Localized Fibrous Tumor of the Pleura: An Unusual Intrathoracic Case

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Abstract

Localized fibrous tumors of the pleura are uncommon and their definitive diagnoses are rarely made preoperatively. These tumors can attain massive sizes before becoming symptomatic. Surgical removal is the treatment of choice. In the present study, a patient with a localized fibrous tumor of the pleura notable for its massive size is reported.

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Introduction

Localized fibrous tumors, previously given a variety of names including fibrous mesothelioma, benign mesothelioma, subpleural fibroma and solitary fibrous tumor of the pleura, are rare. Localized fibrous tumors demonstrate malignant behavior in up to 13% of cases (1). Approximately 800 cases of localized fibrous tumors have been reported (2). In the present study, a patient with a giant localized fibrous tumor of the pleura presenting with progressively increasing difficulty in breathing is reported.

Case Report

A 67-year-old man was admitted to the Department of Thoracic and Cardiovascular Surgery with progressively increasing shortness of breath. On physical examination, decreased breathing sounds and dullness on percussion were observed on the left side together with digital clubbing. On admission, the patient was dyspneic and tachypneic. Chest X-ray revealed almost complete opacification of the left lung, with massive rightward shift of the mediastinum secondary to an effusion, and a suspected mass lesion. The patient had smoked 2 packs of cigarettes per day for 20 years. Urine analysis, complete blood count and blood chemistry results were within normal limits. However, erythrocyte sedimentation rate was elevated. In spirometric

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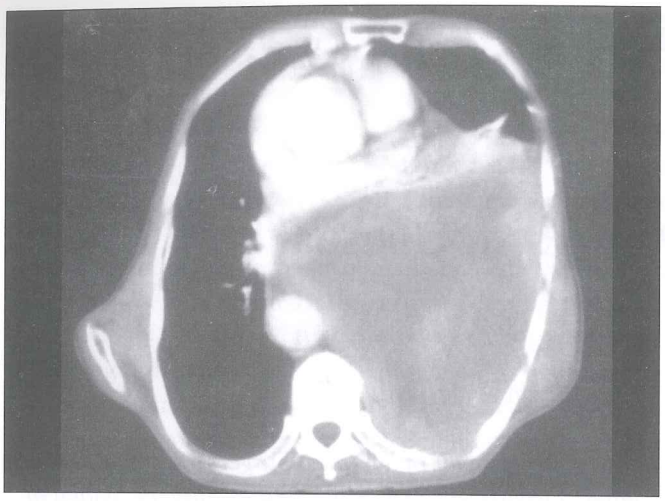

Figure 1. *Computed tomoghraphy of the chest demonstrating a large, well-circumscribed solid mass with displacement of the mediastinal structures.*

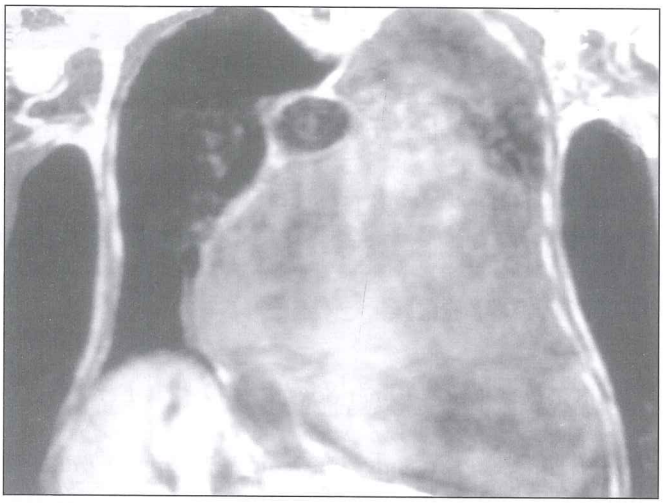

Figure 2. *Magnetic resonance imaging of the chest showing a mass lesion involving almost the entire left hemithorax accompanied by mediastinal shift.*

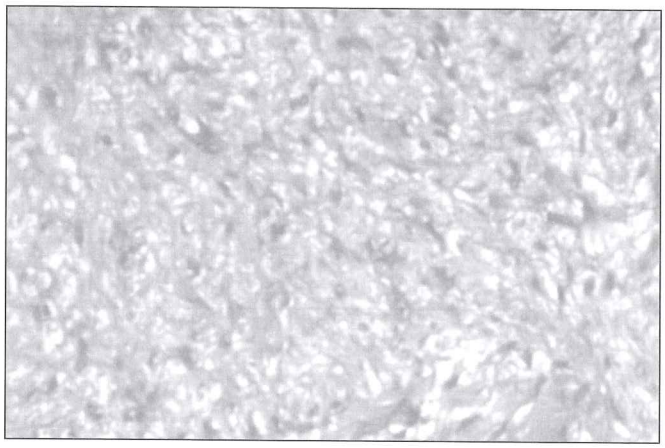

Figure 3. *Pathological specimen shows ovoid or spindle cells without nuclear atypia and sparse mitotic activity scattered haphazardly among strands of collagen fibers.*

evaluation, forced vital capacity (FVC) was 1.35 L (40%), and forced expiratory volume in 1 second (FEV_1) was 0.98 L (37%). Computed tomography of the chest revealed a heterogeneous solitary, well circumscribed, lobulated mass compressing the trachea, heart, lung and other mediastinal structures (Figure 1) with minimal pleural effusion. A large solitary tumor was observed on magnetic resonance imaging of the chest; however, mediastinal and vascular invasion could not be clearly excluded (Figure 2).

