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Giant pleural fibroma: A case report

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Abstract
Malignant tumors of the pleura are more common than benign tumors. The final diagnosis is by histopathological examination and immunohistochemistry. Total surgical resection is generally curative.

Keywords: Fibroma, pleura, giant.

Introduction
Solitary fibrous tumor of the of the pleura is an uncommon pleural tumor. It is often an incidental finding & hence may reach gigantic proportions before being detected.

Case Report
A 46 year old gentlemen presented to the local hospital after a road traffic accident. Chest radiogram was done and a radio opaque lesion was found in the lower half of right hemithorax. Further evaluation by Contrast enhanced Computerized Tomography (CECT) scan (Fig.1) of the thorax showed a well-defined heterogeneously enhancing lesion 18 x 12 x 10 cms with peripherally enhancing thick rim, few enhancing septae, irregularly enhancing soft tissue components, multiple, non-enhancing cystic areas (5-18 HU) and few foci of calcification. The lesion occupied the entire lower half of right hemithorax and displaced the right hemidiaphragm inferiorly. The lesion was closely adherent to the mediastinal pleura and the pleura along the lateral chest wall and compressed the adjacent lower lobe of right lung. The lung parenchyma, cardiac silhouette and major thoracic vessels appeared normal. There was no lymphadenoapthy, pleural or pericardial effusion.

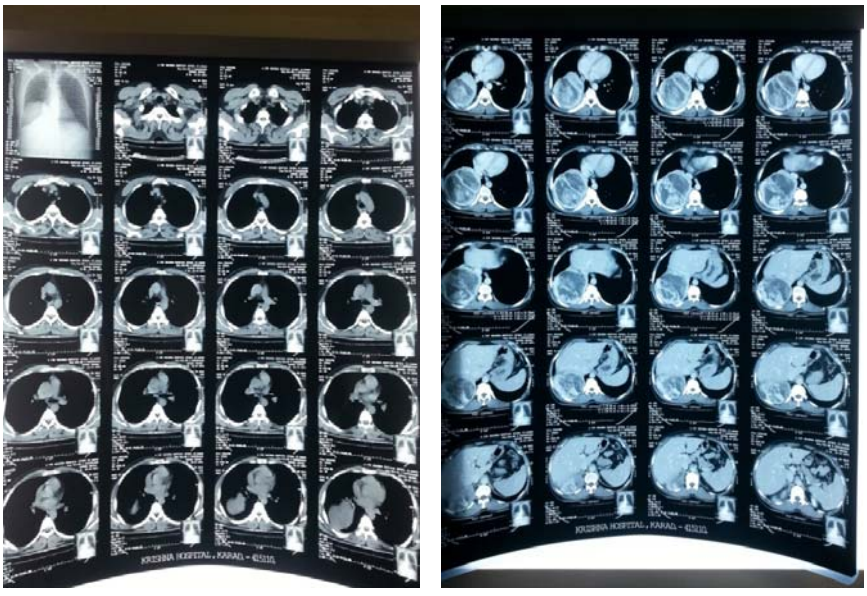


Fig 1: CECT scan showing a well – defined, heterogeneously enhancing mass occupying the lower half of the right hemi thorax.

Elective surgery was done under General anaesthesia through a right posterior thoracotomy via the seventh intercostal space. A well encapsulated, ovoid mass,18 x 10 x 8 cms, weighing 2.5 kilograms was adherent to the right sixth, seventh and eighth intercostal spaces. The lower lobe of right lung was collapsed. Excision biopsy (Fig 2) was done. The histopathological report was a grey- white mass (Fig 3) with variegated appearance and areas of necrosis and hemorrhage. Whorls of elongated spindle cells with eosinophilic cytoplasm; some cells showed nuclear hyperchromasia (Fig 4). Areas of pallisadation present. Peripheral part showed extensive fibrous, desmoplasia, cholesterol clefts and mononuclear cell infilteration. Immunohistochemistry (IHC) (Fig.5) showed that the cells expressed CD-34 antigen and bcl-2 but were immunonegative for cytokeratin (CK) and S-100.

