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Solitary Fibrous Tumor of the Pleura: A Case Report

Plevranın Soliter Fibröz Tümörü: Olgu Sunumu

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Abstract

Solitary fibrous tumors of the pleura are extremely rare mesenchymal tumors that originate from the visceral or parietal pleura, accounting for less than 5% of all pleural tumors. Solitary fibrous tumors are diagnosed based on clinical, radiological and needle biopsy findings. They are often asymptomatic and can reach very large sizes in the thorax, upon which they may produce such symptoms as cough, chest pain, dyspnea and hemoptysis. These tumors are generally benign, but are identified as malignant in 10–20% of cases, and carry the risk of recurrence, making early diagnosis very important. The treatment option is total resection followed by close follow-up. We present here a very rare case with a solitary fibrous tumor of the pleura.

Keywords: *Solitary fibrous tumor, pleura, mesenchymal tumor.*

Öz

Plevranın soliter fibröz tümörleri, visseral veya perietal plevradan köken alan oldukça nadir görülen mezenkimal tümörlerdir. Tüm plevral tümörlerin %5' inden azını oluştururlar. Soliter fibröz tümör tanısı, klinik, radyolojik ve iğne biyopsisi bulguları ile konulur. Sıklıkla asemptomatiktirler ve göğüs kafesinde çok büyük boyutlara ulaşabilirler. Daha büyük boyutlara ulaştıklarında öksürük, göğüs ağrısı, nefes darlığı ve hemoptizi gibi belirtiler gösterebilirler. Genellikle benign olan bu tümörler %10-20 oranında malign olabilmekte ve nüks riski taşımaktadırlar. Bu sebeple erken tanısı çok önemli olup, total rezeksiyonu ve yakın takibi gereklidir. Oldukça nadir rastlanan bu olgumuzu sunmaya değer bulduk.

Anahtar Kelimeler: *Soliter fibröz tümör, plevra, mezenkimal tümör.*

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Solitary fibrous tumors (SFT) are sporadic tumors arising from the mesenchymal tissue beneath the mesothelial layer of the visceral or parietal pleura. These tumors, which can develop throughout the body, constitute less than 5% of all pleural tumors. SFTs have no genetic basis, nor are they related to environmental factors such as tobacco smoking or exposure to asbestos (1). In the early stages, SFTs are usually asymptomatic and noticed by accident on chest X-ray. When growing up in the thoracic cavity, these tumors exert pressure on vital adjacent tissues and large vessels and they may show symptoms such as cough, chest pain, dyspnea, and hemoptysis (2). Although rare, SFT may cause reversible paraneoplastic syndromes with surgical resection, such as hypoglycemic attack (3). Thorax computed tomography (CT) is the most important imaging examination in diagnosis. Magnetic resonance imaging is better than thoracic CT, especially in cases involving larger blood vessels in the thorax, spinal column, or diaphragm (4). Definitive diagnosis of SFTs is made by histopathological and immunohistochemical features. Although mostly benign, approximately 20% of them show malignant features (2). The main treatment for SFTs is complete surgical resection (4,5). After resection, recurrence can be seen in approximately 10-15% of patients with pleural solitary fibrous tumors (6).

CASE

A 55-year-old female patient applied to our polyclinic with a complaint of pain on the right side of her back for one year. She had an interlocutory cough and white sputum. There was no dyspnea. Her smoking history was 35 packs/year. In the physical examination of the respiratory system; on auscultation, there was a decrease in bilateral breath sounds, and on palpation, there was dullness in the lower zone of the right lung. Her blood pressure was 120/80 mmHg, her heart rate was 85 beats per minute, her oxygen saturation on room air was 97% and her body temperature was 36 °C. There were no abnormal findings on electrocardiography. Routine laboratory tests were done. Leukocytes: 8.0x103/μL, hemoglobin: 13.8 g/dL, platelet: 336x103/μL, glucose: 107 mg/dL was detected. Liver and kidney function tests were normal. In the posteroanterior and left lateral chest X-ray of the patient, a sharply demarcated opacity was observed in the basal part of the right lung, adjacent to the diaphragm (Figure 1 a and b). On thorax CT, a hypodense mass lesion of approximately 8.5x6 cm was observed in the basal part of the right hemithorax, at its widest point (Figure 2 a and b). An Ultrasound-guided tru-cut biopsy was performed on the patient by the interventional radiology department. Pathological examination showed spindle-like or epithelioid cells with narrow cytoplasm, hyperchromatic nuclei, and small-caliber vascular structures around them, between areas of fibrosis and collagen bundles in a small area. In the immunohistochemical examination performed in this area, spindle or epithelioid cells showed diffuse positive staining with CD34 and beta-catenin. Ki67 index was seen in the range of 3-5%. Pathological diagnosis was determined as a solitary fibrous tumor. Positron emission tomography/Computed tomography (PET-CT) performed in our patient, a wide pleural-based axial section in the right lung lower lobe posterior basal, 6x10 cm in size, heterogeneous slightly increased FDG uptake in a well-defined hypodense focus (SUVmax:3.75), slightly regressed (SUVmax:3) was therefore considered benign. The case was referred to the Department of Thoracic Surgery after the available evaluations.

