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SHORT COMMUNICATIONS AND CASE REPORTS

Fibrous tumor of the lung: diagnostic and therapeutic considerations apropos of two cases

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Summary

Solitary fibrous tumor (SFT) of the pleura and the lung is an uncommon spindle cell neoplasm arising from the visceral pleura in the majority of the cases. Current diagnostic and therapeutic considerations are discussed apropos of 2 recent cases. 1st case: A 46-year-old male, heavy smoker, presented with nonspecific complaints and a mass in the left posterior mediastinum at the level of the 5th thoracic vertebra. Computed tomography (CT) and magnetic resonance imaging (MRI) of the chest confirmed the position of the mass in proximity to the thoracic wall at that level. Through a left posterolateral thoracotomy, a 4×5×2 cm mass, mushroom-shaped, stalky, completely covered by the lung parenchyma, was resected. Histopathological examination revealed a SFT of the lung. 2nd case: A 54-year-old woman presented with symptoms of respiratory distress and persistent hypoglycaemia. CT scan of the chest showed a huge mass within the right hemithorax, compressing the mediastinum and the contralateral lung. The mass was initially detected on chest radiography and CT scan 3 years before resection. Fine needle biopsy (FNB) results were inconclusive and resection of the mass through a right thoracotomy established the diagnosis of malignant SFT. Resection of the mass resulted in recovery of respiratory function and the paraneoplastic hypoglycaemia.

Radiologic features and inconclusive fine needle aspiration (FNA)/FNB results make preoperative diagnosis of SFTs of the pleura and lung difficult. Diagnosis of SFT will be established with certainty after surgery. Resection with clear margins is the main important prognostic factor.

Key words: fine needle biopsy, localized solitary fibrous tumor of the pleura, solitary fibrous tumor of the lung, solitary fibrous tumor of the pleura, uncommon pleura neoplasm, spindle cell neoplasm

Introduction

SFT is the preferred term for an uncommon (about 800 cases are reported in the medical literature until today), but histomorphologically distinctive spindle cell neoplasm, that was referred in the past to as fibrous mesothelioma, localized fibrous mesothelioma, localized fibrous tumor, localized mesothelioma, pleural fibroma, solitary fibrous mesothelioma, or submesothelial fibroma [1]. Visceral pleura was considered for a long time to be the exclusive location of these slow-growing tumors; occasionally they were also found within the lung parenchyma, without any obvious attachment to the visceral pleura [1,2]. SFTs are now reported to arise also from mediastinal, diaphragmatic or parietal pleura, within a lung fissure and in various extrapleural locations, such as the retroperitoneum, mediastinum, thyroid gland, nasal cavities, meninges and parietal surfaces of the intra-abdominal viscera [1-3].

SFTs of the pleura and the lung are often penduculated masses of variable size with lobulated appearance and easy to remove in the majority of the cases [1-3]. Histologically they are composed of spindle cells with oval nuclei, typically arranged in a "patternless" pattern, separated by thin "strip-like" bundles of collagen. A haemangiopericytoma-like vascular network is typical and 90-95% of SFTs have positive staining for vimentin and the surface receptor cluster of differentiation 34

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(CD34). Bcl-2 and CD99 stainings are also positive in about 50% of SFTs [1-6].

Two recent cases of SFT of the pleura and the lung are reported herein, where the radiographic appearance and preoperative FNB results of the lesions did not establish a preoperative diagnosis, which was finally established after resection. Preoperative radiographic and clinical findings are discussed together with the intraoperative findings during exploratory thoracotomies.

Case presentations

Case 1

A 46-year-old male, heavy smoker, presented with non-specific complaints of malaise and fatigue. His medical history was unremarkable and there was no asbestos exposure. Haematologic and blood chemistry studies were normal. Routine posteroanterior and lateral chest radiography showed a mass in the posterior mediastinum on the left side, at the level of the 5th thoracic vertebra. Helical chest CT scan and MRI of the thorax confirmed the position of the mass in close proximity to the thoracic wall at the level of T4 and T5 thoracic vertebrae (Figure 1).

