Solitary fibrous tumor of pleura: a case report and review of clinical, radiographic and histologic findings

Vishal K. Agarwal[1*], Benjamin E. Plotkin[1], Donny Dumani[2], Samuel W. French[2], Ronald Becker[1], Paul Lee[1]

1. Department of Radiological Sciences, Harbor-UCLA Medical Center, Torrance, California, USA

2. Department of Pathology and Laboratory Medicine at Harbor-UCLA Medical Center, Torrance, California, USA

* **Correspondence:** Vishal K. Agarwal, M.D., Department of Radiological Sciences, Harbor-UCLA Medical Center, 1000 West Carson Street, PO Box 2910, Torrance, CA 90509, USA (UCLAVKAgarwal@gmail.com)

Radiology Case. 2009 May; 3(5):16-20 :: DOI: 10.3941/jrcr.v3i5.200

ABSTRACT

We present a case of solitary fibrous tumor of pleura (SFTP) in a 63-year-old male. Clinical manifestations of this entity, including paraneoplastic syndromes, are discussed, albeit absent in our presented case. Furthermore radiographic findings and pathologic correlations are provided. SFTP remains a rare neoplasm for which radiographic features are suggestive yet nonspecific, and immunohistochemistry remains as the diagnostic method of choice.

CASE REPORT

CASE REPORT

A 63-year-old male, with a history of right middle lobe mass diagnosed at an outside hospital two years previously, presented to the emergency department with worsening right-sided chest pain. The patient's pain began approximately two years prior to this admission and was characterized as intermittent, generally lasting for a few seconds and occasionally radiating to the back. There were no exacerbating or alleviating factors. The patient reported recent onset of associated nausea and decreased appetite, but denied any shortness of breath, cough, or hemoptysis. The patient had no history of thoracic surgery but recalled a transthoracic biopsy of the right middle lobe mass previously, with no diagnosis provided. The patient denied any tobacco, alcohol, or drug use. There was no history of occupational exposure risk, including asbestos.

Physical examination revealed absent breath sounds and dullness to percussion over the anterior inferior right chest wall. There was no cyanosis or clubbing. Laboratory test findings were within normal limits. Plain chest radiographs demonstrated a well-demarcated mass in the region of the right middle lobe (Fig. 1). Computed tomography (CT) of the chest with contrast revealed a well-circumscribed, predominantly homogenously solid, pleural-based mass. The mass had distinct borders and demonstrated obtuse angles with the abutting chest wall. There was no internal calcification, pleural effusion, fibrosis, mediastinal adenopathy, vascular involvement, satellite lesion or mass affect. There was no invasion of adjacent ribs or lung parenchyma (Fig. 2).

Based on the clinical presentation and radiographic findings, a differential diagnosis of benign and malignant pleural lesions was considered, including solitary fibrous tumor of pleura, pleural mesothelioma, and other sarcomatous lesions. Differentiation of these lesions required further analysis via histology and immunohistochemistry. Consideration of bronchogenic carcinoma was largely excluded due to lack of tobacco exposure, benign appearance of the lesion (as described above), and lack of lymphadenopathy, associated atelectasis or local invasion. Metastatic process was excluded as no primary malignancy was identified during the patient's work up.

CT-guided transthoracic core biopsy revealed characteristic "patternless" architecture, varied cellular areas and spindle-shaped cells with scant cytoplasm and ill-defined borders (Fig. 3). Based on the final histology and positive CD34 staining (Fig. 4), a diagnosis of solitary fibrous tumor of pleura was made.

