

Pleural Fibroma: Diagnosis and Management Two Cases Report

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Abstract

Solitary fibrous tumors of the pleura represent less than 5% of all the tumors of the pleura. They are from mesenchymal origins and present a diagnosis difficulty. We report the cases of two patients who presented clinically with a pleural effusion like syndrome. The chest x ray showed a large homogenous opacity of the thorax and the chest CT found a large well defined tumor. Both patients underwent a large surgical resection of the tumor with a satisfying clinical and radiological follow up.

Keywords: *Pleural; Fibrous Tumor; Pathology*

Abbreviations

WHO: World Health Organization; NYHA: New York Heart Association; The chest CT: Computed Tomography; AML : Acute Myeloid Leukemia; HGF: Hepatocyte Growth Factor; IGFII: Insulin-Like Growth Factor II; MRI: The Magnetic Resonance Imaging; CD: Cluster of Differentiation; EMA: Epithelial Membrane Antigen

Introduction

Solitary fibrous tumors are a rare type of mesenchymal neoplasms. The latest World Health Organization (WHO) classification of soft-tissue tumors includes them in the category of tumors originating from fibroblastic/myofibroblastic tissues. Although uncommon, solitary fibrous tumors may occur in various parts of the body including the head, neck, thorax, abdomen, and extremities. Within the thorax, its most common site is the pleura with around 80% originating from the visceral pleura [1].

Cases Report

Case 1: A 47 years old woman without any notable medical history presents with a one year history of progressively worsening left side chest pain, associated to a dry cough and a NYHA stage II dyspnea, without hemoptysis or any other signs except weight loss and asthenia. The physical examination found a left pleural effusion syndrome. The chest X ray (Figure 1) showed a pleural opacity occupying the lower 2/3 of the left thorax. The thoracentesis did not find any pleural effusion. The chest CT (Figure 2a, 2b) showed a large mass measuring 13 x 15 x 18 cm with calcifications, which was in contact with the left main bronchus and upper lobar bronchus with an atelectasis of the upper left lobe. The mass was infiltrating the mediastinal fat, the thoracic aorta and the pulmonary artery without any mediastinal lymphadenopathy. The bronchoscopy showed stenosis of the orifice of the left lower lobar bronchus and the biopsy revealed an inflammatory lesion. A CT guided transthoracic biopsy was performed but the pathology study did not conclude to a diagnosis. The blood tests as well as the abdomen ultrasound did not find any anomaly. A thoracotomy was performed and found a huge mass occupying the lower 2/3 of the left hemithorax which was very hemorrhagic and adherent to the chest wall, the aorta and the inferior pulmonary vein with a limited encroachment of the diaphragm. A complete resection of the tumor was performed as well as 20 cm² of the diaphragm at the phrenic center. The macroscopic study of the surgical specimen found a mass weighing 2120g, measuring 24 x 19 x 10 cm with a whitish aspect. Microscopic analysis concluded a spindle cell proliferation and the immunohistochemistry confirmed the diagnosis of a solitary pleural fibroma. The postoperative follow up was unremarkable; the clinical and radiological control was satisfactory.



Figure 1: Pleural effusion like opacity of the left hemithorax.

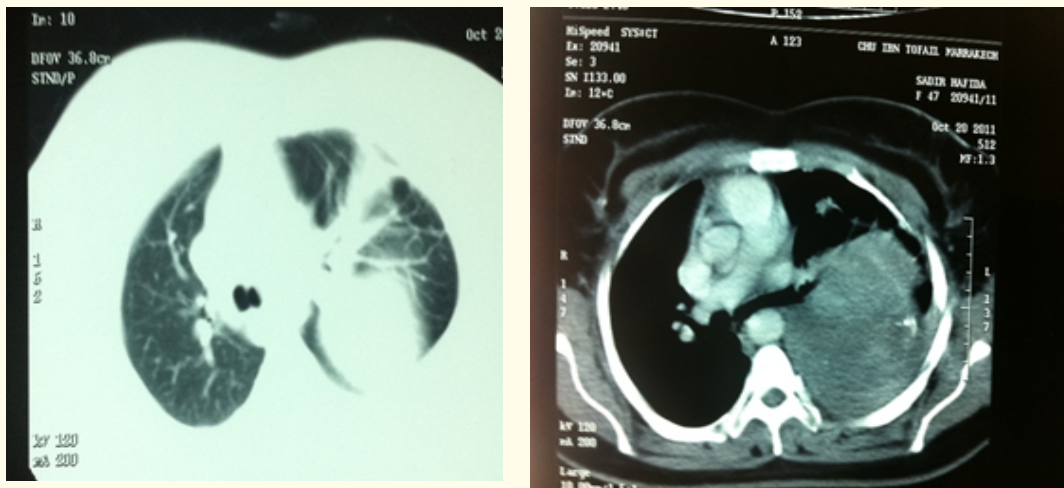


Figure 2a and b: Chest CT scan showing a large tissue mass of the left hemithorax.

