Case Report

Giant Solitary Fibrous Tumour of the Pleura

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ABSTRACT

Solitary fibrous tumour of the pleura is a rare primary pleural neoplasm. These tumours are usually asymptomatic and are incidentally detected. Majority of these neoplasms are benign and surgical excision provides excellent results. With the widespread use of imaging and better diagnostic criteria, this tumour is likely to be detected more frequently. We encountered a patient with a giant solitary fibrous tumour of the pleura. In this report, we describe the case of a patient with a giant solitary fibrous tumour of the pleura, review the literature and present the details of management of this patient. **[Indian J Chest Dis Allied Sci 2012;54:49-52]**

Key words: Pleural, Localised mesothelioma, Solitary fibrous tumour.

INTRODUCTION

Solitary fibrous tumours of the pleura (SFTP) are rare neoplasms of mesenchymal origin. Most of the tumours are slow growing, benign, pedunculated and arise from the visceral pleura.[1] Most of the patients are asymptomatic and the presence of the tumour is usually incidentally detected. Though the number of cases reported is limited, it appears that surgical excision provides good results.[2] We present our experience with a giant SFTP, outline our management and review the literature.

CASE REPORT

A 39-year-old female presented with a swelling in her neck. She was diagnosed to have a diffuse colloid goiter. However, during work-up, her chest radiograph showed a large mass lesion in the right chest. She was referred to our institute for further investigation and management.

On evaluation, the patient had no respiratory symptoms or significant past illness. She had no pallor, clubbing, pedal oedema or lymphadenopathy. Respiratory examination revealed dullness in the right lower hemithorax with decreased air entry. Haemogram and liver function tests were normal. Sputum examination for acid-fast bacilli was negative. The chest radiograph (Figure 1) showed a large homogeneous mass lesion in the right lower zone.

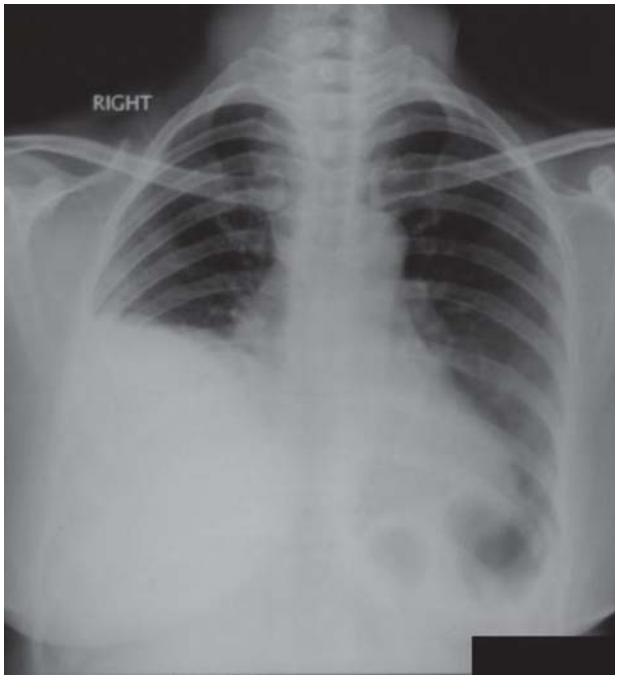

Figure 1. Chest radiograph (postero-anterior view) showing a mass in the right hemithorax.

Contrast enhanced computed tomography of the thorax showed a large mass (24cm x 14cm) in the right mid and the lower hemithorax. The mass appeared heterogeneous, lobulated and had a broad base towards the pleura, and with contrast enhancement suggesting that it had increased

