of life and until the moment no further re-hospitalizations.

**Key words:** *Nasal oxygen high flow therapy. COPD. Respiratory failure.*

### PE 032. COPD NURSING APPOINTMENTS

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**Introduction:** COPD is a current issue and a major worldwide pathology that offers countless challenges in the field of Pneumology, as well as in the associated nursing care. Its exponential growth entails high costs for both patients and the National Health Care System, so more investment in COPD nursing care is need to address the needs and specificities of the patients/families. COPD has a negative impact in the lifestyle of the patient/family, causing a decrease in their autonomy and subsequent change in quality of life.

**Objectives:** to let know the COPD nursing consultation, created to promote patient/family empowerment to deal with the pathology, treatment, and to prevent further complications; to decrease

patient/family anxiety; to encourage a change in behaviour and lifestyle adequate to their condition; to be autonomous in the use of inhalers, and finally to promote self-control of the illness. Nowadays, a nurse is the main link between health care and the community, it cooperates with the multidisciplinary team, formulates nursing diagnosis and plans specific and individualized interventions in order to promote health and prevent disease, improving the quality of life of these patients/families. Nursing consultation encompasses four fundamental phases - initial evaluation, preparation of the care plan, implementation of the interventions, and evaluation of interventions. The initial evaluation is carried out at the first contact with the patient/family and aims to collect all pertinent information about them. The preparation of the plan of care is based on the problems and needs identified, is adjusted to each patient and focuses on the achievement of teachings. To do this, we applied two scales, the modified Medical Research Council (mMRC) for dyspnea assessment and COPD Assessment Test (CAT) for determining the impact of COPD on patients' quality of life. In the execution of the interventions, we highlight the countless teachings carried out, emphasizing three key areas for the patient -the use of inhalers, dyspnea control techniques, and energy conservation techniques. Last but not least, the implemented interventions carried out are evaluated. At this stage, we determine the effectiveness of the teachings and adjust the care plan to continue the process in the next consultation.

**Methods:** They are performed predominantly before the medical appointment and whenever their need is identified. The intervention of the nurse aims a holistic approach by performing: initial and general assessment; information gathering to identify patient/family problems/needs; preparation of individualized care plan; accomplishment of teachings that aim the promotion of health and prevention of the disease and clarification of doubts.

**Results:** From June 2017 to June 30, 2018, we performed 388 COPD Nursing consultations.

**Conclusions:** We verified that the nursing consultations promoted both therapeutic adherence and the reduction of the number of errors associated with the use of inhalers, as well as the well-being of the patient/family with COPD.

**Key words:** COPD. Nursing. Nursing Consultation. Teaching.

### PE 033. WHEN COPD DOESN'T EXPLAIN THE SYMPTOMS - A CLINICAL CASE

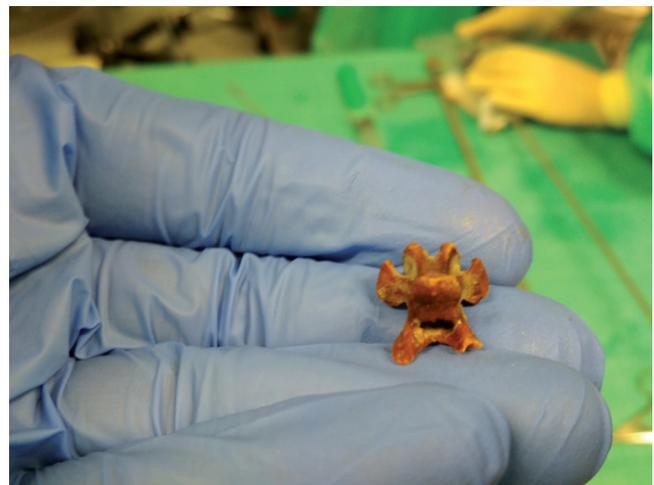
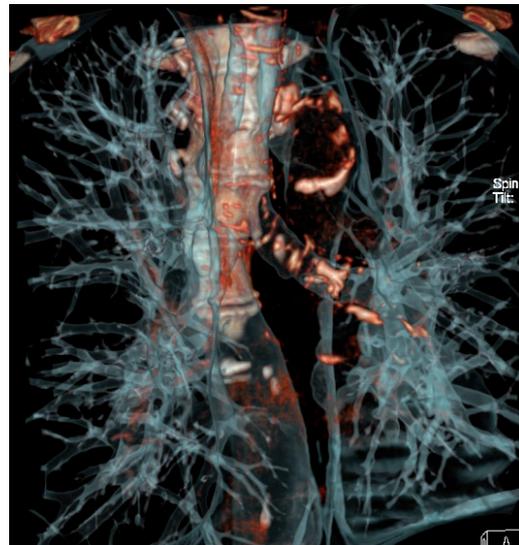
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Hospital de Faro.

**Introduction:** Chronic obstructive pulmonary disease (COPD) is characterized by an airflow limitation and persistent respiratory symptoms. Occasionally, the patients manifest symptoms that are not expected in the natural evolution of the disease, which should make us suspect of a concomitant pathology like a foreign body aspiration. In this case, there is an inhalation of objects or food into the airway, usually resulting in partial or complete respiratory obstruction. An aspiration episode can be a medical emergency with choking hazard but not always is valued by the patient, only being acknowledged afterwards by the manifestations of the foreign body impaction in the airway. 80-90% of the foreign bodies are in the bronchi.

**Case report:** 61 year old patient, male, Caucasian, currently an electrician and a former car mechanic. This patient had a medical history of chronic obstructive pulmonary disease, an ex-smoker (44 pack-year) and dyslipidemia. Followed in pneumology consultation, initially with spirometric values FEV1/FVC 48.42 and FEV1 32.2% post-bronchodilator. The patient had a dyspnea degree mMRC 2 and was medicated with budesonide/formoterol (320/9 µg) bid, acli-

dinium bromide (322 µg) bid, aminophylline (225 µg) bid and salbutamol (100 µg) in SOS. In one of the consultations, the patient had a stridorous respiration, dysphonia and precordial oppression. It was requested collaboration of otorhinolaryngology who opted to perform a laryngoscopy, where no alterations were seen in the visualized structures. In the next consultation there was a worsening in the patient's dysphonia and dyspnea degree (mMRC 3). The new spirometry showed a FEV1/FVC 40.99 and FEV1 25.1% post-bronchodilator. A radiological study was solicited, initially with a chest X-Ray that only showed a diffuse basal and perihilar bilateral infiltrates. Subsequently, after performing a chest 3D CT scan, we found the presence of a perforated foreign body in the left main bronchus, partially obstructing it. The patient performed a rigid bronchoscopy where the foreign body was removed. It was compatible to a chicken's vertebra, measuring 1.5 x 1.5 cm.



**Discussion:** In patients with COPD, even though we should expect a progressive clinical and functional decline, we should always be alert to new symptoms. In this case stridor, precordial oppression and dysphonia appeared which, due to its persistency, led to a more detailed investigation. The fact that this patient already had a severe obstruction may have clinically masked the foreign body aspiration, which led to a more difficult diagnosis, adding the fact that the patient never referred a choking episode.

**Key words:** Chronic obstructive pulmonary disease. Foreign body. Airway.

### PE 034. PROTOCOL FOR A INHALER TECHNIQUE CONSULT IN PRIMARY CARE: A PIONEERING UNDERGOING

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UCSP S. Miguel.

Chronic Respiratory Diseases (CRD) have seen an increase in prevalence, a picture underscored at Primary Care (PC) level in Portugal becoming one of the main causes of morbidity and mortality worldwide. In the Primary Care Medical Center (PCMC) object of this protocol there's already a consult for smoking cessation and a program for spirometry undertaking with a goal to increase early diagnosis and epidemiologic surveillance of CRD. The inhalation pathway is the main mechanism of action for drugs in CRD. There are many inhaler devices, diverging in their physical features, inner workings and way of use, compromising patient adherence and compliance and incurring in a high rate of error associated with its use. An ineffective inhaler technique undermines the control of the CRD, highlighting the significance of learning a correct inhaler technique to the patient with CRD. In the context of PC, the IT instruction is limited by many factors, providing the grounds for the deployment of an IT Consult (ITC) in the PCMC in question. Admission for ITC is established in many institutional routes. ITC's exclusion criteria regards patients undergoing intensive follow-up in the Pneumology Department at the local Hospital. With the main goal of improving the care delivery and health literacy regarding CRD in mind, the specific objectives of ITC are oriented towards the improvement of quality of life (QoL) of the patient with CRD, as well as in the effective control of the pathology by ways of promoting inhaler adherence, starting from a well-understood IT, adjustments of the inhaler device to the patient characteristics and preferences. In the background, the CRD follow-up is analyzed, namely the adequacy of the drugs to the disease, its severity and co-morbidities associated. The patient's characteristics and clinical evolution is quantified for scientific research purposes. Both a nurse and doctor with individual and collective training in the IT division are in charge of the ITC, which is structure in two consecutive pieces. Each segment of the appointment is structure around the patient IT evaluation. The patient IT demonstration starts the appointment in conjunction with placebo devices, with a posterior oral correction associated to a correct physic IT demonstration until the patient repairs the errors made or that these clog the technical teaching. In addition to teaching the motivation of the patient towards the appointment is determined as well as the inhaler device prescription and adherence to the same, the efficiency of the device perceived by the patient, the QoL of CRD, the control of the CRD and the biometric frames, evaluate the functional breathing capacity of the patient, identify the relevant diseases according to the context of the CRD and analyse the follow-up prior to the levels of PC, especially when it comes to the realization of the complementary diagnosis exams and the therapeutic adjustment towards the result of these. The ITC ends with the scheduling of a new appointment according to the needs recomputed and the elaboration of a feedback of the ITC aimed to the Family Doctor of the patient.

**Key words:** Asthma. COPD. Inhaler Technique. Primary Care.

### PE 035. WHEN THE TIGHTS CONFIRM SEVERITY

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**Case report:** We presented a clinical case of a man, 62 years old, current smoker 80 pack-years, with previous diagnosis of chronic obstructive pulmonary disease (COPD) - GOLD 4D with FEV1: 27% (post BD) and DLCO/VA: 52%, under long-term oxygen therapy and noninvasive ventilation. He was admitted at Intensive care unit af-

ter a cardiac arrest with asystole and has a return of spontaneous circulation after 15 minutes of cardiopulmonary resuscitation and he was presented on mechanical ventilation and under noradrenaline perfusion with GCS 6 (O1V1M4). Pulmonary auscultation shown bronchial breath and prolonged expiratory phase. Dermatologic examination revealed circular, and hyperpigmented on the lower thirds of both thighs. These findings were consistent of Dahl's sign. Chest radiograph demonstrated hyperinflation of the lungs and opacification on the right lower lobe suggestive of infectious process. Dahl's sign, also known as Thinker's sign, was first described in a patient with severe COPD in 1963. It characterized by hyperpigmentation caused by persistent pressure from the elbows of patients who spend large amounts of time in the tripod position. This position unloads inspiratory respiratory muscles, especially the diaphragm and leaning forward also increases intra-abdominal pressure, allowing the flattened diaphragm to regain its natural shape improving inspiratory capacity. This condition is reported in not only COPD but also in other conditions including interstitial lung disease, severe asthma, and heart failure.

**Discussion:** Although it is becoming less frequent, this sign can help clinicians to be cautious regarding the severity and chronicity of the obstructive disease.

**Key words:** COPD. Respiratory failure.

### PE 036. PULMONARY HYPERTENSION IN PATIENTS WITH SUSPECTED CHRONIC OBSTRUCTIVE PULMONARY DISEASE

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**Introduction:** Severe pulmonary hypertension (PH) is an uncommon complication in patients with COPD. There are however, other unusual diseases that may prove to be of unexpected importance.

**Case report:** We present a 57-year-old Caucasian woman, teacher. Ex-smoker of 80 units pack/year, with moderate alcohol consumption up to two years before the evaluation. Known diagnoses: benign familial pemphigus and hypothyroidism. She presented a history with two years of evolution of dyspnea for medium effort, fatigue and lethargy. Evaluated for COPD and PH suspected, after hospitalization for cardiac decompensation. However, respiratory functional study did not show exuberant changes (bronchiolar obstruction), with the exception of the diffusion capacity (moderate decrease), with repercussion in the gas exchanges (partial respiratory insufficiency). Laboratory showed altered liver enzymes and liver ultrasound was suggestive of portal hypertension. Computed tomography of the thorax revealed vessels visualized up to less than 1 cm from the pleura, suggestive of dilatation, and it was not possible to exclude the possibility of hepatopulmonary syndrome. Studies of autoimmunity and viral serologies were negative. The patient report prolonged use of erythromycin in relation to treatment of familial pemphigus (to potentiate the development of liver disease?). Right cardiac catheterization revealed pre-capillary hypertension with an increased porto-cava gradient and decreased pulmonary vascular resistance, which is part of the diagnosis of pulmonary arterial hypertension associated with portal hypertension.

**Discussion:** Pulmonary arterial hypertension associated with portal hypertension and hepatopulmonary syndrome are two possible complications of severe liver disease. In the presence of hepatopulmonary syndrome, the balance in favor of vasodilation will have contributed to the intrapulmonary shunt and hypoxemia presented. Dyspnea is a common symptom and non-specific presentation of several pathologies. Due to the high prevalence, COPD tends to be the preferential diagnosis in this presentation (not confirmed in this

patient) when there are risk factors for respiratory disease. However, a detailed diagnostic evaluation is fundamental to exclude other potential causes of dyspnea and to do a correct diagnosis and classification of pulmonary hypertension.

**Key words:** *Dyspnea. Pulmonary hypertension.*

### PE 037. PROGNOSTIC EVALUATION OF EXACERBATION OF CHRONIC OBSTRUCTIVE PULMONARY DISEASE IN A NON-INVASIVE VENTILATION UNIT

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**Introduction:** The APACHE II (Acute Physiology and Chronic Health Evaluation II) scale is used as a predictor of mortality in Intensive Care Units. Higher scores on the APACHE II scale are associated with higher mortality rates. There are also some studies about its use in patients outside the Intensive Care Units and even in exacerbations of Chronic Obstructive Pulmonary Disease (COPD).

**Objectives:** The authors intend to evaluate the role of the APACHE scale in the evaluation of patients with COPD exacerbation admitted to the Non-Invasive Ventilation Unit of the Pulmonology Service of the Centro Hospitalar Lisboa Ocidental from June 2016 to June 2018.

**Methods:** The APACHE II scale was applied retrospectively, primarily evaluating the differences on mortality. Demographic data, previous hospitalizations for COPD exacerbation, nosocomial infection and pCO<sub>2</sub> value were also evaluated. Statistical analysis was performed using SPSS IBM v.25 and Microsoft Excel 2013.

**Results:** Total of 66 patients, mean age of 73.2% ± 9.7, 77.4% of males. The mean APACHE value was 13.9 (values between 9-22). There were 6 deaths, 4 (11.1%) occurred in patients with APACHE scores higher than average. However, the difference was not statistically significant. It was also observed that patients with higher APACHE II scores had higher number of previous hospitalizations due to COPD exacerbation (40.7% vs 25.6%, p = 0.02), higher rate of nosocomial infections (33.3% vs 15.4%, p = 0.04) and higher pCO<sub>2</sub> values (74.1% of patients with higher APACHE scores had pCO<sub>2</sub> ≥ 60 mmHg vs 18.1%, p < 0.01). These differences being statistically significant.

**Conclusions:** The APACHE II scale did not show significant differences regarding mortality in this group of patients. However, the authors admit that this may be due to the small sample size. Previous hospitalizations for exacerbation of COPD and nosocomial infections in patients with higher scores corroborate the overall severity of the underlying disease, and the APACHE scale may be useful in the evaluation of patients with COPD exacerbation. The authors propose the continuity of the application of the APACHE II scale in patients hospitalized in this Unit as an initial assessment tool, and these data can be analyzed in the future with a greater sampling.

**Key words:** *Chronic obstructive pulmonary disease. Exacerbations. Non-invasive ventilation. APACHE II.*

### PE 038. PULMONARY ARTERIAL HYPERTENSION - THE IMPORTANCE OF BELONGING TO THE RIGHT GROUP

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**Introduction:** The etiological diagnosis of pre-capillary pulmonary hypertension may be difficult to establish when multiple pathologies are present. A detailed clinical history is mandatory.

**Case report:** We present the case of a 62 years-old woman, ex-smoker for two years (60 pack-years) with the previous diagnosis of Chronic Obstructive Pulmonary Disease (COPD) with pulmonary centrilobular and panlobular emphysema. Functional assessment revealed a mild obstruction (FEV<sub>1</sub> 1.48 L 81%) and severe DLCO reduction (DLCO 27.5%, DLCO/VA 38.2%). She had partial respiratory failure and was on long term oxygen therapy (4 L/m at rest and 15 L/m on effort). The patient was referred to our Pulmonary Hypertension clinic because of elevated estimated PSAP (67 mmHg) on echocardiogram. She was in functional class III and walked a distance of 240m on 6-minute walk test performed with oxygen (15L/m) with a minimum SpO<sub>2</sub> of 81%. CT angiogram and ventilation/perfusion scan were negative for pulmonary embolism. Sleep study was normal. Right heart catheterization showed a mean pulmonary artery pressure (mPAP) of 46 mmHg, pulmonary capillary wedge pressure (PCWP) of 8 mmHg, diastolic transpulmonary gradient pressure (dTGP) of 24 mmHg, pulmonary vascular resistance (PVR) of 21.8 uWood and cardiac index (CI) of 1.74 L/m/m<sup>2</sup>. Thus, the severity of the pre-capillary pulmonary hypertension was out of proportion considering only a mild obstruction. At this point, the patient revealed she had taken an appetite suppressant (clobenzorex) for one year when she was 20 years old and in her case, pulmonary hypertension was classified as Group 1 and not Group 3. The patient had no symptoms suggestive of connective tissue disease, HIV 1/2 serology was negative. Specific therapy was initiated with two drugs - ambrisentan and sildenafil- with insufficient clinical and hemodynamic improvement: mPAP 38 mmHg, PCWP 4 mmHg, dTGP 18 mmHg, PVR 9.2 uWood, CI 2.4 L/m/m<sup>2</sup>). Selexipag was associated afterwards and the dose was progressively titulated till 3,200 mg/day. Despite triple therapy, the patient remained in functional class II-III with severe hypoxemia and was referred for pulmonary transplantation (on active list at the moment). She was also selected to participate in a phase III randomized, double-blind, placebo controlled clinical study to determine the efficacy, safety and tolerability of the inhaled nitric oxide versus placebo in symptomatic pulmonary arterial hypertension patients.

**Discussion:** In this case, the concomitance of the respiratory disease conditioned the delay of the correct diagnosis and consequently the initiation of the specific therapy, negatively impacting the prognosis. The association between pulmonary arterial hypertension and amphetamine-like appetite suppressants such as clobenzorex has been described since de 1960s, so the history of the use of these drugs should be actively investigated in the patients.

**Key words:** *Pulmonary arterial hypertension. Anorexigenics Group 1.*

### PE 039. RESPIRATORY CARE: TRANSITION FROM ADOLESCENCE TO ADULTHOOD

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*CHUC.*

**Introduction:** The ongoing advances of medical therapies, respiratory home care and greater family support have led to a significant increase in median life expectancy of patients with several chronic diseases diagnosed in pediatric age. Moving from adolescence to adulthood, patients and their families transit from child centred health care to adult oriented, which implies changing hospitals and medical staff. Information exchange is crucial to assure coordinated and uninterrupted health care. The authors aim is to characterize the group of patients transferred, in the past 5 years, from pediatric respiratory care to adult's chronic respiratory insufficiency/sleep disorder consultation in "Hospitais da Universidade de Coimbra (HUC)".

**Methods:** From 1/01/2013 to 1/01/2018, the transfer reports from Paediatric hospital to the respiratory insufficiency/sleep disorder

consultation in Hospitais da Universidade de Coimbra (HUC) were examined regarding patients still attending this appointment.

**Results:** In the considered period, 20 patients were transferred. The identified diagnosis were neuromuscular disorders in 7 patients (35%), various genetic disorders in 9 patients (45%), metabolic diseases in 3 patients (15%) and severe meningitis sequels in 1 patient (5%). Serious cognitive defects were described in 8 patients (40%); 12 patients (60%) had severe mobility difficulties and besides other alterations, 11 patients (55%) had kyphosis. The majority of patients (55%) had cardiac abnormalities. Sleep apnoea disorder was detected in half of the patients for which they were prescribed with non-invasive ventilation. Only one patient was prescribed with long term oxygen therapy.

**Conclusions:** Special health care needs and rare diseases that are less familiar to the pneumologist, were identified in these young adults. Respiratory devices' use was frequent and with a growing tendency as the disease progresses. Clinical comprehension and therapy adhesion may be dependent on the patient's family, since motor and cognitive limitations are often present, making the family-physician relationship essential. These patients' profile is complex, demanding greater effort and dedication from their assistant Pulmonologist, representing a considerable challenge.

**Key words:** Respiratory care. NIV in children. NIV in young adults. Pediatric care transfer.

#### PE 040. NOT AN EZ TASK: TRANSITION TO ADULT CARE OF PATIENTS IN LONG-TERM HOME VENTILATION STARTING IN CHILDHOOD

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**Introduction:** Mechanical ventilation together with other health care and scientific advancements has resulted in more children with life-limiting illnesses, complex chronic conditions and disability reaching adulthood. They represent a high challenge because of the sophisticated technology, financial support needs, and continued health care needs involved. The aim of this study is to characterize children that initiated long-term home ventilation (LTHV) in a pediatric setting and reached 18 years-old or more and to evaluate its evolution and short-term outcomes in the transition process to adult care.

**Methods:** This is a retrospective study with clinical information collected from the clinical charts of all the patients aged more than 17 years-old started on LTHV [CPAP, bilevel non-invasive (NIV) or invasive ventilation (IV)]. In the last 20 years, 191 patients were home ventilated at a single center at a Pediatric Respiratory Unit of a tertiary hospital. Situations are classified as respiratory diseases (Rd) musculoskeletal (MEd), central nervous system (SNCd), genetic syndromes (GS). Descriptive analysis was performed (IBM SPSS, v. 20.0).

**Results:** Thirty-one patients out of 81 (38.3%) that reached adulthood on a program of pediatric home ventilation were successfully transitioned to an adult ventilation clinic, most at the same Hospital. Median age at transition was 21.5 (18-33) years old, 23 (74.2%) were male. Underlying causes were: Rd 10 (32.3%), mostly restrictive disorders, 4 Prader-Willi Syndromes, 1 Klinefelter/Smith Mageinis, 1 cerebral palsy, 2 obesities and 1 bronchiolitis obliterans; MEd in 16 (51.6%) patients, 7 Duchenne muscular dystrophy, 3 spinal muscular atrophy type 2, 2 nemalinic myopathies, 1 myotonic dys-

trophy, 1 central core myopathy, 1 kyphoscoliosis and 1 Albright hereditary osteodystrophy; SNCd in 2 (6.5%) and GS 3 (9.7%). Actually, bilevel NIV was used in 23 (74.2%) patients, CPAP in 4 (12.9%) and IV in 3 (9.7%). Nighttime LTHV was prescribed in 23 (74.2%) patients and 4 (12.9%) were highly dependent patients. The median time on LTHV, since the beginning, from this pool of patients was 9 (2-21) years. In the evolution after transition to adult care 8 (25.8%) discontinued ventilation, 4 patients because of death (1 Duchenne muscular dystrophy, 1 nemalinic myopathy, 2 muscular dystrophies), 2 by non-adhesion, 1 by improvement and 1 by lost to follow up.

**Conclusions:** Despite the challenges LTHV represents, it has been observed that it is possible to maintain this type of respiratory support for long periods. Given the increasing longevity a structured transitional program to adult clinics is mandatory. The preferences, satisfaction and gaps of knowledge to these programs must be accessed.

**Key words:** Long-term home ventilation. Children. Adults. Adult care. Transition.

#### PE 041. SPONTANEOUS PNEUMOTHORAX AS FORM OF COP PRESENTATION

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**Introduction:** Cryptogenic organizing pneumonia (COP) is an unusual condition with an unknown epidemiology. COP has an unspecific clinical presentation and it is frequently confounded with other disorders. Most cases respond well to long term steroid therapy despite the risk of relapse. This case report is about a patient with a secondary spontaneous pneumothorax as form of presentation of COP.

**Case report:** The present clinical case is about a 48-year-old man, smoker (45 pack-years), with history of pulmonary thromboembolism and right-sided spontaneous pneumothorax secondary to emphysema previously drained (years before). He presented to the emergency department with complaints of dyspnea and was diagnosed with left-sided secondary spontaneous pneumothorax. Chest tube drain insertion was complicated with small haemothorax. Control CT scan showed upper lobes emphysema and left apical posterior, lingular and basal pulmonary parenchymal consolidations, with atelectasis, air bronchogram sign and pleural thickening. Despite pneumothorax resolution, he deteriorated his clinical status on the following days with persistent fever, purulent sputum and progressive hypoxemia with no response to 3 courses of broad-spectrum antibiotics. Angio-CT disclosed pulmonary thromboembolism, maintaining the previous alterations. Bronchoscopy with transbronchial biopsies was performed, showing fibroblastic infiltration of the interstitial space, along with the presence of inflammatory cells (polymorphonuclears, lymphocytes and macrophages). Organizing pneumonia was suspected considering the clinical picture, radiological manifestations and absence of response to antibiotics. Associated causes were excluded: the patient had no microbiological findings, drug exposure, pulmonary function tests/laboratory tests were normal (ANA, ANCA, RF, complement and immunoglobulins). Surgical biopsy was not considered before treatment attending the progressive clinical deterioration. Steroid therapy was started with 1mg/Kg/day of oral prednisolone. There was a gradual but significant clinical improvement in the next days with respiratory failure and fever resolution, as well as radiological improvement. 2 months later, CT scan revealed no significant alterations besides known upper lobes emphysema. After 1 year of steroid treatment with slow tapering, he showed full clinical and radiological recovery.

**Discussion:** Pneumothorax is a rare complication/form of presentation of organizing pneumonia. Only 9 cases have been reported so far. This case reveals the importance of considering less frequent

conditions in a patient with clinical deterioration and compatible radiological findings. Organizing pneumonia should be taken into account in these differential diagnosis as long as an attempted therapy leads to good outcomes.

**Key words:** COP. Pneumothorax. Steroids.

#### PE 042. A RARE CAUSE OF SECONDARY ORGANIZING PNEUMONIA

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**Introduction:** Organizing pneumonia (OP) is characterized by the filling of the terminal bronchioles and alveoli by granulation tissue with chronic inflammation. There are numerous causes of secondary OP, including infections, connective tissue diseases and drugs.

**Case report:** A 64-year-old woman, non-smoker, former teacher, with a history of arterial hypertension, valvulopathy with a mechanical mitral valve and chronic sinusitis. No relevant family history or exposition. The patient was admitted twice in the Emergency department complaining of fever, cough and night sweats. She had been treated with antibiotics with a partial clinical response. On February of 2017, the patient was referred to the Emergency department with the same complaints, accompanied with anorexia and weight loss (7% of total body weight) for a three-month period. She was admitted in the hospital ward for etiological investigation. Blood analysis showed CRP of 249.7 mg/L, ESR of 106 mm/h but no leukocytosis or neutrophilia. Chest radiograph presented bilateral heterogeneous infiltrates. Blood cultures and viral serologies were negative. Autoimmune parameters were negative. Infective endocarditis was excluded. Thoracic computer tomography revealed pulmonary micronodules, tree-in-bud lesions, and ground glass consolidation areas with peripheral, basal and bilateral distribution. Sputum culture isolated an *Enterobacter cloacae* complex and the patient was started on piperacillin/tazobactam. Because of the persistence of radiological findings, bronchoscopy was performed with no airway abnormalities. Bronchoalveolar lavage revealed a neutrophilic (33.2%) and lymphocytic (31.2%) alveolitis. BAL microbiological samples were negative, and no malignant cells were identified. Transthoracic pulmonary biopsy showed a pattern suggestive of organizing pneumonia. The patient was started on systemic glucocorticoids (0.5 mg/kg) and was sent to the Pulmonology outpatient clinic. After two months of glucocorticoids, radiological aggravation was noted and it was known the identification of a *Mycobacterium intracellulare* complex (MAC) in the BAL culture. It was assumed the diagnosis of an OP secondary to MAC infection. The patient initiated antimycobacterial therapy with ethambutol, rifampicin and clarithromycin and stopped the glucocorticoids.

