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Solitary Fibrous Tumor of the Pleura: A Rare Cause of Pleural Mass

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Study Design A
Data Collection B
Statistical Analysis C
Data Interpretation D
Manuscript Preparation E
Literature Search F
Funds Collection G

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Conflict of interest: None declared

Patient: Male, 63
Final Diagnosis: Solitary fibrous tumor of the pleura
Symptoms: Chronic cough
Medication: —
Clinical Procedure: CT guided biopsy • surgical resection
Specialty: Pulmonology

Objective: Rare disease

Background: A solitary fibrous tumor of the pleura is a rare but usually benign mesenchymal tumor arising from the pleura. Patients are often asymptomatic, resulting in the majority of tumors being detected incidentally on chest imaging. We present a case of a large solitary pleural tumor and review the typical radiographic and pathologic findings associated with this finding.

Case Report: A 63-year-old white man with chronic obstructive pulmonary disease (COPD) was found to have a large pleural mass on chest radiography during a pre-operative assessment. The tumor was biopsied and findings were consistent with solitary fibrous tumor of the pleura.

Conclusions: SFTPs are generally considered benign tumors although there is a risk of malignant transformation and recurrence. Imaging studies play an important role in identifying the tumor and planes of resection, and histologic diagnosis is critical in differentiating SFTP from other type of pleural masses. Surgical resection is main therapy of choice.

MeSH Keywords: Lung Neoplasms • Pleural Diseases • Solitary Fibrous Tumor, Pleural

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Background

Primary tumors of the pleura are categorized as diffuse or localized malignancies. Malignant pleural mesothelioma is a tumor with diffuse pattern and is the most common primary tumor of the pleura. Patients with malignant mesothelioma generally have a poor prognosis. On the other hand, solitary fibrous tumor of the pleura (SFTP) is a rare and often benign localized fibrous tumor, and accounts for less than 5% of all primary pleural neoplasms [1]. Terms such as localized fibrous tumor, submesothelial fibroma, and localized fibrous mesothelioma have all been used to describe this tumor. Advances in histopathology, specifically electron microscopy, and immunohistochemistry have separated SFTPs as a distinct tumor of mesenchymal origin unrelated to malignant mesothelioma [2]. Patients are often asymptomatic and diagnosed incidentally on radiographic imaging. Imaging studies aid in diagnosing SFTPs, although pathologic examination is required to differentiate benign from malignant disease. Given its rarity, case reports/series have been important in understanding prognosis and developing recommendations on management. We describe a case of SFTP and review the typical radiographic and pathologic findings required for diagnosis.

Case Report

A 63-year-old Caucasian male with a medical history significant for COPD and ischemic cardiomyopathy presented for pre-operative assessment for implantable cardioverter defibrillator (ICD) placement. He reported a chronic cough with clear sputum production and a 9 kg (20 lb) weight loss in the prior 6 months. The patient denied fever, chills, night sweats, shortness of breath, or dyspnea on exertion. He had no known exposure to chemicals or fumes but had worked in construction for 30 years with a 40 pack year smoking history. Vital signs were unremarkable with the patient breathing comfortably on room air. Physical examination was significant for prolonged expiratory phase on chest auscultation with absence of digital clubbing, lymphadenopathy and thyromegaly. Abdominal exam showed no hepatosplenomegaly. Skin exam did not reveal any rashes or nodules.

Chest radiograph revealed a well-circumscribed pleural mass centered at the left lung base (Figure 1). Subsequent computed tomography (CT) scan with intravenous contrast demonstrated an 8.0×9.5 cm well-circumscribed heterogeneous appearing mass arising from the pleura at the posterior left lung base exerting mass effect on surrounding lung parenchyma (Figures 2, 3). Laboratory data was unremarkable. CT guided biopsy of the mass was performed. Tissue pathology demonstrated spindle cells positive for CD34 and negative for epithelial membrane antigen (EMA) and pankeratin

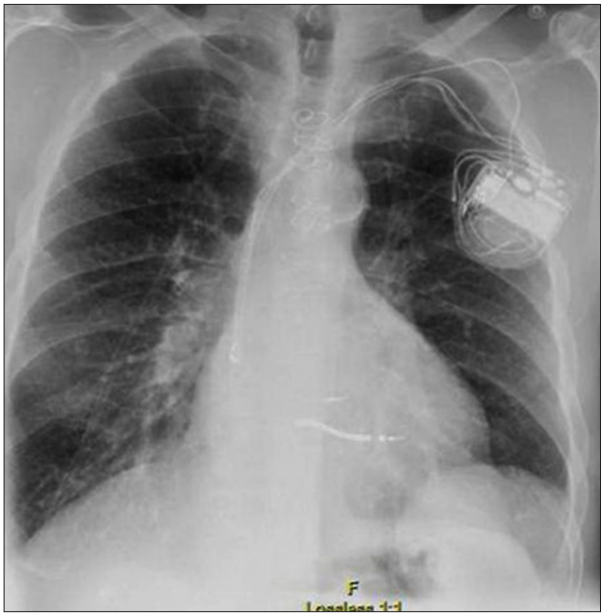

Figure 1. Chest X-ray PA view showed a well-circumscribed pleural mass centered at the left lung base.