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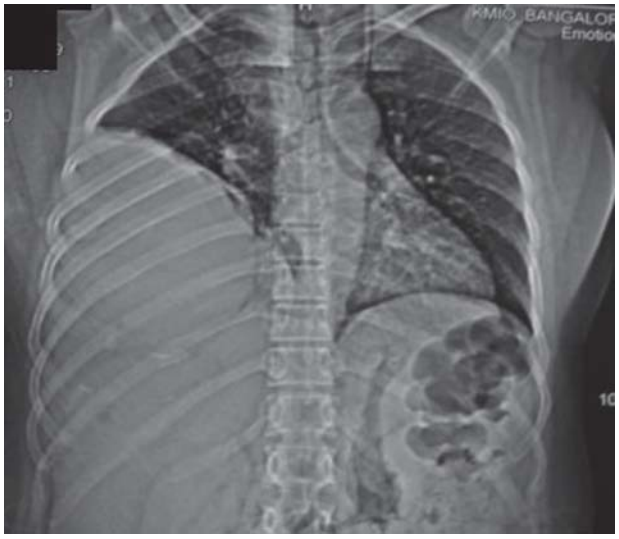
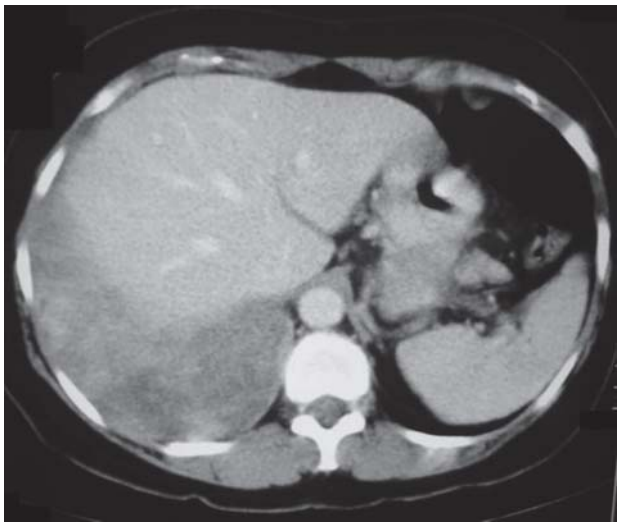
vascularity (Figures 2 and 3). An image guided fine needle aspiration cytology of the thoracic mass showed a few neoplastic round to spindle cells with marked nuclear atypia and scanty cytoplasm. An image guided core-needle biopsy showed predominantly fibrovascular connective tissue with one of the cores showing atypical oval to spindle cells with vesicular nuclei arranged in a diffuse manner.





Figures 2 and 3. Computed tomography of chest showing a heterogeneous soft tissue mass in the right hemithorax.

No native lung parenchyma was seen. On immunohistochemistry (IHC), the neoplastic cells were positive for calretinin and CD34, and negative for cytokeratin and smooth muscle antigen (SMA). A provisional pre-operative diagnosis of SFTP was considered and the patient was worked up for surgery. A right thoracotomy showed a solid, well-encapsulated, non-homogeneous and pedunculated tumour measuring 25cm × 15cm (Figure 4). The mass was arising from the visceral pleura and partly adherent to the right lower lobe. Complete surgical excision along with wedge resection of the right lower lobe was done. The patient had an uneventful recovery and was discharged on the ninth post-operative day.

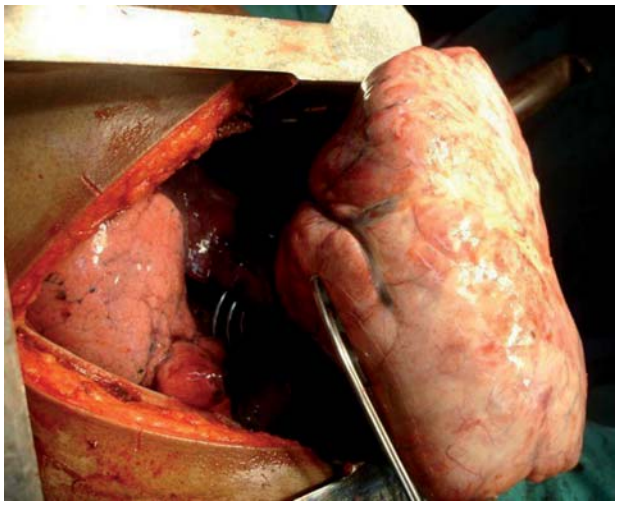


Figure 4. Intra-operative picture showing a large lobulated vascular mass adherent to the lower lobe of the right lung.

The histopathological examination revealed the tumour to be mostly composed of spindle cells in the background of dense collagenous stroma (Figure 5). There were no significant nuclear pleomorphisms or mitosis. These features suggested that the mass was of mesenchymal origin and was benign in nature. The adjacent lung parenchyma was compressed but free of tumour. The IHC of the specimen was positive for calretinin, CD34, CD99 and BCL-2 and negative for cytokeratin and SMA (Figure 5).

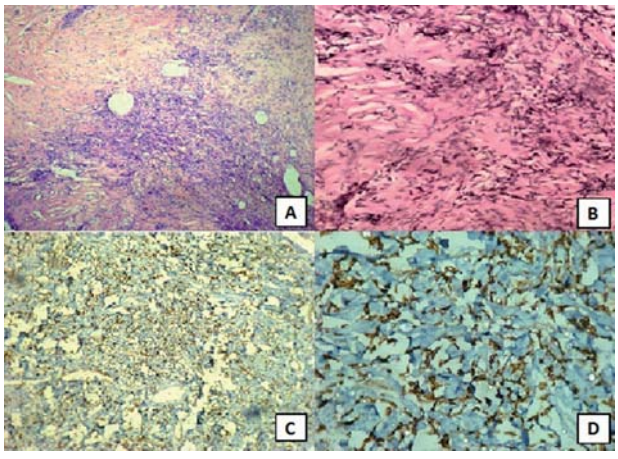


Figure 5. Photomicrograph of the solitary fibrous tumour showing: (A) typical architecture composed of alternating hypocellular (upper left) and hypercellular (lower right) areas (Haematoxylin and eosin ×40); (B) benign spindle cells separated by thick bands of keloid like collagen (Haematoxylin and eosin ×100); (C) diffuse positivity for CD34, DAB (×40); and (D) positivity for MIC2/CD99, DAB (×200).