**Discussion:** The authors describe this case report because of its infrequency and to illustrate how important is to investigate secondary causes of organizing pneumonia, in order to obtain a favorable treatment response.

**Key words:** Organizing pneumonia. *Mycobacterium avium* complex.

#### PE 043. SILICOSIS WITH ENDOBRONCHIAL INVOLVEMENT - CLINICAL CASE

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**Introduction:** Chronic lung silicosis is the most frequent silica-exposure related disease. Diagnosis relies both on the history of exposure and on altered imaging studies, namely micronodules and calcified adenopathies. Endobronchial involvement is rare.

**Case report:** We present the case of a 58 year-old male, non-smoker, with history of occupational exposure to silica, as a stonemason, without the usage of respiratory protection. Non-respiratory background of lipomatosis and gastroesophageal reflux disease, with grade I esophagitis. Silicosis diagnosed in 2005, at the time presenting shortness of breath for moderate efforts and occasional cough. Image studies showed infracentimetric low-density lung nodules and calcified mediastinal adenopathies. Endoscopically presented bronchial mucosa globally flushed and lung function tests were normal. He kept sustained exposure and progressing imaging alterations. In May/2011, due to haemoptysis, high-resolution computerized tomography (HRCT) was performed and showed multiple nodules and micronodules bilaterally, and mediastinal and hilar adenopathies, some calcified. An opacity suggesting middle lobe (ML) atelectasis was identified. Bronchofibroscopy showed oedematous mucosa, mainly on the left B1+2. In January/2012 he repeated BF, showing glandular hypertrophy, oedematous mucosa and ML bronchus (MLB) stenosis. Bronchoalveolar lavage was performed and it showed multirefringent crystalline particles (probably corresponding to silica), negative microbiological examination and no evidence of neoplastic cells. In 2013 he began to use airway protection and in 2016 he ceased exposure. Since 2013 he showed stable lung lesions in image studies. However he exhibited progressively higher susceptibility to viral infections. In July/2018 he presented with gradual malaise and worsening of the respiratory symptoms, namely cough and dyspnoea for moderate-light efforts. In the HRCT there was evidence of growing parenchymal lesions. Therefore BF was performed to exclude infection and/or neoplastic disease. Multiple vegetated, greyish lesions were detected in the right main bronchus, intermediate bronchus and left



upper lobe bronchus, with biopsies made in the latter location. The mucosa was globally flushed. The MLB stenosis was still present, blocking the passage of the BF. Microbiologic study showed normal flora, with negative direct mycobacterial exam. Histologic exam showed bronchial mucosa coated by ciliary cylindrical epithelium without atypia and the underlying chorion was occupied by nodular aggregates of histiocytes with cytoplasmic debris, accompanied by discrete fibrosis and capillary-type ecstasic vessels, aspects compatible with silicotic nodules. No granulomatous tissue was identified. It was not possible to search for birefringent crystals.

**Conclusions:** We present this chronic silicosis case for its evolution to an atypical and rare involvement, with few similar cases published. Some of these few cases also presented with middle lobe syndrome simultaneously, secondary to the endobronchial changes from silicosis. We also highlight this entity as a rare differential diagnosis in this clinical frame, where the most probable aetiology would be either infectious or neoplastic.

**Key words:** *Silicosis. Endobronchial nodules.*

#### PE 044. WHEN LUNG NODULES DECEIVE - SUSPECTED NEOPLASIA THAT TURNED INTO DIAGNOSIS OF PULMONARY SARCOIDOSIS

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**Introduction:** Sarcoidosis is a systemic granulomatous inflammatory disease with pulmonary involvement in most patients. It pres-

ents more frequently in the form of multiple small nodules, however, in 15% to 25% of the cases large nodules can be found mimicking neoplasms. In the early stages irregular dense opacities surrounded by ground glass may also appear on the pulmonary periphery.

**Case report:** 69-year-old female, non-smoker, referred to rapid pneumological diagnosis consultation with a spiculated pulmonary nodule and right pleural effusion identified in abdominopelvic CT performed in the context of low back pain and hemoglobinuria. Patient reported fatigue and dyspnea for medium efforts, dry cough and unquantified weight loss with 1 month of evolution. She was eupneic at rest with peripheral oxygen saturation of 96%, the pulmonary auscultation was normal. History of IgG K monoclonal gammopathy, arterial hypertension, dyslipidemia, renal lithiasis, cholelithiasis, hepatic angioma and uterine myoma with hysterectomy and annexectomy. On chest CT, on the right lower lobe (RLL), there was a 22.5 mm justadiaphragmatic spiculated nodule and a 31.5 mm irregular dense area in the posterior basal segment (Fig. 1). On the left upper lobe (LUL) 2 smaller nodules were observed with adjacent ground glass pattern (Fig. 2), without adenopathies or pleural effusion. Due to the strong suspicion of lung tumour, primitive or secondary, PET/CT was made, revealing hypercaptation of F-18-FDG in the justadiaphragmatic nodule on the RLL (SUVmax 8.84) and in the bigger nodule on the LUL (SUVmax 3.42), supporting the hypothesis of malignant origin. A transthoracic aspiration biopsy was then performed on the nodule of the RLL, but the anatomopathological analysis revealed a chronic granulomatous inflammatory process with areas of necrosis and fibrosis. Suspected aspects of neoplasia were excluded and differential diagnosis of tuberculosis and sarcoidosis was suggested. No microorganisms were observed, namely fungi or mycobacteria. The videobronchofibroscopy, with bronchoal-

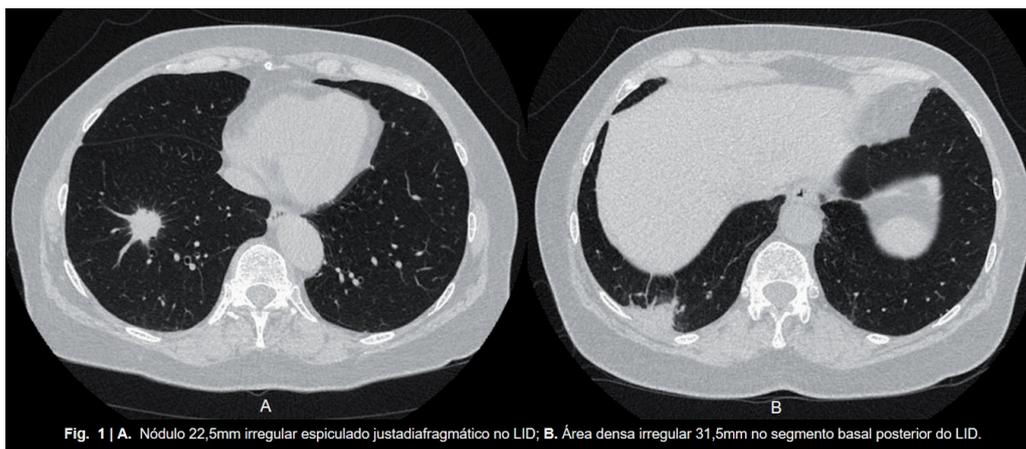


Fig. 1 | A. Nódulo 22,5mm irregular espiculado justadiafrágico no LID; B. Área densa irregular 31,5mm no segmento basal posterior do LID.

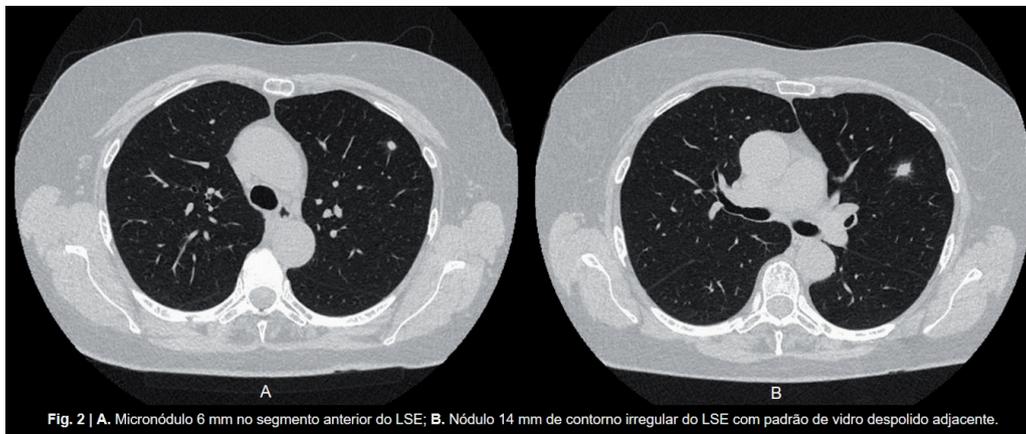


Fig. 2 | A. Micronódulo 6 mm no segmento anterior do LSE; B. Nódulo 14 mm de contorno irregular do LSE com padrão de vidro despolido adjacente.

veolar lavage of the RLL bronchus, revealed 100 cells/uL, mild lymphocytic alveolitis (23%) and CD4/CD8 ratio of 8.1. Ziehl-Neelsen stain, *Mycobacterium tuberculosis* DNA and cultures in liquid and solid Lowenstein-Jensen medium were all negative. Dosing of serum angiotensin converting enzyme was normal (51.6 IU/L). The combination of these data led to the diagnosis of pulmonary sarcoidosis, without lymph node involvement. There was no evidence of involvement of other organs by the disease. Oral steroids were introduced at the initial dose of 60mg of deflazacort, with a monthly reduction of 15 mg. The patient is currently stable and awaits reevaluation by chest CT.

**Discussion:** The clinical and radiological features allow us to narrow the differential diagnosis of pulmonary nodules, however, the histological characterization is necessary to confirm the final diagnosis. We present this clinical case for the pseudotumoral presentation of sarcoidosis that triggered the diagnostic investigation and inevitable concern of the medical team and the patient. The metabolic characterization of the lesions by PET allowed the selection of the ideal biopsy location. The presence of well-formed granulomas, very high CD4/CD8 ratio and exclusion of tuberculosis were factors in favor of the diagnosis of Sarcoidosis.

**Key words:** Lung nodules. Sarcoidosis. Transthoracic biopsy. Bronchoalveolar lavage.

#### PE 045. HEMOPTYSIS: A DIFFICULT APPROACH TO THYMUS LYMPHANGIOMA

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**Introduction:** The main causes of hemoptysis in adults are acute or chronic respiratory infections, bronchiectasis and lung neoplasia. In rare cases they may result from congenital arterio-venous or lymphatic malformations. In most cases, the approach consists of medical therapy.

**Case report:** A 40-year-old woman, former smoker in a social context, with a history of thymus lymphangioma with recurrent chylopericardium submitted to thymectomy and thoracic channel lymphatic at 10 years of age. Referred to for a 3-year history of recurrent hemoptysis with progressive aggravation, not associated with respiratory infections or constitutional symptoms. She also reported wheezing, exertional dyspnea and paroxysmal nocturnal dyspnea. On physical examination, pulse oximetry with peripheral oxygen saturation of 94% in ambient air, decreased vesicular murmur and hypophonic cardiac tones were noted. From the complementary study, viral serologies, as well as direct and cultural examination for sputum mycobacteria, are negative; Thoracic computed tomography (CT) with peri-bronchovascular densification in the pulmonary parenchyma, suggesting a lymphangiomatous process, stenosis of the bronchial emergency due to thickening of the central bronchovascular sheath, anomalous bronchial arteries on the right and calcification of the apparently sequelar pericardium; PET-CT without metabolically active uptake; Bronchofibroscopy with diffuse edema of the mucosa, without bleeding focus. Respiratory functional study compatible with mixed ventilatory disorder (FEV1 23%) and reduction of DLCO (24%). She was medicated with anti-fibrinolytics, antitussives, bronchodilators and inhaled corticosteroids without clinical improvement. After hemoptysis of difficult control, angiography with embolization of the abnormal bronchial arteries was performed, and in the context of an acute constrictive pericarditis, the patient underwent pericardiectomy. Histology of the operative part showed recurrence of mediastinal lymphangioma

with involvement of the pericardium and the middle lobe. Per persistence of recurrent hemoptysis, she initiated diet poor in long chain triglycerides with poor adherence. She is currently medicated with systemic corticosteroid therapy in low dose and monthly intramuscular octreotide having achieved clinical stability.

**Discussion:** Lymphangiomas result from a congenital alteration of lymphatic development. They are most common in childhood and are typically located at the cervical level. In the initial and localized stage, radical resection is usually curative, with recurrence being very rare. In the diffuse forms of the mediastinum, there is no established therapy. The use of somatostatin analogues aims to inhibit lymphatic secretion through specific receptors in the lymphatic vessel and decrease the flux, contributing to the stabilization of this disease.

**Key words:** Recurrent thymus lymphangioma. Haemoptysis. Octreotide.

#### PE 046. PULMONARY INTERSTITIAL DISEASE AS A MANIFESTATION OF COMMON VARIABLE IMMUNODEFICIENCY SYNDROME

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**Case report:** The cases of two patients with lymphocytic interstitial pneumonia (LIP) associated with Common Variable Immunodeficiency Syndrome (SICV) are presented. The first case is of a 48-year-old woman with a previous history of hypertension, non-smoker, and without relevant exposure factors. Referred to the Pulmonology department because of recurrent respiratory infections and exertional dyspnea. Of the study carried out to highlight: 1) Analytical evaluation: IgG and IgA deficiency; 2) PFR: slight restriction and decrease of DLCO (53%); 3) PM6M: walked distance 81% of predicted, minimum SO2 86%; 4) Thorax-CT: Bilateral bronchiolentric micronodular densities, with small consolidations with air bronchogram. Hilar, paratracheal, and subcarinal adenopathies; 5) Bronchofibroscopy without endoscopic alterations, lymphocytic alveolitis (52%); 6) LIP-compatible surgical lung biopsy. She was referred for hematology consultation and the diagnosis of SICV was confirmed. She started treatment with prednisolone 40 mg/day and anti-pneumococcal vaccination. After 5 months of therapy, clinical, functional and imaging improvement were observed, allowing the weaning of the systemic steroid dose. She has been in follow up for for 4 years with a maintenance dose of 10 mg/day of prednisolone due to recurrence of interstitial lung disease at lower doses. Currently, there has been an increase in the frequency of infectious exacerbations and the patient is being evaluated for immunoglobulin replacement therapy. The second case is from a female patient, 41 years of age, diagnosed with asthma since childhood. The patient is referred to Pulmonology for progressive worsening dyspnea and recurrent respiratory infections. Of the study carried out to highlight: 1) Analytical evaluation: IgG and IgA deficiency; 2) PFR within normal range but with reduction of DLCO (51%); 3) PM6M: distance covered 61% of predicted, minimum SO2 95%; 4) CT-thorax: peri-bronchovascular densities in ground glass appearance; 5) Inconclusive bronchofibroscopy; 6) Pulmonary surgical biopsy: compatible with LIP. Immunosuppression with prednisolone 10 mg/day and azathioprine 25 mg/day was initiated and the patient was given anti-pneumococcal vaccination. She was referred to the hematology clinic where the diagnosis of SICV was confirmed and monthly human immunoglobulin was initiated. Improvement in dyspnea and effort tolerance as well as radiological improvement were observed allowing suspension of azathioprine. There was no possibility of reduction of prednisolone dose in the 7 years of follow-up due to

recurrence of interstitial lung disease, similar to the first case. The patient sustained recurrent respiratory infections, including prolonged hospitalization for spontaneous pneumothorax, invasive pulmonary aspergillosis, nosocomial pneumonia with septic shock, and the need for invasive mechanical ventilation.

**Discussion:** LIP is the most common cause of interstitial lung disease in patients with SICV and may affect 10 to 30% of these. In spite of the lack of large studies on this association, some studies point to an important increase in morbidity and reduction of survival compared to patients with non-LIP SCID. Infectious complications constitute a challenge in the management of these patients, contributing to the difficulty in total remission and recurrence of pulmonary interstitial disease, forcing a rigorous follow up.

**Key words:** Pulmonary interstitial disease. Common variable immunodeficiency Syndrome. Lymphocytic. Interstitial pneumonia.

#### PE 047. BLOOD HYPEREOSINOPHILIA: DRUG-INDUCED, VASCULITIS OR BOTH? - CLINICAL CASE

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**Introduction:** The causes of hypereosinophilia (HE) are numerous and are conventionally divided into three main categories: reactive, primary and idiopathic. Blood HE should prompt a thorough evaluation for an underlying cause but this identification sometimes can be difficult. HE can cause serious, potentially life-threatening complications.

**Case report:** The authors describe a clinical case of a 48-year-old male former smoker of 5 pack units/year with allergic rhinosinusitis, bronchial asthma and post-tuberculosis bronchiectasis. In May 2017, he presented hemoptysis that led to hospital admission. Treatment for probable pulmonary tuberculosis (TB) with HRZE was started after Nucleic Acid Amplification Test (NAAT) identification of *Mycobacterium tuberculosis* (Mt) on bronchial secretions (direct and cultural examination was negative). After anti-TB therapy there was development of mild peripheral HE not previously existing. Following the transition to the maintenance phase with HRE (August 2017), the patient presented clinical and radiological deterioration with diarrhea, abdominal pain and limbs paresthesia that led to new hospital admission. Analytically, he presented severe HE ( $26.1 \times 10^9/L$ ), which led to antituberculosis therapy interruption and corticoid therapy. Target organ endpoint with intense peripheral polyneuropathy was confirmed on electromyography and treated with pyridoxine and carbamazepine. Asymptomatic cardiac involvement with elevated Troponin-I, prolongation of QT interval and normal echocardiogram was observed. After hospital discharge, the patient remained under surveillance with maintenance of peripheral HE ( $> 1.0 \times 10^9/L$ ), despite the previous interruption of the antibacterials. Thus, a diagnostic march was started to exclude other possible causes. From the etiological investigation carried out it was verified: negative parasitological examination of feces; negative serologies (HIV, Syphilis, HBV, HCV, CMV); negative autoimmune study except ANCA-proteinase 3; Total IgE 116 IU/ml; normal vitamin B12; normal peripheral blood phenotyping; search for FIP1L1-PDGFR $\alpha$  negative. Of the remaining evaluations of target organ performed, there was a sequential chest radiographs with evidence of migratory infiltrates; videobronchofibrosocopy without endobronchial lesions with direct, cultural and NAAT examination for Mt negative; severe obstruction with positive bronchodilation on lung function test; upper gastrointestinal endoscopy with Barrett esophagus and duodenal ulcer (histology with slight superficial erosion without other changes, *Helicobacter pylori* not identified); normal urinalysis. The diagnosis of probably eosinophilic granulomatosis with poly-

angiitis was admitted after encompassing the clinic, past history (rhinosinusitis, bronchial asthma), presence of pulmonary migratory infiltrates, maintenance of blood HE, and the exclusion of other etiologies. The patient performed a nasal biopsy that revealed vascular congestion without other histological changes. The patient is currently being monitored under corticosteroid therapy.

**Discussion:** The authors describe this case by their difficult diagnosis, initially masked by the possibility of an adverse effect to the antibacterials. The patient presented improvement with corticosteroid therapy, however, maintaining target organ count.

**Key words:** Hypereosinophilia. Vasculitis. Eosinophilic Granulomatosis with polyangiitis.

#### PE 048. TUBERCULOSIS DEMOGRAPHIC DATA, COMORBIDITIES AND DIAGNOSIS IN A RESPIRATORY MEDICINE DEPARTMENT

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**Introduction:** In 2017, tuberculosis incidence in Portugal was 15,4/100.000 inhabitants and 57,3% of the notifications were from Oporto and Lisbon districts.

**Objectives:** To analyze demographic data, comorbidities and biological products used for tuberculosis diagnosis in patients admitted to a Respiratory Medicine Department in a hospital which serves one of the areas with highest tuberculosis incidence in Portugal.

**Methods:** Retrospective analysis of clinical and laboratorial data from patients admitted on a Respiratory Medicine Department for 5 years.

**Results:** The study included 68 patients, the majority male (69%). The mean age was 44 years old (minimum 18 and maximum 87). Águas Livres (16%), Rio de Mouro (12%), Algueirão-Mem Martins (10%), Mina de Água (10%) and Cacém-São Marcos (9%) were the parishes where more cases were identified; 29 patients (43%) were born abroad (8 were living in Portugal for less than 2 years). In the total sample, 16% of the patients were from Angola, 12% from Cape Verde and 9% from Guinea Bissau. 19% of the patients were unemployed, 15% retired, 10% worked in construction, 6% in catering and 6% in shopkeeping. Almost one third (31%) of the patient were active smokers (10% were ex-smokers) and 21% drank more than 2 alcohol units daily. There were two active cannabinoids users and to ex-drug addicts (one heroin and the other cocaine). Other comorbidities included COPD (7 cases, 10%), type 2 diabetes non-insulin dependent (5 cases), chronic kidney disease (3 cases), malignancy (2 cases), one patient with Crohn's disease and another with HIV infection. One patient died due to nosocomial pneumonia. Almost all patients had pulmonary involvement, 2 had only pleural involvement and 12 (18%) had pleural-pulmonary involvement. There were 3 cases of endobronchial involvement and one suspected case of laryngeal involvement. Sputum examination allowed tuberculosis diagnosis in 56% of the patents (positive culture in all, 84% with positive direct examination). Bronchial secretions collected during bronchoscopy confirmed the diagnosis in 32% of the cases; 3 patients underwent bronchial biopsies: one had positive cultural and direct examination and 2 had suggestive histology. Other 10 patients underwent thoracocentesis (negative cultural and direct examination), 9 of which had adenosine-deaminase over 40 U/L (mean value 87 U/L); 6 patients underwent pleural biopsy, 5 evidencing granulomatous pleuritis, 2 of them with positive culture. When analyzing the sensitivity profile, 85% were multiresistant tuberculosis, 2 cases were resistant to isoniazid and one was multidrug-resistant.

**Conclusions:** The studied population had some peculiarities, with a high percentage of immigrants and higher unemployment and alcoholism rates than the national average. More than half of the

patients had positive direct examination; bronchoscopy was necessary in about one third of the cases.

**Key words:** Tuberculosis. Epidemiology. Mycobacteriology.

#### PE 049. TUBERCULOSIS TO THE SKIN SURFACE - A CASE REPORT

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**Introduction:** Despite effective forms of diagnosis and treatment, tuberculosis remains a public health issue. In most cases, the disease courses with pulmonary damage, but it can spread to almost any organ, potentiating a variety of symptoms and clinical manifestations. Tuberculids are rare skin lesions that result from a late hypersensitivity reaction to tuberculin and may have a multiplicity of presentations ranging from indurated papules to necrotic inflammatory lesions. Its diagnosis is based on dermatological manifestations and anatomopathological findings, since cultural examination, molecular detection or Ziehl-Neelsen stain are commonly negative. The following clinical case reports the evolution of a patient with pulmonary tuberculosis and cutaneous lesions compatible with tuberculids.

**Case report:** 60 years old Asian man, non-smoker and with a history of diabetes mellitus and hypertension. Asymptomatic in May 2017, he performs a chest X-ray where a pulmonary infiltrate was identified at the right apex, compatible with tuberculosis. At that time no diagnostic investigation was performed. In May 2018, patient reports a clinic of non-productive cough and weight loss. Repeated chest X-ray, documented a worsening of pulmonary infiltrate. Additionally, he describes the appearance, about 6 months ago, of pruritic cutaneous lesions, with preferential distribution in the chest and limbs, already evaluated by dermatology and persistent despite multiple topical therapies. Due to the high suspicion of tuberculosis, a sample of bronchial secretions is collected but no etiological agents were identified. Diagnostic workup progressed with bronchofibroscopy, evidencing edema-like mucosa and presence of purulent secretions in the right upper lobe, where samples were collected. Regarding the cutaneous lesions, patient was evaluated by dermatology that described a pruritic lichenoid erythematous rash and violaceous papules, and performed a biopsy for complementary study. Obtained positivity in the molecular detection of *Mycobacterium tuberculosis* complex in bronchoalveolar lavage. Anatomopathological study of a cutaneous sample showed an histiocytic infiltration and histochemical evaluation excluded the presence of microorganisms. Patient was oriented to the pneumological diagnosis center where he initiated therapy for tuberculosis. Favorable evolution with improvement of general symptomatology, respiratory symptoms and manifest regression of cutaneous lesions.

**Discussion:** Tuberculosis has a wide variety of presentations and may be present with more or less pronounced manifestations, usually with a dragged evolution. Pulmonary attainment is the most common, with respiratory symptoms dominating the clinic; however, other manifestations may be present and should be explored. Tuberculids are rare lesions that occur in less than 5% of cases of active tuberculosis, of unknown pathogenesis, but commonly considered as the result of cutaneous hypersensitivity reactions to *M. tuberculosis* in patients with high levels of immunity to the agent. As observed in this case, the non-identification of bacilli in cutaneous biopsy is common, but in the presence of active disease with extracutaneous identification of *M. tuberculosis*, the histopathological evidence of granulomas and the improvement of lesions with antibacterial therapy support its diagnosis. Under appropriate treatment, the disease has a favorable evolution and cure.

**Key words:** Tuberculosis. Tuberculids. Hypersensitivity reaction.

#### PE 050. CANDIDA LUSITANIAE - A REAL TROJAN HORSE

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**Introduction:** *Candida* species are isolated with high frequency from cystic fibrosis patients, yet their definitive role in the disease remains unclear. Previously considered to have minimal inherent virulence owing to their commensal ability, the last decade has heralded an increasing recognition of *Candida* infection among patients with cystic fibrosis. *Candida lusitaniae* has been a rare opportunistic pathogen, but when isolated it has caused serious and frequently fatal disease.

**Case report:** A 47 years old female non smoker with a diagnosis of cystic fibrosis since her 43 years old (F508del/V232D), whose pulmonary involvement is manifested by multiple diffuse bronchiectasis, preferentially located in the right upper lobe and middle lobe with intermittent colonization by *PSAE* and *Mycobacterium avium*. She presented with one week history of fever, increased volume and purulence of sputum, with dyspnea for medium efforts. Blood and sputum cultures were collected for microbiological examination and she was empirically medicated with amikacin and meropenem. There was a progressive worsening of the clinical condition with persistent fever, tachycardia, polypnea and hypoxemia with increasing need of oxygen supply (paO<sub>2</sub> 60 with FiO<sub>2</sub> 60%), and the patient was admitted to the ICU where started high-flow nasal cannula (HFNC) oxygen therapy. It was performed a chest CT that revealed "pulmonary parenchyma consolidation in the left lower lobe, other areas of pulmonary parenchyma densification in different lobes with tree in bud pattern" and bronchofibroscopy to collect products for microbiology, showing off-white nodular lesions along the posterior wall of the trachea and purulent secretions in the left bronchial tree. It was isolated both in secretions, bronchoalveolar lavage and in the bronchic biopsies *Candida lusitaniae*. Given the diagnosis of invasive candidiasis to *Candida lusitaniae*, she started treatment with fluconazole with major clinical and radiological improvement, reducing the need for oxygen therapy, allowing the weaning of high-flow nasal cannula oxygen therapy to conventional oxygen therapy at 3L/min by nasal cannula. At discharge, she was on the 7th week of fluconazole therapy, well tolerated after an initial phase with hepatic toxicity that was reversed with a short interruption of therapy.

