Solitary fibrous tumours are mesenchymal tumours that can arise in many tissues and organs.¹ The pleural cavity is the commonest location and most tumours are benign. These tumours have previously been described as benign fibrous mesotheliomas and localized pleural mesotheliomas. Multiple pleural tumours, either synchronous or metachronous, are uncommon, as the term solitary fibrous tumour would suggest. We report a patient who had 3 solitary fibrous tumours and stress the need for close follow-up after the treatment of these benign lesions.

CASE REPORT

An 84-year-old, nonsmoking woman complained of a cough. Thirty years previously a right radical mastectomy had been done for stage II infiltrating ductal carcinoma of the breast. Chest radiography 11 years after her mastectomy showed a right pleural abnormality, for which she underwent thoracotomy at another institution. A mass, 12 cm in dimension, was found arising from the right upper lobe by a narrow pedicle. The resected tumour was reportedly a “benign fibrous mesothelioma.”

Physical examination showed previous surgical scars but no significant abnormalities. Her fingers were not clubbed, and the blood glucose concentration was normal. Chest radiography and computed tomography showed a mass in the right anterior-inferior hemithorax (Fig. 1). Bronchoscopy was unremarkable. Thoracic fine-needle aspiration biopsy showed benign appearing spindle cells. Right thoracotomy was performed at our institution. A lobulated, firm, grey-white tumour, 8.5 cm in dimension, was found arising from the diaphragm on a 1-cm pedicle. It was completely resected, without any operative disruption of the tumour capsule. Initial histologic examination showed benign spindle cells arranged in a storiform pattern suggesting neurilemmoma. However, subsequent review of the pathological findings gave a diagnosis of solitary fibrous tumour of the pleura.²,³

Follow-up chest radiography and computed tomography 4 years later...
showed a right anterior mediastinal mass (Fig. 2). A right thoracotomy was performed. There was no evidence of local recurrence in the region of the previously resected diaphragmatic tumour. A pedicled mass, 8 cm in dimension, was found arising from the parietal pleura, in proximity to the internal mammary vessels (Fig. 3). The resected specimen showed bland proliferation of spindle cells, foci of a hemangiopericytomatic pattern and thin bundles of collagen. Although the average mitotic count was 2 per 10 high-power fields, there was no pleomorphism or necrosis. The histologic features were consistent with a diagnosis of solitary fibrous tumour of the pleura. Immunohistochemical studies with CD34 antibody gave positive results, and S100 antibody studies gave negative results. The diagnosis of solitary fibrous tumour of the pleura was confirmed.

The patient is currently doing well after having 3 solitary fibrous tumours resected from the right chest over 23 years. She undergoes follow-up chest radiography twice yearly.

**DISCUSSION**

Primary tumours of the pleura can be divided into 2 major categories: diffuse and solitary.4 Diffuse pleural tumours are mesotheliomas. They are more common than solitary pleural tumours, arise from mesothelial tissue, and are associated with asbestos exposure and almost always run a fatal course.5 Solitary tumours have been known by a variety of names, which are indicative of their clinical course and controversies surrounding their histogenesis. Previous names included benign mesothelioma, localized mesothelioma, subpleural fibroma and localized fibrous tumour of the pleura.4,6,7 These tumours are now known to be of mesenchymal, as opposed to mesothelial, origin; the preferred term is solitary fibrous tumour.1 They are not related to asbestos exposure and follow a benign course in over 80% of cases.8 When malignancy occurs in a solitary tumour of the pleura, it is characterized by pleomorphism, necrosis and a high mitotic rate (more than 4 per 10 high-power fields).9,10 There is no relationship between solitary fibrous tumours and mesotheliomas.1,4 The pleural space is the usual location for solitary fibrous tumours, but they may arise in a wide range of tissues.1 This is not surprising considering their mesenchymal histogenesis.

Solitary fibrous tumours arise from the visceral pleura more often than the parietal pleura.4 They are typically well circumscribed, pedunculated masses, with large vessels within the tumour pedicle. Radiologically, it is often difficult to tell if the mass is pleural or pulmonary in nature. Bronchoscopy is useful to exclude endobronchial tumours, such as lung cancer. Transthoracic fine-needle aspiration biopsy generally shows benign-appearing spindle cells.

Ultimately, thoracotomy with tumour resection is usually required for diagnosis. Histologic examination shows spindle cells with minimal nuclear pleomorphism and infrequent mitoses. The morphologic appearances are very variable and may simulate a variety of soft-tissue tumours.1,4,11,12 In addition, there are no specific ultrastructural characteristics to separate solitary fibrous tumours from other soft-tissue tumours, such as fibrosarcoma. Until recently, immunohistochemical characteristics (keratin negative, vimentin positive, S-100 negative) were helpful but not diagnostic.8 The diagnosis of solitary fibrous tumour has become more precise since CD34 antibody staining was found to be positive in these tumours;
it is now an essential pathological feature for the diagnosis of solitary fibrous tumours.\textsuperscript{1,2} In retrospect, some intrathoracic soft-tissue tumours previously reported in the medical literature were misdiagnosed; they were actually solitary fibrous tumours of the pleura.\textsuperscript{1,3}

Surgical resection of benign solitary fibrous tumours is usually curative. As with all tumours, complete excision is important. This is easily done with pedicled tumours, but great care is required to obtain negative resection margins in sessile tumours.\textsuperscript{4} In addition to the patient reported herein, there are several reports of tumour recurrence years after seemingly adequate surgical resection of benign solitary fibrous tumours.\textsuperscript{1,6,11} Witkin and Rosai\textsuperscript{11} reported 1 recurrence in a series of 14 patients; it occurred 8 years after tumour resection. Okike, Bernatz and Woolner\textsuperscript{6} described recurrence in 2 of 58 patients with benign solitary fibrous tumours. These recurrences were detected 5 and 17 years after tumour resection. Hanau and Miettinen\textsuperscript{1} reported on a patient with 3 recurrences over a 22-year period.

In some of the cases reported in the literature, the tumour recurred locally in the bed of the previous tumour.\textsuperscript{6} However, other “recurrences” were not necessarily found in proximity to the original tumour. The possibility of tumour disruption at the original surgery with secondary seeding at other intrapleural locations, has been suggested as an explanation. Alternatively, the metachronous occurrence of multiple solitary fibrous tumours within the same hemithorax may indicate a multifocal origin of these tumours.\textsuperscript{6} We believe that our patient’s history and operative findings are most in keeping with this latter hypothesis. Irrespective of the possible reasons for metachronous appearance of multiple tumours, 2 practical clinical issues are worthy of emphasis. First, complete resection of solitary fibrous tumours, without intraoperative tumour disruption, is essential. Second, lifelong follow-up with surveillance chest radiography is required to detect further solitary fibrous tumours of the pleura.

References


