

Enormous thoracic solitary fibrous tumor with inferior vena cava compression: A case report

Inferior vena cava kompresyonlu devasa torasik soliter fibröz tümör: Bir olgu sunumu

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ABSTRACT

Solitary fibrous tumor is a rare fibroblastic tumor that typically involves the pleura and very rarely the pericardium although it has been described in almost all locations in the body. Most intrathoracic solitary fibrous tumors are asymptomatic; however, some have unusual presentations such as inferior vena cava compression which, to our knowledge, was reported only once before. In this article, we report a middle-aged male patient with a huge thoracic solitary fibrous tumor of suspected pericardial origin and inferior vena cava compression who was successfully operated upon.

Keywords: Inferior vena cava; pericardium; pleura; solitary fibrous tumors.

A solitary fibrous tumor (SFT) is a rare fibroblastic tumor that typically arises in the thoracic cavity as a pleural tumor.^[1] However, it has been reported in almost all body locations^[2,3] such as meninges, epiglottis, salivary glands, thyroid, kidneys and breast.^[4] In the past years, the origin of thoracic SFT was controversial. Therefore, terms such as benign or localized fibrous mesothelioma, pleural fibroma, submesothelial fibroma, or localized fibrous tumor were used.^[2-8] Later, immunohistochemical studies revealed that intrathoracic SFTs were derived from

ÖZ

Soliter fibröz tümör vücuttaki neredeyse tüm lokasyonlarda tanımlanmış olsa da tipik olarak plevrayı ve çok nadiren perikardiyumu tutan nadir bir fibroblastik tümördür. Çoğu toraks içi soliter fibröz tümör asemptomatiktir; ancak bazılarında inferior vena cava kompresyonu gibi olağandışı görüntüler vardır ki, bildiğimiz kadarıyla bu daha önce yalnız bir defa bildirilmiştir. Bu yazıda perikardiyal kökenli olmasından şüphelenilen ve inferior vena cava kompresyonlu dev bir torasik soliter fibröz tümörü olup başarıyla ameliyat edilen orta yaşlı bir erkek hasta bildirildi.

Anahtar sözcükler: İnförior vena cava; perikardiyum; plevra; soliter fibröz tümörler.

pluripotent fibroblastic cells in the submesothelial connective tissue rather than from the mesothelial cells.^[7] The term SFT was mentioned for the first time by Wagner in 1870^[7] to distinguish it from the malignant diffuse pleural mesothelioma.

Thoracic SFTs are topographically classified into pleural (visceral/parietal), mediastinal, and pulmonary origins.^[1] Mediastinal SFT may arise in any compartment of the mediastinum^[3] and account for 1 to 8% of intrathoracic SFTs.^[6] Huang et al.^[1] from Taiwan retrospectively reviewed 52 cases of



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intrathoracic SFTs over a 14-year period and found pleural involvement on the top of the list (n=31, 60%) followed by the mediastinum/pericardium (n=12, 23%) and lastly the lung (n=9, 17%). It is noteworthy that there was only a single case of SFT arising from the pericardium indicating the extreme rarity of the tumor.^[1]

Pleural SFTs (PSFTs) account for less than 5% of all primary pleural tumors.^[7] Most modern studies of PSFT report that these tumors develop in one to two patients annually.^[7] There is no relationship between the occurrence of these tumors and exposure to asbestos and cigarette smoking.^[7] Although the tumors may develop within a wide age range (5 to 87 years), they mainly occur in the sixth and seventh decades of life, with equal incidence in both sexes.^[2,3,7]

These tumors often have an asymptomatic clinical course.^[7] Thus tumors tend to grow into huge masses before local compression symptoms develop, especially in patients without routine physical examinations. About 50% of patients reported in the literature are symptomatic.^[7] The most common symptoms are chest pain, cough and shortness of breath.^[4] Larger tumors may have a higher percentage of symptomatology.^[9] The symptoms occur in 75-88% of patients with malignant tumors and in 42-67% of patients with benign tumors.^[7]

Fever, night sweats, and weight loss are rare.^[7] Hypertrophic pulmonary osteoarthropathy (HPO) is a paraneoplastic syndrome that occurs in patients with large SFTs characterized by flu-like illness, arthritis, stiff neck and swelling of joints and ankles resolving several days following tumor resection. Clubbed fingers constitute the most common coexisting physical sign in patients with HPO and PSFT. Although the exact etiology of HPO is unknown, it may be due to short-term ectopic secretion of growth hormone.^[7] Hypoglycemia (Doege-Potter syndrome) is a rare manifestation of the disease, caused by the secretion of tumor insulin-like growth factor 2, which also resolves after excision of the tumor. Pleural effusion associated with PSFT is mostly serous. Fluid in the pleural cavity is more often found in patients with malignant tumors.^[7]

In this article, we report a rare case of a very huge SFT completely filling the left hemithorax and displacing the heart and both lungs to the right side and pressing on the inferior vena cava (IVC), suspected of being of pericardial origin and which was completely excised via a median sternotomy. The relevant literature was reviewed to highlight the management aspects of this rare yet surgically challenging and potentially fatal tumor.

