Solitary fibrous tumour of the pleura: A case report

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CASE REPORT

Solitary fibrous tumours of the pleura are rare pleural neoplasms that are distinct from mesothelioma. Most of them are benign, although some behave aggressively; morphological and pathological features are important in distinguishing them from mesothelioma and in predicting clinical behaviour. Solitary fibrous tumours often grow to a large size before causing symptoms, and are characteristically associated with hypertrophic pulmonary osteoarthropathy in up to 20% of cases. In cases of benign lesions, complete resection is usually curative. A case involving a 62-year-old woman who underwent surgical resection of a solitary fibrous tumour of the pleura measuring 25 cm in size is described.

Key Words: Fibrous tumour; Pleura; Pulmonary osteoarthropathy

CASE PRESENTATION

A 62-year-old woman presented with progressive exertional dyspnea. Her medical history included obesity, sleep apnea and type 2 diabetes. She was a nonsmoker with no history of asbestos exposure. On physical examination, her breath sounds were decreased on the right side; she did not have digital clubbing. Routine laboratory investigations were normal. Her chest x-ray showed a large opacity in the lower one-half of the right lung field. Computed tomography (CT) imaging revealed a large solid mass in the right hemithorax measuring 25.5 cm × 17 cm × 13.5 cm. The mass was homogenous and well circumscribed but seemed to be adherent to the parietal pleura (Figure 1).

Figure 1) A Chest x-ray showing a large opacity in the lower one-half of the right lung field. B Computed tomography showing a large solid tumour in the right hemithorax

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CT-guided core-needle biopsy was consistent with a solitary fibrous tumour of the pleura. The patient was taken to the operating room and underwent a right posterolateral thoracotomy through the sixth intercostal space. Given the large size of the tumour, a counterincision in the eighth intercostal space was necessary for optimal exposure. The tumour was found to originate from the right lower lobe on a pedicle of visceral pleura. It was widely adherent to the adjacent lung as well as to the diaphragmatic and parietal pleurae. The adhesions were very dense and extremely vascular, and the dissection resulted in substantial oozing, with an estimated operative blood loss of 1200 mL. However, once mobilized, the tumour was easily resected by applying a linear stapler across the base of the pedicle (Figure 2). Postoperatively, the patient experienced some transient dyspnea, which subsequently resolved (a thorough workup did not reveal the underlying cause) and she was discharged in good condition on postoperative day 13. Final histological analysis confirmed the diagnosis of a benign solitary fibrous tumour (SFT) of the pleura; immunohistochemistry was positive for CD34.

DISCUSSION

SFTs are rare (1,2). Electron microscopy and immunohistochemistry have confirmed that they originate from mesenchymal cells in the submesothelial tissue of the pleura and not from mesothelial cells themselves, thus distinguishing them from mesothelioma (2,3). Although most SFTs are benign, approximately 12% are malignant (3). Peak incidence occurs in the sixth and seventh decades of life (3), with both sexes equally affected (3). More than 50% of benign tumours are asymptomatic (2), whereas most patients with malignant SFTs have symptoms, with chronic cough, chest pain and dyspnea among the most common complaints (1,2,4). Interestingly, these tumours are associated with pulmonary osteoarthropathy, with or without digital clubbing, in up to 20% of cases (1,4), although the incidence reported in the literature varies widely (2). Hypoglycemia, believed to be due to the secretion of an insulin-like growth factor, occurs in up to 5% of patients (2-4).

Benign tumours usually originate from the visceral pleura, are pedunculated and grow outwardly into the pleural space (1); malignant tumours tend to arise from the parietal pleura, diaphragmatic pleura or within a fissure, and grow into the lung (1). The presence of symptoms, pleural effusion and lack of a pedicle have been shown to have a statistically significant association with the likelihood of malignancy (2).

In the vast majority of patients, the tumours are large and readily identified on standard chest x-ray (2). A mobile mass may be demonstrated, suggestive of its pedunculated nature (1,3). CT scanning shows a homogenous, well-circumscribed lesion, but is nonspecific (2-4). SFTs can be difficult to distinguish from tumours of the mediastinum and chest wall (1,5). Occasionally, the lesion may appear heterogenous on CT imaging because of hemorrhage or necrosis, making it difficult to distinguish from bronchogenic carcinoma (2,3). Magnetic resonance imaging may be helpful by revealing the characteristic fibrous nature of the tumour (2). Fluorodeoxyglucose-positron emission tomography may help identify SFTs with aggressive features, although the data are limited (6). Fine-needle aspiration may not yield the diagnosis in more than 50% of patients (2,4).

Histologically, SFTs exhibit a proliferation of uniform elongated spindle cells intimately intertwined with various amounts of connective tissue arranged in a haphazard distribution or ‘patternless pattern’ (1,3). CD34 is a specific marker that differentiates SFTs from mesothelioma (4). Criteria used to differentiate malignant from benign tumours include high mitotic activity, presence of necrosis and pleomorphism (2,4). However, these are not 100% reliable because in one review, 2% of patients with tumours categorized as benign on the basis of macroscopic and microscopic features succumbed to the disease (3). Complete resection with negative margin remains the treatment of choice in all cases (1-3); this may necessitate en bloc resection of the chest wall, pericardium or diaphragm in cases of invasion (2). There is no established role for conventional chemotherapy or radiotherapy (1,4); however, recent data on the genetic profile of SFTs suggests that they may respond to tyrosine kinase inhibitors (7,8). Video-assisted thoracoscopic resection may be feasible in selected cases, but port site metastasis has been reported, which may be due to attempts at tumour extraction through small incisions (3,4). Vascular adhesions to adjacent visceral or parietal pleurae are frequent and may bleed significantly (1,2); however, formal lobectomy is rarely necessary (1). The prognosis of benign lesions is excellent, although careful follow-up is necessary (1,4).

REFERENCES
