SCIE DOI: 10.14744/scie.2022.53179
South. Clin. Ist. Euras. 2022;33(3):336-340

Case Report

Primary Pulmonary Fibrosarcoma in Giant Mass Appearance: A Rare Case Report

Selime Kahraman,[1] Attila Özdemir,[2] Talha Dogruyol,[2] Mesut Buz,[2] Berk Cimenoglu,[2] Gonca Geçmen,[3] Recep Demirhan[2]

[1]Department of Thoracic Surgery, Dr. Siyami Ersek Thoracic and Cardiovascular Surgery Training and Research Hospital, İstanbul, Türkiye
[2]Department of Thoracic Surgery, Kartal Dr. Lütfi Kırdar City Hospital, İstanbul, Türkiye
[3]Department of Medical Pathology, Kartal Dr. Lütfi Kırdar City Hospital, İstanbul, Türkiye

Submitted: 06.06.2022
Accepted: 20.07.2022

Correspondence: Attila Özdemir, Kartal Dr. Lütfi Kırdar Şehir Hastanesi, Göğüs Cerrahisi Bölümü, İstanbul, Türkiye
E-mail: dr.attilaozdemir@gmail.com

Keywords: Lung fibrosarcoma; primary fibrosarcoma; sarcoma.

ABSTRACT

We planned surgery a fifty six year-old female patient, who applied with the complaint of dyspnea, due to the suspected malignancy. We carried out intrapericardial bilobectomy superior and the final pathological examination showed primary pulmonary fibrosarcoma. Primary pulmonary fibrosarcoma is a very rare type of tumor and surgery is the main choice of treatment. We present this case to contribute to the literature.

INTRODUCTION

The primary sarcomas of the lung constitute 0.5% of all pulmonary malignancies.[1] Fibrosarcomas, which are a subtype of sarcomas and, originate from the mesenchymal cells, may emerge in any part of the body, and may be encountered in any age group.[2] Because they are reported as rare case presentations in the literature, there is no definitive treatment protocol.

Primary pulmonary fibrosarcomas are usually low-grade tumors, but have a high recurrence rate.[3] The survival rate is better in tumors ≤5 cm compared with tumors >5 cm.[4] Early diagnosis is essential to improve survival, and a complete resection with sufficient surgical margins should be carried out.[5]

CASE REPORT

Respiratory sounds could not be detected in the upper and middle zones of the right lung during the physical examination of the 56-year-old female patient, applied with the complaint of dyspnea who had no systemic disease except for hypertension. As the posteroanterior X-ray image of the lung showed a homogeneous opaque mass with regular contours, which filled almost all right lung, the patient was referred to thoracic computed tomography (thoracic CT). Thoracic CT displayed a mass lesion (152×125 mm), which filled the apex and upper zone of the right lung, shifted mediastinum and trachea toward the left, and had necrotic regions in it. Positron emission tomography (PET-CT) and tru-cut biopsy were planned for further examination. In the PET-CT examination, the maximum standardized uptake value (SUV_{max}) of the mass was 4.3. No fluorodeoxyglucose uptake was observed in the mediastinal lymph nodes, and no extrathoracic metastasis was seen. The result of the tru-cut biopsy indicated a chronic non-specific inflammation. An exploratory thoracotomy was planned for the patient. The contrast-en-

hanced cranial magnetic resonance imaging, which was carried out during the preoperative period, did not show any pathological findings.

We performed a right posterolateral thoracotomy and observed a soft-structured mass lesion, which completely filled the upper and middle lobes. We collected samples from the lesion, and a frozen-section analysis was performed. The result of the frozen-section examination was interpreted in favor of malignant mesenchymal tumor consisting of fusiform and epithelioid cells. As the mass lesion involved the vein of the upper lobe, intrapericardial bilobectomy superior and mediastinal lymph node dissection was carried out.

In the final pathology report, the macroscopic dimension of the tumor was 14×7×6 cm. The histopathological examination showed a mesenchymal tumor consisting of monomorphic spindle cells. The stroma contained variable collagen ranging from a sensitive intercellular network to regions with few cells. Immunohistochemically, tumor cells displayed positive staining with vimentin and CD34. No tumor tissue was determined in the surgical margins of bronchus-vessel-parenchyma. Tumor tissue was determined in the pericardial surgical margin. The lymph nodes 3A-7-8-9-10 were considered anthracotic.

No morbidity and mortality were encountered in the postoperative period. The drain was removed on the 8th day, and the patient was discharged with recovery on the 10th day. Radiotherapy was initiated in the postoperative period (Figs. 1–3).