On bronchoscopic examination, rightward deviation of the trachea and extrinsic compression leading to the obstruction of the distal left main bronchus were seen. No endobronchial lesion was present. Bronchoscopic mucosal biopsy, bronchoalveolar lavage fluid cytology and transthoracic needle biopsy were non-diagnostic.

The patient underwent left thoracotomy. The tumor was easily dissected from the mediastinal, diaphragmatic and pulmonary surfaces. It was seen as arising from the parietal pleura on a broad base, and was firmly adherent to the thoracic wall. The tumor was completely excised, together with a 3x8 cm segment of 8th rib and adjacent intercostal muscles and soft tissue. No major bleeding occurred but hypotensive episodes developed during the tumor dissection and mobilization.

Macroscopically, the lobulated yellowish-white mass measured 30x17x15 cm with a weight of 3300 g. Microscopically, spindle and fusiform cells were seen in an abundant collagenous stroma with dispersed vessels (Figure 3). The tumor cells were CD34 positive and cytokeratin (AE1/AE3) negative using monoclonal antibodies with strep-avidin biotin method. Malignant behavior was ruled out due to lack of mitotic activity (<4 mitosis/10 high power fields) and absence of pleomorphism and hemorrhage or necrosis. The mass was totally encapsulated. Histologic examination showed a benign fibrous tumor of the pleura.

The patient had no complications in the postoperative period and was discharged on the 7th postoperative day. He was found to be free of recurrence in the second year following the resection and his clubbing had resolved.

Discussion

Localized fibrous tumors are often initially detected as asymptomatic masses on chest X-rays obtained for other purposes. Symptoms are generally related to the size of the tumor, and include cough, chest pain and dyspnea. The large tumors may be associated with hypoglycaemia in less than 5% of the cases, clubbing and hypertrophic osteoarthropathy in 10-20% of the cases (2). Hypoglycaemia is related to the release of insulin-like growth factor II (IGF-II) (2,3). The

causes of digital clubbing and of hypertrophic pulmonary osteoarthropathy could be an abnormal production of hepatocyte growth factor or of hyaluronic acid by the tumor (2). Our patient had clubbing, but hypoglycemia was not present. It was reported that these paraneoplastic syndromes are regressing after surgical resection of the mass, as also observed in our patient (3-5). These localized pleural tumors tend to enlarge, attain massive sizes before leading to symptoms, are histologically suggestive of low-grade tumors and are amenable to surgical extirpation. Weynand et al. reported that in a patient refusing surgical treatment, the tumor had remained clinically silent despite its volume (estimated by planimetry on chest X-ray) increasing from 65 to 550 cm^3 over a period of 22 months (6).

It is difficult to establish the diagnosis before operation. It has been proposed that a confident preoperative diagnosis can be made by transthoracic fine needle biopsy (6). However, some authors state that preoperative transthoracic needle biopsy is not necessary since it does not exclude malignant variants and does not influence the need for surgical resection (7). In the present case, transthoracic needle biopsy of the mass demonstrated benign fibrous tissue, yet the final diagnosis was obtained upon surgical resection.

Localized fibrous tumors more frequently originate from the visceral rather than the parietal pleura (1,8). Macroscopicaly, most tumors appear as solid, encapsulated, firm and lobulated masses and their cut surface demonstrate a whorled pattern. Tumor sizes range from 1 cm to 39 cm, while massive tumors can weigh up to 3800 g (8). The major concern about localized fibrous tumors is that it is difficult to differentiate their benign or malignant nature. England et al analyzed the histological aspects of localized fibrous tumors and defined criteria of malignancy as high cellularity, nuclear pleomorphism, more than four mitosis per ten high-power fields, presence of necrotic or hemorrhagic areas, and invasion of adjacent structures (8). Immunohistochemical studies demonstrate these tumors to be negative for cytokeratin and positive for vimentin and CD34. This immunoprofile shows that the tumor cells are not mesothelial in origin and supports the fibroblastic origin of these tumors (3).

Despite the large sizes of some of these tumors, they tend to be confined to a small area of the pleura and are often pedinculated (9). Especially in tumors arising from the parietal pleura, localized chest wall involvement is apparent and the involved chest wall should be resected. Although the underlying lung parenchyma can usually be preserved, rarely there is a need for wedge resection during surgical intervention. During the removal of the tumor, hemodynamic changes associated with the decompression or compression of the mediastinal structures can lead to serious cardiopulmonary complications. Okike et al. have reported a patient developing cardiac arrest during the mobilization of the tumor. Moreover, massive bleeding may develop during dissection (4). In the present case, during removal of the mass, cardiac arrest or massive bleeding did not develop despite the occurrence of serious hypotension.

Although histologically benign, localized fibrous tumors of the pleura may undergo malignant transformation (4,7). These tumors may also have recurrences with benign morphological characteristics, similar to those of previously removed tumors (10). Local recurrences have been reported as late as 17 years following surgical excision (10).

Surgical resection is recommended in all cases due to the malignant and expansile potential, and is curative in most of the patients (7). The success of the treatment and survival expectancy is dependent on the adequacy of complete resection. Long-term follow-up is necessary for these tumors, even if the tumor has been completely removed with benign features in histological examination.

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