Case Report

Different clinical and radiological features of solitary fibrous tumor of the pleura: Report of two cases

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ABSTRACT

We report two cases of solitary fibrous tumor of the pleura (SFTP). The first appeared in a young, new mother as a large mass in the upper lobe of the left lung that caused compression of lung parenchyma without significant respiratory symptoms but with polyarticular paraneoplastic syndrome; the other was documented by an occasional chest x-ray in a man affected by chronic obstructive pulmonary disease (COPD) as a small peripheral mass 4 years before and no longer controlled. Both patients underwent surgical resection with quick and full recovery. SFTP is a benign, slow growing neoplasm that is mostly localized. It appears in adult or elderly patients often with few symptoms. The computed tomography (CT) of the chest with contrast medium is important in order to see the shape of the mass and relationships with adjacent structures but only histology can provide the diagnosis. Surgery is the best treatment.

KEY WORDS: Diagnosis, tumor pleura, pleural disease, solitary fibrous tumor

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INTRODUCTION

Primary tumors of the pleura can be benign or malignant and they may manifest as either diffuse or localized neoplasms. Benign tumors are less common than the malignant ones, they are localized and arise from the submesothelial mesenchymal cells with a fibroblastic differentiation.[1] In most cases, the solitary fibrous tumor of the pleura (SFTP) can be asymptomatic; sometimes, it causes symptoms (as dyspnea, cough, chest pain) following the compression of airways and pulmonary parenchyma.[2] The best therapy for SFTP is complete surgical resection with a long-term follow-up.[1,3] We present two cases of SFTP in two different patients, a new mother with a polyarticular paraneoplastic syndrome and an old man with obstructive pulmonary disease who had underestimated the occasional finding on the chest x-ray of a pulmonary small peripheral mass 4 years before.

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CASE REPORTS

Case 1

A 36-year old woman was admitted to the emergency department of our hospital with widespread polyarticular pain and fever (T < 38°C) not responsive to antibiotic therapy that was started 3 weeks earlier. The patient had given birth to a baby 1 week before the onset of the symptoms. With the worsening of fever (T > 39°C), a chest computed tomography (CT) scan was performed revealing [Figure 1 a-b] a large inhomogeneous mass in the upper lobe of the left lung (18 × 11 × 9 cm). The CT scan also showed compression and atelectasis of the lung parenchyma with ipsilateral pleural effusion; there were no significant lymphadenopathies of the main stations of the thorax. Therefore, the patient was allocated to the division of pulmonology. At admission, she did not report dyspnea, cough, or chest pain but persistent polyarticular

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pain and fever; the blood tests [including Waaler-Rose and anti-neutrophil cytoplasmic antibody (ANCA)] were normal except for a mild leukocytosis (13,360 cells/mm^3) and an increase of C-reactive protein (CRP) (16,65 mg/dL) (normal value: <0,5 mg/dL). We decided to perform a CT-guided biopsy of the mass; histological examination revealed the presence of SFTP cells CD34+/B-cell lymphoma 2 (Bcl-2)+ with proliferative activity (expressed with the index of proliferation Ki-67) less than 5. Following this result, diagnosis of SFTP was processed and joint pain and fever were considered as epiphenomena of the tumor (paraneoplastic syndrome). Therefore, the pulmonary lesion was surgically removed with subsequent reexpansion of the lung.

Case 2

A 75-year old man was admitted to our hospital because of a chest x-ray that showed an opacity (5,8 × 4,9 cm) in the left lung with mild dyspnea and productive cough that began 3 months earlier. The patient had a clinical and functional diagnosis of chronic obstructive pulmonary disease (COPD), with a moderate smoking habit (30 packs/year), chronic systemic hypertension, abnormal electrocardiogram findings (Brugada syndrome), diabetes mellitus (DM) (type II). A CT scan, performed 4 years before, had shown a smaller mass (3,5 × 2,5 cm) on the left lung, next to the chest wall in comparison with that documented during hospitalization. At that time, no diagnosis was reached since the patient refused to undergo diagnostic biopsy. Routine tests were normal including blood gas analysis (pH 7,43, $PaCO_2$ 45 mmHg, PaO_2 81 mmHg, and SaO_2 96%). Due to increased mass dimension in comparison with previous CT scan, the patient was asked again to undergo a CT-guided biopsy of that lesion and he agreed [Figure 1 c-d]. Histological examination associated with the presence of cells CD34+/Bcl-2+ was compatible with a SFTP. The thoracic surgeon performed mass resection with a complete recovery of the patient.

