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Solitary Fibrous Tumor of the Pleura: A Case Report

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Abstract: ***Solitary fibrous tumor, formerly called solitary benign fibroma, initially described in the pleura, but can also involve many extrapleural sites (pericardium, meninges, liver, pancreas). It is most often asymptomatic and of incidental radiological discovery. The diagnosis of pleural solitary fibrous tumors (PSFT) is guided by imaging, confirmed by surgery and histology coupled with immunohistochemistry. The evolution of these tumors is unpredictable and the correlation with histology is not strict.***

Keywords: solitary fibrous tumor, pleura, benign, rare tumor.

1. Introduction

Solitary fibrous tumor, formerly called "solitary benign fibroma", is a pleural tumor of mesenchymal origin [1]. It is rare, representing less than 5% of pleural masses with less than 800 cases reported until 2010. Initially described in the pleura, it can also involve many extrapleural sites (pericardium, meninges, liver, pancreas).

These tumors are most often asymptomatic and of incidental radiological finding [2] but can sometimes cause respiratory symptoms as in our situation (dyspnea, chest pain, cough).

We report a case of solitary fibrous tumor.

2. Observation

The patient was 31 years old, never treated for tuberculosis, with no recent tuberculosis infection, no known defects and no toxic habits. He had been presenting for 3 months with progressively worsening dyspnea becoming stage III of the mMRC associated with pleural chest pain on the left side of the body, the whole evolving in a context of fever, night sweats and conservation of the general condition.

The patient underwent a chest X - ray which revealed a pleurisy of medium abundance on the left (Figure 1).

A thoracic CT scan was performed in addition to the X - ray, showing a left basi - thoracic cyst pleurisy with homogeneous hypodense content and pseudo - nodular parenchymatous condensation foci without an air bronchogram opposite (Figure 2).
The pleural effusion was punctured and found to be exudative fluid at 38 g/l with a predominance of lymphocytes, without any evidence of neoplastic cells.

A pleural biopsy was performed as a complement, which came back inconclusive on 4 occasions.

Then the patient underwent a thoracoscopic biopsy, and the anatomo - pathological analysis objectified a mesenchymal proliferation of variable cell density, the tumor cells are spindle - shaped arranged in intersecting bundles around branched and dilated capillaries with an estimated mitotic activity of 1 mitosis ⁄ 10 fields with expression of anti STAT6 positive antibodies

The morphologic and immunohistochemical appearance is in favor of a solitary fibrous tumor (Figure 3 and 4).

3. Discussion

Pleural solitary fibrous tumor was originally described by Klimperer and Rabin as a localized mesothelioma [3]. It is currently described as a mesenchymal tumor that develops from fibroblasts present in the submesothelial connective tissue [4].

It is most often benign but may recur or have malignant potential. Malignant forms represent approximately 12% of cases [5] and are characterized by an increased risk of recurrence.

These tumors are most often asymptomatic and of incidental radiological finding [6] but can sometimes cause respiratory symptoms dominated by cough, chest pain, dyspnea, more rarely hemoptysis and general signs with fever and altered general condition [7].

Paraneoplastic syndromes have been reported in the literature in 10 - 20% of cases [8], hypoglycemia was reported in 4% of cases in the series of Briselli [9].

The diagnosis of pleural solitary fibrous tumors can only be made after an anatomopathological study; the diagnosis is usually made on the excisional specimen, the surgery being both diagnostic and therapeutic. PSFT develops in 67 to 87% of cases from the visceral pleura [8].

Microscopically, PSFT presents as an encapsulated tumor, consisting of cellular sectors made of spindle cells grouped in swirling bundles alternating with sparse collagenized sectors within a vascularized stroma with arborized vessels of hemangiopericytoid appearance. The spindle cells have a sparse cytoplasm and round or oval nuclei, finely nucleolated. They are generally devoid of cytonuclear atypia. Mitotic activity is low and foci of necrosis are rare [10].

Histological examination can give a prognostic indication and suggest malignant evolution when cellular atypia, mitotic activity, or myxoid elements are present [10].

Immunophenotyping is essential for diagnosis. The tumor cells are mesenchymal differentiating cells expressing vimentin, they typically express CD34, CD99 and Bcl2.

Many studies have shown that 80% to 100% of solitary fibrous tumors express CD34 and the expression of this antigen is attenuated in malignant forms [11].

The loss of CD34 expression is probably due to dedifferentiation of the tumor cells during malignant transformation.