**Conclusions:** Although *Candida* is a commensal specie, serious and potentially fatal infections by these microorganisms have been increasing in the last decades. *Candida lusitaniae* should be considered an opportunistic pathogen when isolated in the appropriate clinical setting. Some common risk factors in patients with these infections are: underlying malignancy, prolonged administration of broad-spectrum antibiotics, prolonged hospitalization, intravascular catheters, cytotoxic or corticosteroid therapy or granulocytopenia.

**Key words:** Candida. Invasive candidiasis. Cystic fibrosis. High-flow nasal cannula oxygen therapy. Fluconazole.

#### PE 051. TUBERCULOSIS PLEURAL EFFUSION UNDER ANALYSIS: REAL-LIFE STUDY

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**Introduction:** Tuberculosis (TB) remains one of the most frequent causes of pleural effusion in Portugal. The definitive diagnosis of TB pleural effusions depends on the demonstration of *Mycobacterium*

*tuberculosis* (MT) in pleural fluid or pleural biopsy specimens. Supportive evidence for presumptive diagnosis includes elevated adenosine deaminase (ADA) levels and pleural fluid lymphocytosis (> 50%) as well as demonstration of TB necrotizing granulomas in the pleura.

**Objectives:** Describe aspects related to tuberculosis pleural effusion in our population.

**Methods:** Observational retrospective study of all patients with final diagnosis of pleural tuberculosis after investigation of newly detected pleural effusion referred to our outpatient respiratory/pleural clinic in a time period of five years.

**Results:** The study included 21 patients with pleural tuberculosis. The average age was 48.4 ( $\pm$  26.2) years old and 85.7% of patients were male. Immunosuppression was present on 28.6% of patients. Chest X-ray showed: free pleural fluid on 76.2%, left side effusion on 52.4%, small to moderate effusion size on 95.2%. Thoracentesis: all pleural effusions were exudative. ADA mean value on fluid was 148.8 ( $\pm$  42.9). All patients had pleural fluid lymphocytosis higher than 50%. Nucleic acid amplification tests (NAAT) for MT and/or cultural exam were positive in 57.1%. Closed pleural biopsy: performed on 71.4% of patients, in which 93.3% had necrotizing granuloma. NAAT and cultural exam were positive in 46.5%. Respiratory samples: all patients routinely had sputum analysis for TB. Fiberoptic bronchoscopy was performed on 7 patients (33.3%). NAAT and cultural exam were positive in 6 patients (28.6%). Four patients (19%) had no mycobacteriologic confirmation (negative NAAT and cultural exam) although all had pleural biopsy histology samples compatible with TB. There were no cases of resistance to first line antibiatic drugs in all the patients with positive culture.

**Conclusions:** Thoracentesis and closed pleural biopsy allowed mycobacterial confirmation of pleural TB in 81% of patients. ADA > 70 was present on 85.7% of patients. Six patients (28.6%) had pleuropulmonary TB and one patient (4.8%) presented with disseminated TB. All 4 patients with presumptive diagnosis (no mycobacteriologic confirmation) had histology with necrotizing granulomas and elevated ADA values.

**Key words:** Pleural tuberculosis. Pleural effusion. Thoracentesis. Pleural biopsy.

#### PE 052. (STOP-)LOSS TO FOLLOW-UP: ITS DETERMINANTS AMONG TUBERCULOSIS PATIENTS, IN COIMBRA DISTRICT, 2005-2011

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**Introduction:** Tuberculosis patients who abscond from treatment are more likely to develop Multidrug-Resistant Tuberculosis; as well contributing to persistent disease reservoirs within communities, as well as ongoing transmission. This growing concern has led to the development of this study in order to assess risk factors among Tuberculosis patients lost to follow up (LTFU) notified in the Coimbra District between 2005 and 2011.

**Methods:** A case-control study was designed to compare patients LTFU to those successfully treated. Twenty one LTFU cases were compared to 417 controls. Social and demographic data, co-morbidities, disease location, previous treatment history and drug resistance pattern were analyzed. In order to determine independent risk factors, numeric data was compared by one sample t test, categorical data compared by the chi-square test or the fisher exact test, and included in a logistic regression analysis.

**Results:** Considering LTFU patients, their mean age was 37 ( $\pm$  14.4) years; the majority were men (71.4%); 71.4% had pulmonary disease. Drug, alcohol and tobacco use were common among them (23.8%, 42.9%, and 42.9% respectively). Chronic medical illness included hepatitis C and B virus infection (19% and 14.3%), silicosis

(4.8%) and mental disorders 9.5%. None of the cases had history of incarceration, diabetes, obstructive chronic pulmonary disease or chronic renal failure. Univariate analysis found that LTFU patients were more likely to be foreign born (OR: 3.64; 95%CI 1.25-10.55;  $p$  = 0.03); unemployed (OR: 2.98; 95%CI 1.09-8.08;  $p$  = 0.04); to have a history of drug (OR: 7.83; 95%CI 2.55-24.04;  $p$  < 0.01) and alcohol abuse (OR: 3.02; 95%CI 1.23-7.40;  $p$  = 0.02), HCV and HBV infection. Multivariate analysis found immigrants (aOR: 3.51; 95% CI 1.04-11.84;  $p$  = 0.04) to be the strongest predictor of LTFU.

**Conclusions:** Although the demographic trends of tuberculosis have changed, those patients LTFU remain quite similar in many aspects. In the Coimbra District, its likelihood was strongly associated to social or health problems such as addictive (alcohol, drugs) behaviours and immigrants. It is extremely important that the medical teams are particularly alert to these patients, considering supportive approaches to patients with LTFU risk factors, prioritizing them for intensive follow-up care. The results emphasize the need for stronger coordination with other support services such as mental health, substance abuse programs and tobacco cessation services. Also, healthcare staff must be aware that their attitudes and practices influence patient's willingness to initiate and adhere to treatment. Multidrug-resistant tuberculosis with its growing prevalence, left with few or even none pharmacological options, makes necessary to highlight all of the approaches available and considered appropriated to the prevention and management of these forms of the disease. Those approaches should consider a combination of different strategies, including coercive measures towards recalcitrant patients, particularly when all other less restrictive measures fail, this way contributing to the treatment's completion. This demands an effective legislative response that should appropriately reflect today's globalized 21<sup>st</sup> century and its public health challenges.

**Key words:** Tuberculosis. Lost to follow up. Risk factors.

#### PE 053. LATENT TUBERCULOSIS INFECTION IN THE LAST TEN YEARS IN CENTRO DE DIAGNÓSTICO PNEUMOLÓGICO DA VENDA NOVA (CDPVN)

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**Introduction:** The diagnosis and treatment of latent infection by *Mycobacterium tuberculosis* (*Mt*) reduces the risk for development of active tuberculosis, as well as transmission of *Mt* in the community. The growing utilization, in various pathologies, of biologic therapies (anti-TNF $\alpha$  or anti-interleukines), that rise significantly the risk of developing active tuberculosis (TB) due to reactivation of a latent tuberculosis infection (LTBI), reinforces the importance of diagnosis and treatment of this disease.

**Objectives:** To characterize the population, therapeutic options and adherence to treatment of patients with LTBI in CDPVN.

**Methods:** Retrospective analysis of the demographic and clinical data, introduced in the surveillance system of tuberculosis (SVIG-TB), of the patients with LTBI, assisted in CDPVN between 2007 and 2016. Comparison between the two groups assisted in the first two years of this period (2007/2008) and the last two years (2015/2016), to evaluate possible differences in diagnosis, therapeutic options and results in both periods.

**Results:** During the study period, 1,080 patients with LTBI were assisted in CDPVN, 52.3% women and 20.1% foreigners. The diagnosis was obtained through contact screening in 685 (63.4%) of these, of a total of 1,170 source cases. From the 140 patients diagnosed in 2005/2006, 60.7% were women and 17.1% foreigners, and the diag-

nosis was obtained through screening of contacts in 112 (80%) patients (source cases: 118, total screening: 1,579). In 2015/2016, from a total of 271 patients, 50.6% men and 14.8% foreigners, only 128 (47.2%) were diagnosed in this context (number of source cases in this period: 247, number of screenings 2,461). After the introduction of IGRA (Interferon gamma release assay), in 2009, this started to be used, according to the recommendations, and was positive in the diagnosis of 31 (11.4%) of the patients in 2015/2016. The therapeutic regimens used in 2007/2008 were isoniazid-rifampicin-pyrazinamide during two months (2HRZ) in 65% of the cases and Isoniazide alone during 6 or 9 months (6-9H) in 35% of the patients, while in 2015/2016, the majority of the patients (96.3%) were treated with isoniazid alone, according to the last recommendations. In 2007/2008 the abandon rate of therapy was 10.7%, while in 2015/2016 was 19.2%.

**Conclusions:** The number of patients with diagnosis of LTBI and indication for treatment, has risen in the last 10 years, being this phenomenon in probable relation with the significant rise of patients proposed to treatment with biologic therapies. The treatment regimens recommended nowadays, although more safe, can me implicated in higher abandon rates in the last years, due to longer treatment times.

**Key words:** *Tuberculosis. Latent infection.*

#### PE 054. DON'T FORGET PULMONARY TUBERCULOMA!

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**Introduction:** Tuberculomas are well-defined nodules or masses caused by *Mycobacterium tuberculosis*. Pulmonary tuberculoma is a common radiological finding during life, but its differential diagnosis remains a clinical challenge.

**Objectives:** This study aims to describe the comorbidities, clinical features and radiological findings in patients with lung tuberculoma.

**Methods:** We performed a retrospective data analysis of 44 patients admitted to Pulmonology Diagnostic Center (PDC) at Coimbra, Portugal, between 2007 and 2017 with pulmonary tuberculoma confirmed histopathologically.

**Results:** We obtained a total sample of 451 patients with a diagnosis of pulmonary tuberculosis treated in PDC. From these cases, 44 (9.8%) presented as pulmonary tuberculomas. The patients had a mean age of  $60.4 \pm 14.6$  years and 61.4% were males. Eighteen patients (40.9%) had previous history of cancer (5 lung cancer), 8 (20%) structural lung disease, 6 (13.3%) COPD, 6 (13.3%) immunosuppressive therapy, 9 (20.5%) of diabetes mellitus and 4 (9.1%) had previous history of tuberculosis. In 39 cases (88.6%) the tuberculomas were discovered incidentally during a routine chest X-ray or chest CT-scan. The mean tuberculoma size was  $1.9 \pm 1.2$  cm. In 61.4% of cases, the location was in one of the upper lobes (including lingula), 11.8% cases in the middle lobe and 26.8% in the lower lobes. In 9 cases (20.5%) there was more than one nodule and the maximum number observed was 4 and, in 7 cases (15.95%), they were associated with enlarged lymph nodes. Only 5 patients (11.4%) had clinical manifestations of chest pain, cough or fatigue, which led to diagnostic investigation and imaging tests. In 25 patients, lung cancer or metastasis was suspected based on pathological background and radiological findings: an irregular or spiculate margin, or a pleural indentation in 13 patients (29.6%), calcification in 9 (20.5%), cavitation in one (2.3%) and satellite nodules in two (4.6%).

**Conclusions:** The present study highlights the importance of the tuberculous etiology of pulmonary nodules, which although less

common, should not be undervalued, particularly in countries with a high incidence of mycobacterial infection.

**Key words:** *Tuberculosis. Tuberculoma. Lung nodules.*

#### PE 055. QUALITATIVE INSIGHT ON THE PSYCHOLOGICAL IMPACT OF TUBERCULOSIS AND ITS TREATMENT

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**Introduction:** Tuberculosis and its prolonged treatment predictably impact the psychological and social dynamics of the patients. This impact is usually not evaluated in current clinical care. The authors present a qualitative research study that aimed to characterize the psychological impact of tuberculosis and to identify factors associated with adherence to treatment according to patient's perspectives.

**Methods:** The authors present a qualitative study enrolling tuberculosis patients from 2 medical centres in Portugal. Semi-structured interviews, aimed at evaluating the psycho-social impact of the disease and the treatment as directly observed therapy (DOT), were performed. The data obtained was organized in categories and sub-categories, and a quantitative analysis of the descriptive frequencies of the population answers was performed.

**Results:** The study included 18 patients (mean age of  $51.7 \pm 15.8$  years), predominantly males (83.3%) with pulmonary tuberculosis. Most (72.2%) expressed depressive mood, aggressiveness and anxiety at the time of the diagnosis; and considered that the disease and treatment disrupted daily routines and the relationship with family and friends. Most (77.8%) stated that the emotional support from family and friends did not meet patient's expectations. The most frequent negative determinant of adherence according to patient's perspective was inadequate family support (44.4%).

**Conclusions:** Tuberculosis as a disease and its treatment were associated with a negative impact in patient's daily life and in the relational dynamics with family and friends. This negative mindset experienced by the patients was predominantly a result of an inadequate emotional support from family members and friends according to patient's expectations.

**Key words:** *Psychological impact. Tuberculosis. Adherence.*

#### PE 056. PULMONARY TUBERCULOSIS IN THE PRE-ANTIBACILARY AGE - FOR THE PURPOSE OF A CLINICAL CASE

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**Introduction:** Tuberculosis has been following humanity's history for many years, having been seen as a "mortal enemy" for a long time. As such, the discovery of its treatment had a profound and significant impact on individuals and society. The main breakthrough in the development of tuberculosis treatment came with the discovery of its cause by Robert Koch in 1882, now known as Koch's bacillus. Since then, there have been several advances in the knowledge of tuberculosis. During the time of the sanatoriums and prior to the discovery of antibiotics, the surgical treatment of tuberculosis through collapsed therapy, combined with rest, was the only therapeutic weapon available for pulmonary tuberculosis.

**Case report:** The authors present the case of a 79-year-old male, non-smoker male patient. With a personal history of pulmonary tuberculosis, having undergone surgery in 1962, by collapsing therapy with lucite balls. In 2017, the patient had a consultation due to the appearance of axillary nodular swelling accompanied by worsening dyspnea for medium effort. In the context of the etiological investi-

gation, a CT-thorax was performed that revealed an image with a net content and with nodular lesion in an external location to the rib cage. Given the context and personal history of the patient, allied to the findings of the imaging examination, it was verified that they were lucite balls, used in the surgery of treatment of pulmonary tuberculosis to which it was submitted, with migration to the wall and pleuro-cutaneous. It was later referenced for Thoracic Surgery, for eventual removal surgery of lucite balls. It is then proposed for surgery with thoracoplasty of 7 costal arches and 1/3 lower of homolateral shoulder blade, with removal of collapsing material (21 lucite balls) and drainage of pleural-cutaneous fistula on the left.

**Discussion:** Important advances have been made in the area of tuberculosis diagnosis, treatment and prevention, which have contributed to a reduction in the incidence and mortality of tuberculosis worldwide. This case presents a rare and late complication of a treatment before the development of the antibacterials and that the new generations of doctors no longer knew. It also highlights the importance of knowledge of the historical evolution of treatment and the technological advances that have contributed to an increase in survival and quality of life and, hopefully, to the cure of patients with tuberculosis.

**Key words:** Tuberculosis. Colapsoterapy.

#### PE 057. OCCUPATIONAL EXPOSURE TO INFECTIOUS TUBERCULOSIS PATIENTS IN A UNIVERSITY HOSPITAL - RETROSPECTIVE ANALYSIS OF THE LAST 4 YEARS

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**Introduction:** Tuberculosis is still one of the main biological risks for health professionals (HP) in Portugal. As a means to prevent active tuberculosis, identification and subsequent follow-up of HPs with non-protected occupational exposure to infectious patients without adequate respiratory protection, enables the detection and treatment of new infections.

**Objectives:** Clinical, professional and demographic characterization of HPs with non-protected occupational exposure to *Mycobacterium tuberculosis* (MTB) in the last 4 years, as well as description of follow-up and diagnosis.

**Methods:** Consultation of the registry of non-protected occupational exposures to MTB reported to the OHS-CHLN in the last 4 years and subsequent descriptive analysis of the relevant Clinical, professional and epidemiological variables.

**Results:** During the period of analysis 34 cases of infectious tuberculosis patients were reported and with which 562 HPs contacted without adequate respiratory protection (average 17.1 HP/patient). Of the potentially exposed health professionals, 412 complied with the first consultation (37.5 ± 10.56 years of age; 83.5% women), with an average tuberculin test (TT) induration of 1.6 ± 8.09 mm, 67.1% of them with TT ≥ 10 mm e 46.4% with TT ≥ 15 mm. The 134 with a previous TT test ≥ 10 mm, were submitted to IGRA test, 32.8% of which were positive. Of these, 5 were sent to the CDP (Centro de Diagnóstico Pulmonar). Only 163 HP complied with the second consultation 9 weeks after exposure, 10 of those were found to have latent tuberculosis (positive TT with positive IGR test) and were sent to the CDP. The remaining 153 (93.9%) was discharged with no evidence of inoculation by MTB. In total there were 25 positive TT, 72% of which had a negative IGRA test.

**Conclusions:** Through identifying contacts, active surveillance and chemical prophylaxis of HP diagnosed with latent MTB we've contributed to the decreasing MTB reservoir and cases of active MTB infection among HP.

**Key words:** Health professionals. Pulmonary tuberculosis. Non-protected exposure. Contact surveillance. Tuberculin test. IGRA.

#### PE 058. TUBERCULOSIS SCREENING IN INSTITUTIONS OF SOCIAL SOLIDARITY: THE REALITY OF A SERVICE

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**Introduction:** The diagnosis and treatment of latent tuberculosis infection (LTBI) significantly reduces the risk of developing active disease and of the transmission of the disease in the community. Tuberculosis (TB) screening consists of excluding active disease and assessing the adaptive host immune responses to *Mycobacterium tuberculosis*. Some individuals, such as injecting drug users, those working in prostitution, or those infected with human immunodeficiency virus (HIV), are at a higher risk of progression from LTBI to active TB. Thus, the diagnosis of LTBI is particularly important in those patients. By providing shelter and support to those individuals, institutions of social solidarity play an important role in the community. Considering this, since 2009, the Centro de Diagnóstico Pneumológico de Coimbra (CDPC) has carried out a tuberculosis screening protocol for users and employees of these institutions.

**Objectives:** To analyse the results of the CDPC TB screening in institutions of social solidarity since its implementation in 2009.

**Methods:** Retrospective analysis of the clinical processes of users and employees of the institutions of social solidarity invited to participate in the CDPC TB screening since its implementation. Demographic and clinical data were analysed. Statistical analysis was performed using Microsoft® Excel 2016 and IBM® SPSS® Statistics 25.

**Results:** Of the total of 647 individuals invited for TB screening, from 9 different institutions, 630 (97.4%) performed all screening procedures, of which 371 (58.9%) were men and 259 (41.1%) were women. Eighty-one percent (81%, n = 510) of the individuals had negative screening, 18.2% (n = 115) had LTBI and 0.8% (n = 5) had active TB. Of the LTBI population, 106 patients agreed to start treatment: 30 with isoniazid (H) for 6 or 9 months, and 76 with isoniazid plus rifampicin (HR) for 3 months. In relation to the HR group, 89.5% of the patients successfully completed the treatment; in the H group, only 63.3% concluded treatment. There was a statistically significant association between the treatment regimen and the conclusion, or not, of the treatment (p = 0.006, OR HR/H 5.1).

**Conclusions:** Most of the individuals presented negative screening. Among those with positive screening, most were diagnosed with LTBI. In this population, adherence to treatment was higher with the HR regimen. Screening for TB in institutions of social solidarity is a step forward in addressing this disease. The choice of therapeutic regimen should take into account adherence, ethical aspects and associated side effects. A shorter scheme may have greater adherence in this type of population.

**Key words:** Tuberculosis. Latent tuberculosis infection. Treatment. Preventive.

#### PE 059. TUBERCULOSIS DIAGNOSIS AND TREATMENT DELAY IN GUINEA-BISSAU

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**Introduction:** According with the 2015 Annual Report of the National Plan for the Fight against Tuberculosis in Guinea-Bissau, there

is a notification rate of 135/100.000 habitants and Bissau has de higher proportion of diagnosis (68.2%).

**Objectives:** To analyse causes of tuberculosis diagnosis and treatment delay in Guinea-Bissau.

**Methods:** Study of inpatients in a reference hospital for treatment of TB in Guinea-Bissau in December 2017.

**Results:** From the 68 inpatients, 55.9% (n = 38) were male and mean age was 33.7 ± 1.3 years. 19.1% were illiterate. 52.9% were living in Bissau and the mean household was 9 people and 5.9% didn't have a window. Family income was lower than national minimum wage in 7.4% patients. Initial symptoms were fever (80.6%), cough (71.6%) and night sweats (67.2%). The nearest health center was at a distance of 1-10 km for 47.1% patients and 66.2% had to walk there by foot. HIV was positive in 28.8% but Gene Xpert was only performed in 8.8%. Mean time to look for an official health care center after initial symptoms was 27.8 ± 5.8 days and 54.4% had at least 4 consultations before diagnosis. Time delay for treatment initiation was 131.5 ± 15.2 days. Longer patient delay to seek for a health care facility was associated with a distance superior to 10 km (p = 0.021), illiteracy (p = 0.04) and income lower than the national minimum wage (p = 0.035). HIV positive status was associated with a longer delay to initiate treatment (p = 0.039).

**Conclusions:** There is still a big delay for TB diagnosis and treatment in Guinea-Bissau. It's important to teach the population about TB but above all, there is a urge need for screening projects that could speed up the diagnosis and stop the chain of transmission.

**Key words:** Tuberculosis. Diagnosis delay. Treatment delay.

#### PE 060. MULTIDRUG-RESISTANT PLEURAL TUBERCULOSIS - A CASE REPORT

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**Introduction:** *Mycobacterium tuberculosis* multiresistance (MR) is an emergent and concerning reality in Portugal and a public health issue that requires diligent and adequate treatment.

**Case report:** A 45-year-old autonomous male, usually residing in Angola and with several trips to South America, whose medical records referred to malaria diagnosed and treated in Angola 3 months before, without any other relevant clinical history. Past alcohol intake of ~90 g/day. The patient consulted his family physician for upper airway symptoms and the chest radiography showed a moderate volume right pleural effusion, and was therefore referenced to the Emergency Department. He showed no significant respiratory symptoms; without any evidence of without constitutional syndrome the physical exam revealed poor dentition and abolition of the vesicular murmur in the lower half of the right hemithorax. Diagnostic thoracentesis revealed an exudate, 100% mononuclear cells, ADA < 1 U/L, glucose 90 mg/dL, without identification of malignant cells or and microbiological agents. He started empiric antibiotic treatment and reevaluated after 10 days, with overlapping chest X-ray and clinic. A chest CT scan revealed pre-tracheal and hilar adenopathies, ipsilateral pleural thickening, 2 subpleural nodules of the right lower lobe, and atelectasis associated to the effusion. New diagnostic thoracentesis revealed ADA 101U/L and Glucose 67mg/dL and pleuroscopy only showed inflammatory aspects. The direct microbiological examination of pleural biopsy was negative for mycobacteria, with the use of molecular molecular MR with *rpoB* gene mutation. In the presumption of MR, the patient was electively admitted, sputum samples were collected and bronchofibroscopy investigation was carried out to exclude pulmonary involvement and the patient started antibacilar therapy, based on international standards. According to Kat gene mutations and first line AST, INSA suggested the therapeutic regimen of Z + Am + Ld + Mfx + Ethionamide +

Cycloserine. After the 2nd line AST results, an increase of Mfx dose was required. The patient remained hospitalized until the exclusion of pulmonary involvement and for the management of haematological, cutaneous and articular toxicity, achieved with dose adjustment.

**Discussion:** MR remains a threat, despite all efforts. It is defined by *in vitro* resistance to isoniazid and rifampicin; mutations of *rpoB* and *KatG* genes are very suggestive of MR, in our case confirmed with AST. Pleural tuberculosis represents 3.8% of cases of tuberculosis. The intersection of the two conditions appears as a diagnostic and therapeutic challenge and it requires concerted exercises of the authorities, physician and patient, in order to optimize individual results and public health gains.

**Key words:** Pleural tuberculosis. Multiresistance. *rpoB* gene mutation. *KatG* gene mutation.

#### PE 061. BURKITT'S LYMPHOMA WITH PULMONARY PRESENTATION: A VERY UNUSUAL CASE

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**Introduction:** Burkitt's lymphoma (BL) is an aggressive neoplasm and highly proliferative. It is rarely found in adults and is more common in males. There are three distinct clinical forms: endemic, sporadic and associated with the human immunodeficiency virus (HIV). In Europe, although rare, the sporadic form is the most prevalent and is typically presented by abdominal manifestations.

**Case report:** 18 year-old female student. Previously healthy, only taking oral contraceptives. She presented to primary health care (PHC) with fatigue, fever, cough and purulent sputum. She was treated for Pneumonia with amoxicillin and clavulanic acid (7 days) and deflazacorte. Six weeks later, without a complete symptomatology improvement and on suspicion of allergic pathology, she was given deflazacorte once more. At eight weeks, cough and sputum reappeared, associated to tiredness and posterior thoracic pleuritic pain in the left hemithorax. The thorax X-ray revealed a opacification in the lower two-thirds of the left hemithorax, with an aerial bronchogram and a small pleural effusion. The patient was sent to the Emergency Room (ER). She presented to the ER apyretic, with a peripheral blood oxygen saturation of 98% on room air. Lung sounds were decreased in the lower two-thirds of the left hemithorax, with increased chest vibrations on the second-third. The remaining physical exam was normal. Laboratory findings showed increased inflammatory parameters (leukocytosis 16 k/uL and a C-reactive protein 116 mg/L). A Community-acquired Pneumonia complicated by pleural effusion was diagnosed. She was hospitalized and medicated with levofloxacin (10 days). Despite clinical improvement, on the fourth day, control X-ray suggested an atelectasis of almost all the left lung. Blood serologies for HIV and atypical respiratory infection agents were negative. A bronchofibroscopy (BF) demonstrated an occlusive lesion on the division of the left main bronchus. Computed Tomography (CT) revealed an infiltrative lesion on the middle lobe, involving the homolateral pulmonary artery, with reactive mediastinal adenopathies and hepatomegaly. Histologic results of the BF confirmed the diagnose of BL.

**Discussion:** This case demonstrates that BL, despite rare, can present in a very atypical way, affecting female patients, with predominant respiratory manifestations. It is fundamental to be aware of the atypical manifestations of BL, since the prognosis is dependent on a quick diagnosis and an early treatment.

**Key words:** Burkitt's lymphoma.

## PE 062. PULMONARY NODULES: UNCERTAINTIES AND PROBABILITIES

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**Introduction:** Lung cancer is one of the main causes of morbimortality in the world. Its early diagnosis and treatment is imperative. Pulmonary nodules are common radiological findings which unleash the great challenge of safely establishing whether they are malignant or benign for a correct therapeutic approach.

