Giant Solitary Fibrous Tumour of Pleura
—An Uncommon Intrathoracic Entity—
A Case Report and Review of the Literature

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A 43-year-old woman presented to us with progressive breathlessness, dry cough and weight loss. A chest radiograph showed homogeneous opacification of the entire left hemithorax. A contrast enhanced computed tomography (CECT) scan of the thorax showed a large intrathoracic mass occupying almost the entire left hemithorax and appeared grossly inoperable. A transcutaneous CT guided tru-cut biopsy revealed a solitary fibrous tumour. We reviewed the CT scans based on the biopsy report, and, in retrospect, the mediastinal vessels seemed more stretched and pushed by the tumor rather than directly infiltrated by it. We performed an exploratory thoracotomy and to our surprise, were able to dissect the mass quite easily off the mediastinum. She had an uneventful postoperative recovery, and the final histopathology confirmed a solitary fibrous tumor. We report this case to emphasize that a cursory clinico-radiological interpretation can dissuade surgical intervention in these patients.

Key words: solitary fibrous tumour, benign mesothelioma, Doege Potter syndrome, intrathoracic pleural tumour

Introduction

A solitary fibrous tumour (SFT) is an uncommon entity and represents 4% of all pleural neoplasms. They are also known as benign localized mesothelioma, submesothelioma, or subserosal fibroma. In 1931, Klemperer and Rabin divided primary tumors of the pleura into two categories: diffuse mesothelioma and localized mesothelioma. In 1942, Stout and Murray, on the basis of in vitro cell culture findings, proposed mesothelial cells as the source of the so-called localized mesothelioma which was confirmed by other studies with the help of immunohistochemical staining. SFTs differ from diffuse mesotheliomas in that they are not associated with asbestos exposure and usually show a good prognosis, whereas the latter are related to asbestos exposure and behave aggressively. These tumors usually arise from visceral pleura (in 80% of the cases), but they may also derive from the parietal pleura and other serosal membranes, such as the peritoneum, pericardium and nonserosal sites, such as the pulmonary parenchyma, the mediastinum, the nose, and the paranasal sinuses.

Case Report

A 43-year-old woman presented to us with progressive breathlessness on exertion, dry cough and weight loss of four months duration. The clinical examination was unremarkable, except for a decreased air entry in the left hemithorax. Her chest radiograph showed a homogeneous opacity in the left lower and mid zones with a mediastinal shift to the right. The CT scan revealed a large mass showing heterogeneous enhancement more in the periphery in the left hemithorax measuring $13.1 \times 15.6 \times 18.9$ cm in its antero posterior (AP), transverse and vertical dimensions respectively (Fig. 1). The mass had well demarcated margins and displaced the
There was a subcentimeter sized lymph node in the left prevascular region with focal intense FDG uptake (max SUV 4.2). Fibreoptic bronchoscopy showed extrinsic compression of the left main bronchus with no endobronchial growth.

A CT guided transthoracic biopsy revealed a collagen forming low grade spindle cell lesion (Fig. 3). The tumour cells were negative for smooth muscle antigen (SMA), Desmin, S-100 and CD34 (Fig. 4). Her 2D echocardiography showed normal cardiac function. Pulmonary function tests showed a restrictive pattern. In view of the extensive neovascularisation seen on CT scan, angioembolisation via a transfemoral route was done prior to surgery. A left posterolateral thoracotomy through the fifth intercostal space was performed which revealed a large, well encapsulated intrathoracic mass 15 x 14 x 10.2 cms which had compressed the entire left lung. The tumor was dissected off the surrounding structures relatively easily and excised along with a left lower lobectomy. She had an uneventful postoperative recovery.

.externally, the mass was well encapsulated; on serial slicing, the tumour was firm, solid and predominantly homogeneous and grey and was seen abutting the capsule. Multiple sections from the mediastinal mass showed features of a low grade spindle cell tumour, with a hemangiopericytoma-like vascular pattern and areas of hyalinisation. Tumour cells were positive for CD34 and negative for Human Melanoma Black 45 (HMB-45), smooth muscle antigen and desmin. Histopathological features were consistent with a solitary fibrous tumor. She had an uneventful postoperative recovery.

Discussion

Solitary fibrous tumors of the pleura are relatively uncommon with only about 600 cases having been reported previously. Independent collected reviews by Briselli and England (223 cases) described the clinicopathologic features of this neoplasm. The latter also defined criteria for malignancy, which include abundant cellularity, more than four mitoses per ten high-power fields, cytonuclear atypia, large necrotic or hemorrhagic areas, an associated pleural effusion, atypical location, and invasion of adjacent structures. Using these criteria, 12% to 33% of solitary fibrous tumors of pleura were considered to be malignant. SFTs are not associated with asbestos.
exposure and usually have a good prognosis, whereas diffuse mesotheliomas are related to asbestos exposure and behave aggressively. Only one case of a solitary fibrous pleural tumor reported in the medical literature has suggested a genetic component. This familial case could be attributed to a genetically transmitted germline mutation or exposure to a common agent. Solitary fibrous tumors most often originate in the visceral pleura. Cardillo et al found that 87% of solitary fibrous tumors in their series originated in the visceral pleura and only 13% in the parietal pleura. The size of the tumors can vary greatly between 1 cm and 36 cm (mean, 6 cm) in diameter. Many large tumors are pedunculated on pleural-based pedicles with hypertrophic vasculature. Numerous thin-walled vessels may be present in larger tumors.

