

## Case Reports

# Solitary Fibrous Tumor Arising from the Mediastinal Pleura: Case Report

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### Introduction

Solitary fibrous tumor is a rare mesenchymal tumor affecting mainly the pleura, the visceral pleura is the most commonly affected. It is usually a benign tumor but may have an unpredictable behavior and a malignant potential, complete excision is the standard of treatment. We present a case of solitary fibrous tumor of the mediastinal pleura which is a very rare incidence. Also, we reviewed the literature related to the condition.

**Keywords:** SFT (Solitary Fibrous Tumor), CT (Computed Tomography Scan).

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### Case Report

A 55 year female, known to have hypertension, hypothyroidism, renal stones and common bile duct stones. She complained of shortness of breath on exertion. Chest X-ray showed a chest mass in relation to the left lower lobe of the lung (Figure1). A CT scan of the chest showed the mass with secondary collapse of the left lower lobe (Figure2). Bronchoscopy showed a bulge from outside on a basal segment. CT guided biopsy of the mass showed Solitary Fibrous Tumor (SFT) with no evidence of malignancy. The patient underwent a left thoracotomy, intraoperatively the mass was found to be arising from the mediastinal pleura was and attached to the pericardium medially and the upper and lower lobes laterally. It was an encapsulated, well circumscribed and large mass about 12\*10\*8 cm in size and weighs 414 gm.

Step sectioning of the mass showed multiple small cysts and bland looking spindle cells haphazardly arranged, immunohistochemical staining was positive for CD34, BCG, and CD99 and negative for Claretine and Cytokeratine. The diagnosis of benign solitary fibrous tumor was confirmed.



**Figure (1):** Chest X ray of the patient showing the left sided chest mass.

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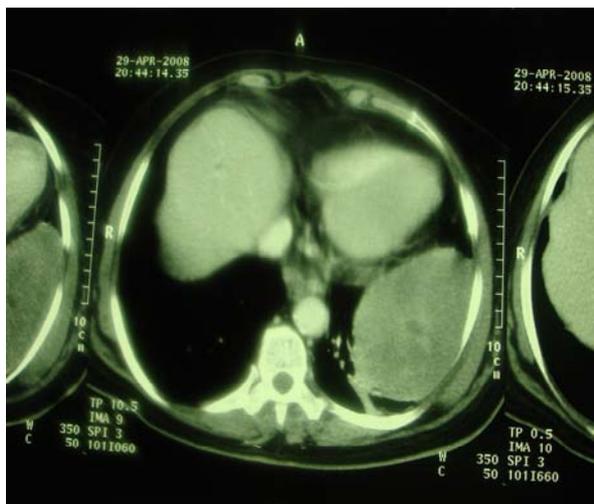
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**Figure (2):** CT chest of the patient showing the intrathoracic tumor and the left lower lobe collapse. The mass is large in size and is related to the mediastinum and chest wall.

### Discussion

SFT was first described by Klemperer and Rabin in 1931 as a form of mesothelioma. In the early seventies, Hernandez and Fernandez proved the SFT<sup>1</sup> are derived from the fibroblasts adjacent to the lining of the visceral pleura. They are unrelated to mesothelioma or asbestos exposure. Solitary benign fibrous tumors of the pleura arise as a solitary mass from the visceral pleura in 80% of patients, and from parietal pleura in 20%.<sup>2</sup> SFTs have been reported to arise in other sites including mediastinum, lung parenchyma, thyroid, liver, retroperitoneum, orbit, and paranasal sinuses.<sup>3,4</sup>

SFTs have been described in all age groups, but they peak in the sixth and seventh decades of life; they have an even distribution between men and women.<sup>2, 5</sup> These tumors are usually an incidental finding in 50 % of cases.<sup>6</sup> Digital clubbing and hypertrophic pulmonary osteoarthropathy (Pierre- Marie- Bamberg syndrome) have been described in 10% to

20% of patients with SFT.<sup>2, 8</sup> In less than 5% of patients, SFT can cause refractory hypoglycemia (Doege- Potter syndrome) due to secretion of insulin-like growth factor II.<sup>2,9</sup>

Malignant SFTs account for approximately 12% of all fibrous tumors of the pleura. They are rarely pedunculated and may arise from the visceral or parietal pleura.<sup>8</sup> Their size is usually greater than 8 cm, and the tumors often present in atypical locations such as the parietal or mediastinal pleuras, lung fissures, and intraparenchymal sites.<sup>5, 10</sup> Histological features usually include increased cellularity, pleomorphism, and more than four mitosis per 10 HPF.<sup>2</sup> All solitary fibrous tumors of the pleura were strongly positive for vimentine and CD34, but remained negative for keratin.<sup>6</sup> De Perrot et al. (2002) classified SFTs into five stages (stage 0 - IV) by the morphologic and histological indicators<sup>11</sup> (table 1).

**Table (1):** Classification of Solitary Fibrous Tumors of the Pleura.

<u>Stage</u>	<u>Characters</u>
<b>Stage 0</b>	<i>Pedunculated tumor without signs of Malignancy</i>
<b>Stage I</b>	<i>Sessile or "inverted" tumor without signs of malignancy</i>
<b>Stage II</b>	<i>Pedunculated tumor with histological signs of malignancy</i>
<b>Stage III</b>	<i>Sessile or "inverted" tumor with histological signs of malignancy</i>
<b>Stage IV</b>	<i>Multiple synchronous metastatic tumors</i>

Benign SFT may rapidly enlarge, and may transform into the malignant form. A complete excision is recommended for all patients. Broad-based neoplasm requires a lobectomy if originating from the visceral pleural and a chest wall resection if originating from the parietal pleura. Pedunculated lesions can be safely treated with a wedge resection if originating from the visceral pleura or with a local resection with

extrapleural dissection if originating from the parietal pleura, provided that the surgical margins are negative.<sup>12</sup> Postoperative adjuvant therapy with radiotherapy, chemotherapy, or both has been sporadically used, but its benefit remains unproved.<sup>7</sup>

Video Assisted Thoracoscopic Surgery (VATS) is useful to excise pedunculated tumors of the visceral pleura and to obtain a more precise view of the resection margin of tumors of the parietal pleura.<sup>13</sup>

Recurrence after complete resection is possible in benign and malignant variants; therefore, long-term annual follow-up with chest roentgenograms is highly recommended.

### **Conclusion**

SFT of the pleura is a rare tumor, most commonly benign arising from the visceral pleura; tumors arising from the mediastinal pleura are extremely rare. The proposed staging is dependent on histological and morphological features. Benign tumors may have a malignant potential. The treatment is complete surgical excision, and VATS is increasingly being recommended. Neoadjuvant and adjuvant chemotherapy and radiotherapy are not well studied because of the limited number of cases.

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## تقرير حالة ورم ليفي مفرد نشأ من الجنبه المنصفية

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### الملخص

الورم الليفي المفرد عبارة عن ورم ينشأ من اللحمية المتوسطة ويصيب عادة غشاء الجنبه، والجنبه الحشوية هي الاكثر اصابة. وهذا الورم يكون حميداً في العادة، الا ان سلوكه غير متنبأ به ويمكن ان يتحول الورم الحميد الى خبيث. ويعتبر الاستئصال الكامل هو العلاج الاساسي. في هذا التقرير نقدم حالة ورم ليفي مفرد اصاب الجنبه المنصفية وهذا نادر الحدوث. وقد قمنا بمراجعة الدراسات والابحاث المتعلقة بالحالة المرضية.

الكلمات الدالة: ورم ليفي مفرد ، تصوير مقطعي محوسب.