Case 2: A 42 years old woman with no notable medical history presented to our department with an 8 months history of a right side chest pain. She experienced dyspnea but did not present any hemoptysis or any other respiratory sign. The physical examination found a right pleural effusion syndrome. The chest X ray showed a homogenous opacity occupying the lower 2/3 of the right thorax (Figure 3). The chest CT showed a large mass measuring 145x 140 x 120 mm with calcifications and in contact with the diaphragm (Figure 4). The blood tests as well as the abdominal sonogram did not find any anomaly.

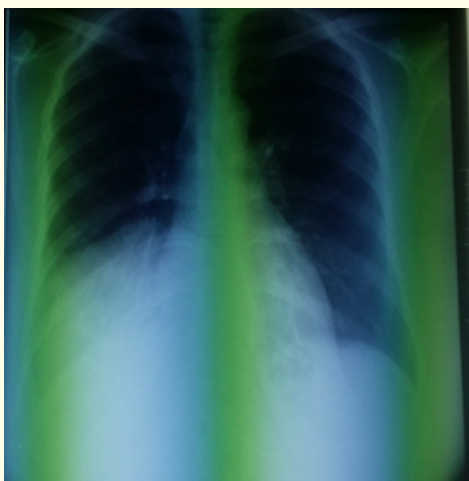


Figure 3: Chest X-ray showing an opacity of the lower right hemithorax.

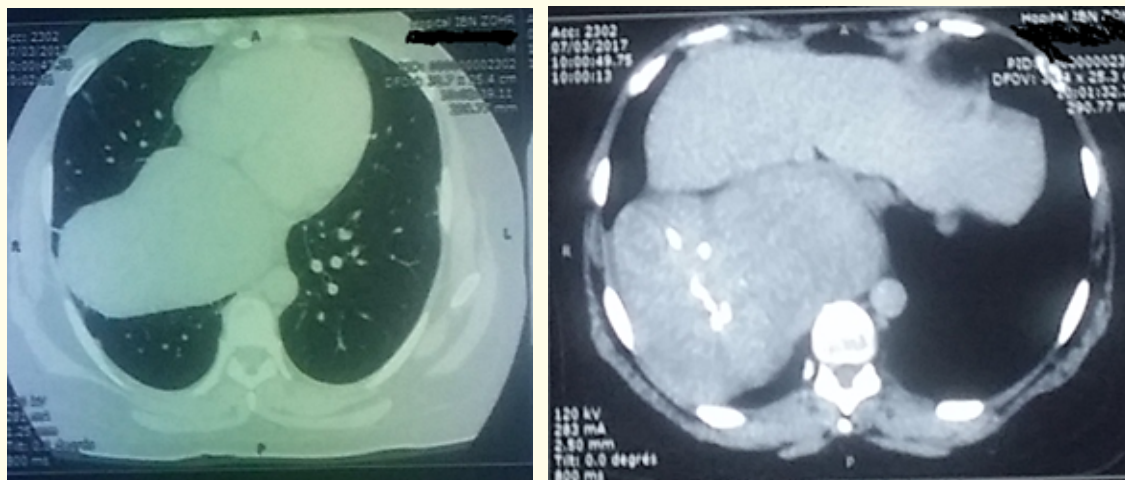


Figure 4: Chest CT showing a large mass of the right hemithorax.

A thoracotomy was performed and found a huge mass occupying the lower 2/3 of the right hemithorax with a limited encroachment of the diaphragm. A complete resection of the tumor was performed (Figure 5). The macroscopic study of the surgical specimen found a mass weighing 1223,5 g, measuring 16 x 10 x 7 cm with a whitish aspect (Figure 6). The pathology study concluded to a spindle cell proliferation and the immunohistochemistry confirmed the diagnosis of a solitary pleural fibroma (Figure 7). The clinical and imagery follow up was unremarkable.

