Solitary fibrous tumors of the pleura: not always a benign entity

Dear Editor,

Solitary fibrous tumor of the pleura (SFTP) is a rare tumor originating in mesenchymal cells; it represents less than 5% of all pleural tumors. It is generally benign but 10–30% of SFTP are malignant.

We report the case of a 61-year-old man, with a past history of bronchiectasis and severe restrictive ventilatory defect due to thoracic scoliosis (Fig. 1 A), who came to the pulmonology outpatients department with complaints of asthenia over the last year. He was an occasional pipe smoker and the physical examination showed marked thoracic deformity which limited chest expansion. A chest X-ray showed a large retrosternal opacity (Fig. 1B) and the chest computed tomography (CT) confirmed the presence of a retrosternal lobulated mass, 10.2 cm × 3.7 cm, in the right hemithorax (Fig. 1C and D). A percutaneous CT-guided biopsy was carried out and the histology showed evidence of mesenchymal neoplasm without necrosis, significant pleomorphism or mitotic figures. The tumor cell population stained diffusely for CD 34, vimentin, bcl-2 and CD 99 and negative for S-100. Ki-67 was <5%. Although a benign SFTP was diagnosed based on the histology, a right thoracotomy was performed for diagnosis and treatment. A pedunculated tumoural mass was complete resected with segmental resection of the right upper lobe. Immunohistochemistry showed positivity for CD-34 and bcl-2 confirming SFTP. The pathological examination revealed multiple mitotic figures (12 mitoses per 10 high-power fields), mild pleomorphism, hypercellularity, focal hemorrhage and several areas of necrosis and the resection margins were tumor-free. According to these features the tumor was classified as malignant. The post-operative period was uneventful and the patient was discharged home. The patient is now in the third month after surgery without any sign of local recurrence or metastasis.

SFTP is a rare tumor with generally an indolent course and good prognosis with a 10-year survival rate up to 98%. It occurs mainly in individuals in their sixth or seventh decades of life without gender bias. There is no association with tobacco, asbestos or other exposures. More than 50% of the patients are asymptomatic and the tumor is frequently an incidental finding on a routine chest X-ray. When symptomatic, the patients usually present non-specific respiratory symptoms such as dyspnea, cough and chest pain, and more rarely systemic symptoms. Occasionally, SFTP can manifest as paraneoplastic syndromes, such as hypertrophic pulmonary osteoarthropathy (Pierre Marie-Bamberger syndrome) and refractory hypoglycemia (Doerge-Potter syndrome). The frequency of these syndromes increases with tumor size.

Malignant SFTP is uncommon and its incidence varies from 7% to 60%. It rarely arises from a pre-existing SFTP undergoing malignant transformation. The role of percutaneous CT-guided biopsy is not yet established because its
diagnostic accuracy is low\(^5\) and it can underestimate the degree of malignancy given the size and histological heterogeneity of these tumors. Therefore, a pre-surgical diagnosis should not affect the decision to perform surgery for benign and malignant SFTP. According to the world Health Organization Classification for soft tissue tumors, the malignant SFTP is diagnosed if one or more of the following histologic features are present: hypercellularity, pleomorphism, tumor necrosis, more than 4 mitoses per ten high-power fields and infiltrative margins. Immunohistochemistry may be useful in differentiating the SFTP from mesotheliomas and intrapleural sarcomas. Generally SFTP is vimentin, CD34, CD99 and \(bcl-2\) positive and cytokeratin negative.

Complete resection of tumor is the treatment of choice, the only demonstrated effective treatment and the most important prognostic factor.\(^5\) The choice of surgical approach (video-assisted thoracoscopic surgery (VATS) and standard thoracotomy) is essentially based on tumor size and the difficulty of removal. The role of adjuvant radio and chemotherapy in malignant SFTP remains unclear since there is no systematic assessment due to the rarity of the tumor. Despite this, there are some reports showing good response to adjuvant radiotherapy in tumors with incomplete resection.\(^5\) Recurrence rates for completely resected malignant SFTP range from 14% to 63%\(^5\) in pedunculated and sessile tumors, respectively, and occur mainly in the first 24 months after surgery. Despite a complete resection, malignant SFTP can have a poor prognosis with a 5-year rate survival of 45,5%.\(^5\) Recurrent tumors should be resected and combined chemotherapy with temozolomide and bevacizumab can be considered in locally advanced, recurrent or unresectable malignant SFTP.\(^7\)

We report a case of a malignant SFTP, a rare pleural tumor that although completely resected and potentially with good prognosis, should have long-term follow-up due to the high risk of local and metastatic recurrence.