on immunohistochemistry. Findings were consistent with SFTP (Figures 4, 5). Patient was referred for surgical resection of the mass. A complete surgical resection of the mass with clear margins was done. Final pathology also confirmed the diagnosis of solitary fibrous tumor of pleura.

Discussion

First described in 1931 by Klemperer and Rabin, SFTP is a rare mesenchymal tumor with about 900 cases reported in literature [3]. In contrast, over 3,000 new cases of diffuse mesothelioma are diagnosed yearly in the United States [4]. Peak incidence of SFTPs is between 50 and 60 years of age with equal sex predilection [5]. Environmental and occupational exposure, including tobacco abuse and asbestos, are unrelated to development of SFTP [6,7]. This tumor is usually benign, but up to 20% of cases can be malignant [8]. Further patients are often asymptomatic and when symptoms are present, they are nonspecific (chest pain, cough or shortness of breath). Asymptomatic patients are detected by incidental radiographic studies [9]. SFTP can be associated with paraneoplastic syndromes. Hypertrophic pulmonary osteoarthropathy (Pierre Marie-Bamberger syndrome) is the most common paraneoplastic syndrome in SFTPs and occurs in 20% of patients [6]. Patients with hypertrophic pulmonary osteoarthropathy often have larger tumors (diameter greater than 7 cm), with associated bilateral arthralgia and gynecomastia [10]. Labile hypoglycemia (Doege-Potter syndrome) occurs in less than 5% of patients and is due to tumor production of insulin like growth factors [6]. Rapid relief of hypoglycemia occurs

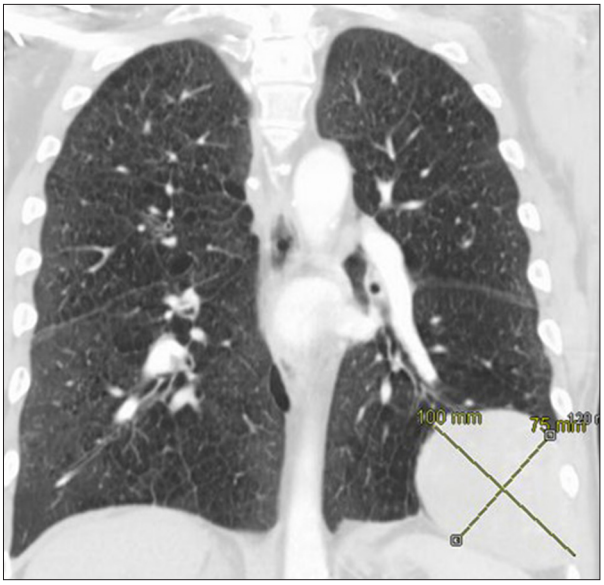

Figure 2. CT chest showing left pleural mass.

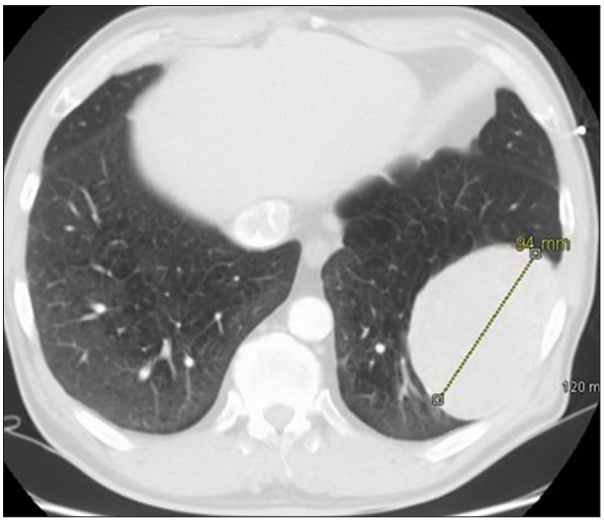

Figure 3. CT chest showing left pleural mass (coronal view).

after tumor resection. Clubbing is a frequent physical exam finding found in patients with hypertrophic pulmonary osteoarthropathy [6]. A large tumor may also cause wheezing or diminished breath sounds in the respective lung fields.