CASE REPORT

A 51-year-old Iraqi male patient was admitted to a private hospital in Sulaimaniyah/Iraq on June 15th 2014 because of worsening exertional dyspnea, bilateral leg edema and mild puffiness of the face of two-year duration. His past medical history was unremarkable and he was not a smoker. One year earlier, he was admitted to an Indian hospital for resection of a huge left-sided intrathoracic mass. However, the procedure was shortly terminated as the patient sustained an endotracheal intubation injury with right-sided pneumothorax for which a tube thoracostomy was necessary.

On examination, the patient had a moderate shortness of breath and tachypnea with an oxygen saturation rate of 75%. The pulse rate was 99 beat/minute and the blood pressure was 100/65 mmHg. The trachea was displaced to the right side. Breath sounds were audible only over the right upper chest while the heart sounds could be heard over the right chest. He had no digital clubbing or lymphadenopathy. The legs were moderately swollen after walking.

The patient underwent a detailed diagnostic workup. Routine chemistry and hematology lab test results were within the normal range with hemoglobin (Hb) of 14.2 g/dL. The chest radiograph (Figure 1a) showed homogenous opacity of the entire left chest with severe tracheal and mediastinal shift to the right side and right-sided heart. Computed tomography (CT) of the chest (Figure 1b) showed a huge lobulated well-defined markedly enhanced heterogeneous soft tissue mass; 22x24x17 cm in size occupying the left chest and crossing the midline but with no evidence of local invasion or mediastinal lymphadenopathy. Fiberoptic bronchoscopy revealed near total obliteration of left main bronchus by external compression. Pulmonary function tests showed restrictive ventilation. Echocardiography was normal apart from the heart being dislocated into the right chest, with an ejection fraction of 58%. Percutaneous trans-thoracic fine needle aspiration cytology (FNAC) was repeatedly inconclusive.

Surgical intervention was deemed too risky by a local thoracic surgeon who advised the patient chemotherapy for this huge "inoperable" tumor. However, careful review of the chest CT scans and particularly the presence of a well-defined mass and lack of invasion encouraged us to consider surgery.

The operation was performed under general anesthesia via a small caliber endotracheal tube. Median sternotomy was chosen to allow access to

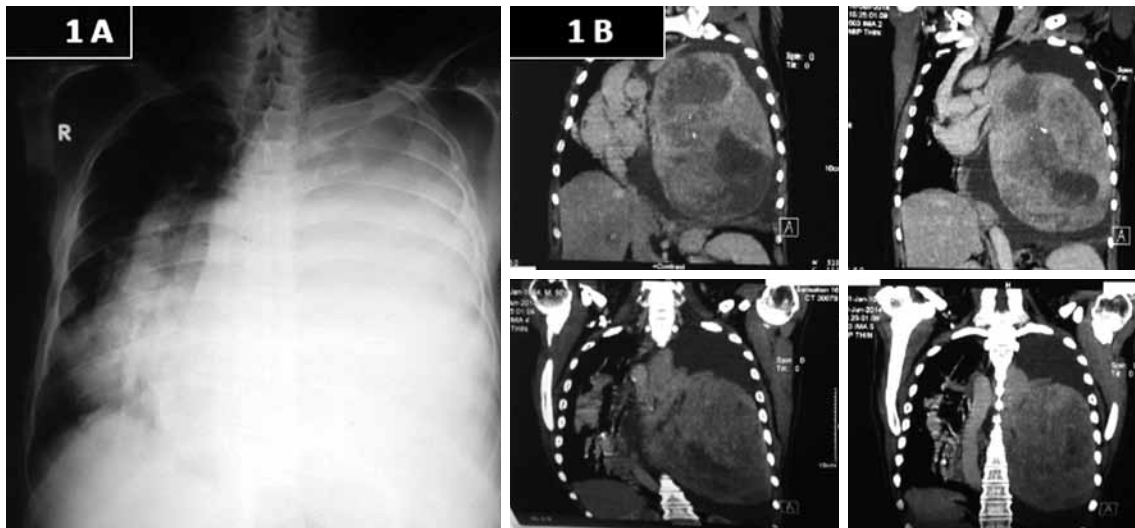


Figure 1. (a) Plain chest radiograph showing total opacity of left hemithorax with tracheal and mediastinal shift to right side. (b) Preoperative computed tomography scans of the chest.

