CASE REPORT

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Intrathoracic Giant Mass: Solitary Fibrous Tumor

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ABSTRACT Solitary fibrous tumors of the pleura are less than 5% of the pleural tumors. Eighty percent of the cases are caused by visceral pleura and 20% by parietal pleura. They are usually asymptomatic. These tumors, which can reach very large dimensions, can cause coughing and shortness of breath due to compression. Hypoglycemia and hypertrophic pulmonary osteoarthropathy are other symptoms. Definitive diagnosis is usually made after tumor excision. Abundant cellular structure with histologically dense and overlapping nuclei, more than 4 mitotic activities per site, pleomorphism with cytonuclear atypia, presence of large necrotic and hemorrhagic areas support malignancy. In addition, pleural effusion coexistence and atypical localization of the lesion are other findings supporting malignancy. Complete resection of the tumor is the most effective treatment and at the same time the most effective way to prevent recurrence.

Keywords: Solitary fibrous tumor; thoracic surgery; pleural neoplasms

Solitary fibrous tumors of the pleura (SFTPs) are rarely seen mesenchymal tumors. They account for less than 5% of all pleural tumors.[1] They were first described by Lieutaud in 1767.[2] Klemperer and Rabin published as localized mesothelioma in 1932. In addition to this term, SFTPs have been named as localized fibrous mesothelioma, fibrous mesothelioma, solitary fibrous mesothelioma, pleural fibroma, subserosal fibroma or submesothelial fibroma in the literature.[3] SFTPs are thought to be caused by subpleural mesenchymal fibroblast or myofibroblast differentiation.[3] Eighty percent are of visceral pleural origin and 20% are of parietal pleural origin.[4] These tumors, which are mostly asymptomatic, can grow substantiall large.[5] Most SFTPs are benign but can be malignant.

CASE REPORTS

CASE 1

A 44-year-old female patient presented to our outpatient clinic with dyspnea and cough continuing for 4 months. Posteroanterior (PA) chest X-ray (CXR) showed a marked opacity with the appearance of a mass and fluid in the right hemithorax. A small portion of the right lung was vented at the apex (Figure 1a). Computed tomography (CT) and magnetic resonance imaging (MRI) were performed. A solid mass measuring approximately 165×125×190 mm, filling the right hemithorax almost completely and pushing the heart and mediastinal structures to the left, was observed (Figure 2). There was a large mass with necrotic areas and intense vascularization within the mass and pleural effusion reaching a depth of 2.5 cm on the right. No signs of invasion into the thoracic wall and mediastinal structures were detected. Laboratory parameters were normal. Trans-thoracic needle biopsy was performed. Biopsy result was reported as "mesenchymal tumor". The patient was operated and the mass that completely filled the right hemithorax was removed. Pathology results revealed solitary fibrous tumor. Histopathologic examination revealed less than 4 mitoses per site (2-3). There was no significant atypia stained with CD34.

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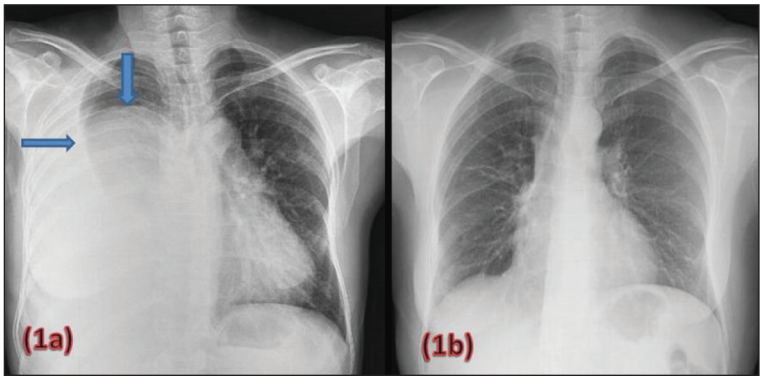

FIGURE 1: a) A small portion of the right lung was vented at the apex, **b)** The patient was in good condition during postoperative follow-up.