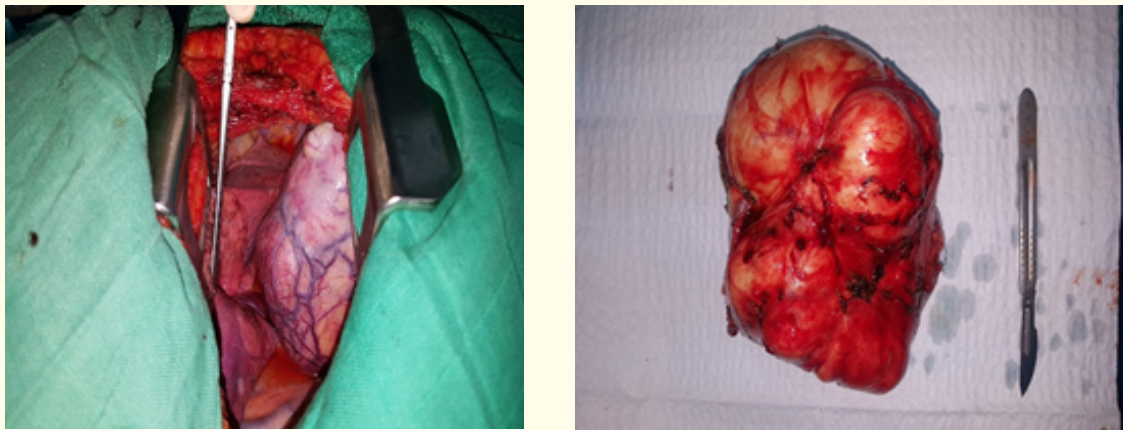


Figure 5: The thoracotomy resected a large tumor weighting 1,22 Kg.

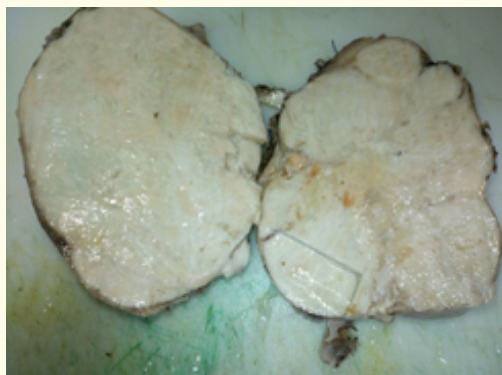
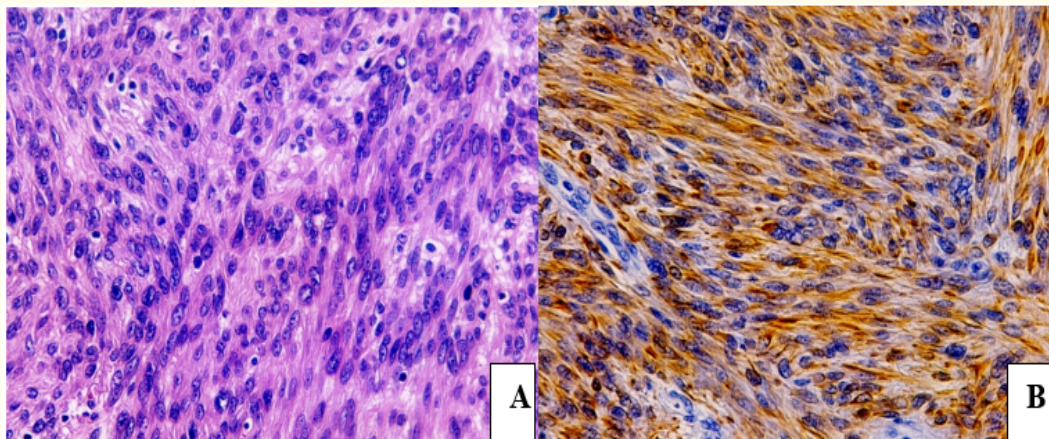


Figure 6: Macroscopic aspect of the tumor.



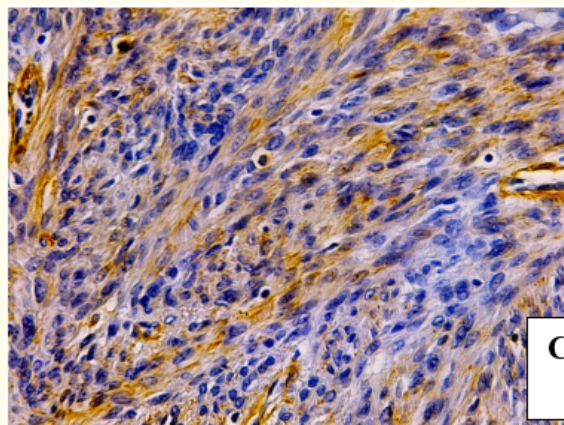


Figure 7: A benign leiomyomatous proliferation with oval nuclei and no abnormal mitosis. B: cytoplasmic expression of anti-desmine antibody by tumoral cells. C: Cytoplasmic expression of AML anti-body.