Diagnosis of an SFTP was offered and a desmoplastic mesothelioma was suggested as a differential diagnosis. The patient has been on regular follow-up for the past two years and has no evidence of recurrence (Figure 6).

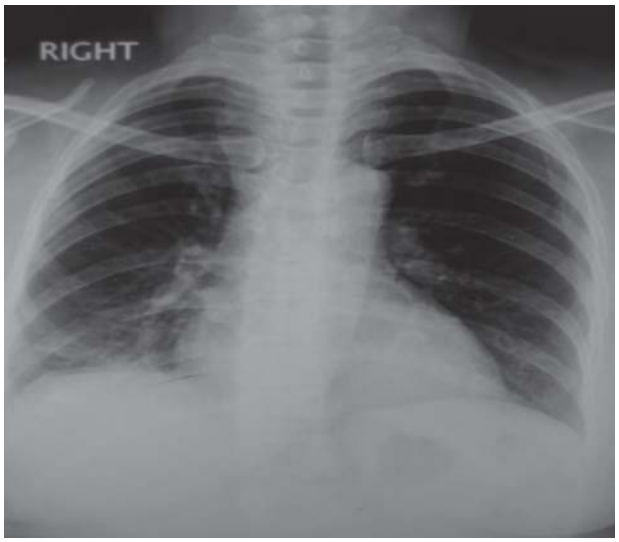


Figure 6. Follow-up chest radiograph (postero-anterior view) showing no evidence of recurrence.

DISCUSSION

Primary tumours of the pleura can broadly be divided into diffuse and localised types. Diffuse tumours are usually associated with asbestos exposure, are commonly malignant, and hence, carry a poor prognosis. On the contrary, the localised pleural tumours now also known as SFTP, have no association with environmental factors, are usually benign and carry a good prognosis.[3] SFTPs are rare, accounting for 5% of all pleural neoplasms, and only about 900 cases have been reported in the literature.[2]

SFTP was first described as a separate pathological entity by Wagner in 1870, but it was Klemperer and Rabin in 1931 who suggested that these tumours were of a submesothelial rather than of mesothelial origin.[4] SFTP in the past has been referred to by various terms including localised mesothelioma, fibrous mesothelioma, solitary fibrous mesothelioma, submesothelial fibroma etc to name a few.[2]

SFTP commonly presents in the sixth decade of life and most series suggest an equal predilection between the genders.[3] All large series in the literature report that upto 50% of patients were asymptomatic and diagnosed incidentally.[2-4] Patients usually become symptomatic because of large tumours or malignant neoplasms. Imaging investigation of choice appears to be contrast enhanced CT of the thorax.[2-4] Though Cardillo *et al*[4] suggest that benign nature of these tumours can be made out on CT, this is not accepted in the published reports.[2,3] Image guided FNAC has low sensitivity in most series and certain centres do not recommend their use in operable lesions.[2] The objective method of differentiating SFTP from diffuse tumours is by IHC. SFTP stains positive for CD34, CD99, BCL-2 and stains negative for cytokeratin.[4]

The mainstay of treatment appears to be surgical resection with preservation of as much lung parenchyma as possible. Frozen section examination is recommended to ensure the completeness of the resection.[4] Smaller lesions are amenable to resection by video assisted thoracoscopic surgery (VATS). Giant SFTPs are usually vascular, with adhesions and requires a thoracotomy to assist their removal. Guo *et al*[5] have reported their experience with pre-operative embolisation of these lesions to ensure decreased bleeding during their extirpation and were successful in all their five cases. Though there have been reports of adjuvant therapies for malignant and recurrent tumours their role remains undefined because of the rarity of these tumours. Benign tumours have a very good outcome with recurrence rates of less than 8%. The malignant tumours, on the other hand, have a high recurrence rate with most deaths occurring within 24 months.[3]

Our patient was asymptomatic and incidentally detected to have a large thoracic mass. The pre-operative core biopsy of the lesion was suggestive of SFTP. Because of the large size and increased vascularity, a conventional thoracotomy and excision of the mass was performed. The pathological examination was suggestive of a benign mesenchymal lesion and the IHC confirmed the diagnosis to be SFTP.

SFTPs are rare primary localised neoplasm of the pleura. Majority of the lesions are benign. These are usually asymptomatic and definitive pre-operative diagnosis may not be possible in most of the patients. Surgery appears to be the best treatment modality and can result in cure in majority of the patients. With the widespread use of imaging and better investigational modalities, such as, IHC and application of diagnostic criteria, these tumours are likely to be detected more frequently. Therefore, the thoracic surgeon should be aware about SFTPs and consider them in the differential diagnosis of soft tissue tumours of the thorax.

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