DISCUSSION

Solitary fibrous tumor of pleura (SFTP) (previously known as benign or localized mesothelioma; subserosal, submesothelial, or pleural fibroma) is a rare neoplasm occurring in approximately 2.8 cases per 100,000 persons, as recorded by the Mayo Clinic Registry (1). Recent advances in electron microscopy and immunohistochemistry demonstrate a mesenchymal rather than mesothelial origin. While the majority of solitary fibrous tumors occur within the pleura, such tumors have also been described in other sites with mesenchymal tissue, including subcutaneous tissues, head and neck (orbit, salivary glands, thyroid), thorax (lung, pericardium), retroperitoneum (adrenal), and abdominal cavity (liver, GI tract, urinary bladder) (1,2,3). No link has been established between smoking or environmental exposure (including asbestos), although cases of SFTP developing after radiation therapy to the chest wall have been reported (4).

Although SFTP has been described in all age groups, peak incidence is in the sixth and seventh decades of life with equal gender and ethnic distribution. The majority of patients are asymptomatic, with diagnosis based on incidental finding. Larger benign lesions may result in compression of adjacent structures, vague chest pain, dyspnea or cough. Rarely, patients may present with associated pleural effusion, hemoptysis and obstructive pneumonitis. Digital clubbing and hypertrophic pulmonary osteoarthropathy (collectively referred to as Pierre-Marie-Bamberg syndrome) and refractory hypoglycemia (Doege-Potter syndrome) have been reported to occur in ten to twenty percent and five percent of patients, respectively (5,6). These paraneoplastic syndromes, due to abnormal productions in hepatocyte growth factor or excessive release of hyaluronic acid and the secretion of insulin-like-growth factor II, respectively, typically resolve following resection (1,7).

Radiographically, benign and malignant SFTP typically appear as well-defined, homogeneous, and spherical masses on plain radiographs. These tumors may occasionally be attached to pleura by a narrow pedicle, a finding considered pathognomonic. If pedunculated, SFTP lesions may change intrapleural location with changes in patient positioning. Pleural effusions may be rarely associated with malignant forms of SFTP. Classically, pleural-based lesions should form obtuse angles with the chest wall. Large or pedunculated lesions, however, may demonstrate acute angles and distinction from intrapulmonary masses may be difficult (5,8,9).

Computed tomography (CT) usually demonstrates a smoothly marginated, pleural based soft tissue mass with either uniform soft tissue attenuation or inhomogeneous enhancement secondary to myxoid degeneration, hemorrhage or necrosis. Regardless of benign of malignant histology, calcifications may be present, and are not specific. While not utilized in this case, magnetic resonance imaging (MRI) may provide some utility in differentiating benign and malignant forms of SFTP. Typically, fibrous tissue in either malignant or benign SFTP should exhibit low signal intensity on T1-weighted sequencing. On T2-weighted imaging, however, mature fibrous tissue containing few cells and abundant collagen stroma has low intensity, whereas malignant degeneration demonstrates high signal intensity secondary to increased edema, cellularity and vascularity. Unfortunately, benign SFTP may have areas of increased T2-weighted signal due to myxoid degeneration, hemorrhage or necrosis, and may not be differentiable from malignant SFTP based on magnetic resonance imaging (9). Despite advances in imaging modalities, the radiographic characteristics of SFTP remain nonspecific, and further assistance from definitive immunohistochemistry is needed.

Microscopically, SFTP demonstrates a "patternless" architecture consisting of alternating hypocellular and hypercellular areas separated by bands of hyalinized collagen and vessels resembling those of hemangiopericytoma. The individualized tumor cells are spindle-shaped with scant cytoplasm, indistinct borders, and dispersed chromatin in a vesicular nucleus. Mitoses are rarely seen, exceeding no more than three mitoses per 10 high-power fields (2). Malignant SFTP are predominately hypercellular showing focally moderate to marked cellular atypia, tumor necrosis, mitoses greater than four per 10 high-power field, and +/- infiltrative margins (10). Unfortunately, malignant potential may not always correlate with histologic findings, and some cases of SFTP may progress to the malignant type of disease (11).

Immunohistochemistry is essential in differentiating SFTP from mesotheliomas and other sarcomatous lesions. By definition, SFTP is vimentin positive and keratin negative. Furthermore, in contrast to mesotheliomas and other sarcomatous lesions, confirmation of SFTP should be gained from specific immunohistochemical stains, including CD34 (90-95%) and CD99 (70%) positivity, regardless of benign or malignant nature. Of note, a small percentage of malignant SFTP may be CD34 negative, and additional tumor markers may be utilized for diagnosis (2).

