Solitary Fibrous Tumor of Pleura Arising from Diaphragmatic Pleura: A Rare Disorder

Syed Mahmood ul Haq[1], Jawed Ahmed Memon[1], Mariya Muhammad Saleem[1*], Rimsha Ashqeen[1] and Shayan Anwar[1]

[1]*Thoracic Surgery Department, Liaquat National Hospital & Medical College, Karachi, Pakistan*

Abstract

Solitary fibrous tumors (SFTs) of the pleura and lung are not very common primary tumors that originate from the sub-mesothelial tissue and usually show a benign clinical course. The diagnosis is made through immunohistochemical analysis. We have reviewed our experience to find a better and improved understanding of this disease.

Keywords: *Solitary fibrous tumor, pleura, soft tissue tumor.*

INTRODUCTION

Solitary fibrous tumors (SFTs) are rare soft tissue tumors. Initially, only pleura-based tumors were reported. However, it is clear that SFTs also occur in other sites including lung, liver, orbit, nasal passages, skin, thyroid, and gastrointestinal tract. These tumors are mesenchymal in origin and positive for CD34 and bcl-2 and negative for cytoplasmic keratin. Sometimes in malignant SFTs, CD34 and bcl-2 can be negative. These tumors are more common in middle-aged adults and each gender is equally susceptible. The majority of tumors present as slow-growing, painless masses [1]. In cases of benign lesions, the treatment is resection. We report the case of a 62-year-old woman who underwent surgical resection of a solitary fibrous tumor of the pleura.

CASE REPORT

A 62-year-old female, known to have hypertension, presented with cough and shortness of breath for the last 3 months. The patient had no history of addiction or exposure to chemical substances. On examination, the breath sounds were reduced in the right middle and lower zones. Chest x-ray showed large opacity in the right hemithorax **(Fig. 1)**. A Computerized Tomography scan showed heterogeneous soft-tissue mass originating from pleura in the right hemithorax causing a mass effect on the right lung **(Fig. 2)**. Ultrasound-guided biopsy was inconclusive, and the open biopsy showed spindle cells dispersed along with thin parallel collagen bands, with no increased mitosis or atypia. The immuno-histochemistry showed positive staining for CD34 but negative for ASMA, Desmin, and S-100. A diagnosis of benign SFT was made. The mass was excised with a right posterolateral thoracotomy approach. On opening, a large right-sided encapsulated pleural mass was found

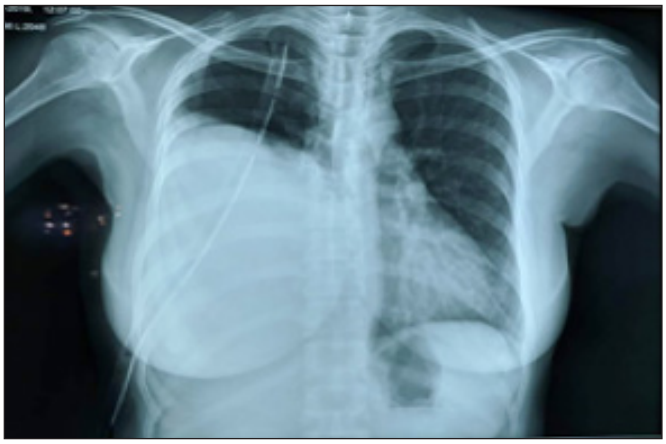

Fig. (1): Chest X-ray showing large opacity in right hemi thorax.

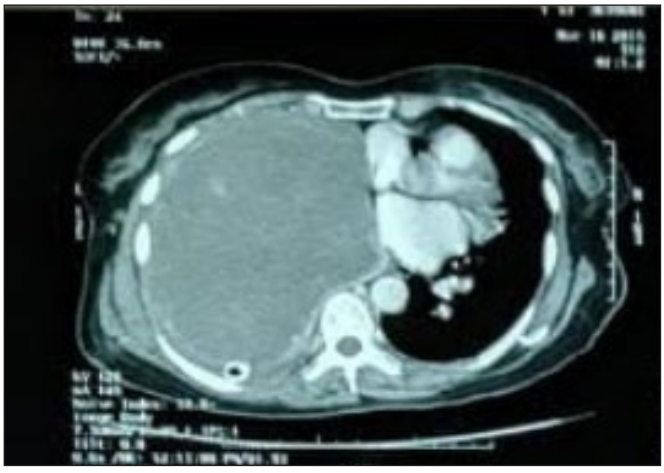

Fig. (2): CT-scan showing heterogeneous soft tissue mass.

which was well-circumscribed and highly vascular with no involvement of the lung. Grossly, it measured 16 x 15 x 10cm **(Fig. 3)**. It was attached to the diaphragm inferiorly and the lung base superiorly. It was completely excised **(Fig. 4)**. The postoperative course was uneventful and she completely recovered **(Fig. 5)**.

The final histopathology report showed features consistent with benign SFT with tumor-free resection

**Corresponding author: Mariya Muhammad Saleem, Thoracic Surgery Department, Liaquat National Hospital & Medical College, Karachi, Pakistan; Email: mm_s043@hotmail.com*

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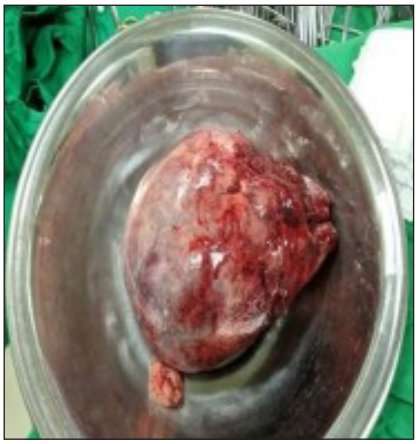

Fig. (3): Well-circumscribed Pleural mass.