**Case report:** The authors present the case of a 62 years old man, ex-smoker of 30 pack-year, with history of dyslipidaemia and hypertension. During a cardiological follow-up he performed a coronary arteries angio-CT that showed a pulmonary nodule in the left upper lobe (LUL) and another in the middle lobe (ML). To characterize them a thoracic-CT was performed: heterogeneous nodule, spiculate (38.7 mm) in the LUL, nodule (5 mm) in the ML and centrilobular emphysema. The investigation proceeded in Pulmonology consultation. He denied respiratory/constitutional symptoms. The laboratory work-up was normal except for an elevated NSE and AFP. Abdominal ultrasound and head-CT were normal. Thoracic-CT was repeated: "semi-solid nodule (38.7 mm) in the LUL, predominantly ground glass with increased solid component, suggestive of minimally invasive/invasive adenocarcinoma; ML nodule unchanged". Bone scintigraphy: discrete gain in the right temporal bone, excluded metastization. PET-CT: LUL mass without significant uptake (SUV max = 1.97) and 5mm nodule in ML without metabolic expression; examination without clear evidence of malignancy (attention to reduced sensitivity in the detection of certain histological types)". PFTs revealed mild obstruction. The case and radiological imaging were discussed at a multidisciplinary meeting, being consensual that it was, most likely lung adenocarcinoma. The patient refused bronchofibroscopy and transthoracic biopsy. Proposed direct surgical approach assuming a stage Ib and after being observed in Thoracic Surgery consultation. He was submitted to 2 atypical resections on the LUL and a ganglionic excision in a private institution - extemporaneous examination revealing no malignancy. Radiological reassessment 2 months later revealed: parenchymal opacity adjacent to the fissure with pleural extension in the LUL and homolateral pleural effusion compatible with post-surgical alterations; spiculated parenchymal opacity with calcifications in the RUL and a nodule in the ML. Three months later radiological imaging remained the same. He maintained follow-up accordingly with thoracic surgery. Twelve months after surgery the CT-scan revealed: "poorly circumscribed area near the fissure in the LUL with air bronchogram, extending to the hilum (recurrence?) and a ground glass nodular image of lobulated contours in the RUL, increasing in size (25 mm vs 20 mm), suggestive of pulmonary neoformation, with a similar image (12 mm) in the adjacent parenchyma, more expressive than in the previous examination. ML nodule, probable granuloma or intrapulmonary lymph node, remained the same.

**Discussion:** 2 RUL nodules suggestive of lung neoformation. To clarify these lesions, tumor markers were repeated (normal) and PET-CT: bilateral pulmonary alterations, with greater structural expression and low metabolic activity - unspecific (scarring admitted), unsuspected malignancy. He maintains follow up and surveillance in Pulmonology Consultation. The authors emphasize, the importance of obtaining a histological diagnosis of suspicious pulmonary nodules and the pursuit of a rigorous follow-up. It's fundamental to take advantage of the different diagnostic methods available, complementing them with multidisciplinary meetings to obtain structured decisions.

**Key words:** *Pulmonary nodules. Diagnosis. Multidisciplinary team.*

## PE 063. SMALL-CELL LUNG CARCINOMA: NON-HODGKIN'S LYMPHOMA AS A RISK FACTOR

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**Introduction:** Small-cell lung carcinoma (SCLC) is a neuroendocrine cancer with aggressive behaviour due to its rapid growth and early spread to distant sites. It represents about 13% of all lung cancers and is associated with smoking history in approximately 98% of the patients.

**Case report:** A 68 years old female patient with a previous smoking history (43 pack-year) was sent to the Pneumology appointment due to a lung nodule at the peripheral right superior lobe (RSL) with 14 × 10 mm observed in a chest CT two months previously (April 2018). She was diagnosed with B-cell diffuse non-Hodgkin's lymphoma (NHL) and treated with rituximab, cyclophosphamide-hydroxydaunomycin-ondansetron-prednisone (R-CHOP) in 2005, being in total remission. She also had renal insufficiency, anaemia, hypothyroidism, chronic gastritis, depressive syndrome, dyslipidemia and had had pulmonary tuberculosis in her infancy. At the Pneumology appointment, she presented with fatigue, without significant weight loss or other symptoms. She repeated the chest CT (June 2018) and it was observed a dimensional growth of the RSL nodule (17 mm), new mediastinal and hilar right adenopathies (the biggest with 25 mm) and there were new multiple suspicious metastatic liver nodules (the biggest with 30 mm). She underwent a videobronchoscopy (July 2018) that revealed a reduction of the anterior right superior lobar bronchus, enlargement of the right main bronchus bifurcation and reduction of the middle lobe bronchus because of external compression. Bronchoalveolar lavage and bronchial brushing were performed and were negative for cancer cells. Bronchial biopsies were positive for SCLC. Endobronchial ultrasound endoscopy (EBUS) allowed the realization of fine needle aspiration biopsies of the 7th (13 mm) and 4R (20 mm) stations, that revealed being SCLC metastasis. She was sent to an Oncology appointment, because of stage IV SCLC (T1bN2M1c).

**Discussion:** Over the last few years evolving therapies, mainly by using alkalinizing drugs, have led to improved long-term survival for patients with hematopoietic cancers. NHL is the 10<sup>th</sup> most common cancer and is associated with the development of secondary cancers, probably caused by chemotherapy and/or radiotherapy or because of some genetic pleiotropy. The authors present this case to emphasize the importance of hematopoietic cancers as a risk factor for lung cancers and its need for investigation, as well as alert for the rapid growth of neuroendocrine cancers.

**Key words:** *Small-cell lung carcinoma. Non-Hodgkin's lymphoma. Risk factor.*

## PE 064. EGRF TYROSINE KINASE INHIBITORS IN LUNG ADENOCARCINOMA - A SUCCESSFUL CASE

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**Case report:** The authors describe the case of a 42-year-old non-smoker female, with no relevant previous medical history and no usual medication. She was admitted in the Emergency Department due to wheezing, dyspnea, thoracalgia and cough with mucous expectoration, lasting for a month. From the exams performed for etiological investigation, we highlight: 1) the chest X-ray with bilateral pulmonary condensations, 2) an analytical blood test showing with increased inflammatory parameters and 3) arterial blood gas analysis in keeping with severe partial respiratory failure. The patient was diagnosed with bilateral hypoxemic pneumonia and empirical antibiotic therapy was initiated. Although, due to absence of clinical and radiological improvement, she underwent a chest CT.

It revealed an extensive lobar condensation with air bronchogram in the left lower lobe and widespread groundglass pattern of the right lung. Subsequently, the patient underwent bronchofibroscope, which revealed no endobronchial abnormalities, but the cytological examination of the bronchial secretions was suggestive of adenocarcinoma, whose anatomical origin would need to be clarified. The histological result of guided transthoracic aspiration biopsy (BATT) confirmed lung adenocarcinoma. She was staged class IV lung adenocarcinoma with bilateral lung involvement. She started carboplatin and paclitaxel based-chemotherapy (CT). Due to clinical worsening after the third CT cycle, we repeated the chest CT, which revealed extensive bilateral condensations suggestive of disease progression. The molecular study of the lung biopsy (BATT) showed a mutation in exon 19 of the EGFR gene, a mutation particularly increasing responsiveness to erlotinib. The drug was started and led to regression of the tumor mass. One year after the diagnosis, due to disease progression, BATT was repeated revealing a new tumor mutation conferring resistance of the neoplasm to erlotinib and, therefore, osimertinib was initiated. The patient was successfully treated and she is currently clinically asymptomatic with an active professional life. The current chest X-ray findings are irrelevant.

**Discussion:** This clinical case intends to highlight the relevance and high efficacy of EGFR tyrosine kinase inhibitors in the treatment of lung adenocarcinoma and the role of immunohistochemical in that diagnosis.

**Key words:** Lung Adenocarcinoma. EGFR. Erlotinib. Osimertinib.

#### PE 065. CLEAR CELL ADENOCARCINOMA OF THE LUNG: A CASE REPORT

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**Introduction:** Lungs tumors frequently show clear cell histology. The clear appearance results from intracellular accumulation of glycogen in absence of mucus production. Most clear cell tumors are metastases of renal or ovarian tumors. Carcinomas entirely composed of clear cells are rare, especially adenocarcinomas. The authors present a case of clear cells adenocarcinoma and its therapeutic approach.

**Case report:** A 60-year-old man, former smoker of 55 pack-year history and status performance = 0. His past medical history was remarkable for chronic obstructive pulmonary disease (COPD) - GOLD stage 2A followed at Respiratory consultation since 2017 for chronic cough and dyspnea mMRC = 1. Computed tomography (CT) of the chest showed the presence of a 24 × 10 mm nodule and with 6 mm of solid component in the upper lobe of the right lung. The Positron-Emission Tomography (PET-CT) was performed and the nodule showed metabolic activity with SUV maximum of 1.68 and 2.32 at latter images. No lymph node or other lesions were detected. The patient underwent, a CT-guided biopsy of the lung to determine the nature of the lesion. The histology confirmed malignant neoplasia with glandular epithelium composed of clear cells, with a positive immunohistochemical profile for CK7 + and TTF1 + and negative for CD10 -, compatible clear cell adenocarcinoma primary of the lung. The patient was referred to our Oncology daily clinic where staging of the neoplasia was completed: stage IA3 - T1cN0M0. The patient was discussed at a Multidisciplinary reunion and was decided the surgical approach - right upper lobectomy and mediastinal lymphadenectomy by videothoracoscopy. Primary clear cell carcinoma of the lung is a rare disease of unknown etiology, with few cases described in the medical literature. It is a rare subtype of large cell lung cancer and adenocarcinoma. The scarcity of studies in terms of clinical significance

and prognosis led World Health Organization to its removal from pulmonary tumors classification in 2011.

**Discussion:** This case is didactic due to the rarity of this histological subtype of adenocarcinoma of the lung and emphasizes the importance of establishing the differential diagnosis of the primitive tumor, with therapeutic implications, in the presence of clear cells.

**Key words:** Lungs tumors. Clear cell.

#### PE 066. PRIMARY PULMONARY DIFFUSE LARGE B CELL LYMPHOMA - A CASE REPORT

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**Introduction:** Primary pulmonary diffuse large B cell lymphoma (PP-DLBCL) is an extremely rare neoplasm representing only 0.5-1% of primary pulmonary malignancies. Lymphoma involving the lung is defined as primary if the lymphoma is confined to the lung with or without hilar lymph node involvement that has been confirmed by clinical, radiologic, and pathologic assessment at the time of diagnosis or in the next three months. Most (70-90%) primary pulmonary lymphomas are low-grade mucosa-associated lymphoid tissue (MALT) type, and only about 10% are primary diffuse large B cell lymphomas (DLBCL). The prognosis of PPDLBCL is worse than pulmonary MALT lymphoma with a median survival of 3 to 5 years despite combined chemotherapy regimens.

**Case report:** A 69 year old woman presented with a 1.5 year history of progressive dyspnea and asthenia. She didn't present weight loss, night sweats or other constitutional symptoms. The patient underwent radiological evaluation including computed tomography (CT) of chest that showed bilateral nodular lesions and mediastinal adenopathies and positron emission tomography-computed tomography (PET-CT) that was positive in bilateral pulmonary lesions with standardized uptake value SUV max 13. Fiberoptic bronchoscopy with subsidiary techniques was performed with visualization of exophytic lesion in the external segment of the left basal pyramid and total occlusion of the posterior segment of the left basal pyramid by mucosal infiltration. The pulmonary diffuse large B-cell Non-Hodgkin's Lymphoma diagnosis was done through transbronchial pulmonary biopsy. The patient was referred to the Hematology Service, where it was performed bone marrow biopsy and myelogram without infiltration by clonal lymphocytes and started treatment with R-CHOP (rituximab, cyclophosphamide, doxorubicin, vincristine, prednisolone). This treatment was well tolerated having been completed 8 cycles with full remission.

**Discussion:** With its non-specific presentation, the diagnosis of primary pulmonary DLBCL is very challenging and often leads to misdiagnosis or delayed diagnosis. Therefore, it is important to increase awareness of this rare disease, as the correct characterization of the tumors has therapeutic and prognostic implications.

**Key words:** Pulmonary lymphoma. B cell. Lung neoplasm. R-CHOP.

#### PE 067. THERAPEUTIC OPTIONS IN ADENOID CYSTIC CARCINOMA OF THE TRACHEA - A CASE REPORT

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**Introduction:** Adenoid cystic carcinoma of the trachea is a rare malignant tumor that arises from the epithelium of mucous glands.

It usually presents as a slow-growing tumor, with large dimensions at the time of diagnosis and obstruction of the tracheal lumen. Surgical excision is conditioned by the tumor location and extent, with radical radiotherapy being a therapeutic option.

**Case report:** A 31 year old female patient, white, non-smoker, without known medical history. Asymptomatic until pregnancy, at which point she developed productive cough with mucous expectoration, dyspnea for exertion and wheezing. After giving birth, she developed stridor and underwent bronchofibroscopy, which revealed extensive mass of the left and anterior wall of the entire trachea, conditioning lumen obstruction, extending to the carina and emergence of the left main bronchus (LMB). Chest and neck computed tomography showed parietal thickening of the trachea, from the thoracic operculum to its bifurcation, with extension to the initial segment of the LMB and to the adjacent mediastinal structures. Endoscopic laser therapy was performed, with repermeabilization of the tracheal lumen. Histological diagnosis was adenoid cystic carcinoma. The patient was referred to the Hospital de Dia de Pneumologia Oncológica (HDPO) of the Hospital Pulido Valente (HPV) and, since the tumor was inoperable, the patient underwent external 3D conformal radiotherapy (total dose of 66 Gy) resulting in mass reduction. She remained without significant symptoms, with stable disease for approximately 15 months. Then she developed complex ring stenosis secondary to radiotherapy with 1 cm of extension, at 2 cm of the carina. Underwent mechanical dilation by rigid bronchoscopy, with posterior need for a tracheal prosthesis (Dumon® ST). Later, it was verified an increase in the primary lesion and abnormal uptake in PET-CT. Ultrasonographic echoendoscopy puncture confirmed disease progression. Control bronchofibroscopy showed tumor endoluminal growth with infiltration of the distal third of the trachea and carina, with the opening orifice of LMB reduced to a slit. Tracheal prosthesis was removed and argon plasma therapy was performed, resulting in good patency of the tracheal lumen and the right bronchial tree. Due to clinical worsening, with the need for invasive mechanical ventilation, it was decided to place a prosthesis in the LMB, due to total atelectasis on the left, with a positive outcome. The patient was referred to a surgical center in Vienna, underwent surgery and died in the postoperative period.

**Discussion:** Although adenoid cystic carcinoma of the trachea presents a slow growth, its diagnosis is usually made at an advanced stage of the disease, not allowing the surgical approach. External radiotherapy and endoscopic therapies allow us to control progression and relieve the symptoms associated with endoluminal obstruction.

**Key words:** Adenoid cystic carcinoma. Tracheal tumors.

#### PE 068. A CHALLENGE IN PNEUMOLOGICAL ONCOLOGY IMAGING

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**Introduction:** Primary pulmonary mucinous adenocarcinoma is a rare histological subtype, being diffuse pulmonary distribution an uncommon form of presentation.

**Case report:** A 61-year-old female secretary, former smoker for 30 years (15 pack-year history) and without known medical history, presented to the emergency department with cough with sputum production, asthenia and weight loss over the last 5 months. At physical examination pulmonary auscultation disclosed rude vesicular murmur bilaterally. Laboratory investigation with positive ANA (1/640 titulation) with a granular pattern and positive Anti-SSA/Ro (60KD) e Anti-SSB. The patient underwent chest X-ray which showed small and

multiple densities in the middle and lower thirds of both hemithoraces. Subsequently, CT scan of the chest was performed which showed multiple lung cavity lesions, with air content, thickened and irregular walls, more expressive in the lower lobes. The patient was referred to pneumology and underwent bronchoscopy which demonstrated no morphological alterations, with negative tests for mycobacteria and negative cytology for neoplastic cells. Pulmonary function testing with moderately severe restrictive ventilatory alterations and mild decrease in diffusing capacity for carbon monoxide. A multidisciplinary team was consulted, and the patient underwent video-assisted thoracoscopic (VATS) lung biopsy. The anatomopathological examination was compatible with mucinous adenocarcinoma. From the staging tests performed, the PET/CT scan found evidence of bilateral diffuse pulmonary anomalous uptake, classifying as stage IVA. There was no evidence of gastrointestinal origin. After obtaining a diagnosis, the patient was referred to pneumological oncology.

**Discussion:** Mucinous adenocarcinoma is the rarest subtype of pulmonary adenocarcinoma, representing a diagnostic and imaging challenge. The mucus secreted by the cancerous cells, is clinically presented by abundant mucus expectoration, being consistent with the clinical presentation of the patient. The clinical case described is of particular interest, given the exuberant imaging presentation that initially motivated the exhaustive investigation of other etiologies, namely, autoimmune, infections (fungi, disseminated pulmonary tuberculosis), systemic granulomatous disease, and metastatic lung disease. The high diagnostic yield of the VATS lung biopsy is highlighted.

**Key words:** Lung cancer. Lung adenocarcinoma. Pulmonary mucinous adenocarcinoma.

#### PE 069. LUNG CANCER AND THROMBOEMBOLISM - PATHOPHYSIOLOGICAL AND THERAPEUTIC SYNERGISMS

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**Introduction:** In non-small cell lung cancer (NSCLC) the incidence of stroke is 2.9%. In these, cerebral metastasis is frequent and constitutes an independent risk factor, increasing the incidence of stroke to 6.3%. Average survival rate is 36 days and overall survival at one-year is 6.7%. Tyrosine kinase inhibitors (TKIs) have demonstrated applicability in metastatic NSCLCs. Osimertinib is a 3rd generation TKIs applicable in metastatic CPNPC with EGFR T790M positive mutation, with a response rate of 61%. We present a case of lung adenocarcinoma with EGFR mutation and an atypical brain involvement evolution.

**Case report:** Woman, 64 years old, self-employed, lawyer, divorced, living alone, non-smoker, controlled hypertension, depressive syndrome, fibromyalgia and chronic erosive gastritis. In November 2016, it was diagnosed an adenocarcinoma of the right pulmonary lobe, with EGFR L858R mutation in exon 21, with cervical adenopathy and hepatic and bone metastasis of the acetabulum and the cranial calotte (cT4N3M1b - stage IV) in the context of non-productive cough and right coxalgia with 3 months of evolution. Was made external radiation therapy under the right acetabulum (30 gray in 10 fractions) and under skull cap (20 gray in 5 fractions). She started erlotinib in January 2017 with partial response. It was switched to gefitinib after 6 months due to important cutaneous iatrogenia. Nine months after switching, there was progression of the disease mainly by increased hepatic metastasis. It was performed a liquid biopsy, negative for EGFR mutation (L858R and T790M). Subsequently, it was performed a liver biopsy and, while waiting for the result, she started a self-limiting headache, dizziness and visual changes (left hemicamp), gait imbalance and dorsalgia with no respond to her usual opioid medication. CT-CE did not show space occupying lesions and it was proposed CE-MRI and she was hospitalized because of neurological deteriora-

tion with psychomotor deterioration, photophobia and somnolence with evolution to coma and upper left limb hemiplegia (grade 3/5). The diagnostic hypotheses of leptomeningeal dissemination or iatrogenic damage to radiation therapy were placed. Visual disturbance was attributed to partial epileptic seizures with vascular cause confirmed by EEG that showed serious and diffuse electrogenesis alteration, and she started levetiracetam 500 mg bid. CE-MRI showed multi-ischemic strokes with petechial hemorrhagic transformation in some lesions, and therefore she initiated therapeutic hypocoagulation with enoxaparin sodium. Meanwhile, liver biopsy showed T790M resistance mutation and osimertinib was started. A gradual recovery of neurological symptomatology was observed, and she was discharged. After starting osimertinib she recovered from most symptoms, except mild paresis of the left upper limb. The imaging reevaluation showed partial response, according to RECIST criteria.

**Discussion:** Neurological abnormalities require a comprehensive differential diagnosis, particularly in cancer patients, in whom metastization or paraneoplastic phenomenon cannot be dismissed. It is important to make a tumor reassessment if there is disease progression. If EGFR is mutated, the use of a third generation TKI (osimertinib) is possible, even in elderying or patients with worsening of clinical status.

**Key words:** *Osimertinib. Lung cancer. Non-small cell lung cancer. Tyrosine kinase inhibitors. Stroke.*

#### PE 070. THE POSITIVE SIDE OF LUNG CANCER - A CLINICAL CASE

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**Introduction:** Lung cancer is one of the leading causes of mortality in the world, with an overall 5 year survival rate of about 16%. The prognosis of patients with lung cancer is generally very reserved, however the appearance of new therapies targeting molecular and immunological targets has shown a new path.

**Case report:** The authors present the case of a 61 year-old patient, a smoker with a smoking load of 40 UMA. Personal background of hepatitis C under TARV, and high blood pressure. In December 2017, the patient goes to a pneumology appointment due to a lung mass at the right upper lobe level. At the initial evaluation, performance status 1 was presented, with no findings relevant to the objective exam, except for alteration to pulmonary auscultation in the upper 1/3 of the right hemithorax. In this context, a thoracic CT scan revealed solid mass with irregular contours in the posterior segment of the right lung of 6 × 3.5 cm and small pretracheal and prevascular vascular mediastinal adenopathies associated with a thin layer of ipsilateral pleural effusion. From the initial analytical evaluation it was highlighted CA 125 (236.6 U) and NSE (27.5 U). Following the etiologic characterization, bronchofibroscopy revealed a tracheobronchial tree without alterations, with negative cytology as well as negative bronchial brushing. For this reason, the patient took a trans-thoracic aspiration biopsy that was compatible with lung adenocarcinoma (TTF1 +), showing PDL1 expression of approximately 75%. To complete the study, the patient performed PET-CT, obtaining staging T3N2M1a - stage IV. In March 2018 the patient started 1st line chemotherapy with pembrolizumab, and tumor reduction was documented shortly after the 1st session, despite G1 toxicity, at the intestinal level, with rectal bleeding. Currently, with clinical stability, in PS 1, criteria of partial remission and analytical negativity of tumor markers.

**Discussion:** This case is an example of a positive and early response that highlights the growing importance of recognition of new molecular targets and consequently the use of targeted therapy.

**Key words:** *Lung cancer. Pembrolizumab.*

#### PE 071. WHAT IS HIDDEN BY BRONCHIECTASIS?

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**Introduction:** Carcinoid tumorlets are small collections of Kulchitsky cells, which are hyperplastic neuroendocrine cells in bronchial and bronchiolar mucosa. They differ from typical carcinoid tumors because of their smaller diameter (< 5 mm), being typically multifocal and associated to lung diseases as bronchiectasis, lung abscesses and fibrosis. Usually, carcinoid tumorlets are casually diagnosed, reason why its incidence is difficult to evaluate. However, the reported cases are rare. They predominate in older women and are considered as a benign process which commonly have a peripheric location.

**Case report:** The authors present a clinical case of a patient, male, 40 years old, Romanian nationality, construction worker, current smoker (25 pack per year), history of pulmonary tuberculosis in childhood (treated), who went to the Emergency Room (ER) because of hemoptysis in moderate amount with hours of evolution, without other symptoms. Analytically to point out the increase of hepatic enzymes (ALT and AST were 4 and 3 times the upper limit of normal, respectively, and GGT was 2 times the upper limit of normal), after that, with the proceeding study we diagnosed Hepatitis C Virus infection which was treated. Chest radiography showed an areolar design at the middle lobe (ML). He did a lung CT-angiography with cylindric bronchiectasis at the ML, associated to some peribronchial ground glass opacification micronodular densities and 3 micronodules, the biggest with 4.8 mm of diameter (2 in the inferior lobes and 1 in the right upper lobe). We did a bronchofibroscopy which revealed blood coming from the ML. The smear and cultural mycobacteriologic exams of sputum and bronchial secretions were negative. The bronchial secretions cytology was suspicious. Few days later he returned to the ER with a new hemoptysis episode in moderate amount, needing aminocaproic acid, reason why we repeated the bronchofibroscopy, which was normal, with a new request of cytology and smear and cultural mycobacteriologic exams of bronchial secretions, which were negative. The nucleic acid amplification test on bronchial secretions was negative for *Mycobacterium tuberculosis*. His pulmonary function was normal and he didn't have any analytical abnormality. He was proposed to a middle lobectomy and at the pathological examination of the pulmonary resected tissue it was identified bronchiectasis with some inflammation, associated to several carcinoid tumorlets spreaded, peribronchial, interstitial and subpleural, the biggest one with a 4mm diameter. Since then the patient started to be followed at Pulmonology appointment to do a regular imaging follow-up, being tiredness the only complaint till now.

**Discussion:** Summarizing, carcinoid tumorlets are associated to lung structural changes, being diagnosed as incidental findings at the pathological examination of pulmonary tissue surgically excised or at autopsy. Usually patients are asymptomatic, but sometimes they may have hemoptysis or bronchiectasis related symptoms. Despite they are considered benign, few cases of metastasis to peribronchial lymph nodes have been already reported, and because of this reason it's crucial to keep imaging surveillance in these patients.

**Key words:** *Bronchiectasis. Tumorlet.*

#### PE 072. CHALLENGES IN THE DIAGNOSIS OF BRONCHIECTASIS - A RARE DIAGNOSIS

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**Introduction:** The differential diagnosis of bronchiectasis may be a challenge in clinical practice, implying an extensive diagnostic gait.

However, the etiological diagnosis is fundamental, as it may have specific therapeutic implications in the future. The presence of bronchiectasis in members of the same family and at young age puts the hypothesis of a common cause, namely genetic alterations.

**Case report:** We present the case of 2 siblings with bronchiectasis and recurrent respiratory infections, non-smokers, followed in a Pulmonology Consultation. The case is a female patient, 27 years old, with cylindrical and cystic bronchiectasis bilaterally, with repetitive respiratory infections with several multisensitive microbiological isolations. Recurrent episodes of rhinosinusitis exacerbation also with multiple microbiological isolations, including fungi and anaerobes. Selective IgA deficiency. Functional respiratory examination with moderate obstruction, without respiratory insufficiency or history of hospitalizations. Dosage of alpha 1 antitrypsin and sweat test within normality. Without situs inversus. An anatomopathological study carried out with scanning electron microscopy was compatible with the diagnosis of complex ciliary dyskinesia. A 25-year-old brother diagnosed with bronchiectasis with recurrent lower respiratory airway infections without perinasal sinus involvement, with a history of hospitalizations in 2011 and 2016 and transient partial respiratory failure in the same context. The diagnosis of primary ciliary dyskinesia was also confirmed by electron microscopy. The authors present the distinguishing clinical characteristics of both clinical cases. Primary ciliary dyskinesia is a rare clinical entity caused by genetic mutations of autosomal recessive transmission that can cause structural or functional changes in the ciliary membrane, compromising mucociliary clearance. This pathology can affect several organ systems, but mainly involves the airways causing long-term pulmonary changes, bronchiectasis being the most common alteration. Despite the absence of a standard gold test, definitive diagnosis is usually made by electron microscopy. To this date there is no specific therapy for this pathology, despite the greater knowledge about it and some promising studies in this area, especially the genetic alterations that cause it. The treatment is directed essentially to the management of its complications, especially the infectious interurrences. Pulmonary transplantation may be an option to consider in these patients.

**Key words:** Primary ciliary dyskinesia. Bronchiectasis. Family history.

### PE 073. HEMOPTYSIS INVESTIGATION, ETIOLOGY AND TREATMENT

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**Introduction:** There are a numerous possible etiologies for hemoptysis, namely infection (tuberculosis or not), bronchiectasis or malignancy. More recent data suggest that cryptogenic hemoptysis incidence is rising.

**Objectives:** To analyze comorbidities, imaging and bronchoscopy findings, etiology and endoscopic treatment of hemoptysis.

**Methods:** Retrospective analysis of clinical data from patients admitted on a Respiratory Medicine Department due to hemoptysis for 5 years.