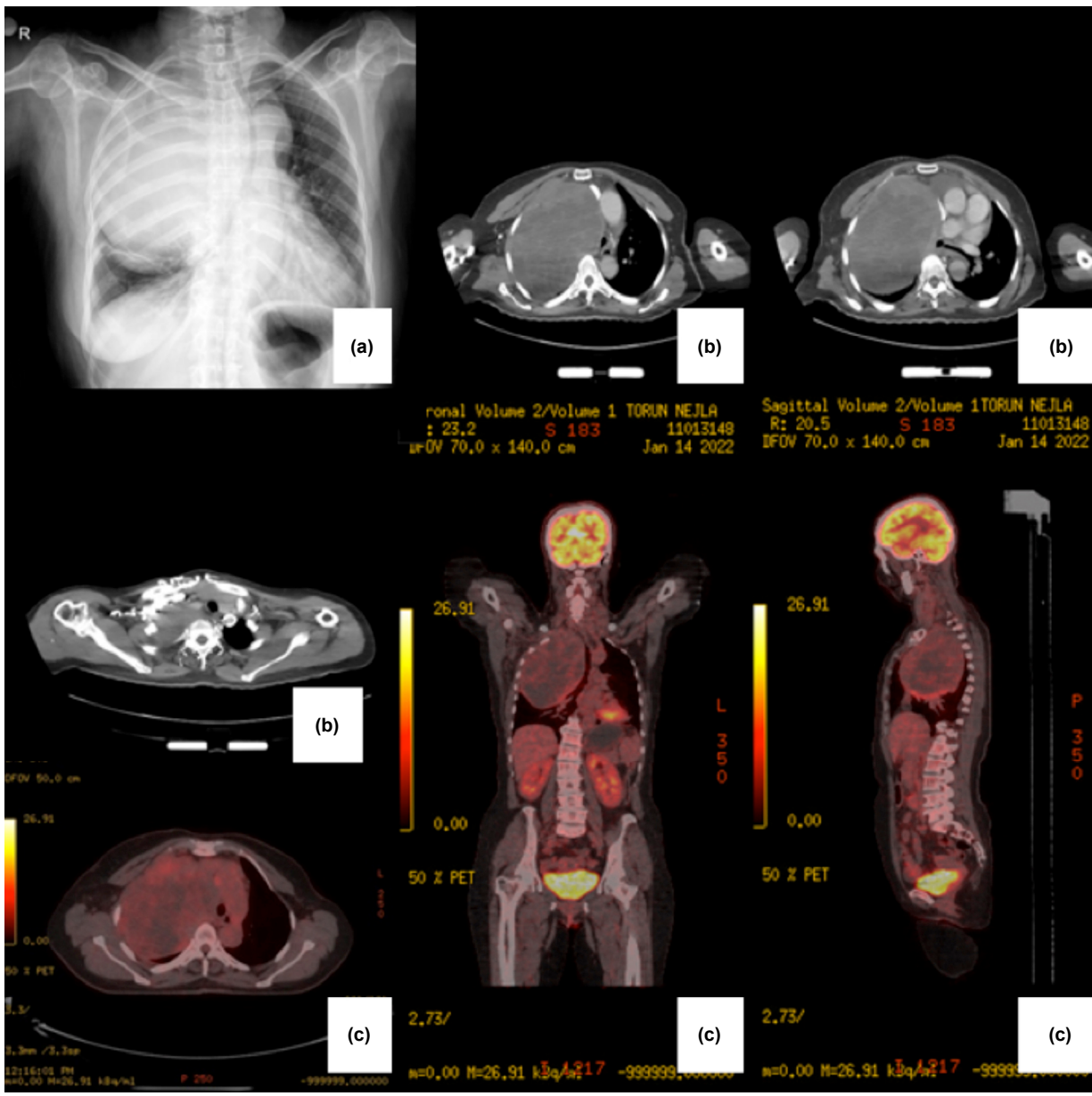

Figure 1. Preoperative radiological evaluation of the patient: **(a)** Chest X-ray, **(b)** axial images in computed tomography, and **(c)** axial, coronal, and sagittal images in positron emission tomography.

