Case Report

Solitary Fibrous Tumour of Lung

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ABSTRACT

Fibrous tumours arising entirely within the substance of the lung are rare. We report one such rare case in whom the diagnosis was established after surgical removal. **[Indian J Chest Dis Allied Sci 2013;55:171-173]**

Key words: Fibrous tumour, Lung neoplasm.

INTRODUCTION

Localised fibrous tumours of the lung are neoplasms that usually arise from the pleura, especially viscera.[1-3] Most of these tumours are pedunculated and project into the pleural cavity. Their histologically benign features are indicative of a favourable clinical course.[2] Inward tumour growth into the lung parenchyma is infrequent while totally intra-pulmonary localised fibrous tumours without histological continuity with visceral pleura have been described rarely.[4,5] We present here a rare case of entirely localised an intra-pulmonary fibrous tumour.

CASE REPORT

A 37-year-old housewife from rural area presented with a left-sided, non-radiating chest pain and dry cough for the last one month. She was a non-smoker with no history of asbestos exposure. Respiratory system examination revealed a diffuse dull note, decreased breath sounds and vocal resonance over the left side with a shift of the mediastinum to the right side. General and other systemic examination was un-remarkable. Investigations revealed no abnormality in the haemogram, blood biochemistry, electrocardiogram and echocardiography.

The chest radiograph showed a homogeneous opacity in the left mid- and lower-zones with mediastinum pushed to the right side, suggestive of a massive pleural lesion. Ultrasound of the chest showed a solid mass lesion occupying the left hemithorax with minimal pleural effusion. Ultrasound of the abdomen revealed no abnormality. Contrast enhanced computed tomography (CECT) of the thorax showed a homogeneous opacity (140mm × 108mm) in the apico-posterior segment of the left upper lobe, lingula and part of the lower lobe. The mass showed areas of enhancement. Calcification was seen in the periphery. The left main bronchus was compressed and occluded. Air-bronchogram sign was present in the mid zone and heart was pushed to the right. Consolidation was seen in apical segment of the left lower lobe, and there was minimal pleural effusion in the left hemithorax (Figure 1).

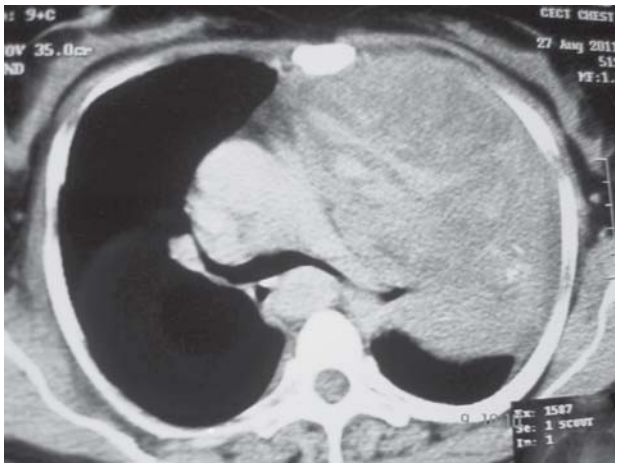

Figure 1. Contrast enhanced computed tomography of chest showing consolidation in left lower lobe with minimal pleural effusion.

A fine needle aspiration was carried out. The cytological appearance was suggestive of a low-grade, spindle-cell neoplasm with close resemblance

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to spindle-cell carcinoid tumour. The patient underwent surgery with excision of the tumour and a left upper lobectomy. Intra-operative findings revealed a large tumour (10cm×8cm), involving the left middle and upper lobes and anterior chest wall. Hilar and para-oesophageal lymphadenopathy was present. Grossly, the mass showed a grey-brown nodular mass measuring 13cm×11cm×7.5cm along with the attached lobe of the lung measuring 9cm×8cm×1.5cm with congestion of the external surface. Cut-section showed necrotic and haemorrhagic areas. Micro-section showed histological features of solitary fibrous tumour (Figure 2) with extensive areas of necrosis, haemorrhage and myxoid change.

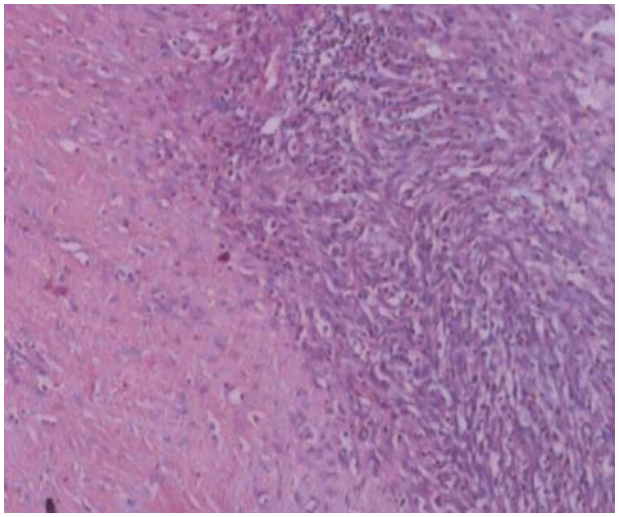

Figure 2. Histological photograph showing pattern-less arrangement of fibroblast cells (Haemotoxylin and Eosin×100).

Immunohistochemistry was positive for Bcl, CD34; negative for EMA, SMA, calretinin and focally positive for desmin (Figures 3 and 4). Sections from lung showed focal areas of chronic inflammatory cell infiltrate in the alveoli. The post-operative period was uneventful.