**Results:** The study included 66 patients, two thirds male. The mean age was 59 years old (minimum 22 and maximum 90); 42% were active smokers and 26% ex-smokers. The most common comorbidities were COPD (17%), malignancy (8%, one of them extra-pulmonary), bronchiectasis (5%) and chronic kidney disease (5%); 21% had history of tuberculosis (2 were still under treatment). At home, 14% were medicated with platelet anti-aggregants (one on double anti-platelet therapy) and 12% were under oral anticoagulant therapy. The majority of the patients (65%) presented mild to moderate hemoptysis, 32% hemoptoic sputum and 2 patients (3%) massive hemoptysis. Laboratory tests revealed that 15% had an elevated

prothrombin time. The most common findings on chest CT scan were ground glass opacities (41%), consolidation (39%, half of which with cavitation) and pulmonary mass (11%). Flexible bronchoscopy was performed in 89% of the patients, showing mainly blood clots (34%), active hemorrhage (25%) and inflammation (17%). Rigid bronchoscopy was performed in 30% of the patients, with 50% of them undergoing blood clots removal, 25% laser photocoagulation therapy and 25% stent placing. The more commonly identified etiologies were community acquired pneumonia (35%, 4 of them necrotizing and 7 in association with bronchiectasis), bronchiectasis (26%), pulmonary malignancy (17%) and tuberculosis (12%). There were 7 cases of hemoptysis assumed to be cryptogenic (6 of those patients had smoking story), one was under oral anticoagulant therapy and other under anti-platelet therapy. One patient underwent arteriography which did not reveal pathological findings and another patient had already undergone the same procedure (also normal). None of the patients needed bronchial embolization. Lobectomy was performed on 2 patients (one due to aspergilloma and the other due to stage IB pulmonary adenocarcinoma); 2 patients died, one due to stage IV malignancy progression and another (elderly patient) due to pneumonia.

**Conclusions:** The most common etiologies were community acquired pneumonia, bronchiectasis, pulmonary malignancy and tuberculosis. Almost all patients presenting with hemoptysis assumed to be cryptogenic had smoking habits. Although there were only few cases of massive hemoptysis, rigid bronchoscopy was necessary in a significant percentage of the cases. Endoscopic therapy allowed resolution of all cases in which it was performed.

**Key words:** Hemoptysis. Bronchoscopy.

### PE 074. TRACHEAL COMPRESSION DUE TO ESOPHAGEAL DISTENSION, AN UNCOMMON CAUSE OF AIRWAY OBSTRUCTION

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**Introduction:** Acute respiratory failure is a medical emergency. There are several known causes, including airway obstruction, which may be intrinsic or extrinsic.

**Case report:** Female patient, 87 years old. Background of spastic paraparesis after medullary infarction for about 20 years, bedridden, dependent in activities of daily living with intact cognitive ability. Patient with recent hospitalization for community-acquired pneumonia, medicated with amoxicillin/clavulanic acid and azithromycin, is admitted to the emergency room due to altered level of consciousness with a Glasgow Coma Scale of 3 and bilaterally miotic and reactive pupils, with low peripheral saturation and respiratory sounds frankly decreased bilaterally. Hemodynamically stable, hypothermic and with severe hypercapnia (pCO<sub>2</sub> 131 mmHg) and respiratory acidosis, the patient started noninvasive ventilation (NIV) and bronchodilator therapy after refusal of invasive ventilation by Intensive Care Medicine. Chest X-ray had no condensations suggestive of pneumonia, pleural effusion or congestion, and head-CT did not show relevant changes. Analytically, there were no relevant changes except for leukocytosis of 21,000 with PCR 10.8. The urinalysis was suggestive of infection, so ceftazidime was started. Given the poor response and low tidal volumes despite high IPAP, the parameters were adjusted, starting AVAPS mode with residual improvement of the general state, with reactivity of pupils and to pain. Small gasometric improvement, pulmonary auscultation with audible respiratory sounds although still much diminished and accompanied now by apparent stridor. In view of the described picture and scarce

improvement of the respiratory failure, chest CTA was performed for exclusion of pulmonary embolism and evaluation of airway permeability. During the mobilization after the exam, the patient presented with vomiting and was placed in lateral decubitus and gastric contents were aspirated. The patient reacted by exhibiting cough and purulent respiratory secretions were aspirated. A nasogastric tube was placed resulting in the abundant drainage of air and gastric contents. There was a marked improvement of the neurological state with recovery of the state of consciousness (Glasgow 15), and eupnea in ambient air, with peripheral saturations of 60%. NIV and oxygen therapy have now been restarted with effective ventilation and progressive clinical improvement. Images of chest CTA demonstrated marked gastroesophageal distension with compression of the airway responsible for severe hypoventilation, and pneumonia that may justify severe gastroparesis.

**Discussion:** In the literature, some cases of respiratory failure due to extrinsic compression of the trachea due to mega-esophagus secondary to achalasia have been described. In this case, the patient presented a gastro-esophageal distension with airway compression and consequent respiratory failure that reverted after resolution of the digestive problem. This clinical case demonstrates the importance of considering less common causes of airway obstruction.

**Key words:** *Respiratory failure. Airway obstruction.*

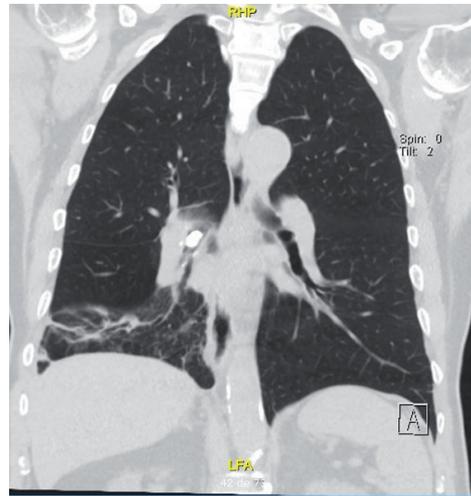
#### PE 075. THE MISSING TOOTH - A CASE OF OBSTRUCTIVE PNEUMONIA

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*Hospital Egas Moniz.*

**Case report:** A.A.M, an 85-year-old male, with a history of prostate neoplasia, reflux disease as well as a 10 pack-year smoking history, was sent to a Pulmonology consultation by the general practitioner. He presented a persistent cough with brown coloured sputum and exertional dyspnea since a recent admission to the hospital in March 2017 due to a right lower lobe Pneumonia with respiratory failure. He denied fever, weight loss or hemoptysis. On auscultation, wheezing from the right hemithorax was audible. Pulmonary function tests (PFT) and a chest CT-scan were ordered. In the 2<sup>nd</sup> consultation the patient had failed to do the CT-scan, but brought the results from the PFT, which demonstrated a small obstructive alteration, especially in the small airways, as well as air trapping. He mentioned a recent respiratory tract infection, initially medicated with azithromycin but since prescribed levofloxacin due to therapeutic failure. Thorax-CT Scan showed a high density foreign body, with approximately 10 × 7 mm, located in the right lower lobe, exactly below the bifurcation of the lower and medium lobes. In the right lower lobe was an area of consolidation as well as traction bronchiectasis and pulmonary volume loss, corresponding to infectious/inflammatory aspects. The patient then recalled that in March of 2017, previously to his admission to the hospital, he had been submitted to a dental extraction where one of the dental pieces had broke and he supposedly swallowed it. At the time, he was evaluated at Hospital da Luz, eventually being discharged. A rigid bronchoscopy was performed in May 2018 with removal of the foreign body, corresponding, as expected, to a dental fragment, with good endoscopic results and repermeabilization of the right lower lobe. Since then, the patient has been well, on auscultation with symmetrical breath sounds and no adventitious respiratory sounds.

**Discussion:** Foreign body aspirations that are not recognized at the time can lead to insidious symptoms, presenting a diagnostic challenge when clinical suspicion is low. They can lead to numerous complications, the most common being obstructive pneumonia. The treatment for obstructive pneumonia includes not only the removal of the obstruction, but also antibiotic therapy for the infection.



**Key words:** *Persistent cough. Obstructive pneumonia. Foreign body. Tooth.*

#### PE 076. TRACHEOBRONCHOPATHIA OSTEOCHONDROPLASTICA - AN INCIDENTAL DIAGNOSIS CASE REPORT AND LITERATURE REVIEW

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**Introduction:** Tracheobronchopathia osteochondroplastica is a rare entity of unknown cause and pathophysiology. The precise incidence is uncertain, but it may vary between 1:200-1:1,000. It was initially described by Rokitansky in 1855, based on a postmortem biopsy. It is a benign disease usually characterized by multiple bone and cartilaginous nodular formations on the submucosa of the trachea and bronchi, characteristically on the luminal surface of the anterior and lateral walls. The diagnosis is usually made over 50 years old and the spectrum of clinical manifestations is diverse, with the majority of patients being asymptomatic. The most common clinical manifestations are chronic dyspnea and chronic cough. The gold standard of diagnosis is bronchoscopy, which reveals typical changes, namely the presence of nodules of hard consistency, with a classic pattern such as “rock-garden” or “cobblestone”. Although biopsy is not essential for diagnosis, it is important to rule out other conditions such as endobronchial neoplasm, amyloidosis, sarcoidosis, tuberculosis, re-lapsing polychondritis and papillomatosis. The treatment is merely symptomatic as a specific and effective therapy is yet to be determined. The disease prognosis is usually good, although in a minority of cases serious complications can occur.

**Case report:** We present the case of a 68-year-old Caucasian woman, previously a smoker, with prior atherosclerotic disease diagnosis

and submitted to aortobifemoral prosthesis placement. This patient also had chronic kidney disease of obstructive etiology, causing multiple hospital admissions due to urosepsis. During the course of the diagnosis and after a computed tomography of the thorax, irregularities suggestive of polypoid lesions were documented in the luminal contour of the trachea and the left main bronchus, especially in the anterior wall. Bronchofibroscope revealed the presence of multiple nipple lesions, of hard consistency and white color, indicative of tracheobronchopathia osteochondroplastica, previously unknown and asymptomatic. From the aetiological study, the main pathologies that make differential diagnosis were excluded, such as amyloidosis, tuberculosis and neoplasms.



**Discussion:** With this clinical case we intend to approach a rare benign pathology, to emphasize that the majority of the patients in the presentation are asymptomatic and that the diagnosis may be incidental in the course of the extensive diagnostic investigation of other pathologies.

**Key words:** *Tracheobronchopathia osteochondroplastica. Trachea. Bronchi.*

#### PE 077. ACUTE AIRWAY OBSTRUCTION DUE TO MULTINODULAR GOITRE

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*Centro Hospitalar e Universitário de Coimbra-Medicina Interna A.*

**Introduction:** Multinodular goitre is a common illness. When accompanied by obstructive symptoms, such as dyspnoea, it carries

an indication for surgery. For the benign cases of multinodular goitre, airway obstruction is rare.

**Case report:** An 88-year-old female patient is admitted to the emergency room presenting with acute dyspnoea. She has a history of progressive exertional dyspnoea and fatigue throughout the previous year. The patient had prior known history of aortic stenosis, systemic arterial hypertension, and dyslipidaemia. At observation, she was afebrile, polypnoeic, tachycardic, and had a peripheral oxygen saturation of 88% despite high-flow oxygen supplementation. She exhibited stridor and, on both lung fields, crackles and wheezing. Arterial blood gas sampling showed hypoxaemia (pO<sub>2</sub> 59.5 mmHg) absent of hypercapnoea. Chest radiography showed marked widening of the upper mediastinum. Electrocardiography showed sinus rhythm, a heart rate of 120 bpm, along with a left branch block pattern. Cytometry and biochemical studies were inconspicuous and included BNP (157 pg/mL), TSH (0.044 uIU/mL, reference range: 0.4-4.0), and free T<sub>4</sub> (1.1, reference range: 0.8-1.9). A chest computed tomography was performed which revealed marked enlargement of the thyroid gland, with an extensive intrathoracic component which impinged on and compressed the superior half of the trachea, which was left with a minimum luminal caliber of 2 mm. Both of the brachycephalic trunks also suffered from extrinsic compression from the thyroid, with marked luminal stenosis. Also present was a left pleural effusion of reduced volume and atelectasis of both inferior pulmonary lobes. No adenopathies were observed. The patient was administered furosemide, adrenaline, bronchodilators and corticosteroids (both inhaled and intravenous), with mild improvement of her symptoms. She was admitted to the enfermary for stabilisation. Throughout her period of admission, she improved from her respiratory failure and her stridor lessened. Despite this, she maintained the necessity for oxygen supplementation and intravenous corticosteroid. She was proposed for total thyroidectomy. The surgery was without complications and the thyroid was fully removed *en bloc*. Despite this, the patient passed in postoperative day 2 from ventricular fibrillation.

**Discussion:** Here we present the case of a patient bearing an illness that usually carries a benign presentation. In the cases of airway obstruction such as the one described above, however, it may carry fatal outcomes. This patient had no previous history of thyroid illness. Six years prior to her admission, she had undergone chest radiography which exhibited superior mediastinal enlargement and that was, at that time, overlooked. Recognition of these cases is important, as they constitute a preventable cause of mortality if timely diagnosed and treated.

**Key words:** *Intrathoracic goiter. Stridor. Tracheal stenosis.*

#### PE 078. THE PNEUMOTHORAX AND THE PEA - UNUSUAL PRESENTATION OF FOREIGN BODY ASPIRATION

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**Introduction:** Foreign body aspiration into the airways in adults is rare, occurring mainly above 75 years. It can cause acute or chronic symptoms or be asymptomatic, requiring a high level of suspicion. In the presence of imaging abnormalities, such as atelectasis or pneumothorax, this hypothesis must be placed. Although it occurs more commonly in the right lower lobar bronchus, it can affect the left bronchial tree in 23% of cases.

**Case report:** 79-year-old man admitted to the emergency department with dyspnea at rest and left thoracalgia with progressive worsening in 3 days. No cough, wheezing, fever or other symptoms and no previous trauma. He presented low peripheral oxygen saturation (88%) and decreased vesicular murmur on the left. History of

persistent allergic asthma and allergic rhinitis, medicated with budesonide/formoterol turbobaler®. Former smoker of 50 pack-years, with exposure to sulphation chemicals, chickens and doves. Laboratory tests showed a slight elevation of the C-reactive protein (1,78 mg/dl). Thoracic radiography presented hypertransparency of the left lung field with visible pleural line, compatible with pneumothorax, and a dense left hilar area due to pulmonary collapse (Fig. 1A). Thoracostomy was performed with insertion of thoracic drain in the 5<sup>th</sup> intercostal space anterior axillary line and underwater drainage, with almost total pulmonary expansion and symptomatic improvement, and the patient was admitted to the ward with oxygen therapy. Due to persistence of the dense left hilar area, suggestive of atelectasis, chest CT was performed confirming pneumothorax and collapse of the left upper lobe (LUL) with obstruction of the LUL bronchus (Fig. 1B). With further questioning the patient reported an episode of choking while eating meat and green peas 5 days before he went to the hospital, not clearly associating the beginning of dyspnea to this event. Videobronchofibroscopy was then executed with observation of total occlusion of the LUL bronchus by a foreign body of green color, probably a pea (Fig. 2A), surrounded by friable mucosa with easy hemorrhage, therefore not

allowing its removal by this technique. However, after the procedure, the patient presented several accesses of productive cough elimination of hemoptoic sputum and solid components, reporting subsequent clinical improvement. The thoracic radiography revealed total resolution of the atelectasis. Videobronchofibroscopy was repeated the next day confirming absence of foreign body and permeable bronchial orifices with local inflammatory signs (Fig. 2B). The bronchial aspirate, brushing and biopsies only identified the presence of *Providencia rettgeri* in culture, and the patient was treated with levofloxacin.

**Discussion:** The presence of organic foreign bodies in the airways may be asymptomatic until a complication arises, as in this case with atelectasis and pneumothorax. The patient does not always recalls or values the episode of aspiration, so a careful clinical history is essential. The presentation with pneumothorax is very rare and in this case it may have had a multifactorial origin, due to intense coughing and maintained bronchial obstruction with lobar atelectasis.

**Key words:** Foreign body. Pneumothorax. Atelectasis. Videobronchofibroscopy.

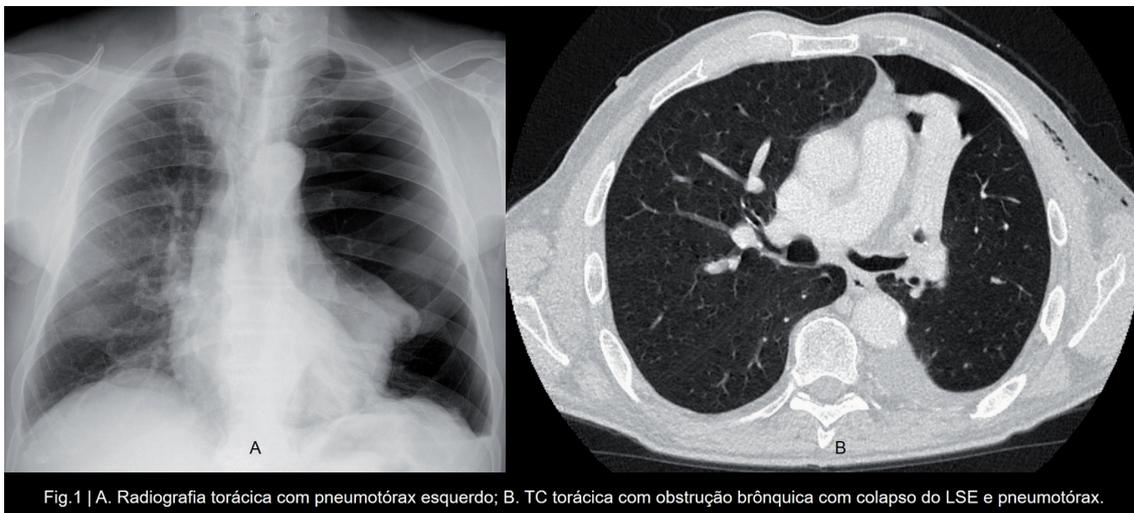


Fig. 1 | A. Radiografia torácica com pneumotórax esquerdo; B. TC torácica com obstrução brônquica com colapso do LSE e pneumotórax.

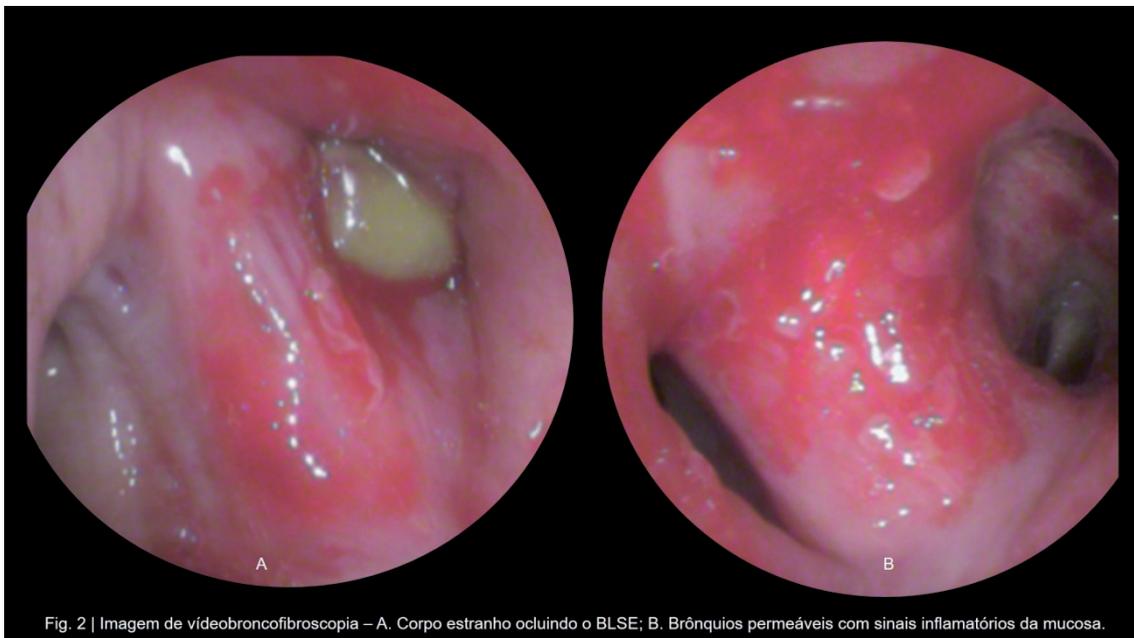


Fig. 2 | Imagem de videobronchofibroscopia – A. Corpo estranho ocluindo o BLSE; B. Brônquios permeáveis com sinais inflamatórios da mucosa.

### PE 079. AN EXUBERANTE CASE OF TRACHEOBRONCHOPATIA OSTEOCHONDROPLASTICA

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**Introduction:** Tracheobronchopatia osteochondroplastica (TO) is a rare disorder of unknown origin of the large airways, characterized by an abnormal growth of numerous cartilaginous and/or bony submucosal nodules, protruding into the tracheal lumen. Patients are commonly asymptomatic, but TO can present with wheezing, dyspnea, hemoptysis, cough or obstructive pneumonia. Computerized tomography (CT) and bronchoscopic examination with biopsy of the lesions are essential for a correct diagnosis.

**Case report:** It's a case of a 75-year-old male, admitted to the hospital for a Community Acquired Pneumonia, presenting with pleuritic chest pain and cough with hemoptoic sputum. During his stay in the hospital he underwent a thoracic CT scan, which showed an exuberant, diffuse irregularity of the trachea and both main bronchi. Examination by bronchofibroscopy revealed a marked decreased of the tracheal lumen caused by extense proliferation of polypoid lesions, with apparently normal mucosa. Bronchial biopsies showed unspecific inflammation and epidermoid metaplasia. The patient was then submitted to a rigid bronchoscopy, where larger biopsies of the polypoid lesions were obtained. Histological examination showed cartilage and mature bone fragments, confirming the diagnosis of tracheobronchopatia osteochondroplastica. Treatment of the underlying Pneumonia resulted in the resolution of all the symptoms. No direct treatment was necessary on the upper airway and the patient remains asymptomatic, under clinical vigilance.

**Discussion:** The verification of these lesions on bronchoscopy is accept as sufficient to confirm diagnosis, without the need for a histology study, however, ideally biopsies are performed with visualization of bone or calcification of the submucosa. The disease course is usually benign and slow, and complications or such an exuberant form are extremely unusual.

**Key words:** Tracheobronchopatia Osteochondroplastica. Bronchoscopy.

### PE 080. TRACHEOBRONCOPATHIA OSTEOCHONDROPLASTICA - A PICTURE IS WORTH A THOUSAND WORDS

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**Introduction:** Tracheobronchopatia osteochondroplastica (TO) is a benign rare disease of unknown pathogenesis and etiology. It is characterized by the presence of multiple submucosal osteocartilaginous nodules that involve luminal surfaces of anterior and lateral walls of the tracheobronchial tree, sparing the posterior wall. It is usually asymptomatic, but it can present with respiratory symptoms like chronic cough, dyspnea, wheezing and occasionally hemoptysis. In most cases, it is an accidental finding in bronchofibroscopy to investigate other symptoms or to exclude other diagnostic hypotheses.

**Case report:** A 68 year old woman, former smoker and with multiple cardiovascular comorbidities, was admitted for urosepsis and performed a chest-abdomen-pelvic computed tomography to exclude obstructive uropathy. The thoracic CT-scan documented tracheal wall irregularity, mostly anterior, with polypoid lesions, and similar images on the left bronchi. Although the patient had no respiratory symptoms, it was performed a videobronchoscopy which revealed the presence of multiple protrusions of hard consistency and light color along the anterior wall of the tracheal mucosa that extended to the left main bronchus. This appearance was compatible with the endoscopic diagnosis of tracheobronchopatia osteochondroplastica. Several biopsies of the lesions were performed confirming the diagnosis through histopathologic examination.

**Discussion:** Tracheobronchopatia osteochondroplastica is a rare, idiopathic, benign and often asymptomatic nosological entity, therefore its incidence *in vivo* may be underestimated. Bronchoscopic images are themselves often diagnostic of the disease and should be easily identified by pulmonologists and bronchologists. Nevertheless, biopsies are relevant to the exclusion of other differential diagnoses. The prognosis of patients with tracheobronchopatia osteochondroplastica is favorable and treatment is aimed at symptoms relieve or when patients develop serious complications like tracheal stenosis and/or bronchial and hemoptysis.

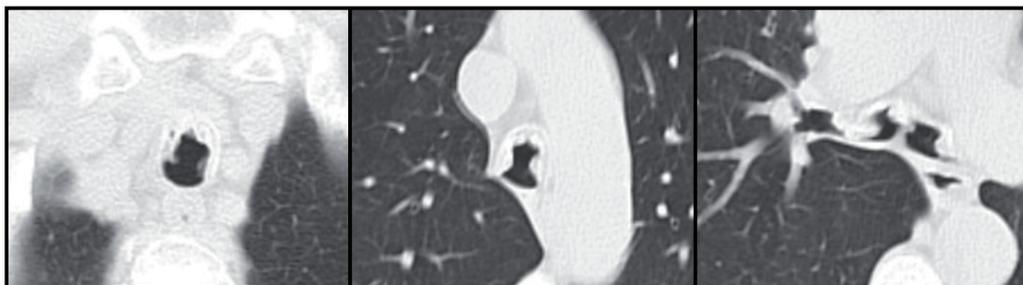


Figure 1 PE 079. Trachea and main bronchi.



Figure 2 PE 079. Endoscopic images.

**Key words:** *Tracheobronchopathia osteochondroplastica. Traqueobroncopatia osteocondroplástica. Computed tomography. Bronchoscopy.*

### PE 081. PULMONARY EMBOLISM WITH HEMODYNAMIC COMPROMISE: WHAT FUTURE?

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**Introduction:** The incidence of pulmonary embolism (PE) has increased in Portugal although the mortality of this pathology decreased but still remains at high levels. Risk stratification and early management of this condition with individualized treatment for each situation are of major importance. In hemodynamic shock situations the need for immediate action should not overcome a correct global evaluation of the patient so the right procedures can be chosen and adapted to each patient. We describe a clinical case of PE with hemodynamic compromise where initial intervention was postponed due to active bleeding; after patient stabilization it was used interventional thrombus fragmentation and aspiration technique instead of most frequent used systemic thrombolytic therapy.

**Case report:** A 64-year-old man, Caucasian, autonomous until 2 months ago. Has a known history of hypertension, gout, compressive spondylitis myelopathy with functional limitation since the last 2 months. He was taken to Portalegre's Hospital Emergency room because of thoracic pain, mental confusion and lost of movement in the legs since the day before. During observation the patient was in polypnea and tachycardia, with no pulse in the lower limbs, hypotension in the right arm and not measurable blood pressure in the left arm. Gasometry in normal air showed respiratory alkalosis and severe hypoxemia (PaO<sub>2</sub> of 47 mmHg). Electrocardiography with ST elevation from V1 to V3 and myocardial necrosis biomarkers increment in laboratory analysis. CT angiogram was performed and revealed main pulmonary arteries obstruction, right ventricle and pulmonary artery dilatation, complete aortoiliac artery bifurcation obstruction and spleen infarct. Massive PE was assumed, central venous catheter (CVC) was introduced and non-fractionated heparin was given. After this the patient was sent to Santa Marta Hospital. In this Hospital we verified an increase in hemodynamic instability and massive right cervical hematoma. CT scan proved a right internal jugular laceration, probably due to iatrogenesis, as source of the bleeding. In patient with massive PE, aorta embolism and lower limbs ischemia we chose to postpone the management of these conditions due to active bleeding and risk of major complication and surgery for jugular repair was performed. After 3 days in the intensive care unit (ICU), with aminergic support, and because of major complication risk for thrombolytic therapy, the patient was submitted to thrombus fragmentation technique. In the days after the procedure the patient evidenced major hemodynamic, clinical and gasometry improvement, so he was discharged from the ICU.