Approximately ten to fifteen percent of SFTP are malignant, eventually recurring locally or via metastatic disease. Metastases typically spread hematogenously, and are located (in order of frequency) in the lung, liver, central nervous system, spleen, peritoneum, adrenal gland, gastrointestinal tract, kidney and bone (5,8). Clinically, patients with metastatic or malignant forms of SFTP are more likely than benign forms to present with symptoms as described previously. CT or MR heterogeneity, large tumor diameter or an associated pleural effusion may be suggestive of malignancy. These findings, however, have been identified in benign lesions as well (8,9). Unfortunately, malignant transformation is unpredictable, and may not always correlate with histologic findings. Complete surgical excision followed by pathologic examination are required for differentiation between benign and malignant forms.

Complete surgical resection remains the treatment of choice for benign and malignant SFTP. Preoperative imaging, namely CT and MRI, provides critical information in regards to tumor characteristics, location, invasion into adjacent structures and tumor vascularity. Whereas pedunculated tumors may be safely resected, larger sessile tumors may pose

more difficult due to extensive adhesions. These may require lobectomy or pneumonectomy in order to achieve adequate resection. Adjuvant therapy has not been well defined due to the limited number of cases, but is thought to be of use in cases of malignant SFTP. Recurrence has been reported in up to 17 years post resection, and carries a high risk in patients with malignant sessile SFTP. For malignant tumors, recommendations for post resection follow-up include half-yearly plain film or computed tomography scan of the chest for the first two years, followed by yearly examinations (1,12).

TEACHING POINT

The diagnosis of solitary fibrous tumor of pleura, albeit rare, may be suggested by clinical and radiographic findings. Since radiographic features of these tumors are nonspecific, confirmation should be performed using immunohistochemistry, as this remains the diagnostic method of choice.

ABBREVIATIONS

SFTP = Solitary fibrous tumor of pleura
CT = Computed tomography
MRI = Magnetic resonance imaging

REFERENCES

1. Perrot M, Fischer S, Brundler MA, Sekine Y, Keshavjee S. Solitary fibrous tumor of the pleura. Ann Thorac Surg. 2002; 74:285-293.
2. Brunnemann RB, Ro JY, Ordonez NG, Mooney J, El-Naggar AK, Avala AG. Extrapleural solitary fibrous tumor: a clinicopathologic study of 24 cases. Mod Pathol, 1999; 12:1034-1042.
3. Mentzel T. Solitary fibrous tumor: clinicopathological, immunohistochemical, and ultrastructural analysis of 12 cases arising in soft tissues, nasal cavity, and nasopharynx, urinary bladder and prostate. Virchows Arch. 1997; 430:445-453.
4. Bilbey J, Müller N, Miller R. Localized fibrous mesothelioma of pleura following external ionizing radiation therapy. Chest 1988; 94:1291-1292.
5. England D.M., Hochholzer L., McCarthy M.J. Localized benign and malignant fibrous tumors of the pleura: a clinicopathologic review of 223 cases. Am J Surg Pathol 1989; 13:640-658.
6. Perrot M, Kurt AM, Robert JH, Borisch B, Spiliopoulos A. Clinical behavior of solitary tumors of the pleura. Ann Thorac Surg. 1999; 67:1456-1459.
7. Briselli M., Mark E.J., Dickersin G.R. Solitary fibrous tumors of the pleura: eight new cases and review of 360 cases in the literature. Cancer 1981; 47:2678-2689.
8. Vallat-Decouvelaere AV, Dry SM, Fletcher CDM. Atypical and malignant solitary fibrous tumors in extrathoracic locations: evidence of their comparability to intra-thoracic tumors. Am J Surg Pathol. 1998; 22:1501-1511.
9. Rosado-de-Christenson ML, Abbot GF, McAdams HP, Franks TJ, Galvin JR. From the Archives of the AFIP: Localized Fibrous Tumors of the Pleura. Radiographics. 2003; 23:759-783.
10. Fukunaga M, Naganuma H, Ushigome S, Endo Y, Ishikawa E. Malignant solitary fibrous tumor of the peritoneum. Histopathology. 1996;28: 463-466.
11. Hasegawa T, Matsuno Y, Shimoda T, Hasegawa F, Sano T, Hirohashi S. Extrathoracic solitary fibrous tumor: their histological variability and potentially aggressive behavior. Hum Pathol 1999; 30:1464-1473.
12. Rena O, Filosso P, Papalia E, et al. Solitary fibrous tumor of the pleura: surgical treatment. Eur J Cardiothorac Surg 2001;19:185–189.