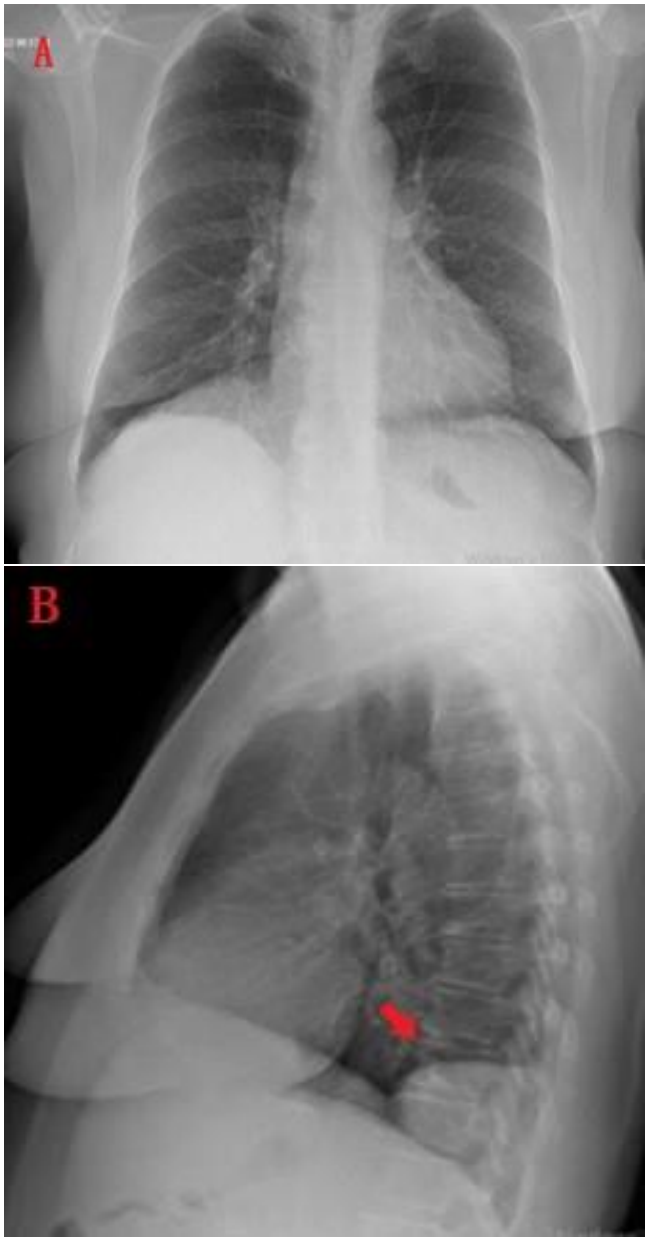

Figure 1a and b: A sharply demarcated opacity in the lower lobe of the right lung was observed adjacent to the diaphragm on posteroanterior chest X-ray (a) and lateral chest X-ray (b)

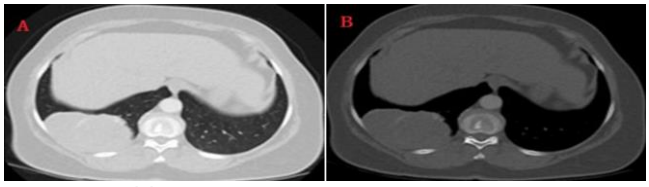

Figure 2a and b: A hypodense mass lesion measuring approximately 8.5x6 cm was observed on computed tomography parenchyma window views (a) and mediastinal window views (b) in the basal part of the right hemithorax, at its widest point

