Case Report

# Primary sarcoma of the lung: A very rare diagnosis and poor prognosis

**Rhizlane Belbaraka, Nabil Ismaili**

Department of Medical Oncology, CHU Mohamed VI, FMPM, Cadi Ayyad University, Marrakech, Morocco

## ABSTRACT

Primary lung sarcoma is a very rare malignant tumor, accounting for less than 0.5% of all lung tumors. We describe the case of a 50-year-old man who presented with primary pulmonary sarcoma. X-ray chest and thoracic computed tomography (CT) revealed a 40 mm × 60 mm mass in the right upper lobe. Biopsy was reported as fusiform sarcoma. Extensive clinical examination, followed by full body CT scan was performed to exclude primary synovial sarcoma located peripherally and distant metastases. Hence, surgical excision was planned after three cycles of chemotherapy, but the patient died after the second cycle. Because a sarcoma of the lung is rare, data regarding its natural history and published cases are limited. Further investigation and data collection are required to optimize the treatment of this group of rare and aggressive tumors.

**Key words:** Chemotherapy, fatal evolution, lung sarcoma, primary sarcoma

## INTRODUCTION

Primary lung sarcoma is a very rare malignant tumor, accounting for less than 0.5% of all lung tumors.[1] The variety of soft tissue sarcomas reflects the range of the mesenchymal tissues present in the lung. The three most common sarcomas include leiomyosarcoma, malignant fibrous histiocytoma and synovial sarcoma.[2] Metastases from the extrapulmonary sarcomas are undoubtedly more common than primary pulmonary sarcomas. Therefore, they must be considered before the diagnosis of primary lung sarcoma is accepted.

We report a case of a primary pleomorphic sarcoma of the lung with fatal evolution in a 50-year-old man.

## CASE REPORT

A 50-year-old Caucasian male presented to our hospital with a 5-month history of progressively increasing shortness of breath and cough. He had smoked one to two packs of cigarettes everyday for 15 years, i.e., he was exposed to 30 pack-year smoking. Physical examination revealed a patient in moderate distress with a heart rate of 110 and respiratory rate of 25, with voice hoarseness. His performance status was equal to 2. Examination of the abdomen did not reveal any lymphadenopathy, ascites and hepatosplenomegaly. Other systems were within normal limits.

Complete hemogram and blood biochemistries were within the normal limit.

Chest X-ray showed an opacity in the right upper lobe. A computed tomography (CT) was performed revealing a large, 40 mm x 60 mm mass in the right upper lobe, compressing the mediastinal structures including the esophagus and careen [Figure 1], with emphysema. This was followed by a CT-guided needle biopsy. There was no evidence of tumor elsewhere on the abdominal and bone scintigraphy.

The biopsy showed a spindle cell tumor with marked nuclear pleomorphism and numerous mitotic figures [Figure 2a]. There was no evidence of epithelial differentiation. Immunohistochemical staining was positive for vimentin and negative for multiple high and low molecular cytokeratins, epithelial-specific antigen, epithelial membrane antigen, actin, smooth muscle actin, desmin, S-100 protein and CD34. CD117 immunostaining showed

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**Address for correspondence:** Dr. Rhizlane Belbaraka, Department of Medical Oncology, CHU Mohammed VI, FMPM, Cadi Ayyad University, Marrakech, Morocco. E-mail: belbaraka.r@gmail.com

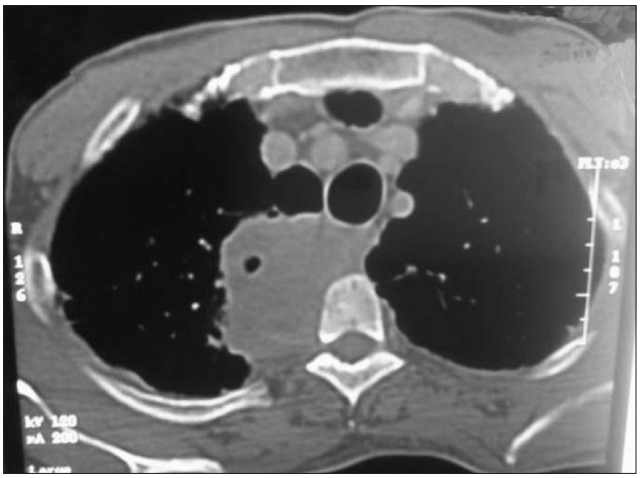

**Figure 1:** Computed tomography scan showing a compressing mass of the mediastinal structures including esophagus and carene