The mass was inaccessible to FNB and the patient underwent a left posterolateral thoracotomy. A 4×5×2 cm mushroom-shaped, stalky mass was found in continuity with the parenchyma of the apicoposterior segment of the left upper lobe. A wedge resection of the mass was performed (Figure 2). Gross examination revealed a well circumscribed, non-encapsulated mass, with whitish and firm appearance (Figure 2). At histology, numerous dilated vessels (haemangiopericytoma-like vessels) were found within the stroma of the tumor. The tumor cells showed round to spindle appearance, with little cytoplasm, indistinct cell borders and dispersed chromatin with vesicular nuclei, and they displayed strong immunoreactivity for desmin, vimentin and CD34. The final diagnosis was benign SFT of the lung. The patient had an uneventful recovery and he was discharged on the 6th postoperative day. He is doing well 3 years after resection, without any sign of local recurrence.

Case 2

A 58-year-old woman, without smoking history and asbestos exposure, was admitted with a huge mass in the right hemithorax that compressed the surrounding lung parenchyma, the mediastinum and the contralateral

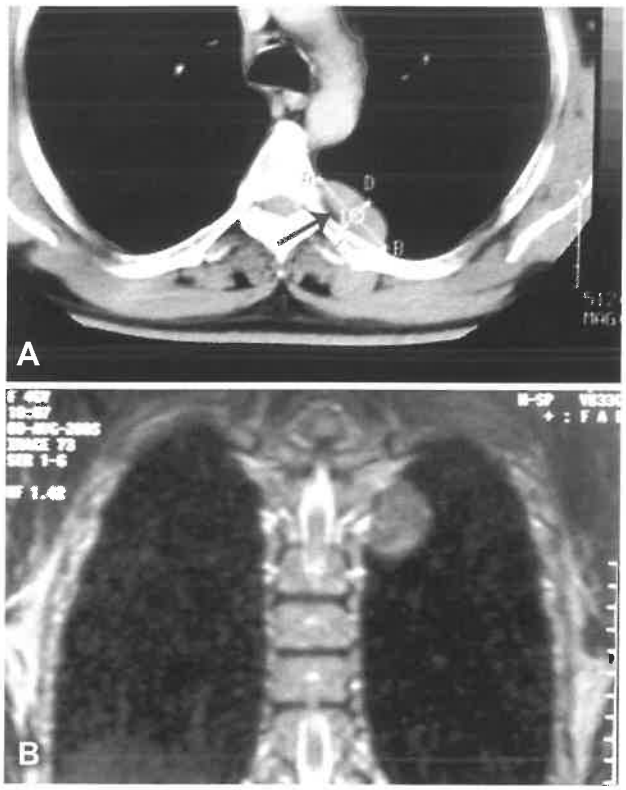


Figure 1. A: CT scan of the chest: well circumscribed, soft tissue, paraspinal (left T4-T5) mass. Note (arrow) the margin between pleura and tumor (hypodense area mimicking air or fat). This fact was appreciated only postoperatively (lung vs. pleural tumor). B: Chest MRI (sagittal view): extrapulmonary, well circumscribed, broad-based mass in the mediastinum at the T5 level (left) without bony changes of the ribs or vertebrae.

lung, resulting in dyspnea on minimal exertion (Figure 3). The mass was initially detected on chest radiography and CT scan 3 years before, but the patient refused to undergo further evaluation, as she was completely asymptomatic at that time (Figure 4). Arterial blood gases on room air at last admission were: pO_2 = 49 mm Hg, pCO_2 = 32 mm Hg. The mass was associated with persistent hypoglycaemia (blood glucose levels < 50 mg/dl), despite continuous intravenous glucose administration. FNB of the mass was inconclusive and the patient underwent right anterolateral thoracotomy for resection of the mass. A huge, multilobulated mass (14×12×10 cm, 1500 g weight) was found, that compressed the parenchyma of all 3 lobes. The mass had large serpigineous feeding vessels on its surface. By dividing the feeding vessels between haemostatic clips the mass was delivered out by sharp and blind dissection from the surrounding structures. Wedge resection of the attached to the mass segments of the upper and middle lobe accomplished the operation, to obtain clearance of the tumor. The patient had a 6-day uneventful recovery. Decompression of the lung parenchyma re-established