FIGURES

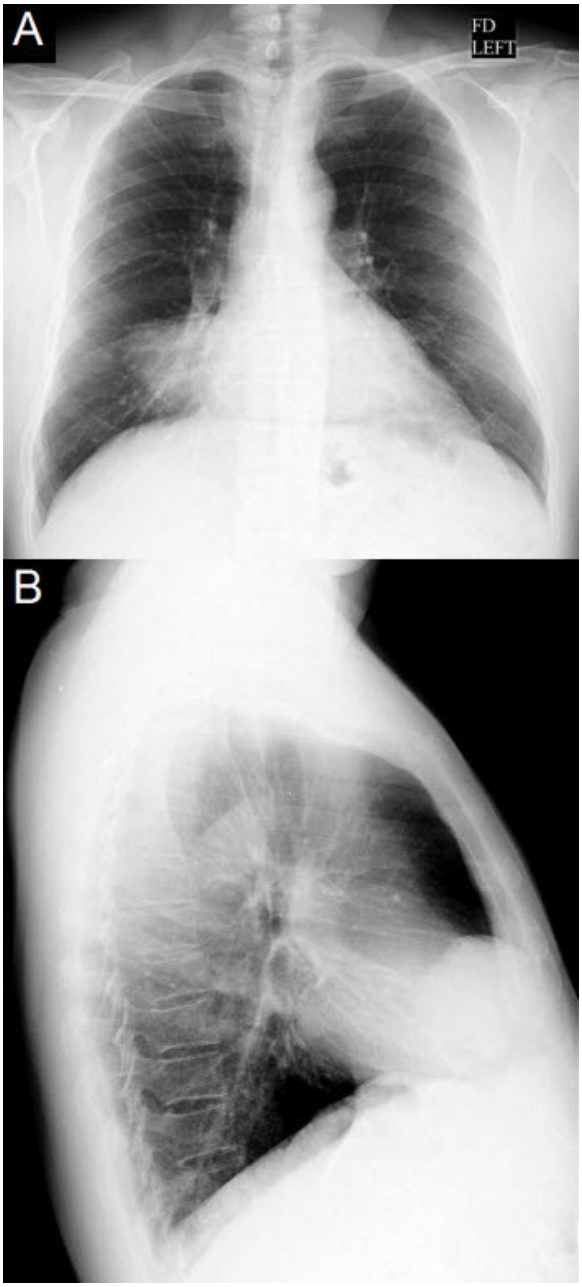

Figure 1: 63-year-old male with solitary fibrous tumor of pleura. PA (A) and lateral (B) chest radiographs demonstrate a well-circumscribed, homogenous density with smooth margins abutting the anterior chest wall without silhouetting of the right heart border. On lateral view (B), the mass appears to form an acute angle with the adjacent chest wall.

