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A Giant Fibrous Tumor of the Lung: A Case Report

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Abstract | **Case Report**

A giant solitary fibrous tumor is a rare mesenchymal tumor of fibroblastic differentiation that is mainly located intrathoracically. It is not associated with specific risk factors. It has nonspecific clinical features and slow growth. The diagnosis is usually incidental and in large-size cases, representing a major surgical challenge. We present a case of a 47-year-old woman with no toxic habits, with no pathological history, who presented four days before admission with worsening dyspnea that had been progressively developing over the past five years, reaching Sadoul stage V without cough, sputum, hemoptysis, or chest pain. All of this occurred in the context of apyrexia and deterioration of her general condition. Her chest X-ray showed a near-complete opacification of the left hemithorax, with displacement of the mediastinum towards the right. This study was supplemented by a computed tomography (CT), which demonstrated a well-circumscribed, non-homogenous mass, occupying the entirety of the left hemithorax. An ultrasound-guided biopsy with pathological examination revealed a fusiform tumor proliferation of mesenchymal appearance. Complete surgical resection of the tumor is the only treatment with curative potential.

Keywords: Case Report, Fibrous Tumor, Mesenchymal Tumor, Intrathoracically, Benign Tumor, Surgical Challenge.

INTRODUCTION

Solitary fibrous tumors belong to a unique subtype of mesenchymal tumors and are presumed to be of fibroblastic differentiation. They can occur in any anatomical region and mainly affect adults between the fifth and seventh decades of life, although they can occur at any age. Cases have been reported at different anatomical levels, such as the pleura, pericardium, peritoneum, mediastinum, and even the paranasal sinuses and nose [1]. Solitary fibrous tumors are classified as having intermediate biological potential, meaning that they generally have a low risk of metastasis and an indolent course; however, depending on their location, they can cause symptoms due to the mass effect they exert on other organs most patients remain asymptomatic for years [2]. The diagnosis is usually incidental, or by compressive symptoms, in large-size cases, representing a major surgical challenge.

We present a case of a 47-year-old woman, who presented with a five years history of worsening dyspnea, and was found to have a large, non-homogenous mass, encompassing the entirety of the left hemithorax and compressing the adjacent lung parenchyma.

CASE REPORT

Our patient was a 47-year-old woman with no toxic habits, never treated for pulmonary tuberculosis and with no other pathological history, who presented four days before admission with worsening dyspnea that had been progressively developing over the past five years, reaching Sadoul stage V without cough, sputum, hemoptysis, or chest pain. All of this occurred in the context of apyrexia and deterioration of her general condition.

The clinical examination found a patient with saturation= 80% in open air, polypneic, signs of respiratory distress, with fluid effusion syndrome in the entire left hemithorax. Cardiovascular, lymph node, gynecological, thyroid, and abdominal examinations were normal. A chest X-ray (Figure 1) was performed, revealing dense, homogeneous opacity of the entire left hemithorax, obscuring the diaphragmatic couple and filling the costodiaphragmatic cul-de-sac, with displacement of the trachea and heart to the right.

