Solitary Fibrous Tumor of the Mediastinum: A Rare Tumor at a Rare Site

Niaz Hussain Soomro[1], Salwa Pervaiz[2], Ammad Hussain[3], Syed Khalid Ali[4]

[1]*Assistant Professor, Department of Thoracic Surgery, Ojha Institute of Chest Diseases & Dow University Hospital, Dow University of Health Sciences, Karachi, Pakistan*
[2]*House Officer, Department of Thoracic Surgery, Ojha Institute of Chest Diseases & Dow University Hospital, Dow University of Health Sciences, Karachi, Pakistan*
[3]*Resident, Department of Thoracic Surgery, Ojha Institute of Chest Diseases & Dow University Hospital, Dow University of Health Sciences, Karachi, Pakistan*
[4]*Senior Medical Officer, Department of Thoracic Surgery, Ojha Institute of Chest Diseases & Dow University Hospital, Dow University of Health Sciences, Karachi, Pakistan*

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Correspondence to: Dr Niaz Hussain Soomro

Address: Department of Thoracic Surgery, Ojha Institute of Chest Diseases & Dow University Hospital, Dow University of Health Sciences, Karachi, Pakistan

E-mail: soomroniaz@yahoo.com, niaz.hussain@duhs.edu.pk,

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ABSTRACT

45 year old male presented with cough, chest pain and dyspnea and was found to have a mediastinal mass extending into the left hemi-thorax. At thoracotomy a 30 x 32 x 10 cm well-circumscribed mass was found consisting of spindle cells. Immuno-histochemical stains were positive for Cluster of Differentiation (CD34), Anti Smooth Muscle Antibody(ASMA) patchy, but were negative for cytokeratin AEl/AEB, desmin, S-100, Cluster of Differentiation (CD31), Epithelial Membrane Antigen (EMA). Thus a diagnosis of solitary fibrous tumor was confirmed. The differential diagnosis of a mediastinal tumor are diverse while the clinical presentations are predominantly similar.

Keywords: Mediastinal Tumor; Mesenchymal Tumors; Solitary Fibrous Tumor

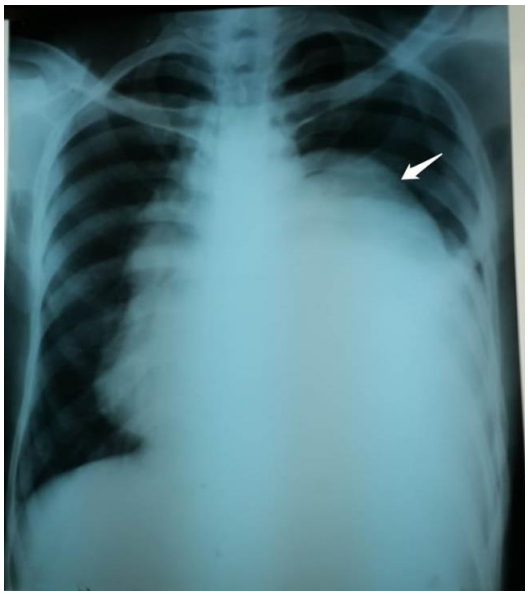
INTRODUCTION

The Uncommon neoplasms (<10%) of the mediastinum include neuroendocrine carcinomas, primary carcinoma of the thymus, lymphomas, germ-cell tumors, and neurogenic, endocrine, and mesenchymal tumors [1]. Solitary fibrous tumors (SFTs) of the mediastinum are rare neoplasms first reported by Witkin and Rosai [2] in 1989. Solitary fibrous tumors are usually of pleural origin but may be found in various parts of the body and are not limited to mesothelial-lined surfaces [3]. SFTs rarely metastasize [4] but need early recognition because wide and complete surgical excision may be curative in most cases. We present a rare case of giant solitary fibrous tumor of the mediastinum in a 45 year old male.

CASE REPORT

A 45 year old male presented in the out-patient clinic with complaints of progressive exercise-induced dyspnea, dry cough and constant, diffuse, left-sided chest pain for one year.
On physical examination, patient had reduced left chest movements with a slightly right-deviated trachea. He had reduced tactile vocal fremitus on the left chest, as well as reduced breath sounds and vocal resonance in the middle and lower parts of anterior, lateral and posterior chest wall.
Apart from hemoglobin of 9.2mg/dL and an Mean Corpuscular Volume(MCV) of 73 fl, his cell counts, electrolytes and coagulation profile were normal, as was his blood glucose level. His chest radiograph (Figure 1) showed a homogenous opacity in the left middle and lower zone, right-sided mediastinal shift, and obliterated costo- and cardio-phrenic angles. Computed tomography of the chest (Figure 2) showed marked left sided widening of the mediastinum with presence of predominantly heterogeneous mass lesion demonstrating extension into the left thoracic cavity up to the left lateral chest wall. The mass measured 14.5 x l6.7cm in its axial dimension with a craniocaudal extension of 25.1 cm, extending from the upper border of the manubrium sterni to and abutting the left hemidiaphragm with its eversion, causing right displacement of the mediastinum. It caused compression of the aortic arch, pulmonary trunk and veins, and the left atrium and ventricle. There was no pleural effusion, pleural thickening or plaque pneumothorax hilar, mediastinal,