**Discussion:** Some centres have discussed the implementation of an PE "green line" in emergency room. This should include a patient's risk stratification and a multidisciplinary approach to achieve therapeutic optimization in emergency situations. It is expectant that some arterial intervention techniques, approved for other pathologies, would also be approved for the treatment of EP in a near future. The use of catheter fragmentation and local thrombolysis are important tools for the intervention decision in cases, as the described, where thrombolytic systemic therapy, regularly used, is too dangerous.

**Key words:** *Pulmonary embolism. Thrombus fragmentation.*

### PE 082. THE IMPORTANCE OF THE BERG BALANCE SCALE - SHORT AND LONG VERSION - ON THE FALL RISK ASSESSMENT AND THE STRATEGY TO IMPLEMENT IN A HOME-BASED PULMONARY REHABILITATION PROGRAM - REABILITAR

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**Introduction:** The change in balance control is known as a significant secondary manifestation in individuals with COPD and a major predictor of the fall risk, especially among older adults, which can lead to functional limitations and decreased mobility. In the context of a home-based pulmonary rehabilitation program (PRP), the functional assessment of balance is crucial in the clinical evaluation needed for the decision making related to the safety and personalization of the program.

**Objectives:** 1) To understand the importance of the Berg Balance Scale (BBS) on the balance and the fall risk assessments for the inclusion of COPD individuals to a home-based PRP - reabilitAR. 2) To compare the scores obtained with the short and the long versions of the BBS to evaluate if there is an increased value in administering the long version for the safety of the home exercise program.

**Methods:** Two individuals referred to the reabilitAR program performed the baseline evaluation, including the administration of the short version of BBS. In the program's routine evaluation, the scale is used to evaluate, quickly and with simple resources, the safety of a self-managed exercise program in a home-based setting and to assess the need to include specific balance exercises to the program. According to the low scores obtained with the short version, scores associated with a high risk of fall, the long version was administered to the 2 individuals with the objective of describing the alteration for their balance with additional and more detailed information. This additional information was necessary for the inclusion of the individuals to the program and to target the possible adjustments that needed to be done to the program.

**Results:** The results obtained with the short version showed a high risk of fall (18 and 8 points/28, for individual A and B respectively). After, with the interpretation of scores of the long version (42 and 27 points/56, for individual A and B respectively) the balance impairments were confirmed. The analysis of the scores showed that the safety of the exercise program, as it is usually executed, was compromised, which led to the necessity of developing specific program adaptations. For individual A, the higher score allowed his inclusion in the program with the exercise self-management. However, specific and supervised balance exercises were added. The individual B, with the lowest score, was oriented to an exercise program that was subjected to major changes involving an increased number of days with the physiotherapist combined with specific balance exercises without the exercise self-management component until a new evaluation of the clinical condition is done.

**Conclusions:** The use of the long version of the BBS can present important advantages for the clinical evaluation of people with balance alterations assessed with the short version as it is more sensitive to discriminate in detail these alterations. The long version allows more precise information about the balance impairment, allowing the elaboration of a personalized strategy and adequate supervision in order to assure the security (lower fall risk), quality and efficacy of a home-based PRP (reabilitAR Program).

**Key words:** *Fall risk. Berg balance scale. Home-based pulmonary rehabilitation.*

### PE 083. INSPIRATORY MUSCLE TRAINING IN CERVICAL SPINAL CORD INJURY

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**Introduction:** Respiratory dysfunction post cervical spinal cord injury (SCI) is characterized by weak or paralyzed respiratory muscles, resulting in reduced lung volume, ineffective cough and increased respiratory tract infections. Inspiratory muscle training (IMT) has been shown to increase inspiratory muscle strength and endurance on people with chronic lung conditions. It has been suggested that increasing strength of inspiratory muscles through specific training in people with SCI could potentially improve pulmonary function and cough capacity.

**Objectives:** The objective of the present study was to assess the immediate effects of IMT in addition to usual rehabilitation care in cervical SCI patients with inspiratory muscle weakness.

**Methods:** Retrospective analysis of patients with cervical SCI, admitted for inpatient rehabilitation program between 2016 and 2018, with a maximum period of 12 weeks, who had impaired inspiratory muscle function (maximum inspiratory pressure < 60 cm/H<sub>2</sub>O). The IMT protocol consisted of two daily sessions, using a Threshold IMT, with a training load of 60% of MIP. Each session comprised 3 sets of 10 breaths. The evaluations were performed at admission and at the time of discharge. Primary outcome measures were MIP and peak cough flow (PCF). Inferential statistics included Wilcoxon signed-rank tests.

**Results:** A total of 9 patients (78% male; 55 ±19 years), with an average time of injury of 55 months (SD = 66) to date of admission, ASIA Impairment Scale (AIS): A (22%), B (12%), C (11%), D (55%) and mean of forced vital capacity of 2.04 liters (SD = 0.95). A significant improvement in MIP was observed (admission: median 52 cmH<sub>2</sub>O, interquartile range [38.5-58] vs discharge: median 68 cm/H<sub>2</sub>O [54-88.5]; p = 0.008). Despite the positive effects on PCF, these were not considered statistically significant (admission: median 200 l/min [105-325] vs discharge: median 260 l/min [130-330]; p = 0.123). It should be noted the absence of adverse effect related to IMT protocol.

**Conclusions:** Despite the limitations of the study related to the small sample size, IMT has a positive effect on inspiratory muscle function in people with cervical SCI who have impaired inspiratory muscles during an inpatient rehabilitation program.

**Key words:** *Spinal cord injury. Inspiratory Muscle training.*

### PE 084. PULMONARY REHABILITATION IN PATIENT WITH CHRONIC OBSTRUCTIVE PULMONARY DISEASE

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**Introduction:** Pulmonary rehabilitation is an evidence-based, multidisciplinary, and embracing intervention for patients with chronic respiratory diseases who are symptomatic and have decreased daily life activities, being designed to reduce symptoms, optimize functional status and reduce health care costs, stabilizing or reversing systemic manifestations of the disease.

**Objectives:** To evaluate if pulmonary rehabilitation is an effective intervention in chronic respiratory disease, particularly COPD.

**Methods:** Research on systematic reviews (SR), clinical trials (CT) and retrospective studies (RS), in the last 5 years, in Portuguese and English, of the terms "pulmonary rehabilitation" and "COPD".

**Results:** We gotten 24 articles and 2 RS, 4 CT and 3 RS were selected. Respiratory rehabilitation is indicated in any patient with

chronic respiratory disease who presents dyspnea, with low tolerance to effort and restricted in their daily activities, despite optimized medical therapy. Pulmonary rehabilitation relieves dyspnea and fatigue, improves emotional function and enhances the sense of control that individuals have over their condition. Respiratory rehabilitation reduces hospital readmissions (moderate - quality evidence) and improves the quality of life and the ability to exercise (high - quality evidence), however, such improvements do not translate into a reduction in mortality.

**Conclusions:** Pulmonary rehabilitation plays an important role in COPD, and it is beneficial in improving the quality of life and the ability to exercise, with scores to evaluate it as the Short Physical Performance Battery, that can be used before and after a respiratory rehabilitation program to try to understand if there was an increase in tolerance to physical exercise. Pulmonary rehabilitation is also a safe intervention in patients with COPD following the onset of an exacerbation. This can be done either in a clinical environment or at home, which gives it importance and versatility as a therapeutic measure. Future research studies should focus on identifying which components of pulmonary rehabilitation are essential, such as, ideal length, the degree of supervision and intensity of training required and how long treatment effects persist.

**Key words:** *Pulmonary rehabilitation. COPD.*

### PE 085. EFFECTS OF A COMMUNITY-BASED EXERCISE TRAINING IN COPD PATIENTS

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**Introduction:** The benefits of pulmonary rehabilitation in COPD patients are considerable. Not all hospitals have conditions to provide rehabilitation to all patients who would benefit from that.

**Objectives:** Evaluate the effects of community-based exercise training on exacerbations, symptomatology and quality of life on COPD patients.

**Methods:** A near-experimental study was carried out with a physical exercise program at the patient's local health center with a decrease in the number of supervised sessions and total 6 months duration (3 weekly sessions for 2 months, 1 weekly session for 1 month and 1 monthly session for 3 months). Each session has an aerobic exercise component and a limb strength training supervised by a physiotherapist. The *modified Medical Research Council (mMRC)*, the *COPD Assessment Test (CAT)*, the *Hospital Anxiety and Depression Scale (HADS)*, the *St George's Respiratory Questionnaire (SGRQ)* and a 6 minutes walking test (WT6M) were applied initially, at 2, 3 and 6 months.

**Results:** Were included 11 patients with a male predominance (72.2%) and an average age of 67.18 years (standard deviation 6.88). Most were ex-smokers (63.6%) and had an obstruction with a post bronchodilation forced expiratory volume in the 1st second of 52.81% (standard deviation 19.86). At 2 months, scores were improved in all questionnaires applied with statistical significance at mMRC and SGRQ (Impact and Total). There was also a clinically significant improvement in the 6-minute walk distance in 7 of the 11 patients. At 3 months of the program (after decreasing supervision) there was a maintenance of the gains obtained with the exception of the HADS (Depression) score that increased to pre-program values.

At the end of the program, patients who had a significant gain in WT6M decreased their performance, although only 1 returned to pre-program values. There was also an aggravation on CAT questionnaire score as well as the SGRQ (Impact).

**Conclusions:** The developed program was feasible and clinically effective at 3 months, however, it was verified loss of majority gains after the end of the sessions followed by a professional.

**Key words:** COPD. Community-based exercise training.

#### PE 086. PULMONARY TROMBOEMBOLISM: MULTIFACTORIAL ETIOLOGY AND THE IMPORTANCE OF AN INDIVIDUALIZED THERAPEUTIC

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**Introduction:** Pulmonary thromboembolism (PTE) has a wide clinical profile in its presentation, being one of the most important causes of morbidity and mortality and hospitalization. Its etiology is multifactorial, its clinical suspicion and diagnosis is sometimes a challenge and the early therapeutic approach is determinant.

**Case report:** The authors present the case of a 56-year-old male, smoker (40-pack year), with ethanolic and toxicophilic habits (cocaine, hashish and marijuana) marked. The patient is referred to the Emergency Service (ES) for lipotoma, chest tightness, sweating and sudden onset dyspnea, with less than 24 hours of evolution. In the ES it was hypotensive, normocardial, under 6 L/min oxygen therapy. From the complementary diagnostic tests carried out, an increase of the D-Dimers 2.7 ug/mL is highlighted in the laboratory evaluation. On suspicion of PE, he performed an angioTC of the chest that revealed extensive reperfusion defects involving the distal segment of the left pulmonary artery and several lobular and segmental arteries of both lungs, main trunk of the pulmonary artery of normal caliber, but with right ventricular dilatation and a transthoracic echocardiogram with right ventricular dilatation, left ventricular motion of the basal segment of the interventricular septum, slight tricuspid regurgitation and inferior vena cava 19 mm with a variation of less than 50% with respiration. After confirming the diagnosis of bilateral PTE, the patient was admitted to an intensive care unit and started anticoagulation with unfractionated heparin with a switch to warfarin and later to rivaroxaban. The study of thrombophilias revealed an antithrombin III deficiency (70%). Upon hospital discharge, the patient had an indication for referral to the General Pneumology, Smoking Cessation and Psychiatry Consultation (having refused), of Immunohemotherapy. In the reassessment in General Pneumology Consultation, the patient maintained the inhaled tobacco and toxicophilic habits. Transthoracic revascularization echocardiography performed at the outpatient clinic showed a slight tricuspid regurgitation.

**Conclusions:** We highlight the importance of the multifactorial etiology of this clinical case (antithrombin III deficiency and smoking) presenting a probable alteration of the pulmonary vasculature resulting from the chronic consumption of cocaine. The individualized therapeutic approach used is also highlighted.

**Key words:** Pulmonary thromboembolism. Smoking. Antithrombin deficiency III. Rivaroxaban.

#### PE 087. THE IMPORTANCE OF VARIATIONS IN MINIMAL EXPIRATORY PRESSION IN THE ADHERENCE AND COMFORT OF THE PATIENT WITH OBSTRUCTIVE SLEEP APNEA SYNDROME | ABOUT A CLINICAL CASE

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**Introduction:** The obstructive sleep apnea syndrome (OSAS) is characterized by a partial or total interruption of the upper airways that

consequently leads to a reduction of the airflow, leading to desaturation and nocturnal awakenings. Adherence to therapy in the treatment of OSAS is one of the main objectives for rehabilitation. It is considered a total adherence in patient that use daily the therapy, with an average utilization equal or more than 4h/night, at least 70% of the nights. These results are possible when appropriate clinical decision is achieved to conduct the therapy as well as proper adaptation of the interface and technical follow-up (reinforcement on education and clarification of possible questions). The home visits allow a monitoring of the adherence of the patient and should be reported to the medical specialist for possible adjustments.

**Case reports:** Clinical case 1: Male, 65 years, starts therapy with Auto-CPAP with the following parameters: EPAP max: -16 cmH<sub>2</sub>O, EPAP min: - 4 cmH<sub>2</sub>O, Ramp - 45 minutes, Expiratory pressure relief - 3.0 cmH<sub>2</sub>O. In the first home visit of the first month, the patient was non-adherent with the following results: percentage of utilization > 4h = 0%; average value of daily utilization: 3 minutes; P (95): 10.6 cm H<sub>2</sub>O, without significant no-intentional leakage (16.8 L/min); residual AHI: 0.5 events/h and showed complaints of low pressure in the beginning of the therapy which created discomfort and dyspnea in the patient. The healthcare professional reported to the medical specialist about the collected information in the home visit and asked permission to remove the ramp to promote more comfort of the therapy until the next medical evaluation, which it was allowed. The patient was evaluated at the hospital in the following week and it was changed the prescription to EPAP min - 8 cmH<sub>2</sub>O. In the following home visit, after 3 months, the results were: percentage of utilization > 4h = 61.1%; mean value of daily utilization: 04h59 minutes; P (95): 13.0 cmH<sub>2</sub>O, with significant intentional leakage (28.8 L/min) - with adjustment of the mask; residual AHI: 4.6 events/h.

**Discussion:** The increase in EPAP (min) of 4 to 8 cmH<sub>2</sub>O was accompanied by increased adherence. Despite the patient still not being considered adherent, it is considered that there is adherence to non-invasive ventilation if the utilization is more than 4 hours in a period of more than 70% of the nights. The patient has a restful sleep with consequent improvement in activities of daily living and we can anticipate, in a short period of time, that this will influence the adherence. Education and monitoring in Respiratory Care in patients with Obstructive Sleep Apnea Syndrome, with healthcare professionals, should include the motivation for adherence as well as the identification of factors that can influence it.

**Key words:** OSAS. Minimum EPAP. Ramp. Adherence. Comfort.

#### PE 088. OBESITY HYPOVENTILATION SYNDROME: THE INFLUENCE OF INTERDISCIPLINARY MEDICAL APPOINTMENT ON THE PROGNOSIS OF A CLINICAL CASE

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**Introduction:** Monitoring patients undergoing continuous positive airway pressure ventilation treatment aims adherence, maintenance of therapeutic efficacy, clinical improvement and intervention in the adoption of healthy lifestyles. Hospital NIV consultations play a vital role in the follow-up of these patients and new multidisciplinary approaches aim a close partnership between hospital healthcare professionals and home respiratory care providers.

**Case report:** Female patient, 43 years, BMI = 30.8 Kg/m<sup>2</sup>. Diagnosis of obstructive sleep apnea syndrome (OSAS) in 2014 and diagnosis of obesity hypoventilation syndrome in 2016, presenting

several successive hospitalizations. In June 2014, she started ventilation therapy with Auto-CPAP in a sleep consultation ( $P_{min}$ : 6 and  $P_{max}$ : 16) and after switching to a NIV consultation (multidisciplinary consultation) changed therapy, in August 2015, to spontaneous bi-level (S) (EPAP: 8 and IPAP: 16), without ramp and with face mask. BMI = 68.6 Kg/m<sup>2</sup>. Several teaching reinforcements were carried out as well as a joint strategy to encourage weight loss with referral of the patient to psychology and nutrition consultation. In June 2017, during hospital NIV consultation is measured spontaneous/timed bi-level (S/T) with EPAP: 6; IPAP: 14; FR: 14 keeping face mask. BMI = 55.6 Kg/m<sup>2</sup>; Reinforcement of adoption of healthy lifestyles. Highly motivated patient for weight loss, recognizes benefits in the current quality of life and aims at total remission of OHS pathology. Rejoined the computer course, which she had abandoned due to concentration deficit, fatigue and apathy. She maintained therapeutic adherence (average of 9h/night), controlled leakage, controlled residual AHI (residual AHI = 2.4 events/h) and always recognized therapeutic benefits. In July 2018, she returned to the NIV consultation refusing therapy, mentioning that she no longer recognized benefits considering her high mental state and mood, with BMI = 30.8 Kg/m<sup>2</sup>. The doctor then request a sleep study repetition, which took place in July 2018, culminating with suspension of bi-level therapy.

**Discussion:** Successive reductions in BMI were accompanied by a decrease to lower stages of disease severity. The patient began to have a restful sleep with consequent improvement in activities of daily living. Teaching and monitoring of homecare treatments in patients with OHS by healthcare professionals should include motivation for weight loss, reinforcing the importance of physical exercise and the adoption of healthy lifestyle habits. An adequate articulation between the patient, homecare technician and the Hospital healthcare professionals is extremely important for the promotion of adherence to therapy, maintenance of therapeutic efficacy, clinical improvement and change of risk behaviors, the focus of the organization of these interdisciplinary consultations.

**Key words:** Home respiratory care. Therapy monitoring. Interdisciplinary consultations. Weight loss.

#### PE 089. PROMOTION OF ADHERENCE TO VENTILATION THERAPY: IMPORTANCE OF THE HEALTHCARE PROFESSIONAL IN THE AWARENESS OF THE PERSONAL AND SOCIAL RESPONSIBILITY OF THE PROFESSIONAL DRIVER WITH OSAS

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**Introduction:** The OSAS patient (Obstructive Sleep Apnea/Hypopnea Syndrome) does not usually realize that he has the disease being referred to the specialist by the spouse or closer people. Some of the most frequent signs/symptoms are snoring, breathing pauses during sleep, irritability, morning headache. In moderate or severe clinical cases, the Gold Standard of the treatment is CPAP (Continuous Positive Airway Pressure), Auto-CPAP (Automatic Continuous Positive Airway Pressure) or Bi-levels (S or Auto). The control of OSAS through these therapies is not always easy, being frequent the difficulty of adaptation and consequent low adherence. It is considered that there is adherence to ventilation (CPAP/AutoCPAP/Auto Bi-level/Bi-level S) if there is more than 4 hours of use in a period of more than 70% of nights and there is therapeutic efficacy whenever clinical improvement is associated with an index of apnea-hypopnea (AHI) < 5/h.

**Case report:** Male, 60 years, professional driver, ex-smoker, hypertensive, obese (Grade III). In June 2011 he was diagnosed with severe OSAS (35.6 AHI), being prescribed with AutoCPAP (5-11 cmH<sub>2</sub>O) and face mask. Refusing, since the diagnosis, to assume the pathology, at the visit of 1<sup>st</sup>, 3<sup>rd</sup> and 6<sup>th</sup> month, included in the protocol of homecare visits, it was verified that the patient did not follow the therapy. It was communicated to the prescriber and several re-adaptations were made to the therapy, however, only in May 2013 began to truly use it, under insistence of the spouse. Continuing without recognizing the disease, in August 2013 it is again evaluated for consultation, with still low adherence (~ 2h30) and AHI = 17.4. The patient was reassessed and consulted every six months resulting in pressure changes, with the aim of improving adherence. However, there was no progress. After the various motivation attempts, the healthcare professional adopted a more rigorous and assertive stance during a homecare monitoring visit (June 2015), reinforcing not only the benefits of the therapy, already exposed in the initial adaptation, but also the responsibility and consequences that, both personally and socially, could come as result of increase in the severity of the disease, such as loss of driving license, cognitive alterations, etc., resulting in a gradual change of posture and increasing therapeutic adherence.

**Discussion:** Adequate patient awareness by healthcare professionals, sometimes adopting a more assertive stance and with clear reference to the personal and social responsibility of the patient, can be fundamental in regaining adherence when there is no justification for non-compliance, especially in professional drivers with OSAS.

**Key words:** OSAS. Adherence. Professional driver. Home respiratory care professional.

#### PE 090. OBSTRUCTIVE SLEEP APNEA SYNDROME IN A PATIENT WITH HYPOGONADOTROPHIC HYPOGONADISM

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**Introduction:** Obstructive sleep apnea (OSA) is a common chronic sleep disorder characterized by repeated episodes of complete or partial upper airway obstruction during sleep, resulting in oxygen desaturation, sleep fragmentation, and daytime sleepiness. OSA in middle-aged men is often associated with decreased testosterone secretion, together with obesity and aging. On the other hand, androgen therapy may precipitate obstructive sleep apnea in men. Several studies have shown that exogenous testosterone exacerbates OSA symptoms, increases the apnea-hypopnea index and decreases oxygen saturation, as well as promotes alterations in sleep structure with reduction of REM time and its latency. The effect of testosterone on OSA is not exerted by changing the dimensions of the upper airway; instead, testosterone most likely contributes to OSA via central mechanisms.

**Case report:** A 36 years old man, health care technician, with the diagnosis of hypogonadotropic hypogonadism, without other relevant pathological or family antecedents, was referred to Pulmonology department because of snoring and nocturia since two years ago. The patient referred poor sleep quality with frequent awakenings and denied morning headaches or daytime hypersomnia with an Epworth scale of 2. He was supplemented with testosterone without other usual medication. The patient was a never-smoker and denied alcohol consumption. Regarding the general physical examination there were no major changes with an oropharynx of normal configuration (Mallampati scale of I). The body mass index was 24 kg/m<sup>2</sup>. In order to diagnose a sleep disorder, a polysomnography (PSG) level 1 were performed. The PSG revealed a marked

decrease in sleep efficiency (65.9%) in a possible relation with the effect of the first night of sleep in the laboratory, a decrease in deep sleep (4.5%), with an increase in REM latency (287 minutes) and a decrease in sleep time of REM (13.3%). No periodic limb movements in sleep were detected. The respiratory disorder index was 14 per hour, essentially at the cost of hypopnoea and respiratory efforts related to arousals. The patient started auto-CPAP with variable pressures between 6 and 14 cmH<sub>2</sub>O. At the 3 month follow-up visit, the patient had excellent compliance with resolution of respiratory events.

**Discussion:** Development of the signs and symptoms of OSA during testosterone supplementation requires assessment by polysomnography and, potentially, treatment with CPAP. If the patient is unresponsive or cannot tolerate CPAP, the testosterone dose must be reduced or discontinued. One of the potential hypothesis that explain the effect of testosterone in sleep breathing disorders is that fluctuating testosterone levels alters the chemoreceptors response to hypoxia and hypercapnia. Therefore, it is important to diagnose and treat OSA in patients with hypogonadism who are considering testosterone supplementation.

**Key words:** Obstructive sleep apnea syndrome. Testosterone. CPAP.

#### PE 091. IS HOSPITAL WORKERS' QUALITY OF LIFE AND SLEEP LINKED TO AGE?

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**Introduction:** Working in a hospital may be stressful and exhausting, often requiring shiftwork, and burnout symptoms may occur.

**Objectives:** To analyze the differences in quality of life, sleep, anxiety and depression between young adults (YA), ≤ 35 years old, and older adults (OA), > 35 years old, working at a central hospital.

**Methods:** Cross-sectional study in which data were collected using a questionnaire, randomly distributed to hospital workers, based on socio-demographic questions and sleeping habits, as well as the Portuguese versions of the following questionnaires: WHOQOL-Bref; Pittsburgh Sleep Quality Index (PSQI); Epworth Sleepiness Scale (ESS) and Hospital Anxiety and Depression Scale (HADS).

**Results:** A total of 137 workers answered our questionnaire; 59% were YA and 73% were female. There was a statistically significant difference between the two groups in relation to workload: YA had a significantly higher workload (64% worked more than 45 hours a week vs 39% of OA ( $p < 0.05$ )) and more of them worked night shifts (80% of YA worked night shifts compared to 36% of OA ( $p < 0.05$ )). Both groups, albeit without statistical significance, exhibited factors that contributed to a bad sleep quality: daily consumption of energy drinks (77% in both groups) and bad sleep hygiene (in 90% of YA and 78% of OA). In our evaluation of sleep quality, 77% of OA had PSQI scores of ≥ 5 (indicator of bad sleep quality) compared to 60% of YA ( $p < 0.05$ ). OA also had higher ESS scores than YA, but this difference was not statistically significant (18% of OA scored ≥ 11 - indicator of daytime sleepiness - compared to 13% of YA). Regarding quality of life, YA had higher average scores in all WHOQOL-Bref areas (indicating better quality of life), with a statistically significant difference in health satisfaction. There was also a statistically significant difference between YA and OA in terms of HADS scores, with a higher percentage of OA having scores suggesting depression (> 7) (29% of OA scored > 7, compared to only 7% of YA -  $p < 0.05$ )).

**Conclusions:** Even though YA had a higher workload and more of them worked night shifts, OA scored worse on all items evaluated, including: sleep quality, daytime sleepiness, quality of life and de-

pression, suggesting that hospital work is exhausting and leads to progressive exhaustion over time. Even though other factors may contribute to this difference, both groups had bad sleep habits, which may also lead to progressive exhaustion in the future. It is important not only to create better working conditions but also to promote better lifestyle habits so that hospital work does not become another exhausting factor for hospital workers.

**Key words:** Hospital work. Quality of life. Sleep.

#### PE 092. PREVALENCE AND CLINICAL CHARACTERISTICS OF ASTHMA IN CHILDREN 06-07 AND 13-14 YEARS OLD FROM LUANDA, ANGOLA

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**Objectives:** To evaluate the prevalence of asthma and other allergic diseases in children from Angola, and risk factors for asthma, in this population.

**Methods:** This was a cross-sectional study, using the International Study of Asthma and Allergies in Childhood (ISAAC) methodology, in Luanda, Angola between August to November 2014 and March to May 2015, in children from 6-7 and 13-14 years old. Were randomly selected, by municipality, 46 (8%) of 552 primary public schools and 23 (12%) of 186 secondary public schools. Asthma, rhinitis and eczema were defined according to the ISAAC protocol, based on symptoms in 12 months. The comparison of proportions was performed by chi-square test or Fischer exact test. To characterise the role of environmental risk factors for asthma, the Odds Ratio was used and a logistic regression model was developed. Data were analyzed using the SPSS Statistics program, version 24.0 and the statistical significance was of  $p < 0.05$ .

**Results:** The sample consisted of 3,080 children of 6 and 7 years old and 3,128 of 13 and 14 years old with validated questionnaires. The prevalence of asthma was 15.7% in children of 6-7 years, and 13.4% in children of 13-14 years, without significant differences between girls and boys. The assessment of respiratory function (peak flow) showed that more children 6-7 years of 13-14 years had moderate or severe bronchial obstruction (47.3% and 3.3% versus 9.5% and 0.2%, respectively). The prevalence of eczema and rhinitis was higher in children of 13-14 years than the children of 6-7 years: rhinitis - 26.9% versus 19.0%, respectively; eczema - 20.2% versus 18.4%, respectively. There were no significant differences between girls and boys in the group of 6-7 years, but the children of 13-14 years, rhinitis and eczema were more prevalent in girls. Rhinitis was associated with a greater number of episodes of wheezing and a greater number of episodes of night cough in both age groups with asthma. From the studied risk factors the presence of rhinitis, eczema, a split-type air conditioning system at home, the frequent intake of paracetamol, antibiotics in the first year of life, the frequent truck passage in the street of home, the presence of cats and dogs at home, and passive smoking in particular the mother, were associated with the presence of asthma.

