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Primary Synovial Sarcoma of the Lung: A Very Rare Diagnosis with Poor Prognosis

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Abstract

Primary synovial sarcoma of the lung is an extremely rare neoplasm and highly aggressive. The diagnosis is established after extra-thoracic sarcoma and other primary lung malignancies are excluded.

We report the case of a 73-year-old man who presented a well-defined mass. Immunohistochemically was positive for bcl-2, vimentin, S-100 but negative for CD34, cytokeratin, epithelial membrane antigen and calretinin.

Given the rarity of primary synovial sarcoma of the lung, there are no guidelines for its treatment and survival is low although new therapeutic possibilities are upcoming and should be considered in future management.

Keywords: Pulmonary neoplasm • Synovial Sarcoma

Introduction

Primary pulmonary sarcomas (PPS) are a group of highly aggressive and rare tumors, accounting for 0.1-0.3% of all lung malignancies [1].

The term "synovial sarcoma" is a misnomer since they do not originate from the synovial tissue but from mesenchymal tissue either of bronchial wall, vessels or pulmonary stroma. As most lung malignant mesenchymal neoplasms are manifestations of metastatic tumor the diagnosis can only be established after exclusion of sarcoma like primary lung malignancies and metastatic sarcomas [2].

Here be in, we describe PPS due to its rarity and its difficult therapeutic management and highlighting the difficult differential diagnosis and new possible therapeutic options.

Case Presentation

We report a case of a 73-year-old man, former smoker (52 pack-year), who attended the emergency department with progressive shortness of breath and pleuritic chest pain with a one month of evolution. Associated with these symptoms there was fever and sporadic episodes of hoarseness and hemoptoic sputum. At physical examination, the patient was dyspneic, unable to complete sentences, had finger clubbing, decreased vesicular breath sounds and dull percussion of the right hemithorax. ECOG performance status 2. Blood tests results showed an increase of inflammatory parameters (PCR of 104 mg/L). Posteroanterior chest X-ray revealed a homogeneous hypotransparency on the lower two thirds of the right hemithorax. Thoracic computed tomography showed a slight mediastinal left deviation, mediastinal adenomegaly (the largest paratracheal with 17 × 14 mm), right pleural effusion and a well-circumscribed mass (128 × 117 mm) in contact with the right lower lobe (RLL) and right middle lobe (RML) (Figure 1). Thoracentesis revealed an exudate according to Light's criteria without malignant cells and no microbiological agents. A fiberoptic bronchoscopy revealed indirect signs of RLL invasion but bronchial biopsies demostrated inflammatory lymphocytes cells and scarce eosinophils.

As none of the above exams were conclusive, a thoracoscopy was performed on the right side, which demonstrated various superior lobe adhesions and a large pulmonary mass with pleural thickening. Biopsies of the parietal pleura and the mass revealed suggestive cells of monophasic epithelial sarcomatous carcinoma of the lung which immunohistochemistry was positive for bcl-2, vimentitin, S-100 and negative for CD34, cytokeratin, epithelial membrane antigen and calretinin.

In order to exclude a lung metastasis of an extrathoracic sarcoma, the patient was submitted to an 18F-FDG PET which confirmed a large mass on the RLL and RML with 18F-FDG hypermetabolism (SUV max=25.24), right pleural effusion and mediastinal lymph nodes uptake with no other 18F-FDG hypermetabolism (Figure 2). Thereby the definitive diagnosis of PPS was accepted. Due to the rapid clinical decline, palliative care was assumed, and the patient died three months later.

Discussion

Synovial sarcoma is a rare and aggressive tumor which usually appears between the third and fifth decade of life and is slightly more frequent in males. Clinical symptoms are nonspecific and up to 25-50% of patients have no symptoms [3]. A large well-defined but not encapsulated mass, with soft tissue component and necrotic regions is identified, as well as, ipsilateral pleural effusion and invasion of adjacent structures is commonly found on thoracic CT [4].

A tumor biopsy from thoracotomy or thoracoscopy is recommended since other methods have limitations regarding sample size. As PPS are histologically indistinguishable from metastatic sarcomas, a total body survey is necessary to exclude a primary tumor elsewhere. Most sarcomas are positive for Bcl-2, vimentin, cytokeratins 7 and 19 with poor immunoreactivity for S-100, CD34, desmin, actin and vascular tumor markers. A specific translocation t (x, 18) (p11.2; q11.2) can help confirm the diagnosis and is present in 90% of the cases, resulting from the fusion of the SYT gene on chromosome 18 to SSX1 or SSX2 on chromosome X [5] with prognostic significance.

