

Solitary Fibrous Tumor of 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. In this report, we describe the case of a patient with solitary fibrous tumour of the pleura.

Key words: Pleural, Solitary fibrous tumour

Introduction

Solitary fibrous tumor of the pleura (SFTP) is a rare entity that usually arises from the submesothelial mesenchymal cells; its incidence is < 5% of all pleural tumors.^{1,2} Most of the tumours are slow growing benign, pedunculated and arise from the visceral pleura.¹ Most of the patients are asymptomatic and detection of tumour is usually incidental. Though the number of cases reported is limited, it has been seen that surgical excision has provided remarkable results. Here we present a case of solitary fibrous tumour of pleura (SFTP), its management and its brief review of literature.¹

Case Report

A 40 years old female patient presented with history of pain in right lower chest for 4 months. She had no positive past medical history. On physical examination, percussion note at the base of the right lung was dull and on auscultation breath sounds were absent in this area. Rest of the general examination and chest examination were normal. Baseline laboratory investigations (renal and liver function tests, and complete blood counts) were normal except a raised ESR (40mm at the end of 1st hour). Chest radiograph showed a round well defined homogeneous soft tissue mass lesion in right cardiophrenic angle which was obscuring medial half

of right diaphragm. Adjacent parenchyma showed nonhomogenous shadowing. Ultrasound correlation showed it to be a solid mass lesion.

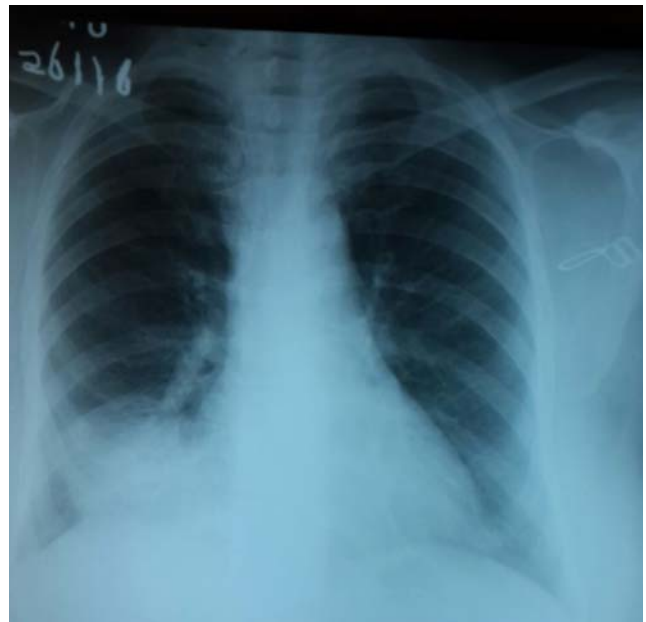


Figure 1. A round well defined homogeneous soft tissue mass lesion in the right cardiophrenic angle

Contrast enhanced CT-scan of chest showed a well margined, ovoid, pleural based benign looking solid mass in medial and lateral segments of right middle and anterior basal segment of right lower lobe, showing mild heterogeneous enhancement. Possibility of SFTP was suggested.

Ultrasound guided biopsy was done, histopathology showed mostly spindle cells with a minor population of plump cells. These cells were in sheets. There was minimal pleomorphism but no mitosis noted. No giant cells or necrosis was seen. A provisional pre-operative diagnosis of SFTP was made and surgery was recommended.

Right thoracotomy was done and showed a large, solid, well encapsulated, non-homogeneous tumor arising from visceral pleura and partly adherent to right lower lobe. Complete surgical excision along with wedge resection of right lower lobe was done. The patient had an uneventful recovery and was discharged on 5th post-operative day.

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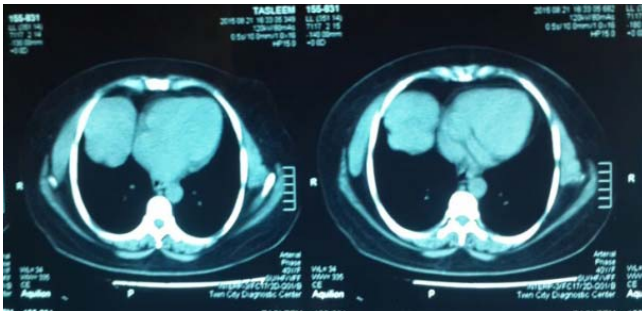


Figure 2. CECT Chest (Mediastinal window) showing a well margined, ovoid, pleural based benign looking solid mass in right hemithorax.

Histopathological examination revealed the tumor to be mostly composed of spindle cells in the background of dense collagenous stroma. There was no significant nuclear pleomorphism or mitosis. These features suggested that the mass was benign in nature. The immunohistochemistry of the specimen was positive for vimentin and negative for ASMA, S 100, Claretinin, CD 117.

Diagnosis of solitary fibrous tumor of pleura (SFTP) was made. The patient was advised regular follow up to see any evidence of recurrence.

Discussion

Primary tumours of the pleura can broadly be divided into diffuse and localized types. Diffuse tumours are usually associated with asbestos exposure. They are commonly malignant, and hence, carry a poor prognosis. On the contrary, the localized pleural tumours now also known as SFTP, have no association with environmental factors. They are usually benign and carry a good prognosis.¹

Their incidence is rare and is predominant in the sixth and seventh decades with no difference in gender distribution. This tumour arises from submesothelial mesenchymal cells. Upto 50% of patients are asymptomatic and diagnosed incidentally.¹ Symptoms include cough, chest pain and dyspnea. Clubbing and hypertrophic osteoarthropathy (Pierre Marie-Bamberger syndrome) have been reported in 10 to 20% of patients. In less than 5% of the patients, the tumor may secrete insulin, which causes intractable hypoglycemia (Doege-Potter's syndrome).³

Contrast Enhanced CT chest is the gold standard for diagnosis. Preoperative U/S/CT-guided biopsy is controversial as its accuracy rate is compromised because SFTPs involve varying cellular areas. Complete resection should be performed for diagnosis and treatment. Unlike benign SFTP, malignant SFTP can have a poor prognosis, which is often associated with tumor size. They can recur or metastasize, and their incidence has been reported to range from 14% to 43%. Their 5-year survival rate has been reported to range from 45-68%.²

Complete surgical resection of fibrous tumors of the pleura is usually curative, but local recurrences have been reported years after surgery. Although the reason for these recurrences is unclear, they may be due to incomplete resection. For tumors arising from the visceral pleura, wedge resection may be performed for complete excision. For those of the parietal pleura, an extrapleural resection may be carried out without any chest wall resection.⁴

Conclusion

Solitary fibrous tumour of pleura is a rare slow growing primary pleural based tumor of mesenchymal cell origin. It can occur at any age with peak incidence seen at 6th decade of life. It is often associated with visceral pleura. In most cases it is discovered incidentally as half of the cases present with no clinical symptoms. Diagnosis is based on histopathological examination. In most cases it is benign with low risk of recurrence and malignant transformation therefore complete surgical resection and long term follow up is recommended in all patients.

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