The prognosis of PSFT depends on several criteria. Perrot proposed a classification for pleural solitary fibrous tumors based on morphologic and histologic appearance into four types: benign pedicle, benign sessile, malignant pedicle [12], and malignant sessile. These types are correlated with significant differences in recurrence rates (2%, 8%, 14%, 63% respectively) and survival, and recurrences can be more or less late, hence the importance of complete surgical resection and prolonged radio - clinical surveillance.

4. Conclusion

Pleural solitary fibrous tumor is most often asymptomatic by chance discovery. When it is symptomatic, it is responsible for non - specific respiratory signs.

The positive diagnosis is based on anatomopathological study which also allows a prognostic evaluation.

Despite the rarity of PSFT and the predominance of benign forms, the evolutionary profile of these tumors remains uncertain.

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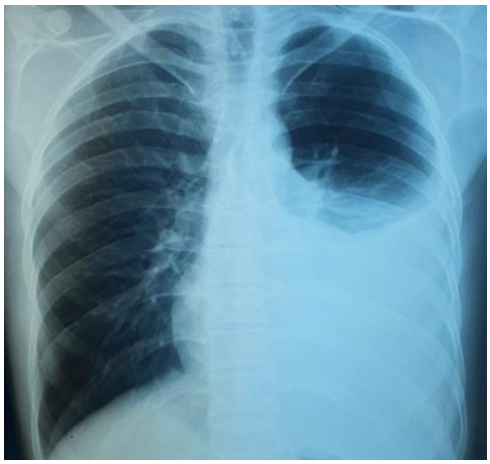
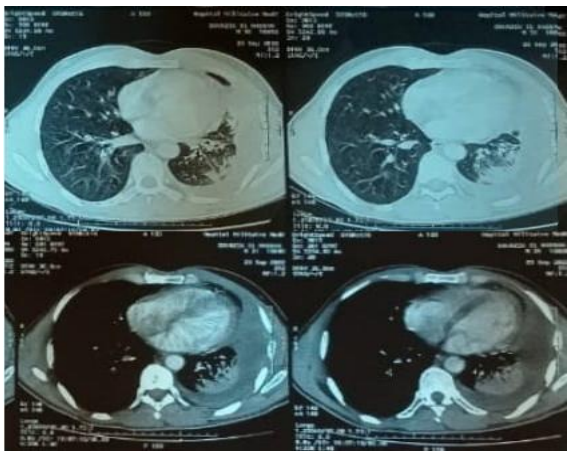


Figure 1: Chest radiograph objectifying a left pleurisy of moderate abundance.



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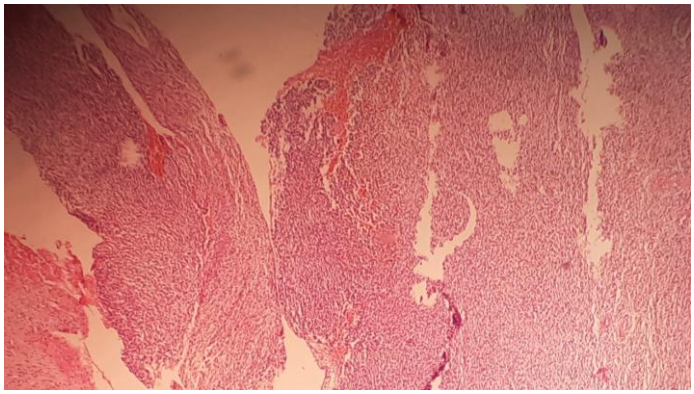
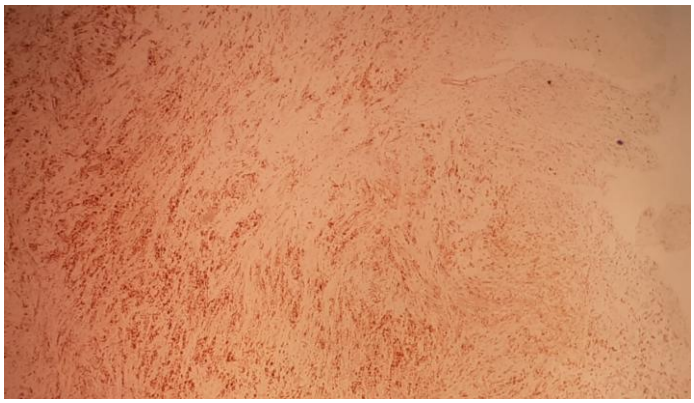


Figure 3: Microscopic image showing a spindle cell tumor proliferation of variable density.



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