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A giant fibrous tumor of the pleura mimics plural effusion: a case report and review of the literature

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Summary

Background: A solitary fibrous tumor of the pleura (SFTP) is very rare with an incidence of less than 5% of all pleural neoplasm. Although some tumors may reach an enormous size, they are often asymptomatic and have sometimes been mistaken as a pleural effusion.

Case Report: Herein we describe a 33 yr-old male patient who diagnosed with a giant SFTP. Chest radiograph revealed a large homogenous opacity filling the right hemithorax that was initially reported as a pleural effusion. Pleural tap was performed twice and turned out to be dry. Computed Tomography) of the chest unexpectedly showed a large hypodense mass filling the right hemithorax. Transthoracic CT guided Tru-Cut biopsy revealed a benign solitary fibrous tumor of pleura that was resected surgically, and the patient made an eventual recovery.

Conclusions: SFTP is a rare tumor. High index of clinical suspicion is essential to reach diagnosis. Long term follow up is important to assess for recurrence.

Key words: **Fibrous tumor • Pleural effusion**

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Background

A solitary fibrous tumor of the pleura (SFTP) is a rare disease, representing less than 5% of the total pleural neoplasm. The majority of these tumors (88%) are benign [1]. It arises in 75% of presenting patients from the submesothelial connective tissue of the visceral pleura, while in the remaining 25% from the parietal pleura [2]. It affects men and women equally [3]. The peak incidence is usually between fourth to sixth decades [2-4]. Forty percent of the patients are asymptomatic at the time of presentation, while the remaining may present with chest pain, cough, dyspnea, fever and/or pleural effusion [5]. Some tumors reach an enormous size and may be mistaken as a pleural effusion [6, 7]. Some tumors have been discovered incidentally either during routine chest radiograph or by CT scan of the chest. Diagnosis is usually obtained after surgical resection of the tumor and rarely reached preoperatively by Tru-cut biopsy [5, 6, 8]. Surgical resection of the tumor is the main line of therapy [9]. Recurrence rate may reach up to 2%, and may require further resection [5, 9]. The aim of this study was to describe an extremely rare case of a huge solitary fibrous tumor of the pleura that was initially mistaken as pleural effusion and to review the relevant literature.

Case Report

A 33 yr old male Yemeni patient, who was working as a salesman, was admitted to King Abdulaziz university hospital (KAUH) through the emergency room (ER) in mid November, 2005. He presented with cough, dyspnea and pleuritic chest pain of 2 months duration that was associated with fever and rigors. There was no history of anorexia, weight loss or hemoptysis. Systemic review was unremarkable. He had no history of smoking, alcohol or drug abuse. *O*n examination: he was febrile with a temperature of 37.8 C and his blood pressure was 110/70 mmHg. His height was 161 cm and weight 63 kg. His oxygen saturation was 97% at room air. He had no clubbing, cyanosis, or palpable lymph nodes. Chest examination showed signs of consolidation / collapse and pleural effusion over the right hemithorax. Examination of the Cardiovascular, abdominal and central nervous systems were unremarkable. Initial investigations showed Hg: 13 gm/dl, WCC: 8.5/cmm. ESR: 3 mm/h, CRP: 150u (N <10u). Chest radiograph revealed a large homogenous opacity filling the right hemithorax and pushing the mediastinal structures anteriorly and to the left that was initially mistakenly reported by a senior radiologist as a massive pleural effusion. He consequently underwent 2 unsuccessful attempts of pleural aspiration. CT scan of the chest was performed and unexpectedly revealed a huge multilobular, oval, well-defined encapsulated hypodense mass compressing the adjacent lower lobe of the right lung and pushing the mediastinum anteriorly. There was no mediastinal lymph nodes enlargement (Fig 1). Tru-cut biopsy was performed and the histopathology showed benign fibrous tissue**.** Bronchoscopy showed narrowing of the right middle and lower bronchial segments mainly due to external compression. The patient underwent surgical resection. The tumor was large oval, encapsulated**,** firm in consistency, grayish in color, measured 25 x 20 x 15cm and weighed 1600 grams and arose from the visceral pleura (Fig 2). The cut section was whorly. It was firmly adjacent to the lower lobe of the right lung but macroscopically had no bronchial or diaphragmatic infiltration. The histopathological diagnosis was localized benign fibrous tumor of the pleura (Fig 3). Immunohistochemistry of the neoplastic cell were positive for CD34 and vimentin and negative for pankeratin (CK, AE1, and