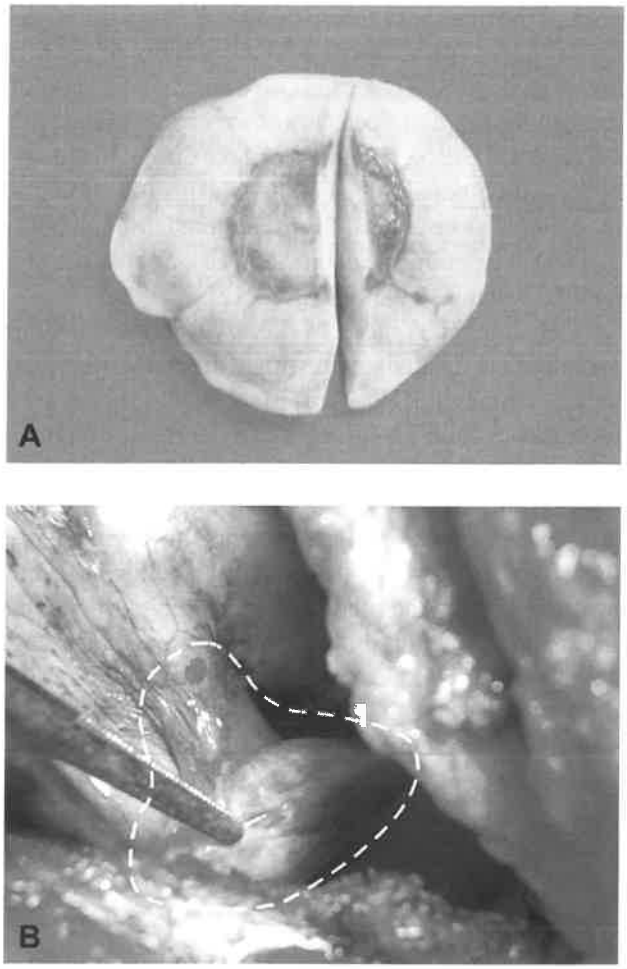


Figure 2. A: 4×5×2 cm mushroom-shaped mass. B: operative picture: stalky mass (dotted line) arising from the apicoposterior segment of the left upper lobe.

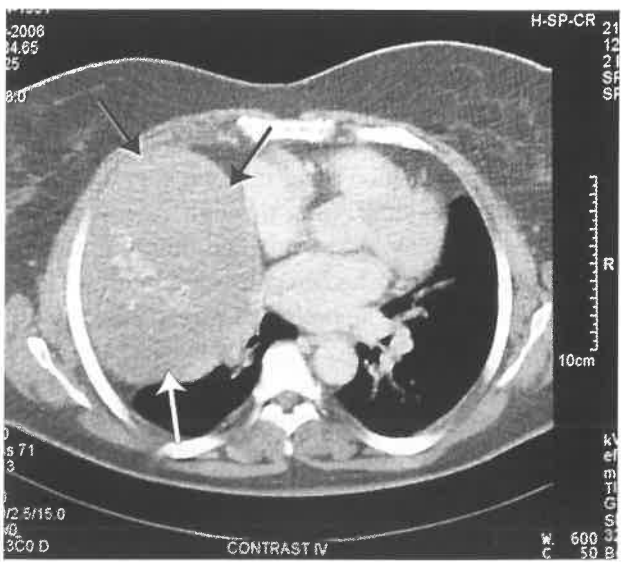


Figure 3. Spiral CT scan of the chest: huge right-sided mass, compressing the mediastinum and the contralateral lung (arrows).