**Conflicts of interest**

The authors have no conflicts of interest to declare.

**References**


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Combined intrapleural therapy in infectious pleural effusion

To the Editor,

Pleural infection often complicates pneumonia, with a mortality rate of approximately 20%.1-3 It can also occur as a complication of pleural management (e.g., surgical procedures) or spontaneously. Surgery is often required in cases that are not resolved by medical therapy and the risks and costs are not negligible. Recent data indicate the effectiveness of intrapleural combined therapy, in which deoxyribonuclease is used in conjunction with a fibrinolytic.4 The authors report two cases of patients with pleural infection who received this treatment, with excellent results.

Case 1

A 54-year-old man with a history of cervical spinal cord injury 28 years ago, with tetraparesis and chronic hypercapnic respiratory failure under nocturnal non-invasive ventilation (NIV), was hospitalized for sympathectomy due hyperhidrosis. The procedure was complicated by a left pneumothorax, and a chest tube was placed. Approximately 24 h after chest tube removal, systemic inflammation was documented, and the chest radiograph showed a left pleural effusion, which thoracic ultrasound proved to be multiloculated (pleural fluid characteristics after thoracentesis were indicative of pleural infection). Despite antibiotics and the insertion of a new chest tube (Fig. 1A), the patient continued to present with fever, and pleural fluid drainage was very limited during subsequent days. Given the high patient surgical risks inherent to his comorbidities, combined intrapleural therapy (CIT) was started using a protocol similar to that explained by Rahman et al., 4 with 5 mg dornase alfa (DNase) diluted in 45 ml NaCl 0.9% and 10 mg alteplase (fibrinolytic), diluted in 40 ml NaCl 0.9% (both administered sequentially twice a day, for 3 consecutive days), clamping the drain for 1 h following administration of each drug, allowing them to act. This resulted in a significant increase in pleural fluid drainage, and rapid clinical and radiological improvement (Fig. 1B). There were no complications associated with this treatment, and the patient was discharged 10 days after beginning CIT, without the need for any additional interventions.

Case 2

A 41-year-old woman with no relevant past medical history, presented with right pleuritic pain, systemic inflammatory syndrome and evidence of loculated right pleural effusion in thoracic ultrasound. Thoracentesis was indicative of pleural infection, so she was given antibiotics, and a chest tube was placed (Fig. 1C). On subsequent days, pleural fluid drainage remained vestigial, and analytic inflammatory markers remained elevated. Thoracic surgery was not found to be emergent; consequently, CIT was undertaken, following a similar protocol as in Case 1, with comparable results (Fig. 1D). The patient was discharged 12 days after beginning intrapleural treatment, and no further drainage was needed. As a complication, she reported intense local pain during the instillation of the drugs, which was suitably controlled with systemic analgesia administered before subsequent procedures.

The efficacy of fibrinolytic therapy in the pleural infection is controversial, as the results obtained in several studies are not consistent.5 Case reports and small observational studies suggest its effectiveness, but it has not been confirmed in larger studies.6,6 In the largest randomized, controlled and double-blind trial, Markell et al. concluded that streptokinase was not superior to placebo in reducing mortality or the need for surgery.6 In a Cochrane meta-analysis,7 which included the previous study, Cameron and Davies concluded that there is a potential benefit of fibrinolytics, although this result should be interpreted with caution. More recently, in another randomized, controlled and double-blind trial, Rahman et al. found that only the combination of alteplase and dornase alfa was effective in increasing drainage, and reducing the length of stay and the need for surgery.8 In a more recent meta-analysis,9 and Janda and Swiston concluded that fibrinolytics are potentially beneficial in pleural infection in adults, and that they should be considered in loculated pleural effusion, as they may reduce the need for surgery.