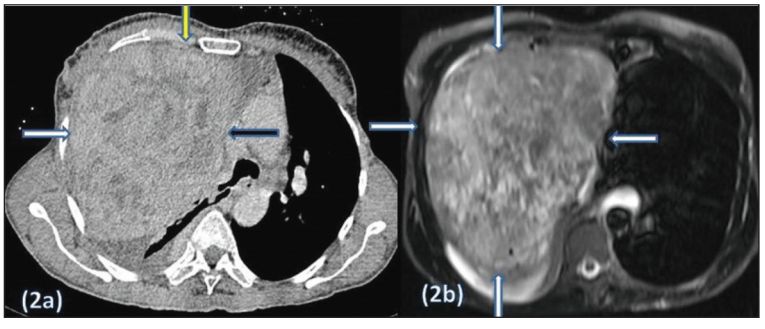

FIGURE 2: A solid mass measuring approximately 165×125×190 mm, filling the right hemithorax almost completely and pushing the heart and mediastinal structures to the left, was observed. **a)** Computed tomographic view of the thorax, **b)** Magnetic resonance view of the thorax.

Foci of hemorrhage and necrosis were present. The mass was originated from the visceral pleura of the lower lobe. The patient was in good condition during postoperative follow-up (Figure 1b).

CASE 2

A 69-year-old male patient presented to our clinic with complaints of labored breathing, fever, and pain in the lower right chest. In the PA CXR, there was opacity and pleural effusion appearing to be a mass that filled half of the right hemithorax (Figure 3a). Pneumonia treatment was initiated. After pneumonia symptoms improved, thoracic CT and MRI were performed (Figure 4a). A 26×18×10 cm lobulated, sharp contoured solid mass in the posterobasal of the right hemithorax extending from the diaphragm neighborhood to the right hilus level was observed. Transthoracic biopsy revealed mesenchymal neoplasia. The patient was operated and total resection was performed. Pathology revealed a solitary fibrous tumor. Histopathological results revealed mitosis (>4/10 high power field): 16 cells, hypercellularity, pleomorphism, necrosis, and hemorrhage. The patient was discharged. The patient's condition was stable in MRI (Figure 4b) and PA CXR (Figure 3b) performed during follow-up.

However, at the 6th postoperative month, it was observed that the lesion occurred again during the control. It was seen that the tumor was growing very fast. The patient was re-operated. The result of the second pathology has been reported as a malignant solitary fibrous tumor (malignant mesenchymal tumor). Immunohistochemically, vimentin, CD34 and bcl-2 diffuse (+), pancytokeratin, melan-a, CD68, d2 40, CD117, CD99, S-100, EMA, desmin, SMA, CD31, calretinin and CD68 (-) and Ki67 index, it was reported to be about 70%. The tumor had 43 mitosis in 10 large magnification areas. The patient was transferred to the oncology clinic for chemotherapy after the operation.

Consent for the studies was obtained from both patients.

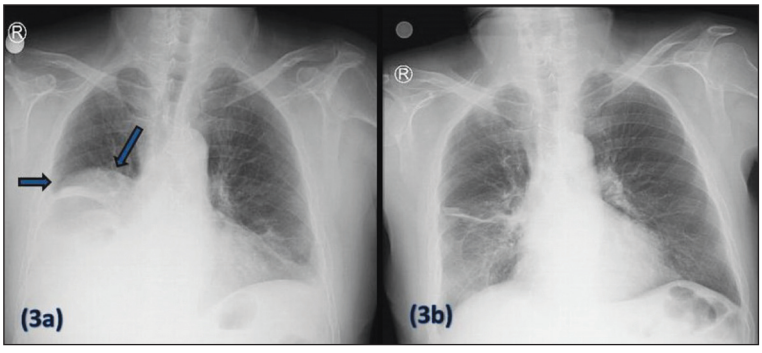

FIGURE 3: a) In the posteroanterior chest X-ray, there was opacity and pleural effusion appearing to be a mass that filled half of the right hemithorax, **b)** Postoperative chest X-ray.

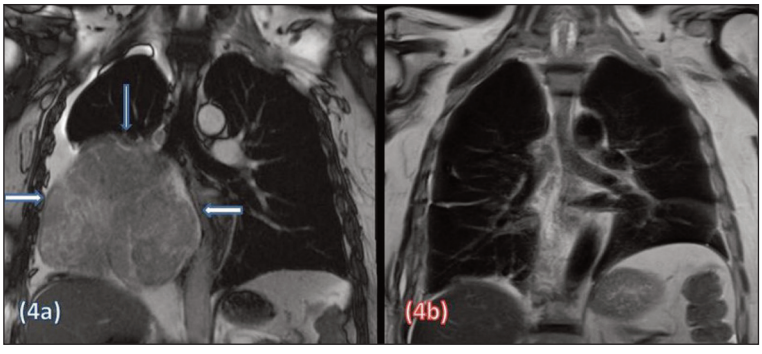

FIGURE 4: a) A 26×18×10 cm lobulated, sharp contoured solid mass in the posterobasal of the right hemithorax extending from the diaphragm neighborhood to the right hilus level was observed, **b)** The patient's condition was stable in magnetic resonance imaging.