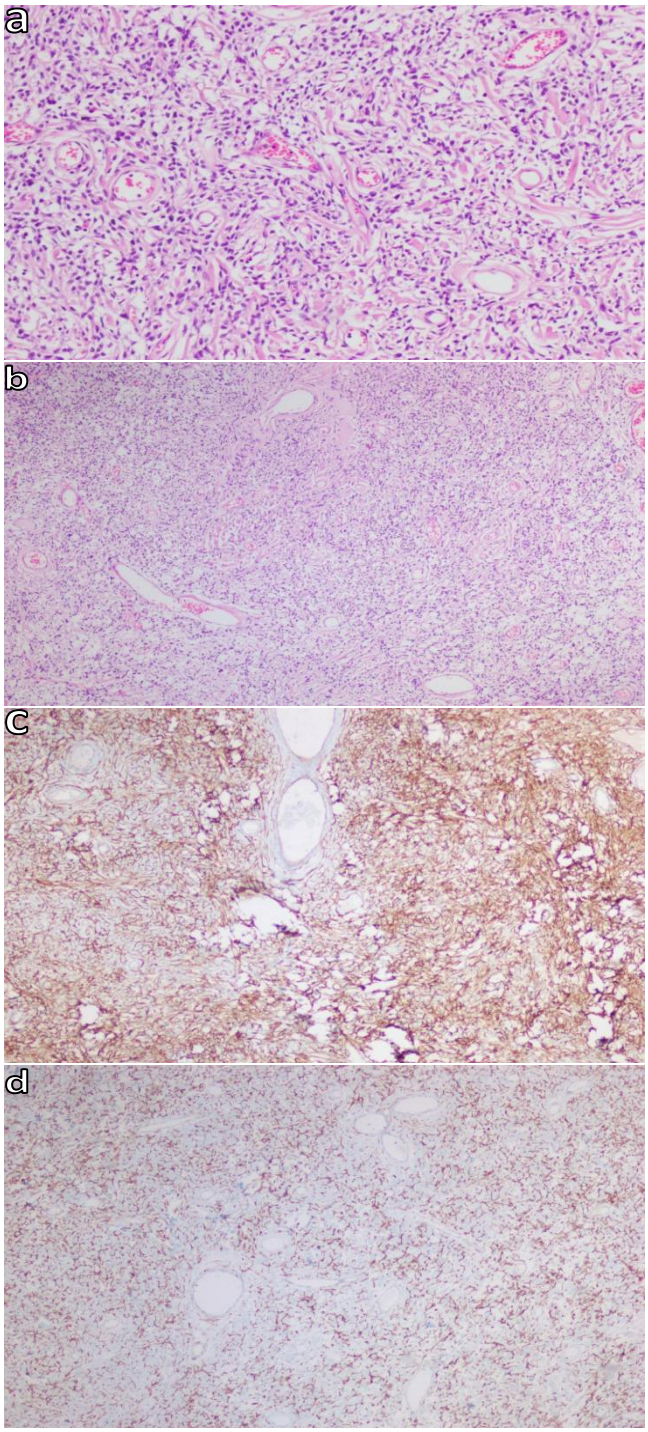

Figure 3a, b, c, and d: Spindle and ovoid cells that do not form a clear pattern in the collagenized stroma (Hematoxylin-eosin x 100) (a); Hemangiopericytimatous vascular pattern (Hematoxylin-eosin x 40) (b); The tumor cells were positive for CD34 (Immunohistochemistry x 100) (c); The tumor cells were positive for BCL 2 (Immunohistochemistry x 40) (d)

The patient underwent thoracoscopic surgery. Right lower lobe excisional biopsy was performed. In pathology examination; A well-circumscribed tumoral lesion measuring 8.5x5 cm in white color with a fibrous appearance; cellularity was moderate, cytological atypia and necrosis was not observed. Mild pleomorphism and <1 mitosis per 10 high power fields, spindle and oval cells forming no clear pattern in the collagenized stroma were observed (Figure 3a). Histopathological features included hemangiopericytimatous vascular pattern (Figure 3b). Immunohistochemical analysis revealed that the tumor cells were positive for CD34 (Figure 3c), CD99, STAT6, vimentin, BCL 2 (Figure 3d), and negative for PAX 8, S-100, NDM2, PanCK, CD31, SMA, ER, CD117, Desmin, SOX10. With these histological features, the diagnosis of solitary fibrous tumor (SFT) was confirmed.

The patient had no postoperative complications and was followed up at 3–6-month intervals after thoracic surgery. In the chest X-ray taken three months after the operation of our patient by the thoracic surgeon and in the thorax CT taken 1 year later, no recurrence occurred during the follow-up period (Figure 4-5 a and b).

DISCUSSION

The solitary fibrous tumor is a rare mesenchymal spindle cell neoplasm mostly originating from the pleura. They usually originate from the visceral pleura and are very rare tumors that make up less than 5% of all pleural tumors (1). SFTs are frequently seen between the ages of 60-70. In our case was a 55-year-old female patient, close to the age range in which SFTs are common.