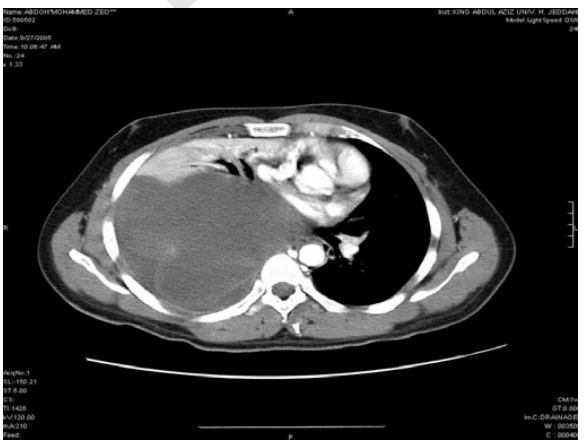

Figure 1. CT scan of the chest showing multi lobular well encapsulated thin walled mass pushing the mediastinum anteriorly.

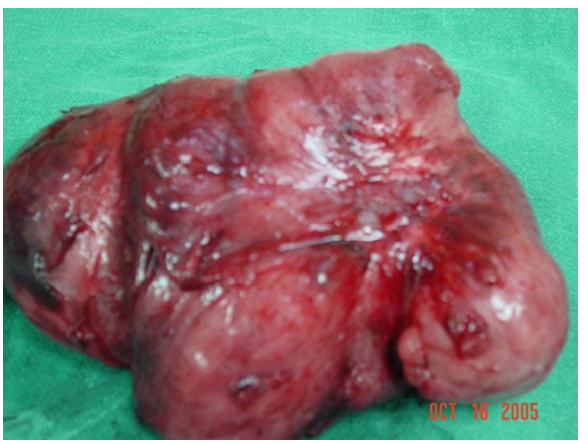

Figure 2. The tumor after resection showed a large multilobulated oval, encapsulated mass that measured 25 x 20 x 15 cm and weighed 1600 grams.

AE3) and calretinin confirming the diagnosis of solitary benign fibrous tumor. Post-operatively, his right lung fully expanded and regained normal size. Two months later, he was seen in the outpatient clinic in good general condition.

Discussion

Localized fibrous tumor of the pleura (LFTP) is rare, accounting for less than 5% of all reported pleural tumors [1, 11]. In the literature, up to 500 cases have been reported [1]. It usually considered benign, but malignant changes may also occur (11). The peak incidence is usually between the fourth to sixth decades of life [1, 8]. However, children may also be affected [1]. In our patient the tumor was diagnosed in his early thirties. Patients usually present with chest pain, dyspnea, and cough and rarely with hemoptysis. Extrathoracic manifestations may include a wide range of symptoms such as weakness, weight loss, nocturnal sweating, fever and chills [1, 11]. Symptoms tend to be more common when a larger tumor is involved [1]. More than 50% of reported cases were found incidentally during routine chest radiographs [5, 10, 11]. In our patient, fever, chills, chest pain, and dyspnea were the main presenting symptoms. Digital clubbing, and or hypertrophic osteoarthropathy have also been reported in up to 20% of the patients [2, 11]. Interestingly, these changes are more common in patients with LFTP than with bronchogenic carcinoma. It is mainly attributed to the fact that tumor cells may produce hyaluronic acid, which has an osteolytic effect [8]. Recurrent episodes of hypoglycemia have also been observed in 2–4% of reported cases [3, 11]. This is due to the production of the insulin-like growth factor II (IGF-II), which causes an increase of glucose utilization and an impairment of growth hormone counter-regulatory response to hypoglycemia [10, 11]. Such symptoms may regress completely after surgical resection [1, 11]. In our patient there was no digital clubbing, hypertrophic osteoarthropathy, or hypoglycemic episodes although the size of the tumor was significantly large. The differential diagnoses of LFTP should include; malignant mesothelioma, metastatic carcinoma, sarcomas, pleural lipoma, bronchogenic cyst, pulmonary abscess, and bronchogenic carcinoma [1, 9]. Chest radiograph is an important diagnostic tool, but may not be specific. Large SFTP may appear as a homogeneous opacity in the chest radiograph that may be mistaken as pleural effusion as occurred with our patient [7]. However, the majority of the patients with SPFT have no pleural effusion [3, 5, 9]. A previous study by England et al. found that approximately 8% of 138 patients with SPFT had a pleural effusion [3].