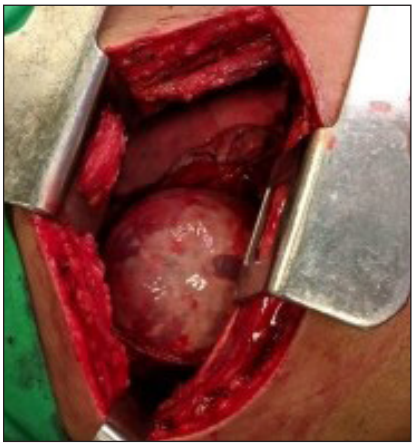

Fig. (4): Mass separated from the lung.

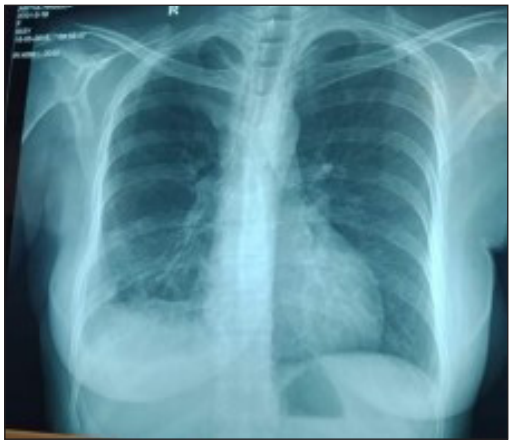

Fig. (5): Post-operative Chest X-ray.

margins. The patient remains asymptomatic and in radiological remission 1 year after diagnosis.

DISCUSSION

SFT originating in the pleura are rare neoplasms. The incidence is less than 3 per 100000 hospital patients. Less than one thousand cases have been described in the literature [2]. A prominent branching, thin-walled, dilated (staghorn) vasculature with NAB2-STAT 6 gene arrangement is a characteristic feature of SFT[3]. The tumors can occur at any anatomical site, including superficial and deep soft tissues, and within visceral organs and bone; however, they are more common at extrapleural locations. SFT of pleura mainly is a tumor of adults and both men and women get affected equally. These tumors are common in the 6th and 7th decades of life. The site, size, and malignant potential of the tumor dictate the clinical picture [4].

SFTPs are usually solitary lesions, but may rarely present as multifocal masses. Most SFTs arise from the visceral pleura. Out of these, around 50% are pedunculated. It is common to find these tumors adherent to adjacent pleural surfaces and the pericardium [5].

Pulmonary osteoarthropathy, with or without digital clubbing, is reported in up to 20% of cases of SFTs, with a widely varied incidence reported. Paraneoplastic syndrome in the form of hypoglycemia occurs in around 5% of patients, presumably because of the secretion of an insulin-like growth factor [6]. Our patient did not have either pulmonary osteoarthropathy or hypoglycemia.

These tumors are incidentally found on chest X-rays performed. The radiographic features of SFTs are non-specific in the form of lung opacity. CT scan demonstrates a well-defined, lobulated mass, isodense to skeletal muscle, with heterogeneous contrast enhancement due to the extensive tumor vasculature [7].

On IHC analysis, vimentin, CD34, CD99, and Bcl2, are positive in SFTs of the pleura. On the other hand, cytokeratin is negative in these. This immunohistochemical marker suggests that SFT originates from mesenchymal cells and not mesothelial cells [8].

The role of postoperative radiotherapy (PORT) for resected SFTs is unclear [9]. The decision for PORT is based on margin status, tumor size, and tumor grade. Low-grade tumors that are completely resected show better outcomes irrespective of tumor size. However, with tumors size exceeding 10 cm, with malignant characteristics, the rate of disease recurrence is higher [10]. PORT should be employed in cases where tumor recurrence can cause significant morbidity [11]. If the SFT is unresectable or the disease is metastatic, RT can be considered as a palliative option for the control of the local disease [12]. Preoperative radiotherapy has often been found to be useful in cases of soft tissue sarcomas for sterilizing the anticipated resection margins and improving local control [12]. Our patient did not receive postoperative treatment as the tumor was a benign SFT, and the surgical margins were also negative.

The prognosis for SFT is variable. Recurrence occurs in 10-30% of SFTs, with 10-40% of recurrences reported after 5 years. Large-sized tumors, malignant histology and certain pathologic characteristics are associated with worse prognosis [13].

Outcomes of SFTs are predicted by histological and morphological characteristics. Generally, malignant SFTs are associated with higher recurrence and mortality rates (up to 30%). Some systemic agents with anti-angiogenic activity (like sunitinib, sorafenib, and bevacizumab) may be useful for patients with unresectable or metastatic tumors [13].

CONCLUSION

Solitary fibrous tumors of the diaphragmatic pleura are rare neoplasms and show an indolent clinical course in most cases. Complete en-bloc resection remains the standard treatment with a good curative rate. The role of conventional chemotherapy and radiotherapy in the management of these tumors is still unclear. The prognosis for benign SFTs is generally favorable but long-term follow-up is still recommended.

DECLARATION OF PATIENT CONSENT

Written informed consent was obtained from the patient for publication of this case report and accompanying images.

CONFLICT OF INTEREST

The authors declare no conflict of interest.

ACNOWLEDGEMENTS

None.

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