Imaging studies aid in diagnosing SFTPs. On chest radiography, these tumors appear as well defined, round/oval, smooth masses, located peripherally with reported changes in shape based on patient positioning or respiration during imaging. SFTP are usually located in the middle or inferior hemithorax. A CT scan with contrast will demonstrate a heterogeneous tumor with larger masses forming acute angles at pleural interface compared to smaller tumors that form obtuse angles [11]. Areas of necrosis can be identified as areas of low attenuation. MRI finding of SFTP usually shows low or intermediate signal

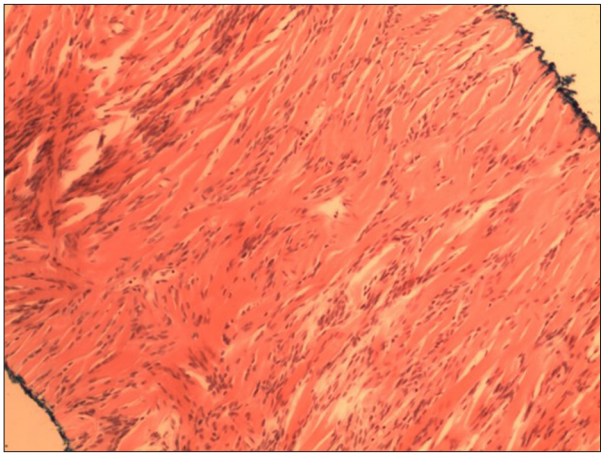

Figure 4. Hematoxylin and Eosin (H&E) stain showing low grade spindle-shaped tumor cell in haphazard pattern (low power).

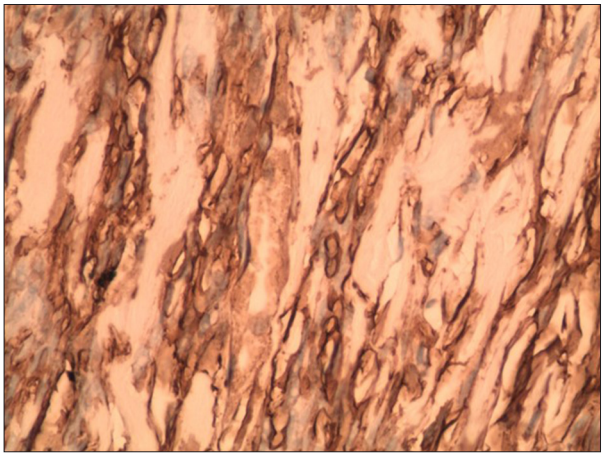

Figure 5. Immunohistochemistry staining showing positive for CD 34.

intensity on both T1 and T2-weighted images, which is correlates with high cellularity and abundant collagen. Angiography aids in evaluating the blood supply to the tumor, which typically enters from pedicles [3]. Lastly, ultrasonography may be helpful for large tumors in the inferior hemithorax. Imaging studies alone cannot distinguish between benign and malignant SFTPs and histologic examination is required for definitive diagnosis. Increased heterogeneity, necrosis, cystic changes, size greater than 10 cm and associated pleural effusion on CT scan may suggest malignant transformation [6,12]. Further, imaging modalities such as ^{18}FDG PET (Fluorodeoxyglucose positron emission tomography) scan, MRI (magnetic resonance imaging) scan or ultrasound add little to identify malignant transformation.

Although imaging studies may suggest the presence of SFTP, pathologic diagnosis is required. SFTP arise from fibroblasts and most are benign. Classic histological characteristic of SFTPs

on microscopic examination reveals low grade spindle-shaped tumor cells that arrange in a haphazard pattern, (Figure 2). Histological features can be used to identify SFTP that are malignant. There are 4 criteria that define SFTP as malignant: (1) histologic presence of greater than 4 mitoses per 10 high power fields (2) evidence of necrosis, (3) nuclear overlapping showing hyper-cellularity (4) nuclear atypia [13]. Further, immunohistochemical staining is crucial for diagnosis. Given their mesenchymal origin, SFTP are positive for vimentin and negative for keratin. Positive CD34, CD99 and bcl-2 markers also support the diagnosis. These staining patterns aid in distinguishing SFTP from other pleural tumors.

Given their rarity, limited data exists on the treatment of SFTPs and management is based on expert opinion. Surgical resection with clear margins remains the predominant mode of therapy. Prognosis for malignant SFTP is favorable with surgical resection, with reported overall 5 year survival of 81% [14]. For recurrent tumors, resection and local radiation in selected candidates should be pursued.

Conclusions

SFTP are generally considered benign tumors although malignant transformation occurs in 20% of patients. Imaging studies play an important role in identifying the tumor and planes of resection, and histologic diagnosis is critical in differentiating SFTP from other type of pleural masses. Surgical resection is main therapy of choice.

Conflicts of interest

The authors have no conflicts of interest.

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