Discussion

The Solitary Fibrous Tumor of the Pleura is very rare. It has gained appropriate recognition in the last two decades as a discrete pathologic entity which represents 5% of the tumors of the pleura [2]. This type of tumor is of mesenchymal origins and not mesothelial ones. It develops from fibroblasts which are present in the sub-mesothelial tissue. It was described for the first time in 1870 by Wagner [3]. It represents 8% of all benign thoracic diseases. Its incidence is estimated at 2.8 /100 000 [4]. There is no known risk factor for this tumor which occurs at any age with a peak incidence between the fourth and sixth decade of life with a sex ratio of 1 in most studies. In 67 to 87% of cases it develops from the visceral pleura, in the two presented cases; it was at the expense of the parietal pleura which is rare (only 20% of cases) [5]. It is an asymptomatic tumor in 50% of cases. For large tumors; the clinical presentation is nonspecific, it is dominated by chest pain, cough, dyspnea, hemoptysis, and it can rarely be responsible for obstructive pneumonia [6]. Few paraneoplastic syndromes have been reported such as clubbing which may be related to an abnormal production of hepatocyte growth factor (HGF) [7], osteoarthropathy of Pierre Marie in 20% of cases (due to the production by the tumor of hyaluronic acid) and hypoglycemia (Doege-Potter syndrome) in approximately 5% of cases which is due to the secretion of an insulin-like type II substance (IGFII) [8]. These syndromes usually regress after the resection of the tumor. None of these signs were found in our patients.

The chest radiograph often shows a rounded, homogeneous and well defined opacity which is often associated with a pleural effusion. Chest CT confirms the pleural origin and the nature of the tumor, specifying its location and connections to the adjacent organs. The tumor is often well defined, homogeneous but a heterogeneous aspect can be observed which indicates the presence of a necrotic or hemorrhagic alterations or a myxoid degeneration. Calcifications can be associated in 5% of cases [9]. In case of diaphragmatic or paracardiac localization; the diagnosis can be difficult, in this case; a chest trans-esophageal sonogram can be useful. The Magnetic resonance imaging (MRI) allows a better topographic localization than the CT and specifies the fibrous nature of the tumor. The diagnosis confirmation is based on the pathological study but the biopsies are inefficient. This was the case of our first patient who had two CT guided transthoracic biopsies which did not confirm the diagnosis. In general the diagnosis is done after the surgical removal of the tumor.

Macroscopically, the tumor is usually single, encapsulated, oval and well defined. At the cutting, the section looks like a pinkish gray, sometimes with pseudo-cystic gaps. The size and the weight are variable.

Cytogenetic studies showed that tumors with a size over 10 cm are associated with chromosomic abnormalities (trisomy 8, trisomy 21) which would explain their scalability and recurrence. Pleural fibroma may be pedunculated or sessile. Sessile tumors have a higher risk of recurrence (63% vs14%) [10].

The microscopic study finds a proliferation of fusiform cells evoking scattered fibroblasts and held by a frame of collagen fibers. Normally, this proliferation doesn't contain any cytonuclear abnormalities, mitotic activity is weak and necrosis is rare.

Typically, the immunohistological study shows diffuse expression of vimentin and CD34, expression of CD99 at a variable intensity and bcl 2 protein, while epithelial markers (cytokeratin, EMA) and the protein S100 are negative [11].

According to these pathological aspects, De Perrot recommended a classification of 4 types [12]: benign pedunculated, benign sessile, malignant pedunculated and malignant sessile.

The prognosis of these tumors remains difficult to assess. England has identified the elements of bad prognosis [13]: weak limitation of the tumor, infiltration of adjacent parenchyma, the invasion of the pedicle and/or the excision margins, hypercellularity, cytonuclear abnormalities, high mitotic activity: more than 4 mitoses in 10 fields and a necrotic tumor.

The ideal treatment is the surgical removal that has to be complete with a large resection of the implantation base which can include: the lung, the chest wall or the diaphragm as was the case of our first case. For invasive forms, treatment may be completed by chemotherapy and/or radiotherapy though it did not prove much benefice [14].

Because of the uncertain evolution profile of this tumor, a clinical and radiological surveillance every 3 months during the 2 first years then on long term basis. The follow up of our patients is satisfying at 2 years for the first case and 6 months for the second one.

Conclusion

Pleural fibroma is a rare tumor. Its diagnosis is usually difficult and is confirmed by the pathology which is helped by the immunohistology. Surgery is the standard treatment; it has to be as complete as possible. The benefice of chemotherapy and radiotherapy is yet to be proven. A close surveillance after surgery is justified because of the recurrence risk.

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