Giant Solitary Fibrous Tumour of Pleura


Abstract

Most pleural neoplasms are metastatic in origin. Primary tumours of the pleura are rare and categorized as diffuse or localized. Diffuse or malignant mesothelioma is more common, related to asbestos exposure, and associated with a poor prognosis. Localized or solitary fibrous tumour (SFT) of the pleura is a less common neoplasm of controversial histogenesis and is unrelated to asbestos exposure.

Surgical resection of benign solitary fibrous tumours is usually curative. We report a rare case of Giant solitary fibrous tumour of pleura with related literature.

Introduction

Solitary fibrous tumour (SFT) of pleura is a rare neoplasm arising from the submesothelial tissue underlying the mesothelial layer of the pleura. Primary tumours of pleura were divided into diffuse and localized form by Klemperer and Rabin in 1937.1

Diffuse pleural tumours or mesothelioma are more common than localized or solitary form. SFT of pleura constitutes less than 5% of pleural tumours, 80% arise from the visceral pleura and the remainder from parietal pleura.2 SFT also affects other sites such as peritoneum, pericardium and in non-serosal sites such as lung parenchyma, upper respiratory tract, orbit, thyroid, parotid gland, or thymus. The origin of these tumours is controversial, and their nomenclature not consistent, with names such as fibroma, fibrosarcoma, localized fibrous mesothelioma, submesothelial fibroma, benign fibrous mesothelioma, etc.3 Generally, there is no apparent genetic predisposition for the tumour and no relationship to exposure to asbestos, tobacco, or any other environmental agent.

Case Report

A 45 year old male presented to us with history of vomiting on and off since 4 years. There was no history of cough, chest pain, dyspnoea, or past history of tuberculosis. X-ray chest showed a large well defined homogenous lesion in left sided lower zone (Fig. 1).

Fig. 1: X-ray chest showing mass lesion in left hemithorax
Computed Tomography (CT) scan revealed large well defined pleural based mass lesion with moderate enhancement with dense areas of calcification and necrosis. There was mass effect in form of diaphragmatic inversion with left lower lobe atelectasis and right mediastinal shift suggestive of pleural fibroma (Fig. 2). Fine Needle Aspiration Cytology (FNAC) was suggestive of fibrous pleural tumour.

Left posterolateral thoracotomy through 5th intercostal space was done.

Intra-operative findings revealed a Giant pleural tumour measuring 22 X 18 X 17 cms filling majority of the lower thoracic cage with a pedicle attached to the lower lobe of left lung. Complete surgical resection was done, following which full lung expansion was seen. Specimen weighed 2600 gms. Grossly it was well encapsulated with smooth capsule, with no capsular breach. On cut surface it had whorled appearance (Fig. 3), most of the areas were firm, grey white, few calcification seen over central area. Histopathologic examination revealed spindle cell fasciculated lesion composed of bland, uniform cell lacking mitosis, atypia or necrosis.

Immunohistochemistry with CD34, Ckit, S100, SMA, Calponin, Desmin, CK, Calretinin, Meso specific antigen were not contributory. Impression on H and E stain was solitary fibrous tumour of pleura. Post-operative recovery was uneventfully. Follow-up of 6 months has shown him to be disease free.

Discussion

The first case of SFT was described by Wagner in 1870 in his article “Das Tuberkelahnliche Lymphadenom”, more than a half century earlier than the classification of primary pleural tumours into diffuse and solitary form. SFT tends to affect mainly adults during the sixth and seventh decades of life. Most patients are asymptomatic, and the lesion is discovered incidentally on chest radiographs. Symptomatic patients may report dyspnoea, cough, or vague chest or shoulder discomfort. Symptoms are usually related to the local mass effect of large lesions or to the associated paraneoplastic phenomena. They range in size from 1-36 cms with a mean of 6 cms. Extrathoracic manifestations include clubbing of fingers in the form of hypertrophic pulmonary osteoarthropathy (HPO) and hypoglycaemia. HPO is reported in up to 22% of patients especially in tumour over 7 cms in diameter. Both paraneoplastic syndromes are more common in tumours larger than 7 cms in size and resolve with surgical resection of the tumour. Because they lack distinctive histological features, immunologic staining has frequently been employed to exclude other neoplasms in the differential diagnosis. Although immunoreactivity with CD34 is

![Fig. 2: CT scan showing giant left pleural tumour with areas of calcification.](image)

![Fig. 3: Gross cut section of giant SFT of pleura.](image)
believed to be highly characteristic of SFT of pleura (80%) and a positive CD34 staining is required for diagnosis, the immuno-histologic staining pattern is not entirely specific. Other entities, such as angiosarcoma and gastrointestinal stromal tumour, also express CD34 and vimentin epitopes. In our case CD34 and other immunohistochemical markers were negative, however H and E stain proved it to be SFT of pleura.

The recommended treatment for SFT is complete resection of tumour and its pedunculated portion along with the site of origin. Five year survival rates as high as 97% for benign tumours, however, with incomplete resection or malignant transformation, the median survival is only 24 to 36 months. The best indicator of a good prognosis is complete surgical resection with favourable histopathology. Clinical and radiological follow up are indicated for both benign and malignant solitary fibrous tumours.

References

TREATMENT OF HYPERTENSION IN PATIENTS 80 YEARS OF AGE OR OLDER

Whether the treatment of patients with hypertension who are 80 years of age or older is beneficial is unclear. It has been suggested that antihypertensive therapy may reduce the risk of stroke, despite possibly increasing the risk of death.

We randomly assigned 3845 patients from Europe, China, Australasia, and Tunisia who were 80 years of age or older ad had a sustained systolic blood pressure of 160 mm Hg or more to receive either the diuretic indapamide (sustained release, 1.5 mg) or matching placebo. The angiotensin-converting-enzyme inhibitor perindopril (2 or 4 mg), or matching placebo, was added, if necessary to achieve the target blood pressure of 150/80 mm Hg.

The results provide evidence that antihypertensive treatment with indapamide (sustained release), with or without perindopril, in persons 80 years of age or older is beneficial.