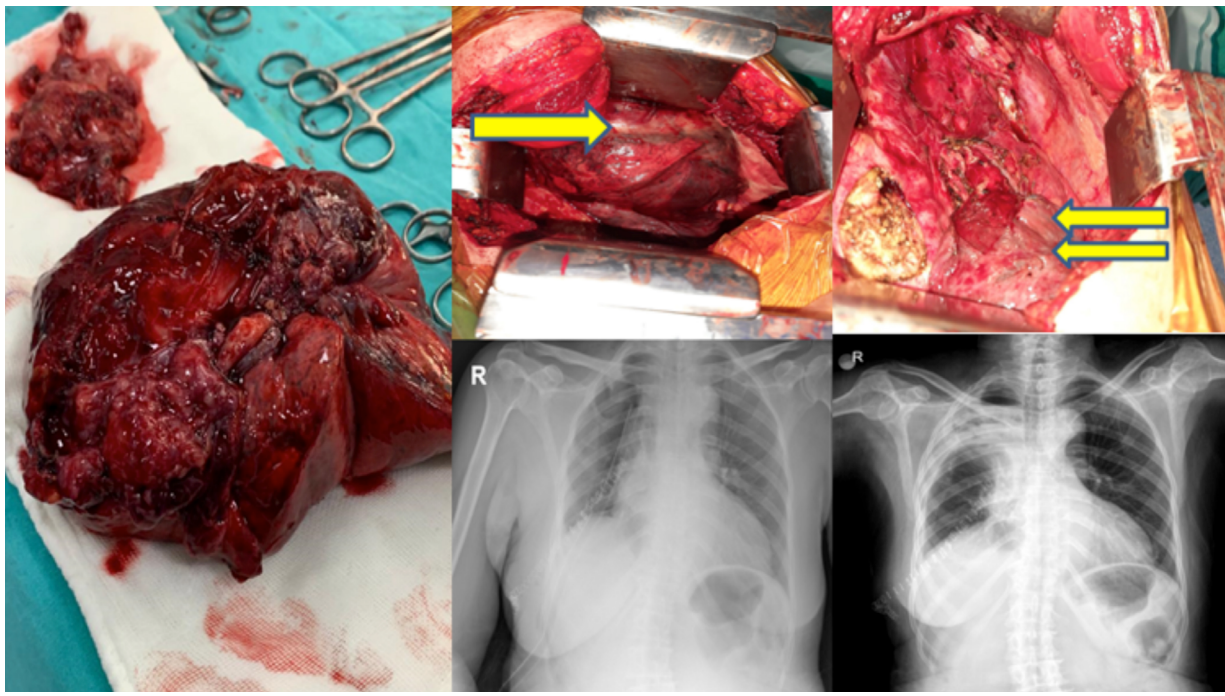

Figure 2. Perioperative images of the patient and pictures of the extracted piece and postoperative PA - lung X-rays. Single arrow: giant mass located in the upper lobe and middle lobe; double arrow: lower lobe.

