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LETTERS TO THE EDITOR

Thinking outside the thorax for patients difficult to wean from non-invasive ventilation: amyotrophic lateral sclerosis diagnosis and management in a Respiratory Intermediate Care Unit

Dear Editor,

Respiratory and upper airway muscles weakness cause insufficient ventilation, ineffective cough and bulbar dysfunction in Amyotrophic Lateral Sclerosis (ALS).[1]

Dyspnea is the first symptom in only 1–3% of cases. Respiratory failure (RF) at the onset requiring mechanical ventilation (MV) is even more unusual.[2,3]

Pulmonology evaluation should occur at the beginning, in parallel with Neurology, in order to detect early signs of respiratory involvement. Unfortunately, in real life this ideal assessment it is not always possible, especially in the hospitalized patient with acute presentation of disease. Non-invasive ventilation (NIV) and pulmonary rehabilitation interventions for clearing airway secretions are recommended, as necessary.[4]

At our pulmonology department, whenever possible, acute patients are started on NIV at the Respiratory Intermediate Care Unit (RICU) to ensure better monitoring and optimize adaptation to NIV. The authors describe three cases of ALS presenting with RF admitted to the RICU of Hospital Pulido Valente – Centro Hospitalar Lisboa Norte in the last 10 years.

Our aim is to call attention to the need for ''non-respiratory'' diagnosis, particularly ALS, in patients dependent on ventilatory support.

Case 1: 64-year-old male, smoker (60 pack years), uncooperative with cough measure, presented with dyspnea, anorexia, weight loss (body mass index (BMI) 17.3 kg/m^2) for 3 months and with no previous history of disease.

There was anemia (9.5 g/dL) and elevated inflammatory parameters on blood tests. Thyroid function was normal. Chest X-ray (CXR) showed no consolidation. Arterial blood gases (ABG) showed global RF (GRF) (FiO_2 21%: pH 7.40; pO_2 = 63 mmHg; pCO_2 = 54 mmHg; HCO_3 = 31 mmol/L; $SatO_2$ = 93%).

Respiratory infection was assumed and treated accordingly. Despite initial improvement, GRF worsened, leading to NIV (pCO_2 > 62 mmHg). Echocardiographic evaluation showed diastolic dysfunction. Clinical and ABG deterioration occurred during daytime weaning attempts. Later, he developed dysphagia. Neurologic exam showed generalized muscle atrophy, hyperreflexia and fasciculations on lower and upper limbs. Electromyography revealed severe diffuse loss of motor units with extensive diaphragmatic involvement, suggestive of ALS. Despite in-exsufflator use, the patient had abundant secretions and needed frequent bronchoscopy for prompt aspiration. In addition, the patient showed difficulties in adapting to the mouth piece and suffered claustrophobia when using the facial mask. After three months, tracheostomy and percutaneous endoscopic gastrostomy were performed. The patient was transferred to a hospital near his address, awaiting home discharge.

Case 2: 70-year-old female, non-smoker, with dyspnea, lower limbs edema, dysphonia and dysarthria for several weeks. She had diabetes mellitus, obesity (BMI 36 kg/m^2), dyslipidemia and depression. Blood tests and CXR were normal. ABG showed GRF (FiO_2 26%: pH 7.35, pCO_2 = 63.1 mmHg, pO_2 = 58 mmHg, HCO_3 = 33.7 mmol/L, $SatO_2$ = 89.4%). The diagnoses of heart failure or obesity-hypoventilation-syndrome were considered. The patient was started on NIV without significant improvement (pCO_2 = 58 mmHg). Functional respiratory assessment showed FVC =22%, 550 mL. Hemoglobin and thyroid function were normal. NTproBNP was slightly elevated (244 pg/mL). Despite treatment optimization, RF worsened after any trial to stop NIV. Neurologic evaluation found paralysis of the tongue, global muscle atrophy, ocular supraversion movement's paresis and hyperreflexia. Electromyography revealed severe diffuse loss of motor units, with respiratory muscle involvement, suggestive of ALS. NMD was rapidly progressive and patient refused tracheostomy. She died one month after hospital admission.

Case 3: 72-year-old female, non-smoker, with dyspnea and orthopnoea for 3 months. She had had pulmonary tuberculosis in childhood, dyslipidemia and aortic valve replacement surgery the previous year. Her BMI was 24.2 kg/m^2. Blood tests were normal. CXR showed a small right pleural effusion and left lower lobe infiltrate. She had GRF (FiO_2 21%: pH 7.41, pCO_2 = 84 mmHg, pO_2 = 64 mmHg, HCO_3 = 53 mmol/L, $SatO_2$ = 92%). The diagnoses of community acquired pneumonia and heart failure were considered and treatment was started. Despite radiological improvement, the patient still required NIV. Spirometry showed FVC = 38%, 910 mL. Patient could not collaborate to assess maximal inspiratory and expiratory pressures.

Sleep polygraphy showed apnea–hipopnea index 6.6/h and min SpO_2 84%. Echocardiographic evaluation revealed

left ventricular hypertrophy with normal systolic function. Aortic prosthetic valve was functional. Neurological evaluation was normal. Electromyography was suggestive of ALS. The patient was discharged home under NIV.

RF as the initial manifestation of ALS is rare and, in such cases, diagnosis is extremely difficult.[5,6]

The Portuguese group of De Carvalho et al.[7] described 3 cases of ALS presenting with acute RF that required IMV.

In our cases, other diagnoses were firstly considered. However, inadequate response to treatment, difficult weaning, problems in eliminating secretions and swallowing dysfunction, raised the hypothesis of NMD.

Diaphragmatic weakness, confirmed in the first two patients, was the major cause of RF, as in the cases reported by De Carvalho et al.[7] Supine evaluation of vital capacity is a highly informative parameter[1] however, this was not assessed in any of the cases reported here which constitutes a limitation of this study. Long term NIV was the solution for one patient. IMV with tracheostomy was the option for the other patient that survived. It is indicated when there is severe bulbar dysfunction and clearing secretions techniques failure.[8,9] When there is total dependence of NIV, tracheostomy is also an option, depending on patient's wishes.[8]

Diagnosis of NMD in RICUs is uncommon. However, despite no previous neurological history, NMD should be investigated in patients difficult to wean from ventilatory support. A multidisciplinary approach to the clinical, psychological and social factors should be available for these patients.

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Conflict of interest

The authors declare that they have no conflict of interest.

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Central alveolar hypoventilation due to progressive multifocal leukoencephalopathy

Dear Editor,

Progressive multifocal leukoencephalopathy (PML) is a demyelinating disease caused by John Cunningham (JC) virus and occurs in immunosuppressed patients. PML affects mainly the cortical and subcortical white matter, includes neurological symptoms such as ataxia, hemiparesis, visual anomalies and can also be accompanied by behavioural alterations. PML can also cause lesions in other central nervous system (CNS) areas, such as the brainstem or the spinal cord.[1,2]

PML may occur in patients with severe brainstem injuries, such as tumours, strokes or infections which might affect the central chemoreceptor zone, causing central alveolar hypoventilation syndrome (CHS).[5–7]

We report the case of a 27-year-old woman who had undergone a double intestinal-kidney transplantation in 2013. Induction immunosuppression consisted of corticoids, tacrolimus and eculizumab. In 2014 she gradually developed diplopia, nystagmus, ataxia and dysphagia. A magnetic resonance imaging (MRI) showed a bilateral asymmetric