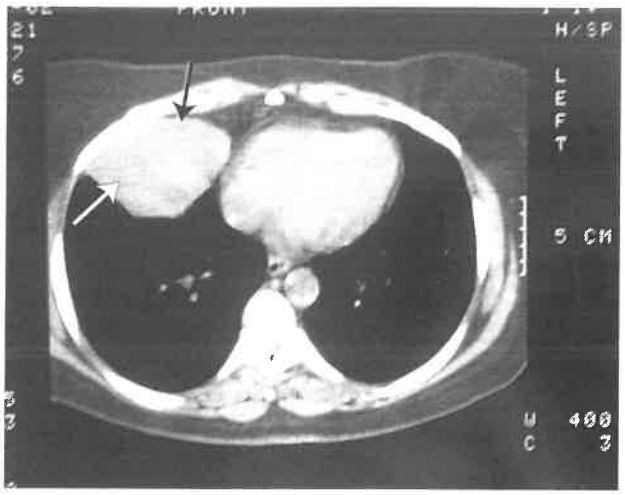


Figure 4. CT scan of the chest: the mass at initial presentation, 3 years before surgery (arrows).

respiratory function and hypoglycaemia disappeared upon resection of the mass. Histology of the mass showed a spindle cell neoplasm with intense vascularity, indistinct cell borders and strong immunoreactivity of tumor cells for vimentin, Bcl-2 and CD34. The final diagnosis was that of SFT with potential malignant behavior, a diagnosis based on the criteria of high mitotic activity (5 mitoses/10 high-power fields), existence of extensive areas of necrosis and maximum diameter of the mass more than 10 cm.

Discussion

SFTs of the pleura and lung are considered to be tumors with favorable clinical outcome [1-6]. SFTs are not related to asbestos exposure and usually are pleural-based or may arise in the lung periphery or, at instances, may be completely intrapulmonary [1-6]. Intrapulmonary location of SFTs is rarely observed, as in the first described case. Possible explanations for these intrapulmonary locations of SFTs include the direct continuity of subpleural mesenchyma with the connective tissue of intralobular pleura or the origin of the tumor from fibroblasts of the pulmonary parenchyma itself or the inversion of a visceral pleura-based tumor into the pulmonary parenchyma, in a "pushing" fashion [7,8].

Radiologically, they are usually found as ovoid or round lobulated nodules or masses, of highly variable size on plain chest radiography (>50% of cases). CT findings are those of a well-defined nodule or mass, showing soft tissue attenuation, with areas of necrosis, myxoid and cystic degeneration (low attenuation areas) present in larger tumors. Heterogeneous enhancement

of the tumor after intravenous contrast administration, which is more intense in the periphery of the tumor, is described to be a common but not pathognomonic CT finding in SFTs of the lung and pleura [1,3-6]. The pattern of tumor's feeding vessels on its surface seen during thoracotomy in the second described patient explains the CT findings reported in the literature. Penduculated tumors may show a marked motility and change their position in serial chest radiographs, an additional radiographic criterion to arise suspicion of SFT if serial radiographs are available [6]. In the first reported case the tumor had radiographically a broad base attachment to the thoracic wall and vertebrae, resembling the image of a pleural or chest wall tumor, while the surgical findings were those of a penduculated mass completely covered by lung parenchyma [3]. Atypical images are reported in most of SFTs in the literature, which makes their radiographic diagnosis problematic.

Small lesions are asymptomatic and they are incidentally discovered during chest radiography performed for other reasons. Symptoms are common in large lesions, including local symptoms (because of compression of adjacent structures) or systemic symptoms (clubbing of the fingers, hypertrophic osteoarthropathy and hypoglycaemia), that are present in about 5% of the cases [4,5,9,10]. Persistent hypoglycaemia should pose the suspicion of SFT in connection with the radiographic image described above. Indeed, hypoglycaemia is a paraneoplastic syndrome observed also in lung cancer and mesothelioma. Primary lung cancer should always be included in the differential diagnosis of any intrathoracic mass associated with hypoglycaemia found in a smoker. Of importance is the event that both local and systemic symptoms produced by SFTs regress after complete resection of the masses [5,10,11].