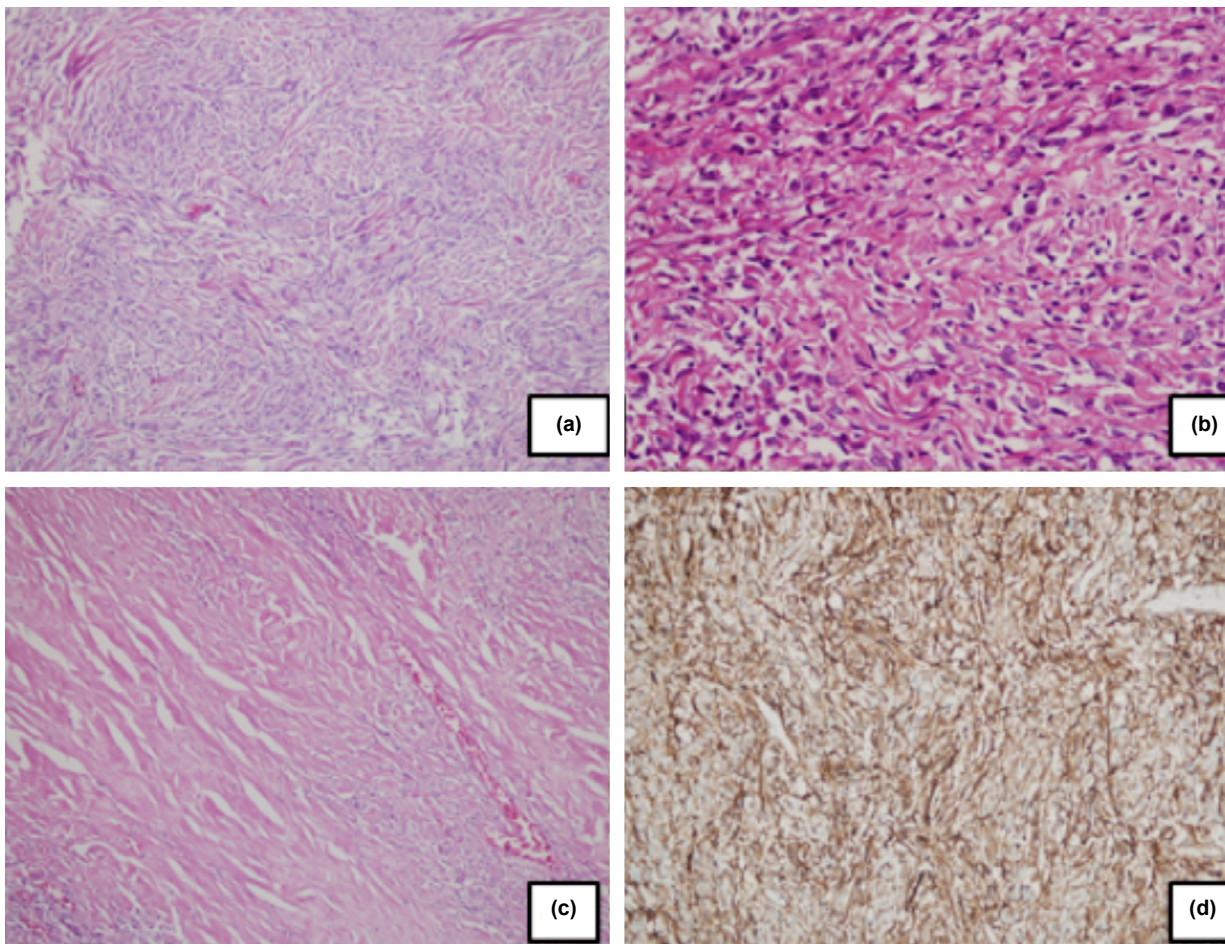

Figure 3. Sections from the patient's pathological specimen. **(a)** Histological examination revealed spindle-shaped cells in the fibrous matrix. **(b)** The stroma has a delicate intercellular network. **(c)** The stroma has particular areas with diffuse sclerosis or hyalinization. **(d)** Diffuse CD34 positivity was observed in tumor cells.

DISCUSSION

The first reports related to pulmonary fibrosarcoma were published in 1931 and 1936.[5,6] In the report of Ball,[5] the cases did not originate from bronchi. In the study by Mallory, one case of pulmonary fibrosarcoma was reported in 8,000 autopsies.[6] The first fibrosarcoma case was published by Pollak et al.[7] in 1941.

The primary sarcomas of the lung constitute 0.5% of all pulmonary malignancies.[1] Primary bronchopulmonary fibrosarcoma is the second most common intrathoracic sarcoma after leiomyosarcoma. Despite this fact, less than 100 cases have been reported worldwide.[8]

The intrathoracic fibrosarcomas tend to emerge as endobronchial masses. Lobar bronchi are involved in children and adolescents, while solidary and multinodular or mass lesions are usually seen in the lungs of adults.[9] When the intrapulmonary fibrosarcomas develop as small endobronchial lesions, they cause cough and hemoptysis. On the other hand, intraparenchymal lesions usually do not cause symptoms.[2] In our case, there was dyspnea depending on the tumor compression, and this complaint was relieved following surgery. As the tumor was located in the intraparenchymal area in our case, we believe that it remained asymptomatic until its size grew to cause compression.

The most common histological subtypes of lung sarcomas are synovial sarcoma, epithelioid hemangioendothelioma, leiomyosarcoma, and malignant peripheral nerve sheath, which are followed by pleomorphic sarcoma, liposarcoma, and rhabdomyosarcoma.[10] When diagnosing primary pulmonary fibrosarcoma in our case, the solitary fibrous tumor was excluded with negative STAT6 staining; and synovial sarcoma with the negative TLE1 staining, and sarcomatoid sarcoma was excluded as epithelial differentiation was not determined with multiple cytokeratins.

The survival rate of primary pulmonary sarcomas is usually worse in the soft tissue sarcomas of the extremities.[11] Endobronchial tumors can be detected earlier, and thus they have a better prognosis.[11] Primary pulmonary fibrosarcomas are usually low-grade tumors but have a high recurrence rate.[3] Early diagnosis of a tumor is essential to improve survival, and a complete resection with sufficient surgical margins should be performed. If the surgical margins are positive and complete resection is not possible, postoperative radiotherapy should be implemented.[12] The largest primary pulmonary fibrosarcoma reported in the literature had a size of 14 cm.[13] In our case, the largest dimension of the tumor was also 14 cm. As the pericardial surgical margin was positive, our case received radiotherapy in the postoperative period. Our case is currently in the 3rd month of the follow-up, and we have not yet observed recurrence or mortality.