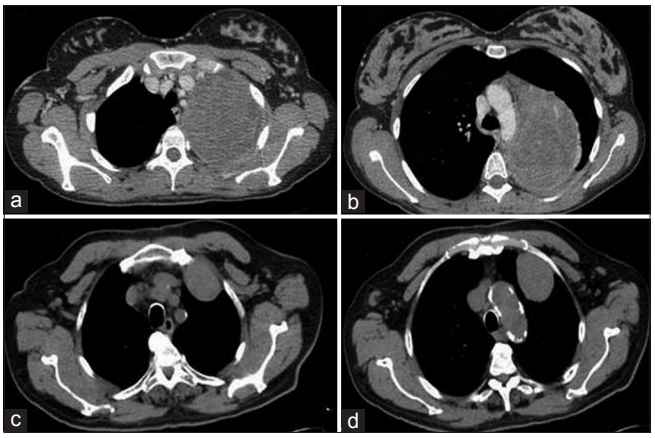

Figure 1: Case 1 - Chest CT scans at the left pulmonary apex level (a) and at the aortic arch level (b); Case 2 - Chest CT scans performed for biopsy show an intraparenchymal neoplasm near the anterior chest wall (c) and at the aortic arch level (d)

DISCUSSION

We have reported two cases of SFTP diagnosed by transthoracic biopsy: The first of a young, new mother with fever and widespread joint pain without significant respiratory symptoms and the other of a 75-year old man with cough and dyspnea attributed to his COPD and who was aware of having a pulmonary neoplasm from 4 years, the nature of which he had never ascertained. Both patients underwent surgical resection with complete recovery and without complications.

SFTP is rare benign tumor and is more frequent in the sixth/seventh decade of life in both the sexes with a modest predominance among females.[1,4]

Generally, solitary fibrous tumors of the pleura are isolated, well-defined masses while conglomerate or multifocal ones are rare presentations. Approximately, half of the masses are pedunculated, with vascular supply maintained by the pedicle.[4,5] The most common symptoms are cough, chest pain, or dyspnea.[2] Patients with implication of parietal pleura manifest chest pain more frequently than the others.[1] Sometimes, large tumors can produce symptomatic atelectasis or in rare cases hemoptysis by compression of a bronchus.[2] Furthermore, large tumors, more frequently than the small ones, produce paraneoplastic syndromes such as digital clubbing and hypertrophic pulmonary osteoarthropathy (Pierre Marie-Bamberg syndrome).[5,6] In patients with hypertrophic pulmonary osteoarthropathy, bilateral arthritis-like symptoms are common findings (e.g. joint pain, pain along the long bones, stiffness or swelling of the joints, and edema of the ankles).[2] The resection of the tumor usually solves the paraneoplastic symptoms (they generally disappear within about 2–5 months or longer after surgery) but they may appear again with recurrence of the tumor.[5,6] Our first patient showed only joint pain and fever that were unresponsive to antibiotics and no respiratory symptoms despite compression of pulmonary parenchyma caused by the large mass; on the contrary, the second patient showed respiratory symptoms that were linked to his COPD. It is worth mentioning that the slow growth of this tumor of the pleura showed a slight increase in size (3,5 × 2,5 cm vs 5,8 × 4,9 cm) in about 4 years.

After an initial approach with chest x-rays, the CT scan is the best procedure in order to study more clearly the size and location of the tumor and to plan surgery in the best way. The CT scan visualizes a small SFTP as well-defined soft tissue mass near the pleural layer,[7] usually lobular in shape, homogeneous in density and noninvasive; larger lesions, on the other hand, are generally heterogeneous and may reveal themselves as diffuse pleural opacities on CT scans.[8]

SFTP cannot be diagnosed only by radiological tools[4] and transthoracic tissue biopsy is essential to reach a diagnosis.[6] It is useful to evaluate the positivity of CD34 expression, a cell marker found in mesenchymal cells (lost in malignant tumors), presence of Bcl-2 (B-cell leukemia/

lymphoma-2 oncogene), vimentin and CD99 (positive in SFTP), cytokeratin (positive in mesothelioma and negative in SFTP).[9] Moreover, the proliferation marker Ki-67 can be used to stratify lesions according to their clinical outcome. Recently, a cutoff level of a proliferation rate of 12% (Ki-67) was suggested to distinguish benign and malignant lesions.[10]

The best and the only demonstrated effective treatment of SFTP is surgical resection removing pulmonary parenchyma compression and allowing the reexpansion of the lung.[1,3]

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Conflicts of interest
There are no conflicts of interest.

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