In our 2nd reported case, FNB results did not establish a preoperative diagnosis despite the large dimensions of the mass, probably because of the extensive areas of necrosis within the tumor. FNA/FNB of a tumor located in the lung periphery or having the radiographic appearance of a pleural mass is always the first step in preoperative work-up, as the mass is easily accessible to transthoracic FNA/FNB and usually not accessible to bronchoscopy. However, the method is considered to be of limited value for the diagnosis of SFTs, as both cytologic and histologic results are usually non-conclusive [4,5,8-10,12]. In 2 large published series, each one including a retrospective analysis of 11 cases of SFTs of the pleura who underwent preoperative FNB, a preoperative diagnosis was established in 45% and 36% of patients, respectively [4,5]. However, in a recent report of 7 patients with fibrous tumor of the lung and pleura, FNA of the mass was considered to be a very useful diagnostic tool [4]. According to the authors of the last mentioned study, FNA can pose a strong suspicion of the diagnosis (spindle cells in a bloody background with interspersed collagen) and even make differential diagnosis from other spindle cell neoplasms (mesotheliomas, synovial sarcomas, fibrosarcomas) through positive CD34 reaction in cell blocks, as it was possible in 5 out of 7 patients of the study [2-4,12].

Histological criteria to classify a SFT of the lung and pleura in the malignant variant are those described by England et al. (1989): a) more than 4 mitoses/10 high-power fields; b) presence of necrosis; c) hypercellularity as detected by nuclear crowding and overlapping; d) nuclear atypia [13]. Tumor size is an important prognostic factor, as malignant variants are usually more than 10 cm in diameter, with areas of haemorrhage and necrosis [5]. On the other hand, favorable prognostic factors are encapsulation, penduculation and resectability with free surgical margins, even in histologically malignant variants. Infiltrated margins are associated with aggressive clinical course and high rates of local recurrences and metastases [3,5,10,11]. In large series the recurrence rate of benign SFTs is reported to be low (1.4%), while the recurrence rate of malignant variants is reported to be higher (9-19%) [2,5]. On the other hand, local recurrences and distant metastasis may occur even 5 years after excision and irrespective of the histological features of the resected tumor. A long-term follow-up is needed, because of the possibility of late recurrence and metastases [2,4,12,14]. Local recurrences detected early are amenable to reoperation and resection, with good long-term results [6,14]. The role of adjuvant therapy in SFTs with poor prognostic factors (size >10 cm and infiltrated margins) still remains unclear [2]. The overall 5- and 10-year survival rate for both benign and malignant variants is reported to be more than 90% [2,6].

Resection with clear margins is considered as the ideal treatment of SFTs of the lung and pleura [4,5, 10,11]. Wedge resection of the tumor is easy to accomplish in small lesions, either through video-assisted thoracic surgery (VATS) or formal thoracotomy. In preoperatively established diagnosis of SFT of the lung and pleura, VATS is the preferred way to resect small tumors. Resection of larger masses should be performed through thoracotomy and may include resection of the adjacent pleura and/or lung parenchyma [4,5,10,11,13]. Special attention should be paid during thoracotomy to excise the tumor intact, as pieces of tumor may become "free bodies" within the pleural cavity and be a possible site of future intrathoracic recurrence [4,5]. Lobectomy and pneumonectomy are rarely necessary. Chest wall resection can be at instances necessary, when

the suspicion of chest wall invasion is made during thoracotomy, in order to achieve radical excision of the tumor [4,5,10,11].

SFT of the lung and pleura is an uncommon neoplasm, where non-specific symptoms, non-pathognomonic radiologic findings and non-conclusive FNA/FNB results cannot establish with certainty a preoperative diagnosis in most of the cases. Resection with clear margins is curative in small, penduculated masses and final diagnosis and prognosis will be defined after surgical resection. Large tumors should be resected intact *en bloc* with the tissues attached to the mass in order to avoid local recurrences.

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