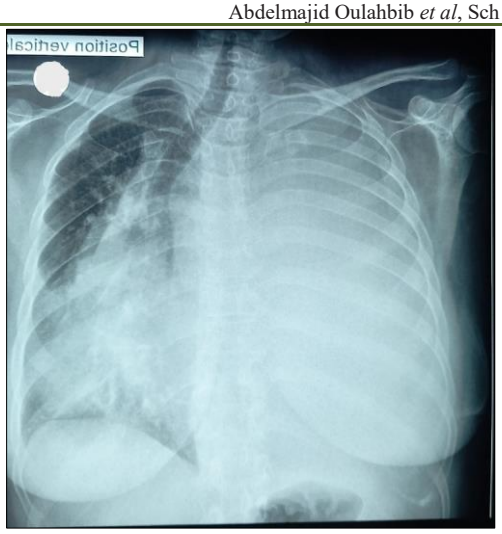
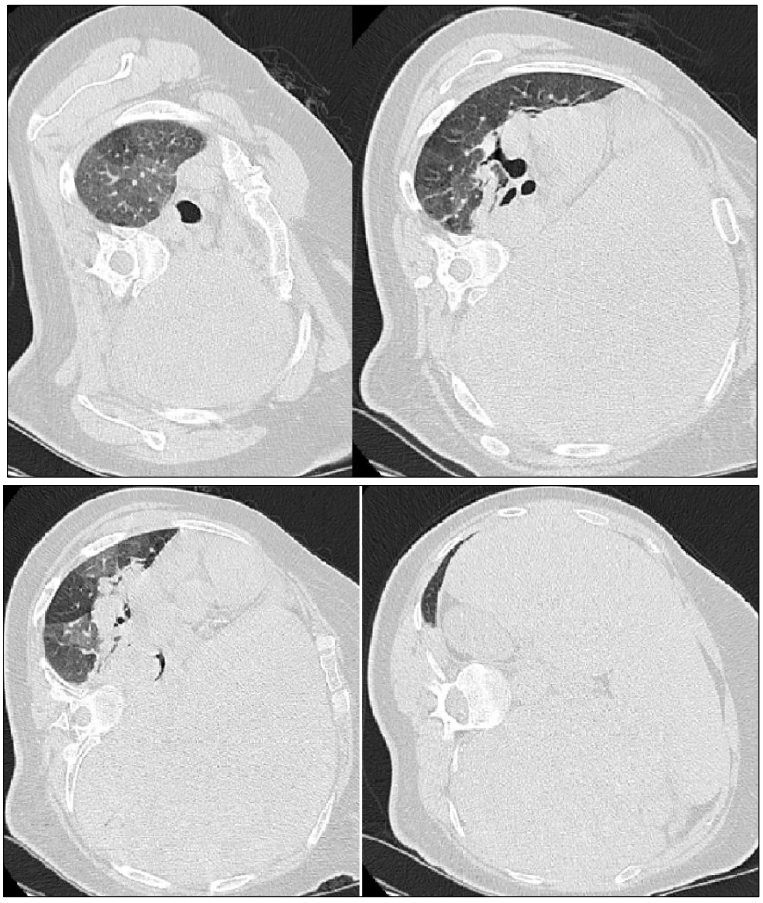


Figure 1: Chest X-ray showing a water-like opacity of the entire left hemithorax with displacement of the trachea and heart to the right

The search for Kock's bacillus in sputum and RT-PCR were negative. Biological tests were normal: D-dimers were at 601 µg/L, no biological inflammatory syndrome CRP=3mg/L, white blood cells were at 5750/mm3, hemoglobin was 14.7 g/dl, platelets were 169,000/mm^3, lymphopenia was 860/mm^3, fasting blood glucose was 0.85 g/l, and kidney and liver function tests were normal.

The chest CT scan revealed a left pleuropulmonary tissue mass encompassing the entire left chest cavity, heterodense, richly vascularized, with heterogeneous enhancement, and diffuse intense nodularity, multilobulated, measuring: 20.2x15.8x28.5 cm, exerting a significant mass effect on the mediastinal structures and on the completely collapsed left lung parenchyma, with mediastinal herniation of the mass to the right (figure2).



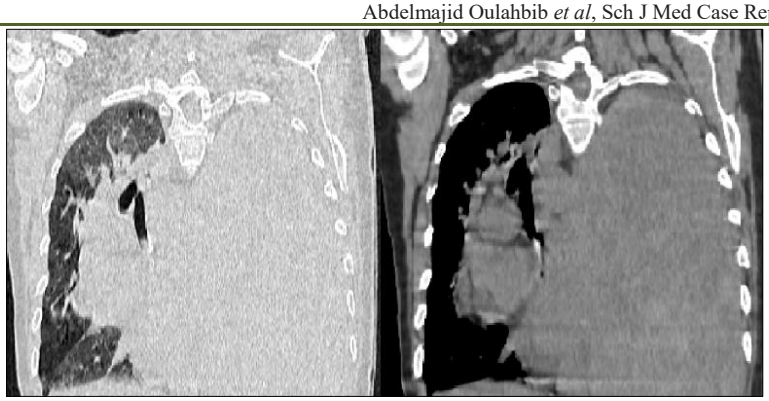


Figure 2: Chest CT scan showing parenchymal and mediastinal windows revealing a compressive left pleuropulmonary tumor formation with mediastinal herniation

