Pulmonary inflammatory myofibroblastic tumour

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ABSTRACT

Inflammatory myofibroblastic tumour is a rare disease and lung is one of the common sites. We report the case of a 36-year-old man with pulmonary inflammatory myofibroblastic tumour with an unusual presentation, an episode of haemoptysis with expectoration of a piece of fleshy tumour.

Keywords: Inflammatory pseudotumour, pulmonary neoplasm, haemoptysis

INTRODUCTION

Inflammatory myofibroblastic tumour is a rare disease and the most commonly affected sites are the lung, mesentery and omentum. Other sites include head and neck, retroperitoneum, liver and bladder. Microscopically, the tumour is characterised by proliferations of spindle cells with myofibroblastic differentiation accompanied by inflammatory infiltrate of plasma cells, lymphocytes, histiocytes and eosinophils. We report the case of a young man with pulmonary inflammatory myofibroblastic tumour with an unusual presentation.

CASE REPORT

A 36-year-old Malay man presented with a sudden episode of haemoptysis and coughing up fleshy tissue. He had a history of unproductive cough in the previous three to four months. He stopped smoking six months prior to presentation. Clinical examination was unremarkable. Chest radiography showed fibrotic scarring and fibrotic strands in the right upper lobe with elevated right hilum. A computed tomography (CT) scan of the thorax reported pin-point centrilobular nodules and tree-in-bud appearances in the apical segment of the right upper lobe.

The macroscopic examination of the fleshy tissue showed a few white to pale brown tissue fragments measuring 2.0 x 0.5 x 0.5cm in aggregate. Microscopically, they consisted of cellular sub-endobronchial epithelial lesion comprising interlacing and whorls of spindle cells with fairly uniform ovoid to elongated nuclei with open chromatin pattern containing small nucleoli. The mitotic activity was very low (3/50 high power field). Moderate amount of lymphoplasmacytic and histiocytic inflammatory infiltrate including a fair number of eosinophils was present scattered amongst the spindle cells.

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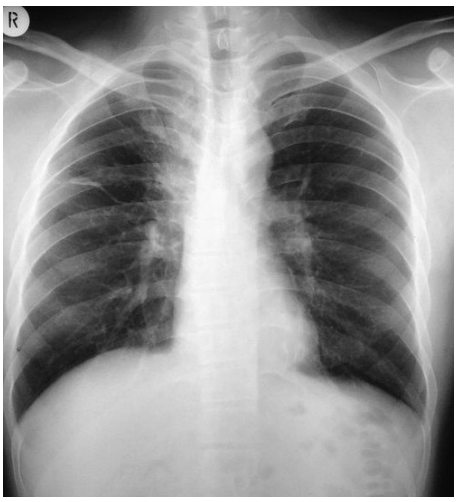


Fig. 1: A chest radiograph showing volume loss in the right hemi thorax. The right hilum is elevated as is the horizontal fissure, in keeping with partial collapse of the right upper lobe. An defined opacity superior to the right hilum in the right paramediastinal region.

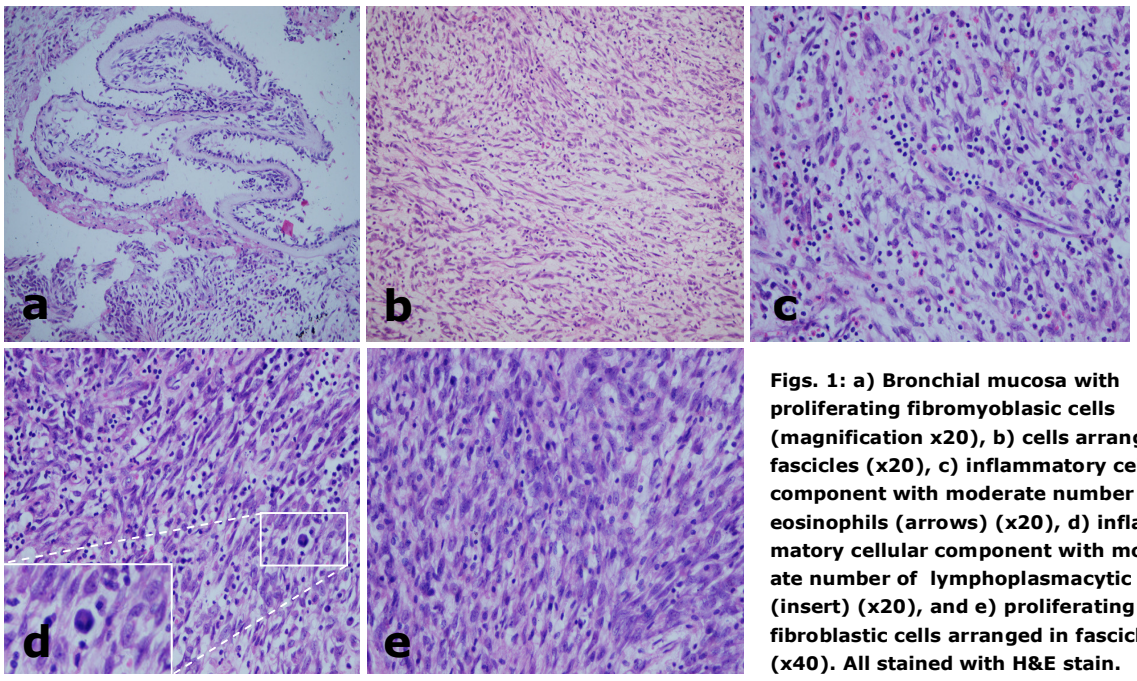
Immunohistochemistry shows the spindle cells are diffusely positive for vimentin, smooth muscle actin and desmin. Pancytokeratin marker CKAE1/3 showed patchy weaker staining. CD34 and ALK protein are negative. A diagnosis of inflammatory myofibroblastic tumour of the lung was made.