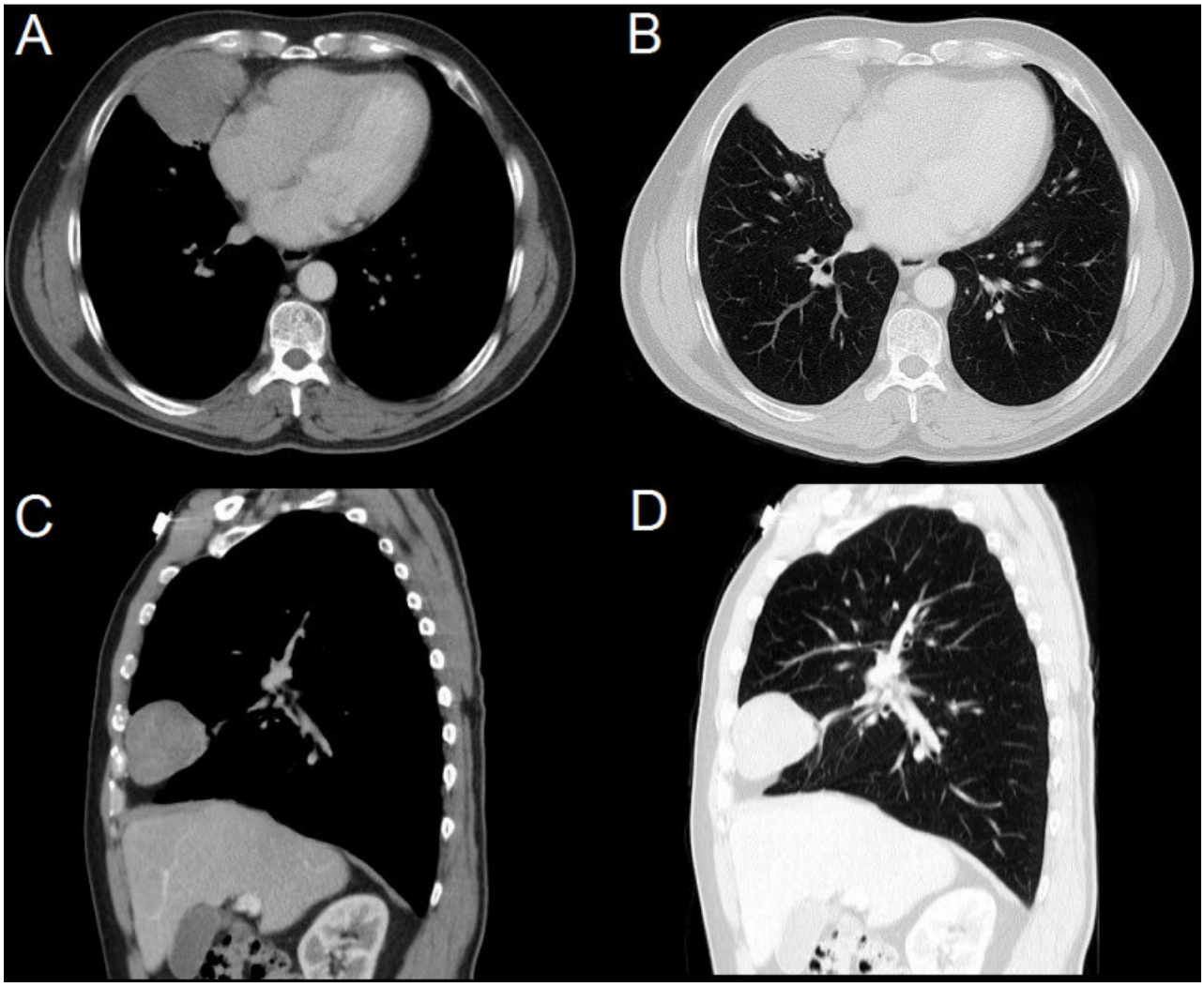

Figure 2: 63-year-old male with solitary fibrous tumor of pleura. Contrast enhanced CT axial (A+C) and sagittal (B+D) images, displayed in both soft tissue and lung windows, illustrate a well-circumscribed, predominantly homogenous, pleural-based mass adjacent to the right heart border. There is minimal peripheral enhancement. Note the distinct borders and sometimes obtuse angles with the chest wall.