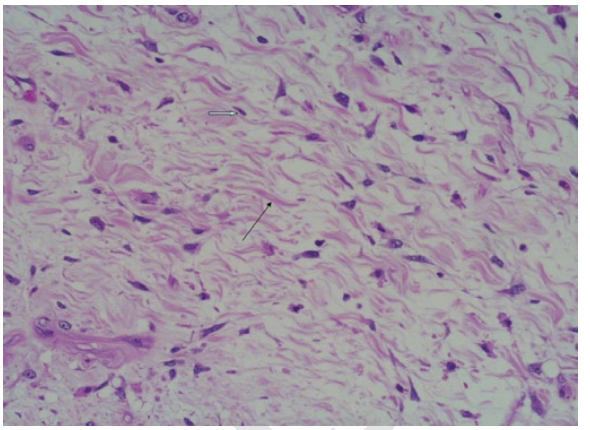

Figure 3. Histopathology of the tumor showed cells scattered haphazardly (upper arrow) among strands of collagen fibers (lower arrow) (HE, ×165).

A CT scan is more sensitive than chest radiograph in diagnosing SPFT. The usual finding is a lobulated mass with variable heterogeneity according to the grade of necrosis with and without calcifications. It is useful in assessing the relationship of the tumor with the adjacent structures including mediastinum and to detect the presence of the pleural effusion, if any [4, 9]. Magnetic Resonance Imaging (MRI) also helps surgeons to define the borders between the tumor and the surrounding structures before resection [11, 12]. The role of Positron Emission Tomography (PET) has not yet been defined [9]. Bronchoscopy also has a minimal role in the diagnosis of such tumors as in our patient [5]. Transthoracic CT-guided biopsy does seem to be a reliable diagnostic method, particularly if good tissue can be obtained [5, 9]. In our patient the diagnosis was reached by Tru-cut biopsy that was consistent with the histopathology of the excised tumor. The tumors appear macroscopically as encapsulated, firm and lobulated masses with a characteristic whorled appearance. The size of the tumors is variable and ranges from small nodular masses within the lung to large pedunculated tumors within the pleural cavity [2, 13], which may weigh up to more than 1500 grams [2, 4]. In our patient the weight of the tumor was 1600 grams. Histologically, the tumors are classified as benign or malignant according to the criteria used by England et al. [3]. Malignancy should be suspected if one or more of the following criteria is present; 1) high mitotic activity, 2) high cellularity with crowding and overlapping of nuclei, 3) presence of necrosis and 4) pleomorfism. The incidence of malignancy in the LFTP is variable in different studies and enncompasses 30% to 38% of all reported cases [5, 11]. The management of SFTP consists of adequate surgical resection during which great care must be taken to avoid bleeding tendency due to high vascularity of the tumor pedicle and surrounding adhesions [1, 5, 11]. The

intra-operative mortality rate is about 12% and mainly due to decompression of the mediastinal structure that leads to fatal cardiopulmonary arrest [4, 7]. The recurrence rate is low, reported as occurring in 2% of patients presenting with benign tumor [9]. The factors increasing the incidence of recurrence include the size of the tumor, presence of necrosis in excised tissue, cell polymorphism, number of mitosis and resectability [3, 8, 13]. Recurrence may occur up to 17 years after initial resection of the primary tumor, and therefore, regular, long term follow up is highly recommended [5, 8, 9, 11].

Conclusions

SFTP is a rare tumor. Recurrence may occur up to 17 years after initial resection of the primary tumor, and therefore, regular and long term follow up is highly recommended [5, 8, 9, 11] to assess for recurrence.

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