Figure 1: Chest radiograph showing a hom -ogenous opacity in the left middle and lower zone (arrow), with the costo and cardio-phrenic angles obliterated. There is shifting of the mediastinum to the right side



supraclavicular or axillary lymphadenopathy. A transcutaneous ultrasound-guided trucut biopsy of the tumor showed spindle cell tumour, consistent with SFT.

After pre-op work-up and consent, left thoracotomy and resection of the tumor was performed and solid tumor, measuring 32 x 30 x 10 cm, was removed in multiple pieces (Figure 3). In contrast to the CT findings, the tumor had not infiltrated adjacent structures and the lung expanded up to the chest wall after resection. On gross appearance, tumor was partially encapsulated multinodular firm with its cut surface being tan white to brown lobulated surface, and areas of hemorrhage and myxoid degeneration. Its microscopic examination revealed spindle cells arranged in short fascicles and storiform pattern. Cells had round to oval hyperchromatic nuclei with scant cytoplasm. The neoplastic cells were arranged in collagenized stroma. Areas of hyalinization were also seen. Approximately 5 mitoses/10 HPF were seen. Special stain PAS+/-D did not highlight glycogen in the neoplastic cells. Immuno-histochemical stains performed were positive for CD34, ASMA (patchy), but were negative for cytokeratin AEl/AEB, desmin, S-100, CD31, and EMA.

Our patient made an uneventful recovery and was discharged on the seventh post-operative day. At six months of follow-up, patient was symptom-free and showed no evidence of recurrence of disease.

DISCUSSION

Two decades ago, extra-pleural SFT were virtually undiagnosed [5,6]. However, SFTs are now increasingly being reported in various sites of the body [3,6] and have become more common than the previously more frequent pleural counterpart [6]. The case at hand was one such tumor, arising from the mediastinum to extend into and occuping most of the left hemi-thorax, as noted on the CT scan.

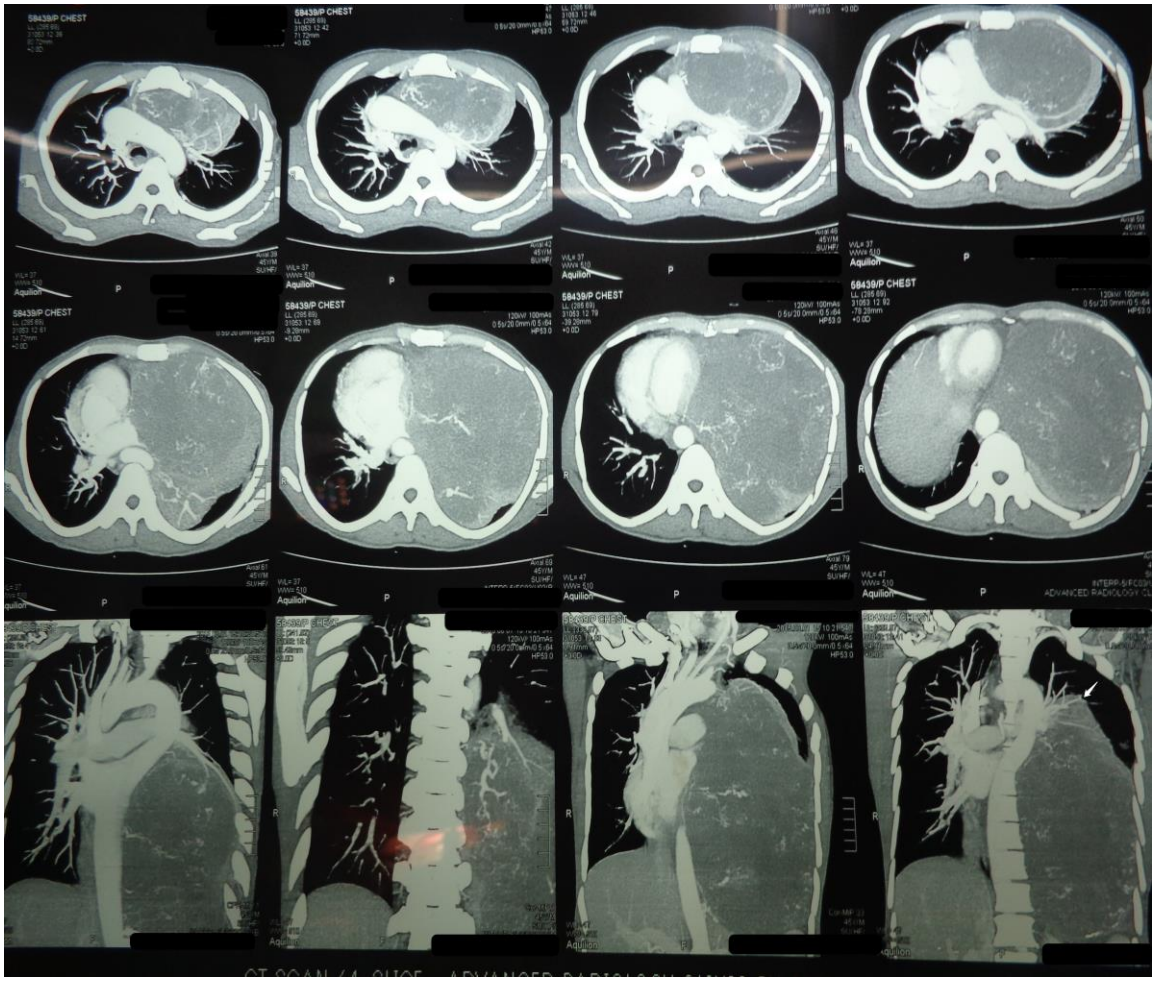
Mediastinal SFTs are rare neoplasms that present with cough, chest pain and dyspnea, but may also be incidental findings in an asymptomatic individual [6].