Subsequent bronchoscopy revealed a small reddish lesion at the entrance of the anterior segment of the right upper lobe bronchus. The biopsy and bronchoalveolar lavage cytology showed similar findings of inflammatory myofibroblastic tumour as that of coughed up material.

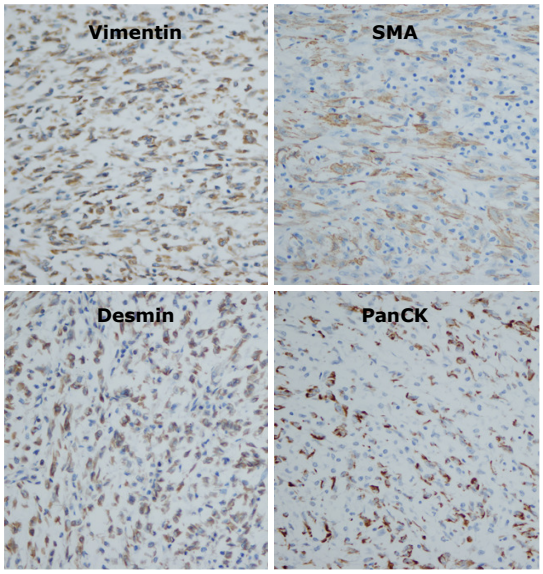
The patient underwent right upper lobectomy. No residual tumour was found macroscopically and microscopically. The patient had an uneventful postoperative recovery. The patient was reviewed at eight months and was well and symptoms free.

DISCUSSION

The incidence of pulmonary inflammatory my-



Figs. 1: a) Bronchial mucosa with proliferating fibromyoblasic cells (magnification x20), b) cells arranged in fascicles (x20), c) inflammatory cellular component with moderate number of eosinophils (arrows) (x20), d) inflammatory cellular component with moderate number of lymphoplasmacytic cells (insert) (x20), and e) proliferating myofibroblastic cells arranged in fascicles (x40). All stained with H&E stain.



Figs. 2: Immunohistochemical profiles; a) Vimentin stain, b) SMA, c) Desmin and d) PanCK.

ofibroblastic tumour is reported to account for between 0.04 and 1% of all pulmonary tumours. It has equal sex distribution and occurs in all ages but more common in patients less than 40 years of age. [1]

In the past, inflammatory myofibroblastic tumour was thought to be a reactive inflammatory lesion simulating a neoplasm, and was referred by different names depending on the predominant cell type: plasma cell granulomas, inflammatory pseudotumours, xanthomatous pseudotumours, fibrous histiocytoma, pseudosarcomatous myofibroblastic tumour and invasive fibrous tumour of the tracheobronchial lesion. Discovery of IgG4 positive plasma cells within some of the lesions suggested an IgG4-related autoimmune aetiology. However, in the recent years inflammatory myofibroblastic tumour has been regarded as low grade malignant neoplasm by many investigators due to the discovery clonal cytogenetic abnormalities involving chromosomes 2p23 that encodes the anaplastic lymphoma kinase (ALK) gene site that typically occurs in children and young adults. [2, 3]

Pulmonary inflammatory myofibroblastic tumour may present as peripheral or central lesions. Peripheral lesions are usually asymptomatic with the lesion being accidentally on radiology but central endobronchial lesions may produce acute symptoms secondary to airway obstruction such as cough, dyspnoea and haemoptysis.

Diagnosis of inflammatory myofibroblastic tumour is usually made on examination of resection specimens. CT scan usually lack specific features to differentiate it from lung cancer. [4] Preoperative transbronchial or transparietal biopsies are often non-diagnostic or unable to provide unequivocal diagnosis. [5] Intraoperative frozen section may also be inconclusive.

Microscopic examination of inflammatory myofibroblastic tumour reveals proliferation of spindle cells with myofibroblastic differentiation accompanied by an inflammatory cell infiltrate consisting lymphocytes, plasma cells, histiocytes and eosinophils. Plasma cell may be prominent and are often associated with lymphoid follicles. Features that may be associated with a poor prognosis include focal invasion, vascular invasion, increased cellularity, nuclear pleomorphism with bizarre giant cells, a high mitotic rate (greater than 3/50

high power field) and necrosis. Immuhohistochemistry usually shows the spindle cells to be diffusely positive to vimentin, smooth muscle actin and ALK protein whilst negative to epithelial membrane antigen (EMA), myoglobin, myogenin, S100, CD117. Focal desmin and cytokeratin staining have been reported.

Surgery is the principal treatment with both diagnostic and therapeutic values. Wedge resection is the first line but lobectomy or pneumonectomy may be undertaken to ensure complete resection. A study of 25 cases of inflammatory myofibroblastic tumour that included cases with local invasion, distant metastasis, recurrence, and sarcomatous degeneration reported a 10 year survival of 89% with complete resection. [3, 6] The use and effectiveness of radiotherapy, chemotherapy and steroid treatment in unresectable, incompletely resected or recurrent tumours are controversial.

In conclusion, inflammatory myofibroblastic tumour is a rare primary lung tumour that is best regarded as a low grade malignant neoplasm. It needs to be included in the differentiated diagnosis of peripheral or central endobronchial lung mass. Surgery has both diagnostic and therapeutic value. Excellent survival is usually achieved with complete resection.

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