Intrapulmonary Malignant Solitary Fibrous Tumour: A Rare Entity

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Citation

Abstract
Solitary fibrous tumours (SFT) of the pleura are neoplasms thought to arise from the mesenchymal tissue. Intrapulmonary malignant fibrous tumours are rare although histologically identical to pleural lesions and should be managed as such. Surgical excision with clear margins is the accepted treatment in the literature, for both benign and malignant disease.

CASE REPORT
A 70 yo female presented with three months of cough, minimal sputum production and haemoptysis. She had a 45 pack year smoking history. The patient reported no weight loss, no fevers and was systemically well. Her CXR showed a 1.5cm opacity in her left upper lobe. CT scan revealed a 1.2 x 1.4cm intrapulmonary nodule in the left upper lobe.

A bronchoscopy was normal, and bronchoalveolar lavage revealed no acid fast bacilli nor malignancy. The patient’s PET scan did not show focal uptake of tracer. A follow-up CT scan was performed at 6 months. The lesion had increased in size to 1.7 x 1.7cm. There was no lymphadenopathy present.

The patient underwent Video Assisted Thoracoscopic Surgery (VATS) and wedge resection. At operation the excised nodule was intra-pulmonary, separate from the pleura, and on section was encapsulated, with pale tan appearance. The histology of the lesion revealed tumour containing thick collagen fibres with spindle cells and haphazardly arranged fascicles. Immunohistochemical staining showed positive stains for CD34, BCL-2 and CD99. It was negative for cytokeratin. This was consistent with a malignant SFT.

Figure 1
Fig 1. Chest X-ray demonstrating a 1.5cm nodule in the left upper lobe and CT image demonstrating the lesion in the left upper lobe with a rim of lung parenchyma.
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Figure 2
Fig 2. H&E Stain. Spindle cells in irregular fascicular arrangement with frequent mitotic figures.

DISCUSSION
Solitary fibrous tumours account for 8% of benign intrathoracic tumours and 10% of pleural tumours. Intrapulmonary fibrous tumours are rare, and are histologically identical to SFTs of the pleura and the literature search draws upon data collected in relation to SFTs of the pleura. It is suggested that the subpleural mesenchyme is continuous with that of the interlobular septa, which can hence give rise to intrapulmonary lesions. The more common pleural SFTs have an incidence of 2.2 cases per 100,000 tumour registries in the Mayo Clinic. In several series, 7% to 37% of SFTs were malignant on histological examination. They have been reported in patients with an age range between 5 to 83 years, with a peak in the fifth decade of life. There is often no history of asbestos exposure.

Up to 48% of patients with SFT are asymptomatic. Briselli et al. reviewed 360 cases in the literature reporting symptoms in 64% of patients, 46% of who presented with cough, 44% with chest pain and 37% with dyspnea. There may be an association with the presence of symptoms and malignant fibrous tumours of the pleura. In a series by P Magdeleinat et al. who reported 22 cases of malignant fibrous tumours 17 of who were symptomatic at presentation (77%). Only 36% patients reviewed with benign disease were symptomatic.

Current treatment of SFT is surgical excision with a clear margin. This has been shown in a number of series to be adequate in definitively treating the majority of cases of pleurally based SFTs both benign and malignant. Patients with completely excised benign lesions showed no recurrence at follow-up (mean 91 months). Those with malignant SFTs with complete surgical excision had recurrence rates between 14% to 42%. In a series of 63 patients by Sung et al. 19 patients had malignant SFTs, and there were 8 reported deaths in this group (42%). All were cancer related and due to metastatic spread of tumour to bone, brain, lung and lymph nodes. Incomplete resection occurred in only 2 of their 19 patients with malignant disease. Their reported mean survival in patients with malignant SFT of the pleura was 24 months. All patients with benign disease were alive by the end of the follow-up period of 29.8 months.

Magdeleinat et al. published a series of 60 patients, 22 (37%) of whom were diagnosed with malignant tumours. In this group 1 died due to post-operative complications, and of the remainder, there were 3 recurrences of tumour. In 2 patients, there was local relapses and were reoperated. Both were disease free at follow-up of 12 and 108 months. They reported only one case of metastatic recurrence with dissemination to the liver. The patient died 32 months after the primary excision of tumour. This group reported 10-year survival rates of 89%. A long follow-up time was recommended given the slow growing nature of the tumour. They concluded that adequate surgical resection was the best prognostic factor in patient survival. There was no data available on the use of adjuvant chemotherapy or radiotherapy.

In conclusion, intrapulmonary malignant solitary fibrous tumours are a rare entity. There is little data available specific to intrapulmonary lesions, the literature being largely limited to a few case reports. However, histochemically they are identical to malignant solitary fibrous tumours of the pleura and are managed as such. Adequate surgical resection with clear margins remains the gold standard in a disease with a small body of evidence to draw upon. With adequate resection, 10-year survival rates of 89% have been reported.

References
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