DISCUSSION

SFTPs can be seen at any age and no gender difference has been reported in terms of prevalence.[2] Outside the thorax, they can also be caused by other serosal membranes such as peritoneum and pericardium.[2] It is also reported in the literature that SFTPs can be seen in non-serosal tissues such as liver, sinuses, lung parenchyma and orbita.[2]

SFTPs are often asymptomatic. Symptoms such as coughing, labored breathing and chest pain may be seen in large ones due to the compression of the mass. Hypertrophic pulmonary osteoarthropathy (Pierre-Maire-Bamberg syndrome) has been reported in 20% and hypoglycemia has been reported in 4% of the patients.[6] These findings are more common in patients with large tumors. Patients with neurological symptoms have also been reported in the literature.[7]

Radiological imaging of the lesion is important during diagnosis. SFTPs may show as a smooth solitary nodule or mass in PA CXR.[8] They can be seen as a homogeneous, well-defined and lobulated soft tissue mass with prominent borders in thoracic CT.[9] Tumors with pedicle involvement may show mobility in the pleural space.[10]

Transthoracic needle biopsy can be performed for the diagnosis of SFTPs, but preoperative diagnosis is rarely made.[11] Even if diagnosis is made with fine needle aspiration biopsy, it is often difficult to make a histological diagnosis. In both of our cases, histological diagnosis was made postoperatively. Macroscopically, SFTPs are large, lobulated, well-defined, vascularized structures with capsules. Microscopically, the "patternless pattern", which is characterized by the dense collagen bundles and the random distribution of spindle cells, stands out.[12] Most SFTPs are benign. In the literature, malignancy rates have been reported to be between 7% and 38%.[12]

de Perrot et al. staged SFTP according to histomorphological features (Table 1).[13]

TABLE 1: de Perrot et al. staged solitary fibrous tumors of the pleura according to histomorphological features.[13]

Stage 0: Pedicle tumor without malignant features.
Stage 1: Tumor that is seated at a wide angele to the pleura or has flat appearance with no malignancy features.
Stage 2: Stalked tumor with malignancy criteria
Stage 3: A tumor with a wide angle to the pleura or a flat appearance with malignancy criteria
Stage 4: Multiple and metastatic tumors

England et al. evaluated the presence of abundant cellular structure with dense and overlapping nuclei, mitotic activity of >4 per site, pleomorphism with cytonuclear atypia, presence of large necrotic and hemorrhagic areas, coexistence of pleural effusion, and atypical localization as malignancy criteria.[5] CD34 positivity is useful in the differential diagnosis of these tumors.[12] Our second case also had CD34 positivity. The tumor has large necrotic and hemorrhagic areas. There were 43 mitosis in 10 large magnification areas. The tumor had grown very quickly in a short time. All of these findings were consistent with the literature information as findings supporting malignancy.

Complete surgical resection of the tumor is the most effective treatment for SFTPs and the most important prognostic factor preventing recurrence.[14]

Use of neoadjuvant chemotherapy is limited in SFTPs due to the difficulty of preoperative histological diagnosis. Adjuvant chemotherapy is used in malignant SFTPs after surgical resection and is recommended especially in recurrent forms.[15]

Prognosis is generally good. However, recurrence or metastasis have also been reported.[11] de Perrot et al. reported a recurrence rate of 2% for benign pedicled tumors, 8% for benign squamous tumor, 14% for malignant pedicled tumors, and 63% for malignant squamous tumors.[13]

SFTPs are rarely seen tumors. These tumors, which are usually asymptomatic, can grow substantially large. They may cause chest pain, cough, and dyspnea by causing pressure on neighboring structures. Most SFTPs, where preoperative histological diagnosis is difficult, are benign. The presence of abundant cellular structure with dense and overlapping nuclei, mitotic activity of more than 4 per site, pleomorphism with cytonuclear atypia, presence of large necrotic and hemorrhagic areas, coexistence of pleural effusion, and atypical localization are evaluated as malignancy criteria. Complete surgical resection of these tumors is the basis of treatment. The most effective way of preventing recurrence is complete resection. Regular follow-up of patients is necessary in case of possible disease recurrence.