CONCLUSION

Primary pulmonary fibrosarcoma is a rare pulmonary malignancy. As only a limited number of cases are reported in the literature, a definitive treatment protocol is not available, but surgery is considered the main treatment choice. We presented this case to contribute to the literature.

Informed Consent

Written informed consent was obtained from the patient for the publication of the case report and the accompanying images.

Peer-review

Internally peer-reviewed.

Authorship Contributions

Concept: S.K., A.Ö.; Design: M.B., B.C.; Supervision: T.D., R.D.; Fundings: S.K., M.B.; Materials: S.K., A.Ö.; Data: B.C., R.D.; Analysis: A.Ö., M.B.; Literature search: B.C., T.D.; Writing: T.D., R.D.; Critical revision: T.D., R.D.

Conflict of Interest

None declared.

REFERENCES

1. Etienne-Mastroianni B, Falchero L, Chalabreysse L, Loire R, Ranchère D, Souquet PJ, et al. Primary sarcomas of the lung: a clinicopathologic study of 12 cases. Lung Cancer 2002;38:283–9.
2. Turan D, Ozgul MA, Cansever L. A Rare Case of Pulmonary Fibrosarcoma Treated by Sleeve Lobectomy. Respir Case Rep 2020;9:125–8. [CrossRef]
3. Ono N, Sato K, Yokomise H, Tamura K, Horikawa S, Suzuki Y, Nishiyama H, Maekawa N, Shizuki K. Primary bronchopulmonary fibrosarcoma: report of a case. Surg Today 1998;28:1313–5.
4. McCormack PM, Martini N. Primary sarcomas and lymphomas of lung. In: Thoracic surgery: frontiers and uncommon neoplasms. St. Louis: Mosby; 1989. p. 269.
5. Ball HA. Primary pulmonary sarcoma: a additional review with a report of an case. Am J Cancer 1931;15:2319–30.
6. Mallory TB. Case 22441: Case records MGH. N England J Med 1936;215:837–9. [CrossRef]
7. Pollak BS, Cohen S, Barrone MG. Primary sarcoma of bronchus. Am J Roentgenol 1941;41:909.
8. Pettinato G, Manivel JC, Saldana MJ. Primary bronchopulmonary fibrosarcoma of childhood and adolescence: reassessment of a low-grade malignancy: clinicopathologic study of five cases and review of the literature. Hum Pathol 1989;20:463–71. [CrossRef]
9. Perez DG, Aubry MC, Molina JR. Primary lung sarcomas. The Mayo Clinic experience. J Clin Oncol 2004;22:7370. [CrossRef]
10. Attanoos RL, Appleton MAC, Gibbs AR. Primary sarcomas of the lung: a clinicopathological and immunohistochemical study of 14 cases. Histopathology 1996;29:29–36. [CrossRef]
11. Matthew BS, Eric B, Ryan B. An analysis of patient characteristics and clinical outcomes in primary pulmonary sarcoma. J Thorac Oncol 2013;8:147–51. [CrossRef]
12. Loh ML, Ahn P, Perez-Atayde AR. Treatment of infantile fibrosarcoma with chemotherapy and surgery: results from the Dana-Farber Cancer Institute and Children's Hospital, Boston. J Pediatr Hematol Oncol 2002;24:722–6. [CrossRef]
13. Porte HL, Metois DG, Leroy X. Surgical treatment of primary sarcoma of the lung. Eur J Cardiothorac Sur 2000;18:136–42.

Dev Kitle Görünümünde Primer Akciğer Fibrosarkomu: Nadir Bir Olgu Sunumu

Nefes darlığı şikayeti ile başvuran elli altı yaşında kadın hastayı malignite şüphesi ile ameliyat etmeyi planladık. İntraperikardiyal bilobektomi superior yaptığımız hastanın nihai patolojisi primer akciğer fibrosarkomu olarak raporlandı. Primer akciğer fibrosarkomu oldukça nadir görülen bir tümör olup tedavisinde cerrahi esas rolü oynamaktadır. Olgumuz litaratüre katkı amacıyla sunulmaktadır.

Anahtar Sözcükler: Akciğer fibrokarsomu; primer fibrosarkom; sarkom.