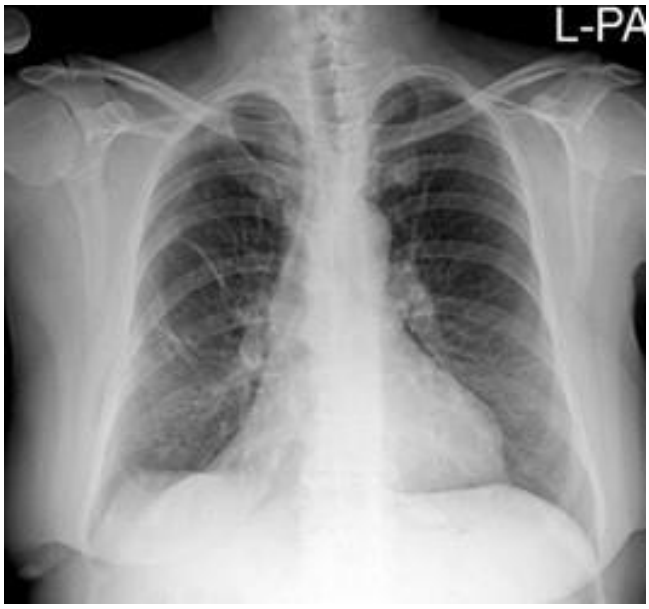

Figure 4: Postoperative 3rd-month control Postero-anterior chest x-ray

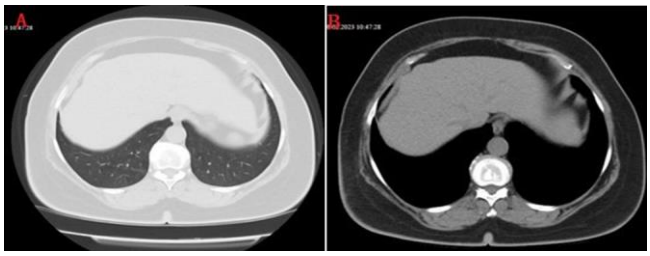

Figure 5a and b: Postoperative 1st-year control computed tomography parenchyma window views (a) and mediastinal window views (b) show no recurrence

In the early stages, SFTs are mostly asymptomatic and are detected incidentally on imaging (4-7). They grow slowly and can be symptomatic when large. These symptoms are mostly atypical chest pain, dyspnea, cough, and hemoptysis (2). They usually develop in the middle-lower hemithorax (8). In our case, a hypodense mass lesion was detected in the lower lobe of the right lung after the examinations performed in her application for back pain.

Thorax CT is an invaluable diagnostic method that clearly shows the location and size of the lesion in patients with SFT. Radiologically, they appear as round masses with smooth borders and are associated with the pleura. In our patient, a well-defined 8.5x6 cm hypodense mass lesion was observed in the basal part of the right hemithorax. These mostly benign tumors can become malignant at a rate of 10-20%. A tumor larger than 10 cm in diameter, infiltrating the chest wall, containing areas of necrosis/hemorrhage, and/or with associated pleural effusion has a greater risk of malignancy (2). Definitive diagnosis is made histopathologically and immunohistochemically (2-8). It is diagnostically important to see a hemangiopericytoma-like vessel pattern accompanied by proliferating spindle cells and thick collagen bands in SFTs. SFT is positive for vimentin, CD34 and Bcl2 (2). Histopathologically, mitotic rate > 4/10 high-power areas, presence of necrosis, atypical nuclei and hypercellularity are criteria for malignancy (9). In our case histopathological examination; cellularity was at a moderate level, cytological atypia and necrosis were not observed. There was mild pleomorphism, mitotic rate <1/10 high-power areas and a hemangiopericytimatous vascular pattern. In immunohistochemical analysis, tumor cells were positive for CD34, STAT6, vimentin, BCL 2. With these findings, we evaluated the case as a solitary fibrous tumor. Although complete surgical resection of the tumor is the only curative treatment, SFT can usually recur in the first two years of resection. Therefore, six-monthly follow-up, chest X-ray, or Thorax CT scans are recommended in the first two years after resection (7). In the chest X-ray taken three months after the operation of our patient by the thoracic surgeon and in the thorax CT taken 1 year later, no findings in favor of recurrence were observed.

In conclusion, solitary fibrous tumors of the pleura are extremely rare. They are usually asymptomatic and detected incidentally. Diagnosis is made radiologically and histopathologically. They are usually benign. The main curative treatment method is surgical resection of benign solitary fibrous tumors. However, they may lead to repeated recurrences and/or malignant transformation. Because of that long-term clinical and radiological follow-up periodically in the postoperative period is recommended for all patients with solitary fibrous tumors.

CONFLICTS OF INTEREST

None declared.

AUTHOR CONTRIBUTIONS

Concept - E.A., M.Ç., A.K., Z.E., N.S.; Planning and Design - E.A., M.Ç., A.K., Z.E., N.S.; Supervision - E.A., M.Ç., A.K., Z.E., N.S.; Funding - E.A., M.Ç., Z.E.; Materials - E.A., M.Ç., A.K.; Data Collection and/or Processing - E.A., M.Ç., A.K.; Analysis and/or Interpretation - E.A., M.Ç.; Literature Review - E.A., M.Ç.; Writing - E.A., M.Ç.; Critical Review - E.A., M.Ç.

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