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Conflict of Interest

No conflicts of interest between the authors and / or family members of the scientific and medical committee members or members of the potential conflicts of interest, counseling, expertise, working conditions, share holding and similar situations in any firm.

Authorship Contributions

This study is entirely author's own work and no other author contribution

REFERENCES

1. Crnjac A, Veingerl B, Vidovic D, Kavalar R, Hojski A. Giant solitary fibrous tumour of the pleura. Case report and review of the literature. Radiol Oncol. 2015;49(4):395-401. [Crossref] [PubMed] [PMC]
2. Akı H, Durak H, Kaynak K, Demirhan Ö, Öz B. Plevranın soliter fibröz tümörü ile izlediğimiz 3 olgu [Solitary fibrous tumor of the pleura]. Cerrahpaşa J Med. 2002;33:127-31. [Link]
3. Robinson LA. Solitary fibrous tumor of the pleura. Cancer Control. 2006;13(4):264-9. [Crossref] [PubMed]
4. Aydemir B, Celik S, Okay T, Doğusoy I. Intrathoracic giant solitary fibrous tumor. Am J Case Rep. 2013;14:91-3. [Crossref] [PubMed] [PMC]
5. England DM, Hochholzer L, McCarthy MJ. Localized benign and malignant fibrous tumors of the pleura. A clinicopathologic review of 223 cases. Am J Surg Pathol. 1989;13(8):640-58. Erratum in: Am J Surg Pathol 1991;15(8):818. [Crossref] [PubMed]
6. Bini A, Brandolini J, Davoli F, Dolci G, Sellitri F, Stella F. Solitary fibrous tumor of the pleura: surgery and clinical course in 18 cases. Asian Cardiovasc Thorac Ann. 2009;17(4):378-81. [Crossref] [PubMed]
7. Santambrogio L, Nosotti M, Palleschi A, Rosso L, Tosi D, De Simone M, et al. Solitary fibrous tumor of the pleura presenting with syncope episodes when coughing. World J Surg Oncol. 2008;6:86. [Crossref] [PubMed] [PMC]
8. Abu Arab W. Solitary fibrous tumours of the pleura. Eur J Cardiothorac Surg. 2012;41(3):587-97. doi: 10.1093/ejcts/ezr009. [Crossref] [PubMed]
9. Chamberlain MH, Taggart DP. Solitary fibrous tumor associated with hypoglycemia: an example of the Doege-Potter syndrome. J Thorac Cardiovasc Surg. 2000;119(1):185-7. [Crossref] [PubMed]
10. Dynes MC, White EM, Fry WA, Ghahremani GG. Imaging manifestations of pleural tumors. Radiographics. 1992;12(6):1191-201. [Crossref] [PubMed]
11. Liu CC, Wang HW, Li FY, Hsu PK, Huang MH, Hsu WH, et al. Solitary fibrous tumors of the pleura: clinicopathological characteristics, immunohistochemical profiles, and surgical outcomes with long-term follow-up. Thorac Cardiovasc Surg. 2008;56(5):291-7. [Crossref] [PubMed]
12. Yücel H, Demirci NY, Erdoğan E, Demirağ F, Yılmaz Ü, Yılmaz A, et al. Yavaş büyüyen bir pulmoner nodül: intrapulmoner soliter fibröz tümör [Slow growing pulmonary nodule: intrapulmonary solitary fibrous tumor]. Respir Case Rep. 2016;5(2):144-8. [Crossref]
13. de Perrot M, Fischer S, Bründler MA, Sekine Y, Keshavjee S. Solitary fibrous tumors of the pleura. Ann Thorac Surg. 2002;74(1):285-93. [Crossref] [PubMed]
14. Veronesi G, Spaggiari L, Mazzarol G, De Pas M, Leo F, Solli P, et al. Huge malignant localized fibrous tumor of the pleura. J Cardiovasc Surg (Torino). 2000;41(5):781-4. [PubMed]
15. Bora G, Colaut F, Segato G, Delsedime L, Oliaro A. Solitary fibrous tumor of the pleura: histology, CT scan images and review of literature over the last twenty years. Biomed J Sci & Tech Res. 2017;1(1):188-92. [Crossref]