Witkin and Rosai have reported in their case series that the age range of patients is from 27 to 70 years, with a median of 54 years [2]. Witkin and Rosai also reported a slight male predominance in their cases. The majority of the patients reported dyspnea, cough and pleurodynia as the presenting symptoms [2, 9]. A rare sign of a mediastinal SFT is hypoglycemia, which is mostly linked to larger SFTs (mean size about 20 cm) - approximately 40% of SFTs associated with hypoglycemia are found to be malignant. The hypoglycemia is likely due to the production of non-suppressible insulin-like active substances and insulin-like growth factors by the tumor. The hypoglycemia resolves after tumor removal [9].

Mediastinal SFTs, like their serosal equivalents, are spindle cell neoplasms that are characterized by a "patternless pattern of hyper- and hypocellular spindle cell regions" [6,8], with hypocellular areas of "keloid hyalinization". Another pattern shown is the staghorn blood vessels in the background. The WHO classification of soft tissue tumors categorizes SFTs as intermediate (rarely metastasizing) neoplasms [4]. Mediastinal SFTs may exhibit an aggressive clinical course. In one series, up to 64% of mediastinal SFT showed aggressive clinical behavior on follow-up while 27% (3 of 11 patients) patients succumbed to disease progression[2].

Vallat-Decouvelaere et al described nuclear atypia, hypercellularity, necrosis and having more than four mitotic figures in each 10 high power microscopic fields [5,10] as suggestive of aggressive clinical behavior but not predictive of it. Our patient's sample showed hyperchromatic nuclei and 5 mitotic figures per 10 HPF, placing

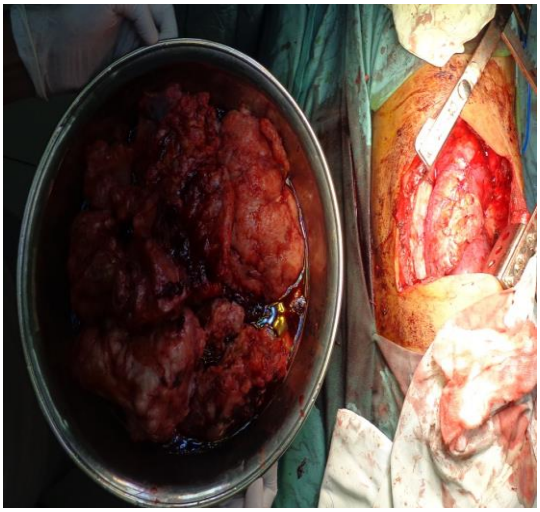
Figure 2: CT scan chest showed presence of heterogeneous mass lesion arising from mediasti--num extending into the left thoracic cavity up to the left lateral chest wall with compression of the aortic arch, pulmonary trunk and veins, and the left atrium and ventricle



our case in the category at risk of an aggressive course of disease. However, he showed no evidence of metastases during work-up and remains free of disease to date. Thus he required no referral to oncology. Moreover, studies show that the results of radiotherapy and chemotherapy for malignant SFT are doubtful and disappointing at best [9,11]. Our patient will be kept on regular follow-up to monitor disease recurrence [11].

In conclusion, mediastinal SFTs are a rare entity whose clinical behavior, although usually benign, can also become aggressive requiring close follow-up and monitoring for disease recurrence.

Figure 3: Operative picture showing a giant, solid tumor, being removed in multiple pieces



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