**Conclusions:** Asthma and related allergic diseases such as rhinitis and eczema, are a public health problem in Luanda, since the prevalence of these diseases in children is relevant. Preventive and control measures should be encouraged.

**Key words:** Asthma. Children. Angola. Prevalence. Risk factors.

### PE 093. CONTROL OF ALLERGIC RHINITIS AND ASTHMA TEST (CARAT): IS TELEPHONE-BASED APPLICATION A RELIABLE AND VALID METHOD?

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**Introduction:** CARAT is a valid tool for assessing asthma and allergic rhinitis control over a 4-week period using paper-based forms. For follow-up purposes, telephone interview may be useful, but validity and reliability of this method has not been studied in this population. The aim of this study was to evaluate the telephone-based application of CARAT in patients with asthma.

**Methods:** Adolescents ( $\geq 15$  y) and adult patients were recruited in 25 secondary care centres (pulmonology, paediatric, and allergy)

between November 2017 and May 2018 in the context of an observational prospective study of the INSPIRERS project (URL: [goo.gl/KdEJ2L](http://goo.gl/KdEJ2L)). Patients filled in the paper-based CARAT during a medical appointment. After 3-10 days (median-M 5), CARAT data were again collected by telephone interview. Total (CARAT-T) (from 0-worst to 30-best), upper airways (CARAT-UA) and lower airways (CARAT-LA) CARAT scores were obtained. A CARAT-T score  $> 24$  indicates controlled asthma. Internal consistency (Cronbach's  $\alpha$ ), test-retest reliability (intraclass correlation coefficient-ICC<sub>2,1</sub>) and agreement (percentage of agreement and Cohen's kappa) were calculated.

**Results:** A total of 245 patients (20% adolescents; 34% male; M = 36 [interquartile range-IQR 32] years) were analysed. The CARAT-T, CARAT-UA and CARAT-LA median scores were 21 [IQR 10 vs 9], 7 [IQR 6] and 15 [IQR 5] using both methods ( $p > 0.05$ ). Cronbach's  $\alpha$  for CARAT-T, CARAT-UA and CARAT-LA was 0.85, 0.80 and 0.82, using paper-based forms and 0.80, 0.74 and 0.77, using telephone interview, respectively. Excellent test-retest reliability levels were obtained, with an ICC of 0.80 (95%CI 0.75-0.84) for CARAT-T, an ICC of 0.77 (95%CI 0.72-0.82) for CARAT-UA, and an ICC of 0.76 (95%CI 0.71-0.81) for CARAT-LA. Classification of asthma control was concordant in 208 (85%) patients, indicating good agreement between the two methods (kappa = 0.66;  $p < 0.001$ ).

**Conclusions:** Telephone-based application of CARAT is a valid and reliable method for assessing asthma control, thereby being an alternative approach to conventional paper-based forms in patients with asthma.

**Key words:** Persistent asthma. Asthma control. Patient-reported outcome measures. Telephone interview. Validation.

### PE 094. PREVALENCE OF OBSTRUCTIVE SLEEP APNEA IN ASTHMATIC PATIENTS AT A RESPIRATORY ALLERGOLOGY OUTPATIENT CLINIC

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**Introduction:** Asthma and obstructive sleep apnea (OSA) are common chronic respiratory disorders worldwide. It is estimated that asthma affects 1-18% of the population, particularly adult women, while OSA affects around 14% of men and 5% of women. Overlap of these two diseases is also common, since asthma increases the predisposition for OSA, with common risk factors.

**Objectives:** Characterize the population with asthma and OSA followed at the Respiratory Allergology outpatient clinic at a central hospital, during 2017.

**Method:** Analytical, cross-sectional, retrospective study of patients with asthma and OSA followed at this clinic. We analyzed the following variables: age, time of asthma evolution and classification, comorbidities, smoking status, pulmonary function tests (PFT), treatment and asthma control, symptoms suggestive of OSA, severity of the diagnosed OSA and therapy and consequent asthma control.

**Results:** We reviewed all the patients followed at this outpatient clinic and identified 47 patients with asthma and OSA, 53% of which were men, with an average age of  $59.62 \pm 12.77$  years; 62% had allergic asthma and 81% adult-onset asthma. Relevant comorbidities: rhinitis (49%) and arterial hypertension (47%). Only 1 patient had a normal body mass index, 60% had obesity class I and II and 9% had morbid obesity. Only 34% had smoking habits (31% current smokers). On PFT 72% of the patients had an obstructive pattern and 28% were normal. At time of OSA diagnosis, 11% of the patients had no asthma treatment, 13% were at GINA's step 1, 2% at step 2, 21% at step 3, 49% at step 4 and 4% at step 5. Asthma was controlled in 45% of the patients already followed at the clinic. Snoring and excessive daytime sleepiness were present at 72% and 66%, respectively, and

justified referral to the Sleep Disorders clinic. All patients had a home sleep apnea study or in-lab polysomnography which diagnosed OSA. OSA was classified according to apnea/hypopnea index (AHI, AASM) in mild (21.3%), moderate (38.3%) and severe (40.4%). Moderate OSA was more frequently diagnosed in women (27.7% vs 12.8%, p-value = 0.014), while severe OSA was more frequent in men (29.8% vs 10.6%, p-value = 0.020). Positive airway pressure was prescribed in 85%; 11% had indication to sleep hygienic-dietary measures and control other respiratory comorbid diseases; and 4% mandibular advancement splint. After implementing these measures, there was an asthma improvement in 35% of cases not previously controlled. **Conclusions:** Severe to moderate OSA was diagnosed in 79% of the patients. Allergic and adult-onset asthma were more prevalent and 53% of the patients were on GINA's step 4 to 5. Obesity was an important comorbidity. Treatment of OSA contributed to better asthma control in 35% of the cases. We can conclude that OSA investigation is important in patients with uncontrolled asthma, in order to achieve a better disease control.

**Key words:** Asthma. Obstructive sleep apnea.

### PE 095. SPECIFIC IMMUNOTHERAPY IN PEDIATRIC ASTHMA TREATMENT

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**Introduction:** asthma is a respiratory tract disease frequently associated with allergies and very common in pediatric age. It consists in excessive IgE production as the result of environmental allergens exposure. Allergen specific immunotherapy appears to be the only treatment capable to change the natural course of allergic disease, reason why it can have an important role in asthma. The goal of this article is to evaluate current knowledge on efficacy and safety of short and long term specific immunotherapy treatment in pediatric population asthma.

**Methods:** Research in some of the most reliable databases of practice guidelines, clinical trials and systematic reviews from the last 5 years in Portuguese and English languages using Mesh terms: "immunotherapy"; "asthma" and "child".

**Results:** 147 articles were obtained and 7 were selected: 1 practice guideline, 2 systematic reviews, 3 clinical trials and 1 retrospective study. Data support that specific immunotherapy is safe and effective for the treatment of allergic asthma in pediatric population. Systemic reactions rate is less than 5% and anaphylactic reactions are rare. Exacerbations prevention is also highlighted. Most articles show reduction in respiratory symptoms and in drug doses needed to control them. However, some data suggest there is no significant reduction in bronchodilator therapy needed to keep disease stable after specific immunotherapy treatment.

**Conclusions:** Allergen specific immunotherapy seems to be an effective alternative to reduce respiratory symptoms and asthma associated therapeutic in pediatric population. It allows an improvement in the quality of life of those patients. Its high safety profile can be a stimulus for an increased clinical use. However results show some inconsistency so more studies with different allergens are required to determine long term safety and efficacy of specific immunotherapy.

**Key words:** Asthma. Treatment. Immunotherapy.

### PE 096. OMALIZUMAB - 10 YEARS OF EXPERIENCE IN SEVERE ASTHMA

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**Introduction:** Approximately 80% of asthmatics are atopic with a one or more positive skin test to common allergens and elevated

plasma contractions of specific immunoglobulin E (IgE). Activation of Helper 2 T lymphocytes by a certain allergen leads to an activation of B lymphocytes with consequent IgE production to that allergen. IgE stimulates mast cells and basophils to release histamine, prostaglandins, and leukotrienes that cause bronchoconstriction and plasma exudation upon exposure to the allergen. Omalizumab is an IgG1 monoclonal antibody that binds with high affinity to IgE, inhibiting its effect. It is a treatment for severe asthma with elevated IgE levels, being a therapeutic weapon in the 5<sup>th</sup> step according to the *Global Initiative for Asthma*, which presents an important evolution in the treatment of severe allergic asthma.

**Objectives:** Evaluate the efficacy of omalizumab in severe asthma patients followed at the severe asthma clinic during the last 10 years.

**Methods:** We retrospectively analyzed the clinical data of all patients who underwent treatment with omalizumab from August 2008 to July 2018. A statistical analysis was performed using the IBM-SPSS v24 resource.

**Results:** Thirty-two patients with a diagnosis of asthma and allergic rhinitis were included, with a mean age of 52.6 years, the youngest being 22 years old and the oldest being 69 years old at the beginning of treatment. 68.6% were female. Regarding symptomatology, the mean value in the Control of Allergic Rhinitis and Asthma Test (CARAT) was 11.6 and the mean number of exacerbations requiring systemic corticosteroid therapy was 3.06/year before the start of Omalizumab. The most commonly used inhaled corticosteroids were fluticasone propionate and budesonide at a mean daily dose of 1,000 µg and 1,200 µg, respectively. All patients were medicated with leukotriene receptor antagonists, 81.25% received muscarinic receptor antagonists and 68.75% xanthines. At the time of analysis 15 patients were on treatment. The mean duration of treatment was 44 [± 26] months. There was a symptomatic improvement after treatment with omalizumab with statistical significance, with a significant increase in CARAT score, and a reduction in the number of exacerbations to 1/year. It was possible to reduce the dose of the inhaled corticosteroid and gain in the forced expiratory volume in the first second (FEV1) with statistical significance. There was a reduction of 28% in patients taking xanthines and 13% in patients receiving muscarinic receptor antagonists, but without statistical significance.

**Conclusions:** This retrospective analysis shows the importance of using omalizumab in reducing exacerbations, reducing exposure to systemic and inhaled corticosteroids, and improving symptom control in patients with severe asthma.

**Key words:** Asthma. Omalizumab.

### PE 097. MEPOLIZUMAB IN SEVERE ASTHMA TREATMENT - A RECENT EXPERIENCE

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**Introduction:** Severe asthma is defined as a patient who needs high dose of inhaled corticosteroids or systemic corticosteroids to maintain control of their asthma or patients who never achieve symptomatic control despite optimal treatment. Treatment of severe asthma requires a multidisciplinary approach to patient education, exposure reduction, treatment of comorbidities, and therapeutic optimization. Eosinophils are an inflammatory cell that play a key role in asthma and are responsible for remodeling the airways. The importance of interleukin 5 (IL-5) in the pathogenesis of asthma has been elucidated, being essential to the terminal differentiation, maturation and migration of eosinophils. Thus, the potential of anti-IL5 therapy in asthma is understood by inhibiting IL-5 signaling by reducing the growth, differentiation, recruitment, activation

and survival of eosinophils. Mepolizumab is a monoclonal antibody to IL-5, indicated for the treatment of severe asthmatic patients with peripheral eosinophils greater than 150  $\mu\text{L}$ , in order to reduce exacerbations in these patients.

**Objectives:** Characterization of the population with severe asthma on treatment with mepolizumab.

**Methods:** All patients with severe asthma who performed mepolizumab until June 30, 2018 were clinically evaluated. Statistical analysis was performed using IBM-SPSS v24.

**Results:** Six patients with a mean age of 55.67 [ $\pm$  10.85] years were included, with 83% being female. Before the onset of mepolizumab the patients had an average asthma control test (ACT) score of 11.5 [ $\pm$  2.8] and a mean number of asthma exacerbations per year of 3 [ $\pm$  0.63]. The number of peripheral eosinophils the mean value was 626/ $\mu\text{L}$  [ $\pm$  113]. At the time of evaluation, the mean duration of treatment was 5 [ $\pm$  3.9] months, with a maximum of 12 months and a minimum of 1 month. Only 2 patients were taking mepolizumab for more than 6 months. In these 2 patients it was possible to observe a reduction in the number of exacerbations, an increase in ACT score and an improvement in forced expiratory volume in the 1<sup>st</sup> second (FEV1) and in pulmonary insufflation in the treatment period.

**Conclusions:** In patients with severe asthma with eosinophilic phenotype the use of mepolizumab showed beneficial effects in reducing exacerbations. Although only 2 patients were receiving treatment for more than 6 months, the effect of reducing exacerbations in these patients was already evident, as was the symptomatic improvement. The future follow-up of these patients, as well as the increase in patients treated, will make it possible to assess in clinical practice the importance of this treatment for severe eosinophilic asthma and to define the optimal duration of treatment.

**Key words:** Asthma. Mepolizumab.

#### PE 098. CHARACTERIZATION OF A POPULATION WITH ASTHMA IN RELATION TO THE PRESENCE OF OBESITY

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**Introduction:** The prevalence of obesity has been increasing in the general population in and in particular in the population with asthma. Both pathologies have an underlying a chronic inflammatory state that seems to exacerbate each other when present in the same individual. The authors characterize a population of patients with asthma regarding the prevalence of obesity; comparing the subgroup of obese with the remaining population regarding symptom control, rate of exacerbations and prevalence of severe asthma.

**Methods:** This retrospective study included 402 consecutive patients. The population in question was characterized as follows: age, gender, body mass index, symptom control assessed by the Asthma Control Test (ACT), rate of exacerbations in the 12 months preceding inclusion in the study and prevalence of severe asthma defined according to the joint consensus of the European Respiratory Society and American Thoracic Society published in 2014. The subgroup of patients with obesity (defined as BMI  $\geq$  30 kg/m<sup>2</sup>) was compared with the rest of the population for the previously named variables. The comparison between groups was done through the student t test for variables with normal distribution or through the Mann-Whitney test for non-parametric variables. Statistically significant difference was considered if  $p < 0.05$ .

**Results:** A total of 402 patients with asthma were included with a predominance of females (71.6%) and mean age of 48  $\pm$  17 years. The prevalence of obesity in this population was 40.5%. Patients with asthma and obesity had the following statistically significant

differences: higher mean age (55  $\pm$  14 vs 44  $\pm$  17  $p = 0.001$ ); greater rate of exacerbations in the last 12 months (3  $\pm$  0.5 vs 1  $\pm$  0.3  $p = 0.04$ ); worst symptom control (13  $\pm$  4 vs 21  $\pm$  3  $p = 0.003$ ); higher prevalence of severe asthma (8.78% vs 2%  $p = 0.005$ ).

**Conclusions:** Obesity in patients with asthma was associated with poorer symptom control, a higher rate of exacerbations and a higher prevalence of severe asthma. Considering these findings, interventions that seek weight reduction in this population may have a significant impact on the reduction of symptoms and rate of exacerbations.

**Key words:** Asthma. Obesity.

#### PE 099. PULMONARY ASPERGILLOSIS OR LESS OBVIOUS DIAGNOSIS

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**Introduction:** Aspergillosis is an infection of the airways, lungs, cutaneous or extrapulmonary spread caused by *Aspergillus* species, most commonly the species of *A. fumigatus*, *A. flavus* and *A. terreus*. *Aspergillus* species are present in nature and the inhalation of infectious conidia is frequent, however the infection occurring mostly in cases of immunosuppression. Risk factors include: severe and prolonged neutropenia, treatment with high doses of glucocorticoids or other medications or conditions that lead to chronically compromised cellular immune responses.

**Case report:** Patient with history of anti-MBG disease (diagnosis performed in the previous month), dependent on dialysis and under a weaning regimen of corticosteroid therapy (20 mg/day) resorted to emergency due to productive cough with pink expiration and dyspnea. From the study carried out in the SU, respiratory failure type 1 and 2 g/dL drop in hemoglobin were associated with increased PCR, leukocytosis with predominance of neutrophils and chest CT images suggestive of alveolar hemorrhage vs infection: "(...) of pericardial effusion. A slight volume of bilateral pleural effusion. In the pulmonary parenchyma, multiple bilateral parenchymal infiltrates of peribroncovascular predominance, areas of bronchoalveolar consolidation and associated depolarized glass sketch, in the upper lobes, middle lobe and lower lobes, sparing the pulmonary vertices and with lower expression in the bases, concomitant air bronchogram. Nodular/micronodular opacities of peribroncovascular distribution. In the context of the antecedents of GoodPasture syndrome and clinical pink expectoration, the hypothesis of alveolar hemorrhage cannot be excluded. Inpatient with diagnosis of right pneumonia and alveolar hemorrhage. Initiated pulses of methylprednisolone, antibiotic therapy with meropenem and realization of plasmaferesis. In D3 of hospitalization altered the corticotherapy for prednisolone 1 mg/kg in progressive weaning. He did not start treatment with cyclophosphamide given the infection picture. In D4 of hospitalization due to persistence of cough, respiratory failure type 1, aggravation of analytical inflammatory parameters and lack of improvement in pulmonary radiographs, despite the measures instituted, bronchofibroscopy revealed: "AB mucosa diffusely swollen with small amount of purulent secretions and scattered hematocrit. "Bronchial aspirate and bronchoalveolar lavage revealed isolation of *Aspergillus fumigatus* and performed TACAR, which revealed: "improvement of parenchymal findings, slight bilateral pleural effusion." Established treatment with Voriconazole and consequent gradual improvement of respiratory insufficiency, hemoptotic expiration and inflammatory analytical parameters. The patient was discharged, externally oriented and medicated with Voriconazole and steroid corticosteroid.

**Discussion:** This clinical case aims to alert to the importance of broad clinical suspicion. In this patient, the most likely cause for hemoptysis would be the alveolar hemorrhage underlying the anti-

MBG disease. However, clinical research has identified aspergillosis as the main and obvious diagnosis, given that it is an immunodepressed patient. The classic triad of immunosuppressed presentation is fever, pleuritic pain, and hemoptysis. The absence of this triad should not rule out the diagnosis before a patient with risk factors for disease. In such patients, the pulmonary image often reveals lung and/or infiltrate nodules.

**Key words:** Hemoptysis. Immunosuppression. Anti-MBG disease. Pulmonary Aspergillosis.

#### PE 100. PNEUMOCYSTOSIS IN INITIAL DIAGNOSIS OF HIV INFECTION - CASE REPORT

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**Introduction:** *Pneumocystis jirovecii* pneumonia is the most common opportunistic infection in patients infected with human immunodeficiency virus (HIV), being considered one of the most common AIDS-defining diseases in Portugal. In HIV-positive patients, pneumocystosis generally occurs when TCD4 + cell counts < 200 cell/mm<sup>3</sup> and especially when < 100 cell/mm<sup>3</sup>. The presumptive diagnosis is based on the presence of respiratory symptoms, hypoxemia, elevated lactic dehydrogenase (LDH) and radiographic changes. Definitive diagnosis implies the detection of *Pneumocystis* in samples of respiratory secretions or bronchoalveolar lavage.

**Case report:** We present the case of a 42-year-old woman, Caucasian, active smoker, airport worker, divorced and with a fixed partner for eight years, apparently healthy. Known history of controlled bronchial asthma with inhaled therapy in SOS and psoriasis. History of hospitalization in February 2018 in the Dermatology service for toxidermia to amoxicillin/clavulanic acid, having been medicated with steroid therapy. Chest X-ray was normal. One month later, she went to the emergency service with fatigue for minor efforts, thoracalgia, dyspnea and cough with mucous sputum, with evolution of 3 weeks, without fever or other symptoms. For presumed respiratory infection, she was treated with levofloxacin without improvement, so she returned to the emergency service now with arthralgia, myalgia and weight loss of 5 kg in 1 month. At the clinical examination she was polypneic at rest, peripheral saturation pO<sub>2</sub> 95% with oxygen at 1 L/min, pulmonary auscultation with rude vesicular murmur, bilateral crackling fervor. No palpable adenopathies or cutaneous lesions. At ambient air she presented respiratory alkalemia, hypocapnia and hypoxemia (PaO<sub>2</sub> 68 mmHg). Blood testes without leukocytosis or neutrophilia, relative lymphopenia 2.5%, LDH 263 U/L, CRP 0.400 mg/dl, SV 54 mm/h, D-dimers 0.17. Chest X-ray with bilateral homogenous consolidation and bilateral hilar reinforcement. Angio-CT thorax without signs of pulmonary thromboembolism, with bilatateral diffuse densification in depolished glass pattern, in central/peri-hilar topography. Detected HIV1 positive serology of February of 2018, that the patient didn't know, confirmed by Western Blot. TCD4 + lymphocyte count 33.9 cel/mm<sup>3</sup>, that together with imaging and hypoxemia, made us put the hypothesis of pneumocystosis and treatment with cotrimoxazole and prednisolone was initiated. Subsequently, the patient reported knowing HIV1 infection of the current partner, diagnosed and treated for 6 years, so that since the diagnosis they had protected sexual relations, but she didn't know she was infected. Investigation included high viral load of HIV-1-RNA 204,387, HBV, HCV, CMV, Adenovirus, Coxsakie virus, Herpes virus, Epstein Barr, Cryptococcus, Toxoplasmosis, syphilis and autoimmunity study, all negative. Negative hemocultures and uroculture. Bronchofibroscopy for bronchoalveolar lavage with negative bacteriological and mycobacteriological tests, with PCR positive for *Pneumocystis jirovecii*, confirming the diagnosis. After 8 days of therapy she presented clinical and imagiolog-

ic improvement and had hospital discharge referred to the immunodeficiency consultation, maintaining treatment.

**Discussion:** This case alerts us to the need for a high level of suspicion of *Pneumocystis jirovecii* infection, even in patients with unknown immunosuppression, remembering that both objective examination and chest X-ray may be initially normal. In these cases empirical treatment should be initiated, even before the definitive diagnosis.

**Key words:** HIV Infection. *Pneumocystis jirovecii*. *Pneumocystis pneumonia*.

#### PE 101. REVIEW OF HOSPITALIZATIONS DUE TO PNEUMONIA IN A PRIVATE HOSPITAL

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**Introduction:** Pneumonias are among the leading causes of hospital admission, but less than 1/3 of patients with pneumonia require hospitalization. The population of patients hospitalized with this diagnosis has particular clinical features. The knowledge of these characteristics, their management during hospitalization and the results obtained allow to adjust attitudes and procedures to improve the prognosis of the patients.

**Objectives:** To analyze the sociodemographic and clinical characteristics, antibiotic prescription patterns, microbial yield of different diagnostic methods and isolated microorganisms in the samples obtained in hospitalized patients with pneumonia.

**Methods:** Retrospective study with data collected from hospital records. Adult patients hospitalized in an Internal Medicine ward with a diagnosis of pneumonia were selected in a 12-month period -from June 2017 to May 2018- and the demographic characterization, study of comorbidities, antibiotic therapy, yield of microbiological studies and treatment outcome were recorded.

**Results:** Eighty-four patients (35 women and 49 men) were included, with a mean age of 78 years, with a total of about 42% being 85 years or older. The age group of 81-90 years included more patients (33% of patients) than the others. The length of hospital stay was, on average, 8 days. An average of 7 patients/month was hospitalized with pneumonia, with a higher incidence in the winter months (December to February). The mean comorbidities per patient were 3, with cardiovascular diseases being the most frequent; structural lung diseases affected about 36% of the total. Approximately 25% of the sample consisted of former smokers. The most commonly prescribed antibiotics were beta-lactams (amoxicillin-clavulanate - 36% - and ceftriaxone - 24%), followed by levofloxacin (23%). In 12% of patients piperacillin-tazobactam was the initial antibiotic. The association with a macrolide occurred in 50% of the cases. The change in initial therapy occurred in 11% of patients, and in 2/3 was due to failure of previous treatment. It was considered that there was no indication for the prescription of antibiotics in 5% of the cases. Blood cultures were collected in 82% of the cases, with a yield of 6.8%. *Streptococcus pneumoniae* urinary antigen testing was performed in 63% of patients, with a yield of 17%. The microbiological study of bronchial secretions was performed in a quarter of the cases, with a profitability of about 33%. Most patients at discharge (55%) were referred for consultation and the mortality rate was 7%.

**Conclusions:** Patients hospitalized for pneumonia are mostly old and very old, with a high number of comorbidities. A high percentage of patients were treated with piperacillin-tazobactam or levofloxacin in the initial regimen because of etiology modifying factors. It is also worth noting the low yield of blood cultures in the sample considered.

**Key words:** *Pneumonia*. *Antibiotics*. *Microbiological study*.

### PE 102. FACTORS ASSOCIATED WITH MORTALITY IN PNEUMOCOCCAL PNEUMONIA - ONE YEAR RETROSPECTIVE ANALYSIS IN A HOSPITAL CENTER

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**Introduction:** Pneumococcal pneumonia is an important cause of morbi-mortality in the Portuguese population, particularly in elderly individuals with certain comorbidities. Although vaccination is considered an effective method to prevent severe forms of disease, studies show that vaccination coverage in this population remains low.

**Objectives:** Identification of factors associated with a higher risk of death in patients diagnosed with community-acquired pneumonia of *Streptococcus pneumoniae* at a Central Hospital Center during the year 2017.

**Methods:** Retrospective study of patients from a Central Hospital Center diagnosed with *Streptococcus pneumoniae* pneumonia during the year 2017. Data collection through the analysis of medical records. Serotypes were obtained by request to the laboratory responsible for their identification. The patients were divided in two groups: survivors and deceased during hospitalization. Statistical analysis to identify possible factors associated with increased risk of death using SPSS Statistics v25<sup>®</sup> and Microsoft Excel 2013<sup>®</sup>.

**Results:** Total sample of 54 patients with mean age of 72 years and 77.8% were males. Sample mortality rate was 16.7%. Deceased patients presented more frequently alteration in mental state (56.6% vs 20%,  $p < 0.05$ ), had higher mean serum urea and total bilirubin levels (84.1 vs 53.7 mg/dL and 1.6 vs 0.9 mg/dL,  $p < 0.05$ ) and lower mean hematocrit values (32.1 vs 37.4%,  $p < 0.05$ ). There was also an increase on admission of this patients in intensive care unit (56% vs 11.1%,  $p < 0.05$ ) and higher mean SAPS-II score (56 vs 40,  $p < 0.05$ ). There were no statistically significant differences between survivors and deceased in relation to age, sex, comorbidities, smoking habits and mean score of APACHE II.

**Conclusions:** The fact that the sample showed no impact of age and comorbidities in mortality may be due to the small sample size. Nevertheless, it was possible to verify the impact on mortality of biochemical factors classically associated with greater severity such as the serum level of urea and total bilirubin.

**Key words:** *Pneumococcal pneumonia. Risk factors. Streptococcus pneumoniae.*

### PE 103. RARE ETIOLOGY OF PLEURAL EFFUSION

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**Introduction:** Multiple myeloma (MM) accounts 1 to 2% of all cancers and 17% of hematologic malignancies. The incidence is 2 to 3 times higher in Blacks, it is more frequent in men and the median age at the diagnosis is 66 years. Extramedullary plasmacytoma is a plasma cell tumor that rises outside the bone marrow, occurring in 7% of MM patients.