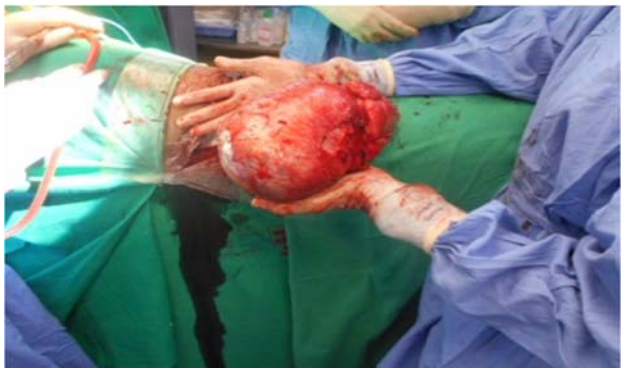


Fig 2: Excised tumor from the right hemi thorax

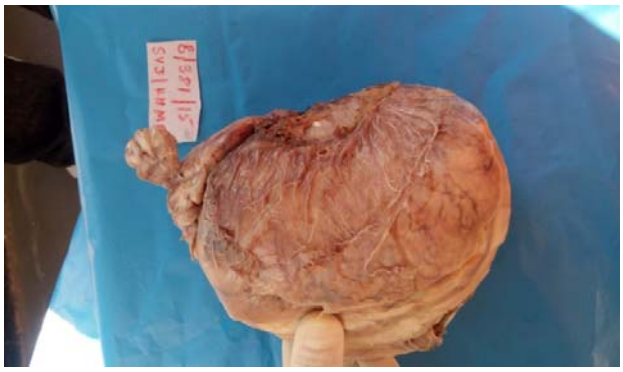
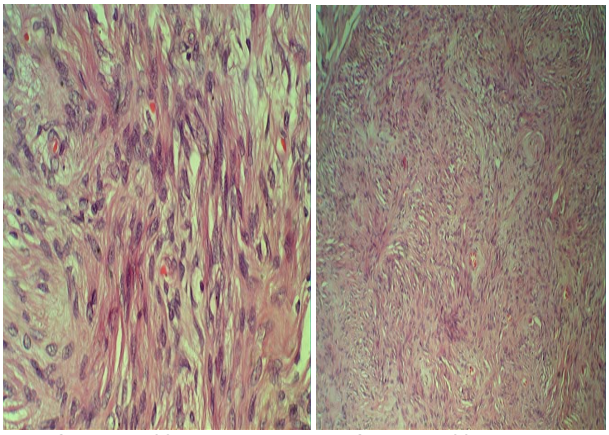


Fig3: Gross pathology specimen of the tumor



H&E stain 100 x H&E stain 400x

Fig 4: Photomicrograph showing diffuse sheath storiform pattern and whorling of Spindle cells (H&E stain)

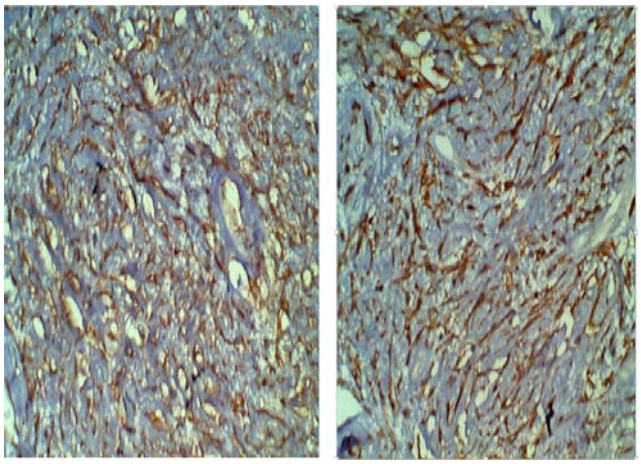


Fig 5: Immunohistochemistry showing tumor cells expressing CD34 and bcl 2

Discussion

Solitary fibrous tumor (SFT) of the pleura is a rare pleural tumor and had also been called benign mesothelioma [1]. Electron Microscopy and IHC have demonstrated that these tumors have a mesenchymal rather than mesothelial origin and hence the term “mesothelioma” has been abdandoned [2]. About 12% are malignant and cause death by local recurrence or metastatic disease [3]. Extraserosal occurrence in the lung, thyroid gland, paranasal sinuses, liver and mediastinum have been reported.

It is usually detected in middle aged individuals. There is no gender predilection. It is often an incidental finding on a chest radiograph. Sometimes, compression of the surrounding structures may cause wheezing, shortness of breath, cough, chest pain. The paraneoplastic syndromes associated with SFT are (i) Hypertrophic Osteoarthropathy (Pierre- Marie- Bamberg syndrome)-occurs as a result of production of ectopic Growth–hormone -like substance and is more common with tumors greater than 7 cms and (ii) hypoglycemia (Doege-Potter syndrome) - attributed to the production of Insulin- like growth factor II.

The imaging modality of choice is contrast enhanced Computerized Tomography (CECT). Biopsy is the gold standard. Hisotologically, the tumor cells shows a whorled appearance with bizarre growth of spindle cells with scanty cytoplasm. Immunohistochemistry shows positivity for CD 34 antigen and negative for Cytokeratin (CK). Surgical resection is the mainstay of treatment. Video assisted throracoscopic surgery is reserved for small tumors. The progonosis is excellent and recurrence rate is low [4].

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