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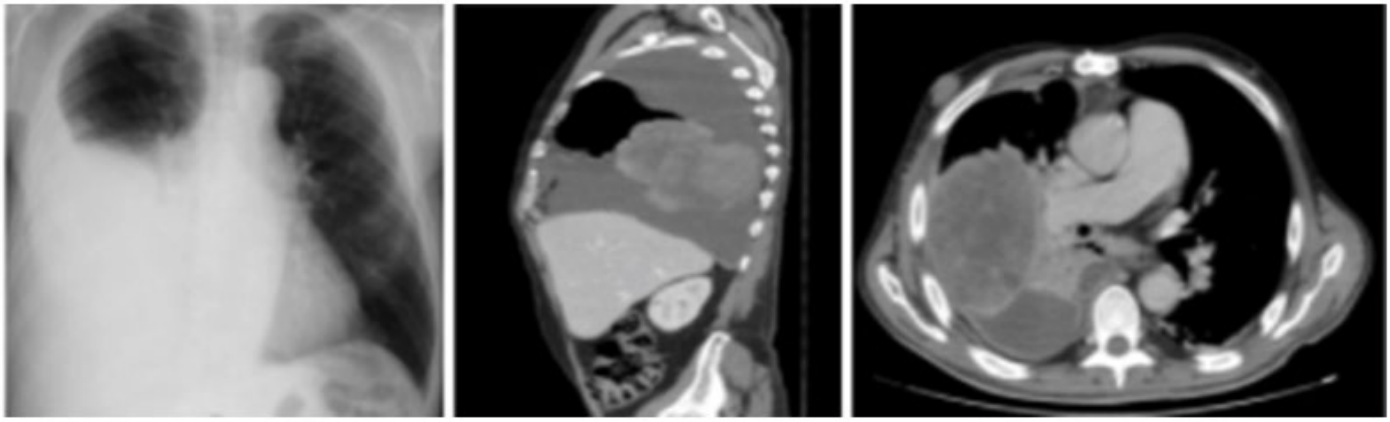

Figure 1. (a) Posteroanterior chest X-ray with homogeneous hypotransparency on the lower two thirds of the right hemithorax, (b) Thoracic Computed Tomography scan reconstructed in the sagittal projection; (c) Thoracic Computed Tomography scan in the axial projection with mediastinal deviation, mediastinal adenomegaly, pleural effusion and a well-circumscribed mass.

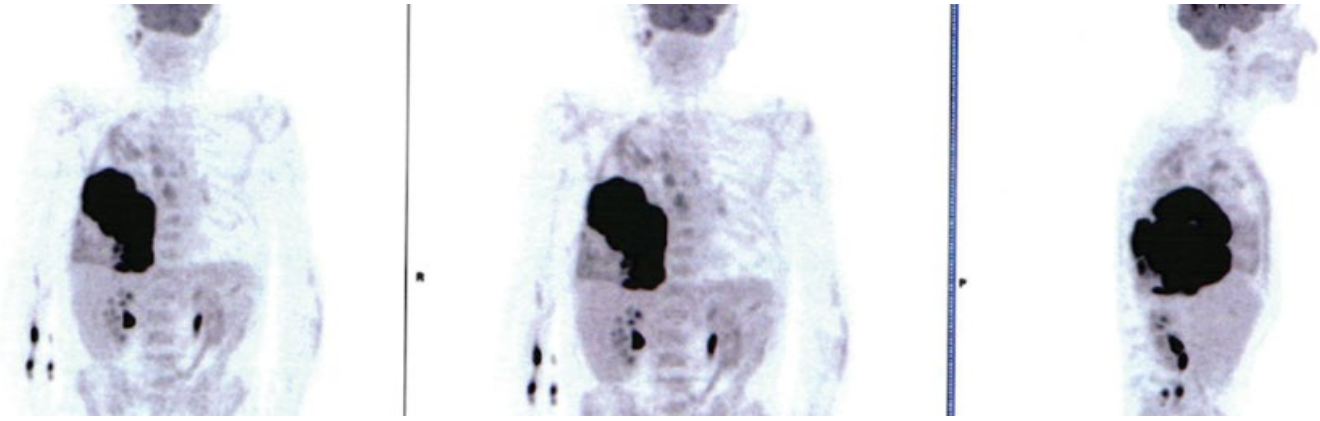

Figure 2. PET-CT scan with 18F-FDG hypermetabolism on the lung mass (SUV max=25.24).

The prognosis is poor with a five-year survival rate of 38%. Currently there is no standardized therapy and although complete surgical resection remains the only strategy associated with survival, only one third of these tumors are resectable at presentation. The role of chemotherapy remains debatable, as treatment with doxorubicin and ifosfamide only provides response in about 20% and does not improve survival [6]. The benefit of immunotherapy is not fully defined since the role of PD-L1 expression remains uncertain although pembrolizumab has shown some promising result [7].

Prognostic factors in PPS are still under investigation as the low number of patients and published data make it difficult to establish repetitive factors. However, it has been shown that complete resection significantly improves prognosis and size >5 cm, male sex, age >20 years, extensive tumor necrosis, large number of mitotic figures (>10/10 high-powered fields), neurovascular invasion and SYT-SSX1 variant are poor prognosis factors [6].

Conclusion

In conclusion, PPS is a rare and aggressive type of tumor with a poor prognosis. The diagnosis is difficult and related not only by the low incidence but also because a definitive diagnosis depends on clinic, imaging but mainly on immunohistochemical staining and molecular tests.

Conflict of Interest

The authors declare that there are no conflicts of interest in carrying out this work.

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