

Monophasic Synovial Sarcoma of the Lung[☆]

Sarcoma sinovial monofásico pulmonar

To the Editor,

Synovial sarcomas are very rare tumours, and primary location is an exceptional finding. We present the case of a 46-year-old patient with a late diagnosis of a primary thoracic monophasic synovial sarcoma.

A 46-year-old man was admitted to hospital due to progressive dyspnoea and a painless axillary mass that he had had for several months (Fig. 1). Histology revealed a spindle cell tumour with mixed cellularity, focal myxoid changes, sparse collagen and mitotic figures. Epithelial membrane antigen was also found. The differential diagnosis included spindle cell tumour variants: liposarcomas, leiomyosarcoma, malignant peripheral nerve sheath tumour, malignant fibrous histiocytoma and monophasic synovial sarcoma (MSS). MSS was confirmed by S-100 negativity and muscle markers. The patient underwent tumour resection, chemotherapy with ifosfamide and radiotherapy, but the tumour returned and he died as a result of pericardial effusion.

Pulmonary sarcomas are a very rare histological variety, and the MSS group is particularly rare. The majority are metastases

from a primary tumour at any site.¹ Primary pulmonary sarcomas represent about 0.5% of lung cancers.² Diagnosis can only be established after all other possible sites explaining that the pulmonary extension has been ruled out by clinical examination and imaging techniques. A detailed immunohistochemistry study is also required to exclude other spindle cell tumours. Leiomyomas, fibrosarcomas and hemangiopericytomas are the most common varieties of primary sarcoma of the lung.³

Synovial sarcoma is a tumour that is well defined morphologically, and occurs most frequently in the soft tissue. This type of tumour represents 10% of all tissue sarcomas. It is classified into four histological subtypes: biphasic, monophasic fibrous, monophasic epithelial and poorly differentiated. However, the monophasic epithelial subtype (presented here) is extremely rare.⁴

Thoracic MSS typically occurs in the form of a mass in the ribcage, although they may also emerge from the lung and the pleura. Generally they occur in adults, and the mean age at diagnosis is 38 years.⁵ Patients with chest wall involvement have chest pain that is difficult to manage even with potent analgesics, dyspnoea and cough.

Treatment of thoracic MSS requires an integral approach, including surgery, radiotherapy and chemotherapy protocols (principally with ifosfamide). Median 5-year survival is 50%.



Fig. 1. Intra and extra-thoracic MSS localization.

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An Asthmatic Patient With Bronchomalacia and Good Response With Continuous Positive Airway Pressure[☆]

Asmática con broncomalacia y buena respuesta al tratamiento con presión positiva continua en la vía aérea

We present the case of an asthmatic patient with incapacitating symptoms that did not improve with optimized asthma treatment but responded well to continuous positive airway pressure (CPAP). The patient is a 71-year-old woman, never-smoker, hypertensive with a 40-year history of anxiety-depression syndrome. At the age of 65 she began to develop episodes of dry cough, dyspnea and wheezing. Spirometry was normal, skin prick tests for airborne allergens were negative and bronchial challenge testing with methacholine showed PC20 of 21 mg/ml. The episodes of cough and dyspnea with wheezing became increasingly frequent and more severe. Spirometry was repeated, revealing mild obstruction and a post-bronchodilator test was positive. Treatment was initiated with high-dose inhaled corticosteroids, bronchodilators and anti-leukotrienes. Despite this, the patient frequently visited the emergency department and required long cycles of oral corticosteroids. She came to our specialized asthma clinic, where a head and neck CT was performed, that was normal. ENT examination ruled out vocal cord dysfunction. On bronchoscopy, the trachea and carina were normal but there was excessive dynamic airway collapse: collapsibility of >50% on expiration in the segmental and subsegmental bronchi of the RLL, RUL, LLL and to a lesser extent in the LUL (Fig. 1). Treatment with nasal CPAP at a pressure of 6 cm H₂O was prescribed during sleep and intermittently during the day. The patient progressed well, with good adaptation to CPAP and

improved symptoms. She has not had to return to the emergency room, has discontinued her systemic corticosteroids, and remains controlled with moderate doses of inhaled corticosteroids.

Bronchomalacia is a defect of the composition of the cartilage of the bronchi, involving a loss of mechanical resistance. This means that the positive pressure developed during expiration narrows the lumen of the bronchi to a variable extent, making the passage of air difficult. This disorder is rare in adults and is usually caused by damage to the bronchial tree from chronic bronchitis, tuberculosis, prolonged intubation, surgery, injury, lung transplant or cancer.¹ Chronic symptoms such as cough, dyspnea and recurrent infections occur. It is diagnosed by assessment of airway dynamics with tomography and fiberoptic bronchoscopy, and classified as mild, moderate or severe, depending on the degree of expiratory collapse.² It can be localized or diffuse, affecting the trachea (tracheomalacia), the bronchi (bronchomalacia) or both (tracheobronchomalacia).^{3,4}

The primary aim of treatment is to manage concomitant disease, and if there is no improvement, the use of CPAP as a pneumatic stent can be considered. There are, however, few reports of adults treated in this way, and controlled studies are required to confirm the benefits of this approach. Intermittent CPAP with a nasal mask during the day and continuous application at night is recommended. There are no specific criteria for selecting the appropriate CPAP pressure in these patients. In some cases, positive pressure stabilizes the patient and serves as a bridging treatment to other alternatives, such as stenting of the airway or surgery.² We believe that long-term CPAP will be suitable for our patient, since, for the moment at least, this is providing satisfactory results.

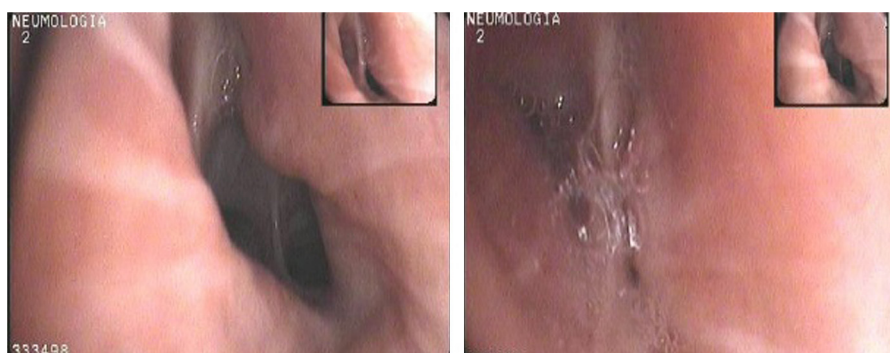


Fig. 1. Bronchoscopic image of the left lower lobe during inspiration (left) and expiration (right).

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