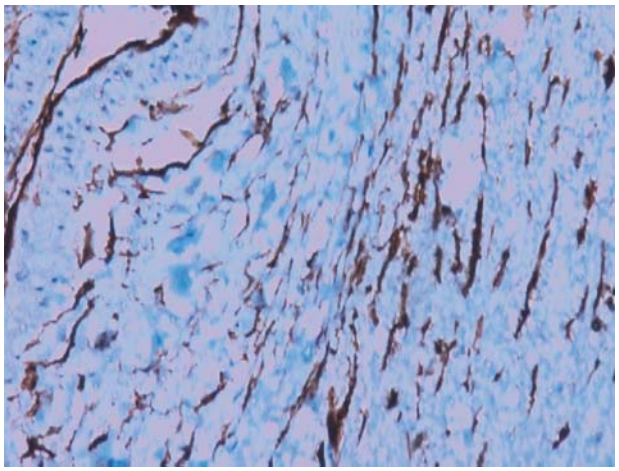

Figure 3. Immunohistochemistry stains for CD34 positive (x200).

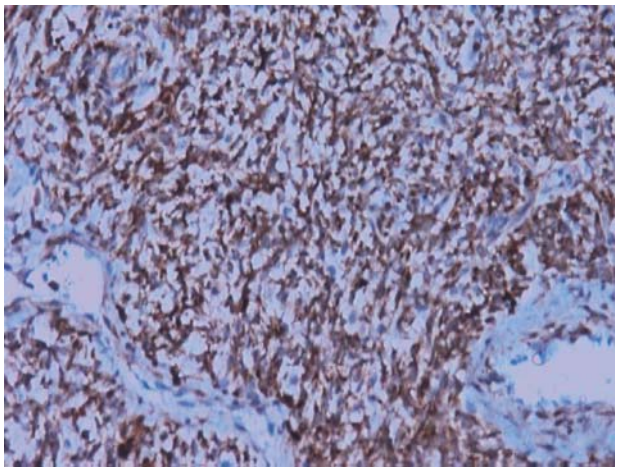

Figure 4. Immunohistochemistry stains for bcl2 (x200).

DISCUSSION

Currently, localised fibrous tumours are recognised as sub-mesothelial in origin based on immuno-histochemical and ultra-structural findings. These usually occur during the fifth and sixth decades of life with no sex predominance.[2,3] These tumours most often involve pleura, especially the viscera with a pedunculated attachment. The histological differential diagnosis includes fibrosarcoma lung, spindle-cell neoplasms, such as leiomyosarcoma, monophasic synovial sarcoma, neurofibrosarcoma and sarcomatoid mesothelioma.

Histologically, the tumour cells are composed of spindle cells with dense bundles of collagen, and the spindle tumour cells are arranged in a so-called "pattern-less pattern" that is characterised by a haphazard distribution of spindle cells and collagen fibres. These are classified as benign or malignant, based on histological findings (e.g., pleomorphism, mitotic activity, invasive growth, or presence of necrosis) although pathologic criteria may slightly differ between observers. As these lack distinctive histological features, immunohistochemical examination is very important for their diagnosis. Immunohistochemically, these tumours have been shown to lack expression of cytoplasmic keratin but express vimentin, a marker of mesenchymal cells.[1-5] In addition, their positive expression of CD34, a transmembrane cell surface glycoprotein has recently been shown in a study[6] and is useful for excluding similar spindle cell neoplasms in differential diagnosis.

Most localised fibrous tumours of the pleura are exophytic pedunculated masses that extend from the visceral pleura into the thoracic cavity. Inward growth into the lung peripheral parenchyma is infrequent; the term "inverted" has been used to describe such pleural derived tumours.[3] In the present case, the tumour did not show histological

continuity with the visceral pleura, although it was likely to be macroscopically adjacent to the visceral pleura. In addition, alveolar pneumocytes and small bronchioles that retained their histologically benign appearances were entrapped within fibrous cells of the tumour. Consequently, we consider this tumour to have arisen from the parenchyma of the lung. In fact, early reports of "fibroadenoma" of the lung may represent this spindle-cell tumour with entrapped alveolar epithelium.[7]

Two main hypothesis for their entirely parenchymal location have been proposed.[4,5] First, the subpleural mesenchyma is in direct continuity with the connective tissue of interlobular septa and intrapulmonary fibromas may arise from the septal mesenchymal or invagination of the visceral pleura. Secondly, these tumours may originate from facultative fibroblastic elements that can be seen in the sub mesothelial area of normal pulmonary parenchyma. It has been reported that these elements have ultrastructural and immunohistochemical features similar to those of sub-pleural connective tissue elements.[8] With such a parenchymal location, the tumour should not be pathologically mistaken for choriocarcinoma or adenocarcinoma with a metaplastic spindle cell component.

There are 12 previously reported cases of intrapulmonary localised fibrous tumours found in the medical literature,[9] ranging in the age from 20 to 82 years, and mostly asymptomatic, without predilection for any site with size ranging from 1cm to 15cm. Histologically, all tumours showed benign features. All of the patients, except one, were treated by complete surgical resection (i.e., a wedge resection to a lobectomy of the lung).[10]

Although prognosis of benign localised fibrous tumours of the pleura is generally good; recurrence has occasionally been reported. The highest risk of recurrence (recurrence rate of 63%) is seen in tumours of pathologically benign and morphologically sessile types, whereas the lowest risk of recurrence (recurrence rate of less than 2%) has been reported in tumours of pathologically benign and morphologically pedunculated types.[2] Morphologically, the sessile type is more likely to recur than pedunculated type.

To conclude, a localised fibrous tumour arising from the lung parenchyma is extremely rare. The parenchymal occurrence is suggested by histological findings of direct continuity between tumour and the visceral pleura; and the presence of entrapped alveolar epithelium within the spindle cell proliferation of the tumour. Complete surgical resection is both diagnostic and curative.

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