A case of giant benign localized fibrous tumor of the pleura

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A 57-year-old man presented with progressively increasing shortness of breath. A chest roentgenogram showed the presence of a giant mass and magnetic resonance of the chest confirmed the mass with an inhomogeneous density in the right hemithorax. The patient underwent a thoracotomy. A tumor was seen to have arisen from the parietal pleura. Size of the tumor was 22x19x15 cm and weighed 2650 g. It was successfully resected. The pathological diagnosis of the tumor was giant benign localized fibrous tumor of the pleura. [Turk J Cancer 2000;30(4):175-180]

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Localized fibrous tumors of the pleura are rare neoplasms. Previously these tumors have mostly been classified as localized mesotheliomas of the pleura, either benign or malignant (1). There is no clear relationship to asbestos exposure. This neoplasm originates from pluripotential mesenchymal cells located in the subserosal connective tissue (2). On the average it measures 5 to 6 cm in diameter (3). Over six hundred cases of localized fibrous tumor of the pleura have been reported in the world literature (4).

We report a case of a patient with benign localized fibrous tumor of the pleura, which is notable for its rare massive size.

Case Report

A 57-year-old man presented with progressively increasing shortness of breath. PA and lateral chest roentgenograms showed presence of a giant mass (Figure 1) and magnetic resonance of the chest confirmed the mass to have an inhomogeneous density in the right hemithorax. There was no history of exposure to asbestos. Hemogram and biochemical findings were normal. Blood glucose level was 69 mg/dl.
FIBROUS TUMOR of the PLEURA

Fig 1. Preoperative chest roentgenogram showing a mass involving the right hemithorax

At physical examination, breath sounds were absent on right lower zone. Bronchoscopy revealed a leftward deviation of the trachea and extrinsic compression causing obstruction of the distal right main bronchus. Right posterolateral thoracotomy was performed for the resection of the tumor. No pleural effusion was encountered at the time of operation. The tumor was arising from the parietal pleura. A chest roentgenogram on the tenth postoperative day showed good reexpansion of the right lung.

Grossly the tumor had a solitary lobulated shape that is covered by a smooth glistening capsule and that had a broad base pedicle. Size of the tumor was 22x19x15 cm and weighed 2650 g (Figure 2).

The cut surface had a nodular, firm, gray-white appearance, small cysts and areas of hemorrhage (Figure 3).

On histologic examination, the tumor were composed of uniform collagen-forming spindle cells, which were arranged in interlacing fascicles and accompanied by the deposition of abundant collagen, and showed no mitotic activity (Figure 4). Some areas had myxoid changes and hyalinization. Immunoreactivity for vimentin was observed (Figure 5). However, there was no immunoreactivity with antibodies to cytokeratins, desmin, epithelial membrane antigen, smooth muscle actin, S100, and factor VIII.

Pathological diagnosis was giant benign localized fibrous tumor. He was doing well at the 8-weeks of follow-up.
Fig 2. Grossly, a solitary lobulated mass covered by a smooth glistening capsule

Fig 3. The cut surface showing a nodular, firm, gray-white appearance
Fig 4. Benign localized fibrous tumor of the pleura. Interlacing fascicles showing tightly packed spindled cells (hematoxylin and eosin, x200)

Fig 5. Vimentin immunoreactivity in the cytoplasm of many spindled tumor cells (labelled streptavidin biotin method, x400)
Benign solitary fibrous tumors, previously called benign mesotheliomas, are rare, with approximately 600 cases reported in the literature (4). The localized form of the fibrous tumors are usually considered benign, but malignant variants of these localized tumors have been reported (5,4). The benign variant is three to four times as common as the malignant one (6). These tumors have slightly higher incidence in females than males. The peak incidence is in the sixth and seventh decades of life (6). Our case was a 57-year-old man.

The usual presentation is an asymptomatic mass discovered incidentally on a chest radiograph (4). However, cough, chest pain, dyspnea, and clubbing are seen in %30 of the patients and are more common with tumors greater than 10 cm and those with malignant cellular features (4,5). At the time of surgery our patient was symptomatic.

Because of release of insulin-like peptide, hypoglycemia occurs in about 5% of the solitary fibrous tumors of the pleura, especially with those of large size (3). Blood glucose level of our patient was 69 mg/dl.

Unlike the malignant form, benign localized fibrous tumor of the pleura appears to be unrelated to asbestos exposure, but rare instances of this association have been described (3). There was no asbestos exposure in our case. Eighty percent originate from the visceral pleura and 20% from the parietal pleura (6,1).

The size of the tumors ranges from 1 cm to a massive tumor measuring 39 cm and weights range from 12 to 3.800 g (7). These tumors are usually less than 10 cm in size and are pedunculated (4). Grossly the tumor manifests as a solitary localized mass that is attached with a richly vascularized pedicle or a broad base to the visceral or parietal pleura that is covered by a smooth glistening capsule (4). The cut surface has a uniform or nodular, firm, gray-white mass with a characteristic whorled appearance or shows a variegated picture with alternating firm and soft myxoid areas, sometimes with cystic spaces and areas of hemorrhage (3).

Although most solitary fibrous tumors are benign, up to 13% of the localized tumors can have malignant characteristics and a more aggressively fatal course (6). The malignant variants can be recognized by their greater cellularity, cellular pleomorphism, and increased mitotic activity, usually more than four mitotic figures per 10 high-power fields (3). There was no mitotic figures in our case.

Infrequently a pleural effusion may be present. England et al. (7) reported that 8% of 138 patients had a pleural effusion. We did not encounter pleural effusion at the time of operation.

Treatment of benign fibrous tumors of the pleura consists of adequate local excision. Great care must be taken at the time of operation because of the highly vascular pedicle (8). The overall operative mortality is reported to be 12% because of hemodynamic changes associated with decompressing the mediastinal structures (6).

Although most tumors are considered histologically benign, local recurrences and rapid enlargement without signs of invasion or metastasis have been reported (9). Perrot et al. (9) reported a case that recurred with
fibrosarcomatous features after 6 years complete resection. Also, local recurrence has been reported as late as 17 years after surgical excision (5).

Survival is directly related to whether the tumor can be resected completely and the presence of malignant changes histologically (4). Long-term annual follow-up with chest roentgenograms is highly recommended (9). After two months follow-up, our patient is healthy and has no sign of recurrence.

References