**Case report:** The authors present a case of a 61-year-old woman from Cape Verde, without relevant antecedents except the inhalation of nasal snuff in the last 20 years. Patient came to Emergency Service presenting lower right back pain with two-months of evolution, fatigue with one-month of duration and decreased lower limb strength and urinary incontinence since the week prior to admission. On clinical examination, the vesicular murmur was abolished in the lower 2/3 of the right hemithorax and the chest X-ray presented homogeneous hypotransparency suggestive of right pleural effusion, she was hospitalized. After thoracentesis pleural fluid was compatible with an exudate, with more than 1,000 leukocytes (53% lymphocytes), ADA

72.8 U/L, negative cytology for neoplastic cells. Analytically with normochromic normocytic anemia, slightly increase in inflammatory parameters, increased sedimentation rate (119 mm/1h), renal insufficiency (creatinine 1.4 mg/dl), hypercalcaemia (10.40 mg/dl), normal protein electrophoresis and increased in  $\beta_2$ -microglobulin (6.97 mg/L). Chest computed tomography (CT) scan: "solid formation with 9.4 x 6.6 cm (...) in the upper lobe of the right lung, with invasion of the mediastinum, right main bronchus, destruction of the 5<sup>th</sup> posterior costal arch, transverse apophysis and D5 vertebral body, with involvement of muscles of the posterior wall of the dorsal region adjacent to the 5<sup>th</sup> costal arch (...) pathological adenopathies in mediastinum, internal mammary and retrocrural chain (...) ". After developing paraplegia in the hospital, the patient underwent a spine CT scan which added: "(...) secondary osteolytic lesions (...) in T4/T5 (...), T10/T11 (...), lytic image of left iliac (...) reduction of the permeability of L5/S1 (...) and L4/L5 conjugation channels, with radicular conflict". Magnetic resonance imaging confirmed the thoracopulmonary cancer, adding "(...) between T10-T12 right paravertebral tumor mass". It was discussed with Oncology, Radiotherapy and Neurosurgery, without surgery indication or conditions for urgent radiotherapy, which could limit histological diagnosis and cancer-directed treatment. Anemia, renal failure and hypoalbuminemia worsened. 24-hour urine presented with nephrotic proteinuria. Urine immunofixation of proteins evidenced Bence-Jones protein positive for lambda light chain and the serum immunofixation of the proteins evidenced a monoclonal band corresponding to lambda light chains, confirming the diagnosis of lambda light chains MM. The myelogram showed an infiltrated bone marrow of cancer cells of plasmacytic lineage (61%), 52% plasmocytes. The patient started the CyBORd protocol. Transthoracic needle aspiration, initially suggestive of lymphoma/carcinoma, confirmed later the diagnosis of pulmonary and vertebral plasmacytoma.

**Discussion:** The presentation of this clinical case evidenced the difficulty to diagnosis a rare disease that emerges at a very advanced stage of its clinical course.

**Key words:** *Pleural effusion. Multiple myeloma. Plasmacytoma.*

### PE 104. INTRAPLEURAL FIBRINOLYSIS - A CASE OF WELL-ESTABLISHED SUCCESS WITH A NOT-SO-WELL-ESTABLISHED APPROACH

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**Case report:** The case presented is that of a 52-year-old woman who went to the Emergency Department for pleuritic chest pain with a week of evolution. There was abolition of the vesicular murmur in the lower 2/3 of the left hemithorax and increased inflammatory parameters. Chest CT showed extensive multiloculated pleural effusion, and thoracentesis was performed. The liquid had characteristics of an exudate and were isolated *Prevotella intermedia* and *Fusobacterium nucleatum*, for which it was initiated directed antibiotic therapy. Because of the persistence of a radiological image of pleural effusion, ultrasound-guided thoracic drainage was placed. However, drainage has proved to be ineffective even under active aspiration. In order to try to avoid surgical intervention, it was decided to initiate intrapleural fibrinolysis with alteplase 10 mg 12/12h and dornase alfa 5 mg 12/12h (4 administrations in total). With fibrinolytic therapy there was progressive clinical and radiological improvement. Pleural infection is an important cause of morbidity and mortality and its incidence is increasing. Its treatment involves drainage and antibiotic therapy but in 1/3 of the patients drainage is ineffective due to the formation of loculations, requiring surgery to perform decortication. Fibrinolysis has been used for more than 50 years in empyema/pleural infections as an alternative to surgery but there is no consensus or established guidelines about its use.

**Discussion:** The present case, which had a favorable outcome, corroborates other similar ones in the literature, showing that this therapeutic option can be beneficial in situations of loculated effusions, minimizing the need for surgery, with the inherent risks and costs. Further studies are needed to establish the most suitable indications, dosage and regimen of intrapleural fibrinolytic therapy.

**Key words:** *Empyema. Fibrinolysis. Alteplase. Dornase.*

### PE 105. YELLOW NAIL SYNDROME - A RARE CAUSE OF PLEURAL EFFUSION

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**Introduction:** The yellow nail syndrome was first described in 1964. It is a rare and probably underdiagnosed syndrome. It usually presents between 40-60 years old and it's characterized by: lymphedema, dystrophic yellow nails and chronic respiratory symptoms (respiratory infections, pleural effusion, chronic cough). Only two of the characteristics need to be present in order to establish the diagnosis. The cause is still uncertain, but it has been described in association with systemic diseases, cancer and immunodeficiency.

**Case report:** 58 year old woman, active smoker of 60 pack unit years, with a clinical history of obesity (BMI 34.6), lymphedema of the lower extremities, chronic cough and sinusitis. She went to the emergency department complaining of dyspnoea and worsening of the oedema of the legs. She referred several respiratory infections in the past 8 years and yellow nails. Physical examination showed an oximetry of 90% in room air, diminished vesicular murmur in the lower half of the right hemithorax, symmetric oedema of the lower extremities and dystrophic yellow nails. The lung radiogram evidenced a opacity of the lower half of the right hemithorax, suggesting pleural effusion. A thoracocentesis was performed. The results were compatible with and exudate with lymphocytic predominance. The cytology was negative for cancer cells. The cultural exams of the pleural liquid were negative. A pleural biopsy was performed, and showed unspecific chronic pleuritis. The rest of the investigations were negative - with a normal protein electrophoresis, negative auto antibodies (antinuclear antibodies, rheumatoid factor, antineutrophil cytoplasmic antibodies), and normal complement. Thus, and having excluded other causes, the diagnosis of yellow nail syndrome was established. The therapeutic approach is symptomatic, nonetheless, the patient needed to perform multiple pleural drainages, for dyspnoea relief. We ended up repeating the pleural biopsies - which came out negative. To ultimately control the symptoms she had performed a pleurodesis.

**Discussion:** The authors present this case because it reflects a rare, underdiagnosed syndrome that may present with unspecific respiratory symptoms. It is an uncommon cause of pleural effusion that, when present, almost always is an exudate with lymphocytic predominance. Pathogeny is nuclear, with recent studies suggesting an important role of impaired lymphatic drainage with protein leak. Long-term prognosis is favorable, depending mainly on the severity of the respiratory manifestations and the patient's overall clinical condition.

**Key words:** *Pleural effusion. Yellow nails.*

### PE 106. PERSISTENT PLEURAL EFFUSION - MANIFESTATION OF RHEUMATOID ARTHRITIS

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**Introduction:** Rheumatoid arthritis (RA) is a systemic inflammatory disorder with autoimmune origin, which primarily affects joints,

causing a symmetrical and progressive destruction of cartilage and bone. There are extra-articular manifestations that contribute significantly to the morbidity and mortality of the disease. Pleural involvement is common and it usually manifests as pleural effusion. This occurs most often in men who have active arthritis and subcutaneous nodules. The majority of pleural effusions are of small to moderate volume, unilateral and asymptomatic. The pleural fluid is typically a sterile exudate, with low pH and glucose plus elevated lactate dehydrogenase levels. Most effusions resolve spontaneously, with no need for specific therapy.

**Case report:** A 57-years-old man with personal history of pulmonary thromboembolism due to genetic thrombophilia, hereditary spherocytosis (splenectomized), IgA nephropathy, rheumatoid arthritis (under immunosuppression), arterial hypertension and dyslipidemia. The patient was sent to the HUC's emergency department, transferred from the residence's area hospital with dyspnea, post-inferior left thoracalgia with pleuritic characteristics and asthenia, with one-week evolution and progressive worsening. He had been treated for 3 days with levofloxacin for community-acquired pneumonia, but remained subfebrile and without clinical improvement. He reported recurrent respiratory infections in the last 6 months. From the study performed at the hospital of origin, there was an increase of the inflammatory parameters and a persistent opacity at the base of the left hemithorax in the chest X-ray. The antibiotic therapy was adjusted to piperacillin-tazobactam and the patient was hospitalized for an etiological study. From the analytical study, we highlight the positivity of the anti-CCP autoantibodies and the increase of the rheumatoid factor (RF) in the serum, and high IgA immunoglobulin, in agreement with personal background already known. Cultures of sputum, urine and blood were negative, as was the urine antigen screening. Arterial blood gas was normal. The thoracic CT showed left pleural effusion in apparent organization and partial collapse of the inferior lobe and lingula, which was probably compressive. He underwent videobronchoscopy with bronchial aspirate, washing and brushing, that did not reveal alterations. In spite of the initial improvement after a complete cycle of antibiotic therapy, there was a new rise of the inflammatory parameters and an increase of the pleural effusion's volume. Subsequently, a linezolid and meropenem treatment was started, with good clinical and analytical response, despite maintaining a moderate volume of the pleural effusion. The patient was submitted to diagnostic and evacuator thoracocentesis. The fluid's study revealed an exudate with no cytology specificity, negative microbiology and high rheumatoid factor (88 IU/mL), which led to a diagnosis of pleural effusion secondary to RA. After thoracocentesis, there was no recurrence of pleural effusion.

**Discussion:** A persistent pleural effusion in a patient with history of RA should not be immediately assumed as a secondary condition. It is essential to rule out more frequent and serious causes, as infections or malignancy. Once the causality is admitted, the differential diagnosis with empyema is still difficult, since it could be a complication of rheumatoid effusion.

**Key words:** *Pleural effusion. Rheumatoid arthritis.*

### PE 107. PNEUMOTHORAX: ONE YEAR EXPERIENCE AT HOSPITAL SANTA MARIA

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**Introduction:** Pneumothorax, defined as the presence of air in the pleural space, has a varied aetiology. It can be categorized as primary spontaneous (PSP), secondary spontaneous (SSP) or traumatic pneumothorax (TP). PSP has no predisposing lung disease and has an incidence of 7.4-18 cases/100,000 per year in men and 1.2-6

cases/100,000 per year in women. SSP results from an underlying lung disease with an incidence of 6.3 cases/100,000 per year in men and 2.0 cases/100,000 per year in women. Because of its incidence and the importance of early diagnosis and intervention, the authors present the experience of the hospital in the year 2016.

**Methods:** A retrospective analysis of clinical processes of hospitalized patients with a diagnosis of pneumothorax was conducted about the year 2016 at the Respiratory Service of Centro Hospitalar Lisboa Norte - Hospital Santa Maria.

**Results:** During 2016, there were 504 hospitalizations in the Respiratory Service, 55 (10.9%) of which due to pneumothorax, corresponding to 50 patients. The majority were men ( $n = 36$ ; 72.0%), with a median age of  $49.6 \pm 22.7$  years. 64.0% of the patients were smokers/ex-smokers with a mean pack year of smoking of  $25.0 \pm 19.0$ . 14.0% of patients had predisposing occupations/hobbies (due to heavy load), 12.0% consumed inhaled cannabinoids/opioids and 2.0% used a wind instrument. Pleuritic chest pain was the most frequent clinical presentation ( $n = 38$ ; 76.0%), followed by breathlessness ( $n = 27$ ; 54.0%). PSP occurred in 48.0% ( $n = 24$ ) of the patients, SSP in 28.0% ( $n = 14$ ) and PT in 24.0% ( $n = 12$ ). 37.5% ( $n = 9$ ) of PSP had blebs or subpleural bullae identified in chest CT. The causes of SSP were as follows: COPD ( $n = 7$ ; 50.0%), bullous emphysema ( $n = 2$ ; 14.3%), lymphangioleiomyomatosis, asthma, suberosis, radiation pneumonitis and lung metastases due to esophageal neoplasm, each being 7.1%. TP were due to Implantofix® or central vein cannulation ( $n = 5$ , 41.7%), after pacemaker insertion, after thoracotomy, after transthoracic biopsy procedure, barotrauma in an invasive ventilated patient, after a fall resulting in rib fractures, in a polytrauma patient and in a patient who suffered from a gunshot, each being 8.3%. Tension pneumothorax occurred in 18 cases (36.0%). Subcutaneous emphysema was present in 26.0% ( $n = 13$ ) of the patients and there were 2 (4.0%) deaths, due to underlying pathology, in a SSP patient and in a TP patient. There was a total recurrence rate of 24%. As for their resolution, 76.0% ( $n = 38$ ) of the patients needed thoracic drainage, 10.0% ( $n = 5$ ) underwent surgical treatment (either videothoracoscopy or thoracotomy with removal of emphysema bullous) and 10.0% ( $n = 5$ ) needed pleurodesis by thoracoscopy or by talc slurry instillation.

**Conclusions:** In the studied population and in accordance to literature, the most frequent risk factors for pneumothorax were male gender, smoking history and/or airway obstructive pathology. Chest CT is important in distinguishing PSP and SSP. The risk of recurrent pneumothorax, mainly in SSP, requires an early therapeutic intervention, either with thoracoscopy and/or surgery.

**Key words:** *Pneumothorax.*

## PE 108. PLEURAL EFFUSIONS AND THE CLUES THEY GIVE US

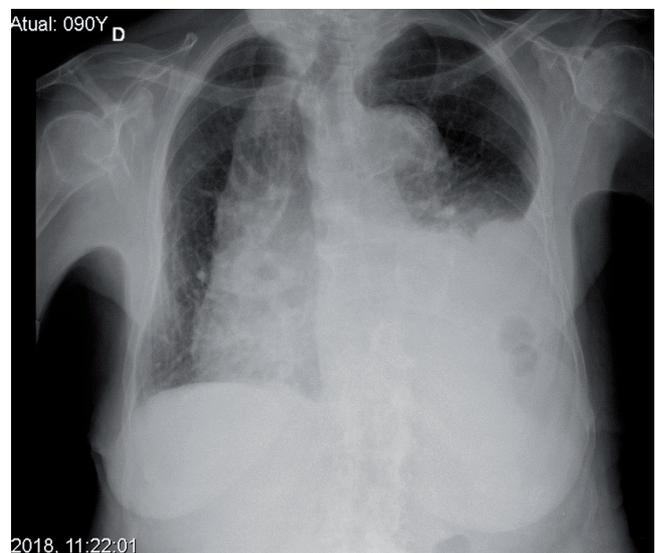
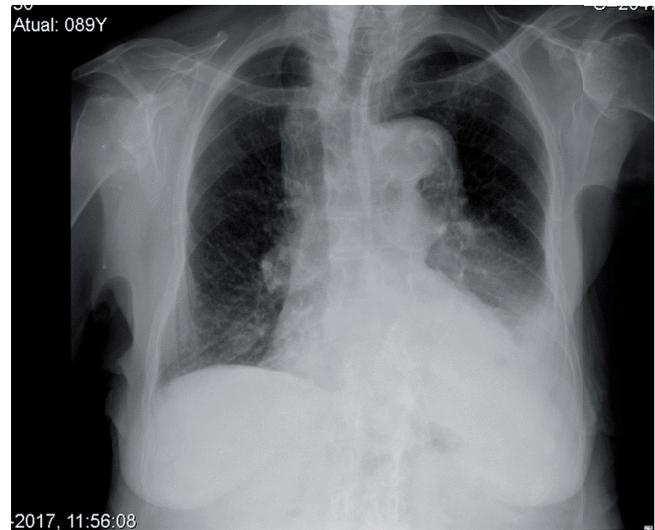
C. Sousa, J. Barbosa, M.L. Carvalho, C. Gomes, C. Bárbara

*Centro Hospitalar Lisboa Norte.*

**Introduction:** Pleural effusion is a common finding in the daily activity of a Pneumologist. Even when it's possible to rule out the more frequent causes, the characteristics of the liquid still remain helpful to guide the differential diagnosis and are determinant to achieve a final etiology.

**Case report:** This case is about a 90 year old female patient, still autonomous on her daily activities, with a history of congestive heart failure, monoclonal gammopathy, chronic kidney disease, and occluded left pleural effusion that has been in study since 2016. In May 2018 the patient was admitted to the hospital due to an upper respiratory infection associated with global respiratory failure. She was submitted to non-invasive ventilation, antibiotics and her heart failure medication was optimized. When compared to previous images, thorax X-ray showed increased volume of pleural effusion. A

thoracic CT scan was done and besides the pleural effusion it also showed a new consolidation with 1 cm. She was submitted to thoracentesis of 40 mL of pleural liquid, with normal pH, exudate, ADA of 40U/L and 69% of lymphocytes with no neoplastic cells. She was also submitted to a bronchofibroscopy, but no biopsies were performed due to spontaneous hemorrhage. Microbiologic and mycological exams, as well as mycobacterium search in sputum and in bronchoalveolar lavage were negative. Cytological study of the lavage was also negative for neoplastic cells. As the patient had displayed monoclonal gammopathy and some adenopathies in an abdominal computerized tomography that she did in 2017, and because of the finding of lymphocytes in the pleural liquid, we asked for an immunophenotyping of the blood cells and we found she had chronic lymphocytic leukemia of B cells (LLC-B).



**Discussion:** LLC is the most frequent leukemia and is a rare cause of pleural effusion. Only 4 to 9% of the patients with LLC-B have this pulmonary presentation. In this case, the suspicion of the diagnosis due to the comorbidities of the patient and the characteristics of the pleural liquid, allowed us to find an etiology for the pleural effusion, showing the importance of the analyses of the liquid on the achievement of a rare diagnosis.

**Key words:** *Pleural effusion. Lymphocytes. Chronic lymphocytic leukemia of B cells.*

### PE 109. UNUSUAL COMPLICATION OF RHEUMATOID ARTHRITIS

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**Case report:** A 51-year-old man was studied for a right pleural effusion found on a routine chest radiography. He was a current smoker (44 pack years) and had had pulmonary tuberculosis at the age of 36, treated with first line agents for one year. There was no other significant past medical or family history and he was not taking any medication. In anamnesis, he revealed pain and swelling of multiple joints. On physical examination, we found clinical features of small right-sided pleural effusion and subcutaneous nodules on his left elbow and wrist. Chest computed tomography (CT) showed a right pleural effusion with pleural thickening, ipsilateral small pneumothorax and a pleural nodule. On the left, changes were consistent with his past of tuberculosis. There was no evidence of infection, nodules, masses, emphysema, pulmonary cysts or mediastinal lymph nodes. Blood tests revealed mild anaemia and mild leucocytosis; erythrocyte sedimentation rate and C-reactive protein were slightly elevated. Rheumatoid factor and anti-cyclic citrullinated peptide antibody were positive. By thoracentesis we obtained a milky fluid with low pH. It was an exudate with high lactate dehydrogenase, low glucose, high adenosine deaminase and cholesterol. Cholesterol crystals were observed. Gram and Ziehl-Neelsen stains were negative. Pleural fluid cytology was innocent and blind pleural biopsies revealed fibrosis and inflammatory infiltrate without granulomas or neoplastic tissue. First of all, empyema was suspected so the patient was treated with chest tube drainage and intravenous antibiotics. However after knowing pleural effusion

features we conclude it was a pseudochylothorax. He was discharged home with no signs of pleural effusion or pneumothorax. According to 2010 American College of Rheumatology/European League Against Rheumatism classification criteria, the diagnosis of rheumatoid arthritis (RA) was established. Additionally, subcutaneous nodules were proved to be RA nodules after surgical excision. Immunosuppressive treatment was initiated by rheumatology team with improvement of articular symptoms. Four years after the diagnosis, a small right pneumothorax recurred and new pleural nodules were identified on chest CT. Since the patient was asymptomatic we decided not to drain the pneumothorax, only keeping clinical and radiological follow-up.

**Discussion:** This case illustrates a very unusual complication of rheumatoid pleural disease suggesting that when pleural effusion is detected and has pseudochylothorax features, RA should be considered, particularly if joint complaints. We consider the association of RA and pseudochylothorax/pneumothorax did not look like a coincidence, since pleural disease is common in RA. We excluded other causes of pseudochylothorax and pneumothorax. Despite little is known why pseudochylothorax occurs in RA, there are some reported cases in the absence of significant pleural thickening, as we found in our case. Also, we did not find a better explanation for pneumothorax, besides the rupture of rheumatoid pleural nodules.

**Key words:** *Pseudochylothorax. Rheumatoid arthritis. Pleural disease.*

### PE 110. 6 MINUTE WALK TEST - THE NECESSARY STANDARDIZATION?

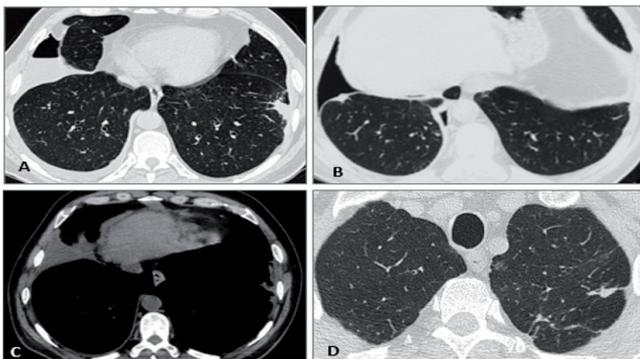
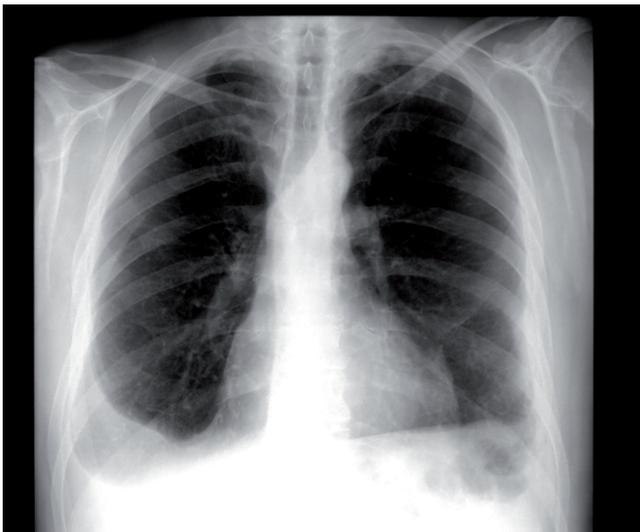
R. Boaventura, L. Almeida, E. Eusébio, P. Amorim, A. Pimentel, J.C. Pipa, A. Neves, P. Teles, M. Jacob, F. Machado, M. van Zeller, M. Drummond

*Centro Hospitalar de São João.*

**Introduction:** The 6-minute walk test (6MWT) is a useful exam in assessing the exercise capacity of respiratory patients, especially in the assessment of low-intensity exercise capacity. The examination should always be carried out under the same conditions as the results are sensitive to the execution method. The authors aim to test a new method of execution of 6MWT that eliminates some of the variability between tests.

**Methods:** A convenience based sample of patients scheduled for 6MWT was selected. All patients underwent traditional 6MWT and 6MWT with headphones, with a 30-minute recovery interval and randomized alternation. The method with headphones consisted of 6MWT whose explanation and encouragement during the examination was provided by means of an MP3 recording, which patients listened to with headphones. In the end, the patients completed a 7-item satisfaction questionnaire that compared the two methods. The study included 43 subjects with 58% (n = 25) males, mean age 60 years [Standard Deviation (SD): 13]. No statistically significant differences were found in terms of final distance traveled and variation of peripheral oxygen saturation (SpO<sub>2</sub>) during the test.

**Results:** In the satisfaction assessment, patients significantly considered the method with headphones more motivating (p = 0.02) and that, if they repeated the exam, they would opt for the headphones method (p = 0.03). It was also noted a preference towards the headphones method with regard to the questions: which method was preferred (n = 19 with headphones and n = 13 both methods); which method was heard better (n = 21 with headphones and n = 11 both methods); which method was better understood (n = 21 with headphones and n = 12 both methods); which method was more comfortable (n = 21 with headphones and n = 9 both methods) and which method was considered more reliable (n = 17 with headphones and n = 9 both methods).



**Conclusions:** The authors conclude that the new 6MWT method was favorably preferred by the patients and demonstrated non-inferiority of diagnostic efficacy in the population tested.

**Key words:** 6 minute walk test.

#### PE 111. PERSISTENT HYPOXEMIA - A CASE THAT REQUIRED AN EVEN MORE PERSISTENT DIAGNOSTIC APPROACH

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**Case report:** The case presented is that of a 77-year-old woman, retired as a cook, without smoking habits, diagnosed with hypertension, degenerative osteoarticular disease, peripheral arterial disease, hypothyroidism and obesity. During the last year she had begun a follow-up in Rheumatology Consultation to investigate a progressive aggravation of arthralgia of the great joints. In November of 2017 she went to the Emergency Department due to mental confusion during the last week. Cranial CT and CSF analysis showed no significant changes. Arterial blood gas analysis in ambient air revealed partial respiratory failure (pO<sub>2</sub> 56.7 mmHg). During hospitalization, she presented several episodes of desaturation, difficult to correct with supplementary O<sub>2</sub>, and NIV was instituted. Thorax angiography and V/Q scintigraphy excluded pulmonary thromboembolism. The transesophageal echocardiogram showed no intracardiac shunt. The pulmonary function tests showed a slight obstruction of the small airways. The polysomnographic sleep study revealed an apnea-hypopnea index 9.6/h, whereby the bi-level was replaced by auto-CPAP. In addition to the respiratory failure, the patient developed a bilateral foot drop, so she underwent electro-

myography, which revealed marked axonal sensorimotor polyneuropathy. The case was discussed with the Rheumatology team, who put the hypothesis of vasculitis associated with ANCA PR3 vs paraneoplastic vasculitis. The brain MRI excluded CNS vasculitis and the thoraco-abdomino-pelvic CT did not show signs of neoplasia, so a 5-day course of immunoglobulin EV + single dose of cyclophosphamide 1g was instituted and the patient was discharged. One month later, she went to the Emergency Department because of generalized myalgias. She had hypoxemia again (pO<sub>2</sub> 45.5 mmHg with O<sub>2</sub> at 2L/min), fever and increased inflammatory parameters, so she underwent piperacillin + tazobactam course, and MSSA was isolated in blood cultures. For etiologic investigation of partial respiratory failure, she repeated transesophageal echocardiography, which revealed permeable ovale foramen (PFO), interauricular septum aneurysm, spontaneous shunt, and easy inversion with Valsalva maneuver. The patient was submitted to percutaneous closure of PFO with good immediate result. She was discharged with improvement of the hypoxemia (pO<sub>2</sub> 68.8 mmHg in ambient air), with a diagnosis of ANCA-associated vasculitis (PR3).

**Discussion:** In such a situation, where the severity of hypoxemia is not explained by other hypotheses, the possibility of a venoarterial shunt should always be present. Notably, right-to-left inter-atrial shunt is a frequent and underdiagnosed situation, curable with simple procedures. This case reminds us that maintaining a high clinical suspicion should motivate the repetition of diagnostic tests, even if the first result is negative. No examination is 100% sensitive and there may be intermittent situations, as in the present case. Finally, while the search for a common cause for multiple system dysfunction is important, patients may have parallel unrelated problems, each deserving the best diagnostic and therapeutic approach.

**Key words:** Patent foramen ovale. Veno-arterial shunt. Hypoxemia. OSA.