both thoracic cavities. Adhesions to chest wall and diaphragm were released by blunt and sharp dissection. Ligation and division of a big vascular pedicle attached to the pericardium allowed en bloc resection of the huge mass (Figure 2a) leaving an empty thoracic cavity apart from a rim of compressed left lung beside the heart. There was no evidence of invasion of the tumor capsule, pulmonary or mediastinal structures. The raw inner chest wall developed severe bleeding following tumor resection and required transfusion of 10 pints of blood intraoperatively and another two pints postoperatively to achieve an hemoglobin of 10 g/dL. The postoperative period was uneventful. The patient's

symptoms were relieved. The postoperative CT scan of the chest (Figure 2b) showed no residual tumor with fully expanded lungs and centrally located heart.

The biopsy result was SFT showing spindle-shaped tumor cells arranged in a haphazard pattern. Immunohistochemistry studies revealed a negative reaction to desmin (Figure 3a) but positive reactions to S 100 (Figure 3b), cluster of differentiation 34 (CD 34) (Figure 3c), cluster of differentiation 99 (CD 99) (Figure 3d), B-cell lymphoma 2 (BCL 2) (Figure 3e) and epithelial membrane antigen (Figure 3f). A written informed consent was obtained from the patient.

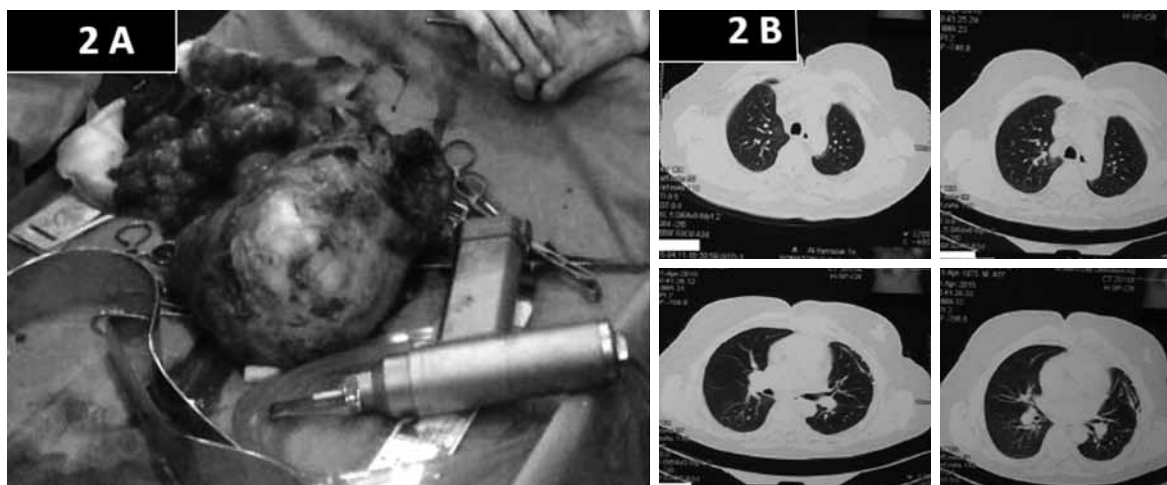


Figure 2. (a) The tumor was well encapsulated; size: 22x24x17 cm. (b) Postoperative chest computed tomography scan.