An ultrasound-guided biopsy with pathological examination revealed mesenchymal proliferation with low cell density. It is arranged in short, intertwined bundles. The cells are fusiform, small to medium in size, with elongated nuclei that are wavy in places. They are free of cytonuclear atypia or abnormal mitoses. This tumor proliferation is rich in blood vessels with thickened, hyalinized walls. The interstitial tissue is fibrous.

Immunohistochemistry revealed benign fibroblastic proliferation consistent with fibromatosis without obvious malignancy in this sample, with expression of anti-AML and ß-catenin antibodies, and a negativity of desmin, PS100 and CD34. Ki67 was expressed by less than 1% of tumor cells.

The patient was placed on high-concentration oxygen via a mask, received 120 mg/day of methylprednisolone intravenously, and was given low molecular weight heparin at a preventive dose.

During her hospitalization, the patient's dyspnea worsened, requiring transfer to an intensive care unit, where she died.

DISCUSSION

Solitary fibrous tumors belong to a unique subtype of mesenchymal tumors and are presumed to be of fibroblastic differentiation. They can occur in any anatomical region and mainly affect adults between the fifth and seventh decades of life, although they can occur at any age. Cases have been reported at different anatomical levels, such as the pleura, pericardium, peritoneum, mediastinum, and even the paranasal sinuses and nose. [1].

There are fewer than 2,000 cases described in the literature, most of them retrospective and from a single center [3, 4]. The distribution is homogeneous between both sexes, usually between 40 and 60 years old. Approximately half of the cases are diagnosed incidentally; the most frequent symptoms are chronic cough and dyspnea resulting from intrathoracic occupation, as the case reported. Chest pain is characteristic of those who depend on parietal pleura, especially if an infiltration of the latter coexists [5].

Giant solitary fibrous tumors can cause paraneoplastic syndromes such as Doege Potter syndrome, which causes symptomatic or refractory hypoglycemia, which responds to surgical treatment. This is caused by the creation of insulin-like growth factor II (IGF-II), which leads to decreased insulin levels and displacement of IGF-1 [6]. Our patient did not show any of these signs.

According to the literature, the finding is incidental in imaging studies, especially in contrast-enhanced CT scans, where they appear as rounded masses, with or without areas of necrosis or myxoid degeneration or hemorrhage, homogeneous and exerting a mass effect on surrounding organs such as the heart and lung [1]. The mass occupies the entire left thoracic cavity, with pericardial involvement and total left lung collapse, causing respiratory symptoms such as dyspnea and coughing, in addition to constitutional symptoms.

The diagnosis is based on histological analysis of the surgical specimen. When resection is not considered, a simple biopsy of the lesion can be performed, sometimes by mediastinoscopy, thoracoscopy or even thoracotomy for intrathoracic tumors.

Complete surgical resection of the tumor is the only treatment with curative potential. However, a conservative approach with watchful waiting may be adopted for smaller, asymptomatic tumors. Recurrence is rare with approximately 2% for pedunculated and 8% for sessile tumors. Surveillance with serial CT scans at 6-monthly intervals is recommended [7, 8].

CONCLUSION

The radiological and clinical signs of a giant pulmonary fibrous tumor can mimic massive pleurisy. Although it is a benign tumor, the challenge with giant tumors lies in the surgical approach and the resection and

extraction technique, given the limited technical facilities in our context, as well as the delay in diagnosis. This makes this type of tumor a poor prognosis in developing countries.

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