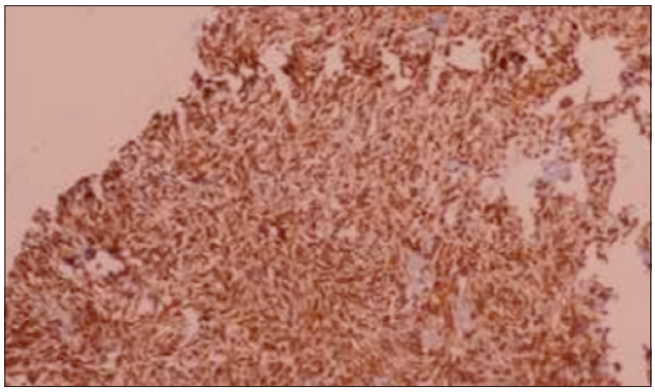

**Figure 2b:** Spindle cell immunoreactive for vimentin

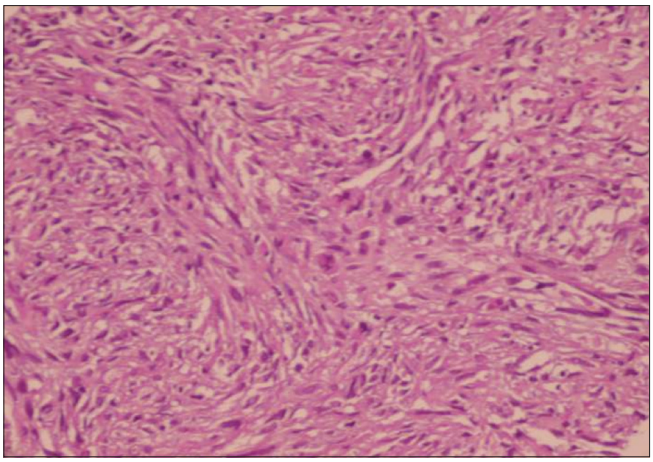

**Figure 2a:** Spindle cell tumor with marked nuclear pleomorphism

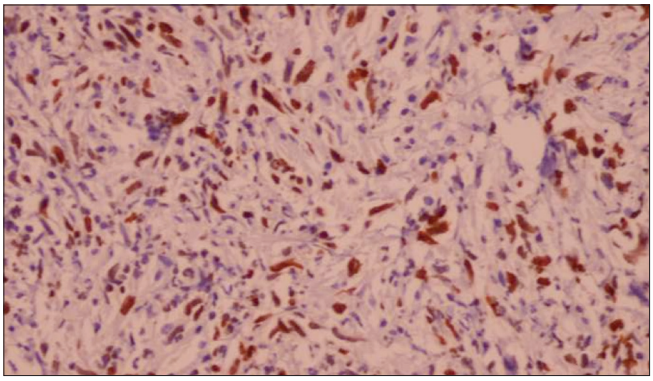

**Figure 2c:** High T-cell proliferation index

no mast cells within the tumor. The cell proliferation index with Ki 67 was 50%. A diagnosis of pleomorphic grade 2 sarcoma was made [Figure 2b and c].

A treatment by chemotherapy was performed with doxorubicine and ifosfamide, but the patient died 2 weeks after the second cycle.

## DISCUSSION

Primary pulmonary sarcomas are rare tumors, representing about 40% of "rare" pulmonary neoplasms and 9% of all sarcomas. To date, approximately 300 cases have been reported in the literature.[3]

Primary sarcomas of the lung usually present as bronchogenic carcinoma. Symptoms and radiographic features are similar to those seen in other epithelial lung tumors and depend more on tumor localization than on their histological characteristics.[4,5]

In this context, the absence of cigarette smoking might alert the physician to the possibility of a sarcoma. They typically occur in middle-aged individuals, with a slight predominance in men. This was true in our case.

The most important differential diagnosis of primary pulmonary sarcoma is metastatic spread from an extrapulmonary sarcoma. It is therefore necessary to obtain a detailed clinical history and to initiate appropriate investigations to address this possibility. Our patient had no evidence of present or past soft tissue neoplasms. Our patient had no history of previous radiation exposure, which has been recognized as a predisposing factor in the development of sarcomas.[6] Other differential diagnoses include bronchogenic carcinoma and malignant melanoma.

Immunohistochemistry has an important role in the classification. A panel of antibodies is necessary to classify soft tissue sarcomas correctly. Immunohistochemistry is also essential for ruling out the much more frequent pulmonary sarcomatoid carcinomas. In our case, antibodies to high and low molecular cytokeratins, epithelial-specific antigen and epithelial membrane antigen ruled out sarcomatoid carcinoma, synovial sarcoma and diffuse malignant mesothelioma, a diagnosis that should be considered when the tumor involves the pleura or mediastinum.

Synovial sarcoma is characterized by mast cells within the tumor, and none were detected in this tumor on staining with CD117.[7] Smooth muscle actin, actin and desmin are seen in leiomyosarcoma, and the latter two in rhabdomyosarcoma, S-100 protein in malignant peripheral nerve sheath and liposarcoma, while malignant solitary fibrous tumor and malignant vascular tumors are immunoreactive for CD34, all of which were negative in our case.

All the tumors that were considered in the histological differential diagnosis are predominantly composed of spindle cells and express vimentin.

Primary pulmonary sarcomas may have an aggressive progression. In our case, as like as in metastatic or advanced sarcomas, chemotherapy with doxorubicine and ifosfamid is the only active treatment, but the response is less than 20%.[2] High-dose chemotherapy is feasible and provides interesting response rates in patients with soft tissue sarcomas, but cannot yet be considered a common practice.

In localized disease, surgical removal is the treatment of choice for all histological types, followed by radiotherapy if the removal is incomplete. Extensive clinical examination, followed by full body CT scan, was performed to exclude primary synovial sarcoma located peripherally and distant metastases. Hence, surgical excision was planned after three cycles of chemotherapy, but the patient died.

We describe a patient with a primary fusiform sarcoma of the lung whose tumor could not be resected and who died after two cycles of chemotherapy. Because a sarcoma of the lung is rare, data regarding its natural history and published cases are limited. Further investigation and data collection are required to optimize the treatment of this group of rare and aggressive tumors.

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