They are as common in females as in males, with peak incidence in the 6th and 7th decades of life. The usual presentation, as described in over 50% of the cases reported in literature, is an asymptomatic mass discovered incidentally on a chest radiograph. Symptoms tend to be more common in larger lesions and include either thoracic (such as chest pain, cough, dyspnea, and more rarely hemoptysis) or extrathoracic (such as weakness, nocturnal sweating, chills, weight loss, digital clubbing, hypertrophic osteoarthropathy, and hypoglycemia) manifestations. When symptomatic, they are more often multiple and regress completely after surgical resection. Hypertrophic osteoarthropathy (Pierre-Marie-Bamberg syndrome) has been reported in 20% of cases and is thought to be due to increased production of hyaluronic acid by the tumor, or an increased production of hepatocyte growth factor. Hypoglycemia, which is observed in up to 2% to 4% of the cases, is attributed to the production of insulin-like growth factor II (IGF-II), which lowers the blood glucose and impairs the growth of insulin-like growth factor II (IGF-II), which lowers the blood glucose and impairs the growth hormone counter-regulatory response to hypoglycemia. Serous pleural effusion associated with this tumor accounts for fewer than 10% of all cases.

Preoperative diagnosis of SFT is difficult with only radio imaging. The imaging modalities of choice are a chest radiograph and CT scan, but these findings lack specificity. Thoracic CT scan shows in most cases a well-circumscribed round tumor with a homogenous density. A CT scan cannot differentiate between benign and malignant fibrous tumours of the pleura. A whole-body FDG PET is a noninvasive technique to readily and accurately differentiate benign from malignant masses, as the uptake in malignant mesothelioma would be very high.

Most authors agree that the CT-guided fine needle aspiration is often inconclusive. Anti-CD34 antibody has been claimed to be specific for solitary fibrous tumors. Although CD34 is expressed by various cell types, such as hematopoietic progenitor cells, endothelial cells, and mesenchymal tumor cells, its detection together with that of vimentin but not of cytokeratin in cells from a pleural tumor excludes the diagnosis of mesothelioma and of most other pleural tumors, such as carcinomas, fibrous histiocytomas, fibromatosis, fibrosarcomas, and synovial sarcomas. They have variable histologic patterns. The pattern-less pattern (mixture of fibroblast like cells and connective tissue cells) is the most common, followed by the hemangiopericytoma pattern. Others include a leiomyoma-like pattern, neurofibroma like pattern and mixed pattern.

Surgery is the mainstay of treatment, and recurrence and the clinical outcome is mainly related to the completeness of the surgical treatment. Important, favorable prognostic factors are the presence of a benign histological variant, a pedunculus, and a limited size. The surgical treatment of choice is local removal by with intra-operative assessment of free surgical margins. Video assisted Thoracoscopic Surgery (VATS) may be attempted for appropriate tumours, but conversion to open technique should be employed if margin clearance cannot be done with VATS. Some authors recommend VATS for lesions less than 5cms. Extended resections such as a chest wall resection, excision of parietal pleura, pericardium and lobectomies have been reported in order to attain a surgically clear margin. In some cases of large sized tumors, with major feeding vessels identified by angiography, preoperative percutaneous embolization has been reported to be useful in reducing perioperative blood loss. The overall operative mortality ranges from 1.5% to 12% because of hemodynamic changes associated with decompressing the mediastinal structures.

De Perrot and colleagues provide a classification for recurrence of SFT according to tumor characteristics and prognosis: (1) benign pedunculated tumors had a 2% recurrence rate, (2) benign sessile tumors had an 8% recurrence rate, (3) malignant pedunculated tumors had a 14% recurrence rate, and (4) malignant sessile tumors had a 63% recurrence rate and 30% mortality, with most deaths occurring within 24 months. However, local recurrence is not as
worrysome in benign lesions, which have an 8% chance of recurrence, compared with malignant lesions, which have a 63% recurrence rate even after complete resection.

CT scans may be used to monitor for recurrence every 6 months for the first 2 years and then yearly.15) Most recurrences, particularly of the sessile malignant tumors, occur within 24 months of the initial resection. Nevertheless, all patients with SFT need long term follow-up of 15 to 20 years due to the possibility of late recurrences. If a local recurrence is detected in any patient, strong consideration should be given for re-resection, which may well be curative.15, 24)

Anecdotal reports describe long-term survivals with postoperative radiotherapy in patients with incomplete resection of the tumor. Responses to ifosfamide and doxorubicin have been reported for recurrent, inoperable SFT. Nevertheless, recurrent benign or malignant tumors should be strongly considered first for a repeat surgical resection. Following the resection, adjuvant therapy should be considered for recurrent tumors, particularly the sessile, malignant variety, although little experience is described in the literature with postoperative treatment.15, 24) The role of adjuvant therapy in the management of SFTs is yet to be established.

Summary

SFT is an uncommon entity and is rarely suspected on a routine clinicoradiological workup. They can present as a diagnostic dilemma, and a core biopsy is usually required for an accurate diagnosis. An FDG PET scan is reliable to differentiate between benign and malignant fibrous tumors. Surgery is the mainstay of treatment and there is no proven role for adjuvant treatment. Radiological impressions in giant SFT may be misleading and every attempt should be made to resect these tumors. Large vascular tumours may benefit from pre-operative angioembolisation which could decrease the anticipated peri-operative blood loss.

References