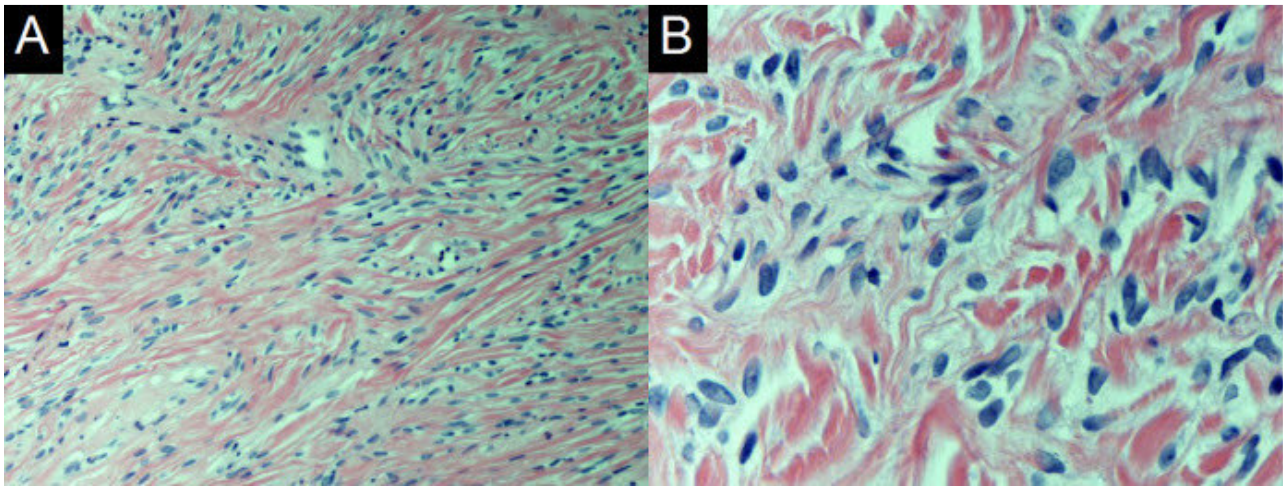

Figure 3: 63-year-old male with solitary fibrous tumor of pleura. Hematoxylin and eosin stains. Low power magnification (347X, A) demonstrates "patternless" architecture with varied cellular areas separated by hyalinized pink collagen. Under high power magnification (1040X, B), spindle-shaped cells with little cytoplasm and ill-defined borders are identified. Note the absence of mitotic figures.

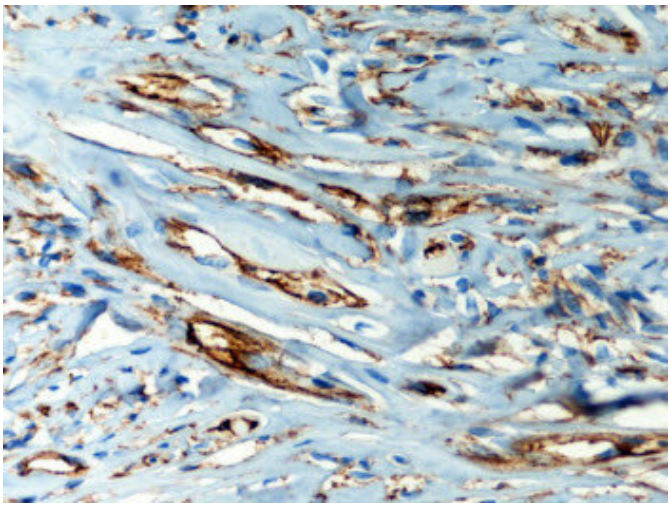

Figure 4: 63-year-old male with solitary fibrous tumor of pleura. CD34 staining demonstrates diffuse positive staining at 1040X high power magnification.

KEYWORDS

Solitary fibrous tumor, pleura, computed tomography (CT), magnetic resonance imaging (MRI), CD34, immunohistochemistry