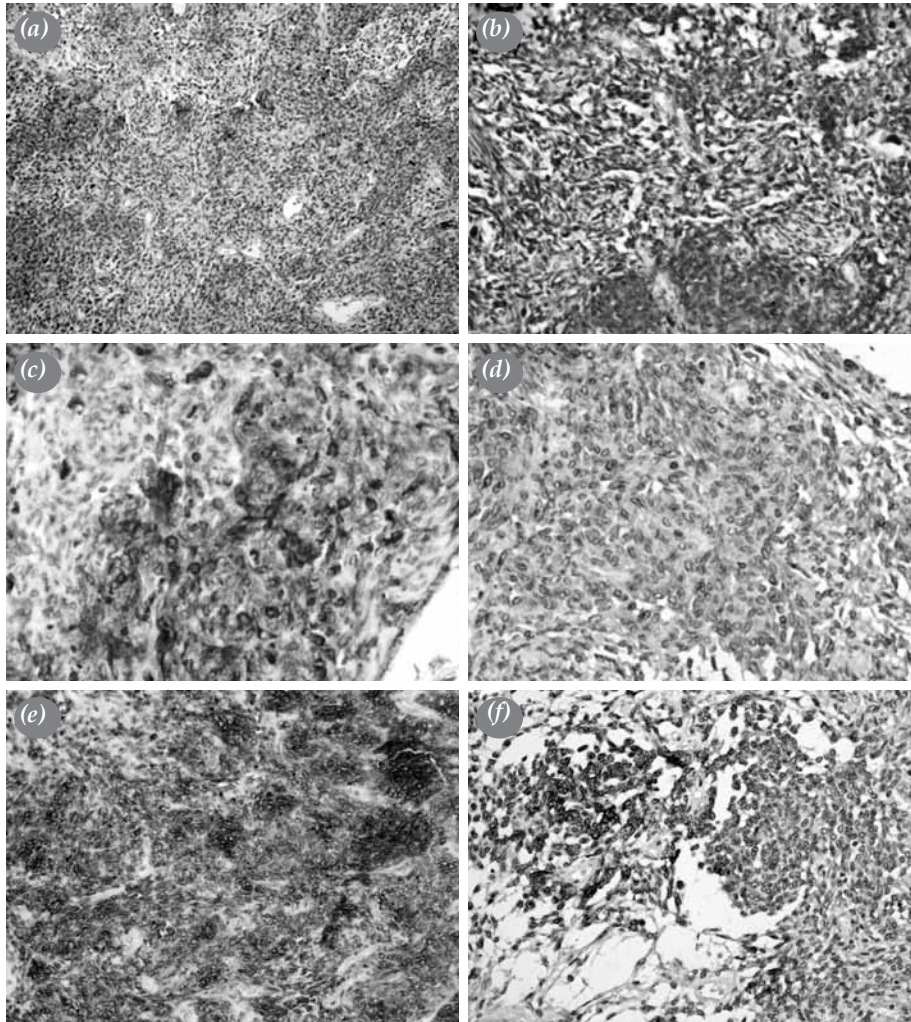


Figure 3. (a) Immunohistochemistry (IHC)-staining is negative to Desmin ($\times 4$). (b) IHC staining is positive to S 100 ($\times 10$). (c) IHC staining is positive to cluster of differentiation 34 ($\times 20$). (d) IHC staining is positive to CD 99 ($\times 20$). (e) IHC staining is positive to B-cell lymphoma 2 ($\times 20$). (f) IHC staining is positive to epithelial membrane antigen (EMA) antibody staining ($\times 20$).

DISCUSSION

Solitary fibrous tumors were first described as a distinct clinical and pathological entity by Klemperer and Rabin in 1931^[4] and by the year 2009, around 900 cases have been reported worldwide.^[5,7] In a previous publication,^[8] we have reported two cases of localized pleural mesothelioma (one benign and one malignant) in two Iraqi ladies, which were both successfully excised and which we think that they were better named as PSFTs.

In our case, the tumor could be of pleural origin as it has displaced the heart and both lungs to the opposite side. Tumors arising from the mediastinum usually grow

into the nearby pleural space while the mediastinum itself remains central.^[4] Moreover, the severe bleeding from the chest wall after excision of the tumor indicates an extensive blood supply of the tumor from intercostal arteries which goes with a pleural origin of the tumor. On the other hand, the presence of a big vascular pedicle attached to the pericardium favors a pericardial origin. The histopathological features of SFTs are the same regardless of their origin. However, SFTs of mediastinal/pericardial origin are thought to have a more aggressive behavior.^[1,3] In our case, regardless of the precise origin of the tumor (pleural or pericardial), surgical removal was challenging due to the huge tumor size, great vascularity and dense adhesions.

Solitary fibrous tumors are usually benign; only 20% are malignant.^[7] Scientists have discovered three histopathological criteria associated with the malignant nature of the tumor: a large number of cells with overlapping nuclei, increased mitotic activity, and nuclear pleomorphism.^[1,3,7] We think that despite the giant size of the SFT in our case, it was benign due to lack of invasion, lymphadenopathy or metastatic spread and good health of the patient^[4] besides lack of the three histopathological criteria of malignancy.

Despite the huge size of the tumor, our patient developed symptoms in the last two years only. The tumor most likely had been growing slowly for many years without producing significant symptoms. The main symptoms were shortness of breath and bilateral lower limbs edema due to cardiac chambers and IVC compression. Other causes of lower limb edema were excluded by clinical examination. Both lower limbs edema and the shortness of breath have disappeared in the early postoperative period. This clinical presentation was similarly reported by Shaker *et al.*^[10] Moreover, there was a minimal serous pleural effusion but no HPO or hypoglycemia.

Chest CT scan is the key imaging study as it clearly shows the size and location of the tumor and aids in surgical planning.^[4] Both the benign and malignant varieties of PSFT usually appear as well-delineated, often lobulated masses. The mass effect of large lesions (like ours) may produce atelectasis and displacement of bronchi and vessels, but there should be no evidence of invasion into the lung or chest wall, nor multiple pleural seeding. The lesions may grow to be very large, almost filling a hemithorax.^[7]

A confident preoperative diagnosis of a fibrous tumor of the pleura can be established by histologic and immunohistochemical analysis of material obtained by trans-thoracic Tru-Cut needle biopsy.^[4,5] However, in most cases, the diagnosis is confirmed only by pathological evaluation of the resected specimen supported by immunoreactivity of neoplastic cells for certain stains.^[4,7] Tumor cells are immunoreactive for CD34 and CD99, variably positive for BCL-2; while they are usually negative to cytokeratins and desmin.^[3,4] Recently, nuclear staining of STAT6 resulting from NAB2-STAT6 gene fusion has been shown to be a more specific marker for SFT.^[1,3] In our case, percutaneous trans-thoracic FNAC was repeatedly inconclusive. Tru-Cut needle biopsy was not carried out, whereas histopathological examination and immunohistochemical staining confirmed the diagnosis. Spindle cells arranged in a “patternless

pattern” without a mitotic activity were consistent with a benign SFT.^[1,3,4,7]

We agree with Liu *et al.*^[5] that every suspected or proven PSFT should be surgically treated because clinical or radiological criteria cannot accurately distinguish benign from malignant forms. Surgery is the gold standard of treatment as neither radiotherapy nor chemotherapy proved to be effective.^[7,11] In our case, excision of the mass was followed by profuse bleeding that required a massive blood transfusion during and after the operation. The surgical excision of giant intra-thoracic SFTs is considered a surgical challenge due to poor exposure, significant tumor blood supply, and heavy adhesions, among other characteristics.^[9,12] Aydemir *et al.*^[12] from Turkey reported a 59-year-old female with a giant intrathoracic SFT that almost completely filled the right hemithorax for whom the operation was terminated after taking a biopsy because of excessive bleeding during dissection. Later on, angiography showed that the tumor was supplied by multiple intercostal arteries as well as an aberrant branch from the celiac trunk. The mass was then resected in a repeat operation; however, the post-resection bleeding could not be controlled even by cardiopulmonary bypass (CPB) and the patient died due to total circulatory arrest. We think that excessive bleeding from the raw chest wall in our patient could be due to a similar blood supply via intercostal arteries and we agree with the authors that CPB and total circulatory arrest are not valid alternatives to control the bleeding. We share the advice of many authors^[9,12-14] to perform a preoperative CT angiography for such huge tumors to reveal the feeding arteries and perform preoperative embolization to reduce the intraoperative blood loss. In addition, we recommend auto-transfusion as a good intraoperative measure to save the patients' lives. Tumors originating from the parietal pleura are more challenging to resect due to the difficulty in obtaining a clear margin along the chest wall.^[7] In our case, the greatest diameter of the tumor was 24 cm, while the greatest reported dimension of these tumors was 27.5 cm.^[1] Unfortunately, the specimen was not weighed.

The huge SFT larger than 10 cm required closer surveillance of our patient.^[2] Solitary fibrous tumors can recur and metastasize after surgical resection particularly if the tumor was bigger than 10 cm or had a histologically malignant component.^[2] Follow-up of our patient revealed no evidence of recurrence two years after surgery as shown by chest CT scans.

In conclusion, the origin from the pericardium, the huge size and inferior vena cava compression were all rare features of the solitary fibrous tumor reported herein. Surgical excision was a challenge due to the enormous size and great vascularity of the tumor, yet it was safely performed as the computed tomography scans of the chest revealed a well-encapsulated mass with no local invasion.

Declaration of conflicting interests

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