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A Rare Case of Pulmonary Fibrosarcoma Treated by Sleeve Lobectomy

Sleeve Lobektomi ile Tedavi Edilen Nadir Bir Pulmoner Fibrosarkom Olgusu

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Abstract

A 28-year-old male presented to the emergency department with hemoptysis. A pulmonary examination revealed diminished lung sound on left side, and a CT scan of the chest showed an endobronchial polipoid lesion in the left main bronchus. A rigid bronchoscopy showed a polipoid lesion in left the main left bronchus, while the orifice of the lower lobe was not visible. The lesion was cut into two pieces with a snare and removed with cryotherapy, and hemostasis was achieved. A pathological examination of the lung bronchial biopsy specimen revealed fibrosarcoma. A left lower sleeve lobectomy was performed, and as surgical margins were tumor free, no chemo/radiotherapy was considered necessary. Primary endobronchial pulmonary fibrosarcomas exhibiting polypoid growths are rare. Surgical excision is the preferred treatment option in such patients.

Key words: *Endobronchial fibrosarcoma, endobronchial therapy, Hemoptysis, lung surgery, snare electrocautery.*

Özet

Yirmi sekiz yaşında erkek hasta, acil servise hemoptizi şikayetiyle müracaat etti. Solunum sistemi muayensinde solda solunum sesleri azalmıştı ve tomografide alt lobda tıkayıcı polipoid bir lezyon ve distalinde atelektazi vardı. Bronkoskopide sol ana bronşa taşan polipoid lezyon görüldü. Lezyonun görünen kısmından snare ile biyopsi alındı ve cryo ile hemostaz sağlandı. Patolojisi fibrosarkom gelen hastaya sleeve sol alt lobektomi uygulandı. Lenf nodları ve cerrahi sınırlar negatif bulundu. Hastaya kemo/radyoterpi tedavi uygulanmadı. Primer pulmoner fibrosarkomların polipoid olarak büyümesi nadir görülmektedir. Bu hastalarda tercih edilen tedavi şekli cerrahi eksizyondur.

Anahtar Sözcükler: *Endobronşial fibrosarkom, endobronşial terapi, hemoptizi, akciğer cerrahisi, elektrokoter.*

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Primary sarcomas of the thorax are rare, but can occur in the lung, mediastinum, pleura and chest wall, while fibrosarcomas usually develop in the chest wall. Although primary thoracic sarcomas commonly manifest as large, heterogeneous masses, they may also develop as solitary pulmonary nodules, central endobronchial tumors and intraluminal masses within the pulmonary arteries (1). Early diagnosis is vital, although it is important to exclude other spindle cell proliferations and sarcomatous neoplasms with a similar morphological appearance. This report describes the case of a primary pulmonary fibrosarcoma presenting as a polypoid endobronchial growth mass, which can be treated surgically.

CASE

A 28-year-old man presented to the emergency department with hemoptysis. He had been in a usual state of health until 2 months prior to presentation, when he experienced hemoptysis. In a physical examination he was found to be in mild respiratory distress. A pulmonary examination revealed diminished lung sounds on the left side. A chest radiography revealed volume reduction in the left lung and a mediastinal shift toward to the left side (Figure 1). A CT scan of the chest showed an endobronchial polypoid lesion in the left main bronchus and atelectasis on the left upper lobe in the apicoposterior segment, the lingular segment and the lower lobe subsegments (Figure 2). A rigid bronchoscopy was carried out. A polypoid lesion was observed in the main left bronchus, and mucosal hypervascularity was present (Figure 3). The lesion was cut into two pieces with a snare and removed with cryotherapy. A pathological examination of lung bronchial biopsy specimens revealed a spindle cell mesenchymal tumor with low malignancy potential. The results of immunohistochemistry staining were: Pancitokeratin (-), Vimentin Clone V9 (+), Desmin (+), CD34 (-), S100 (-), ALK (-), CD 56 (-), Synaptophisin (-) and Kİ 67 (+). A consultation with another pathology service confirmed the diagnosis of spindle cell mesenchymal tumor; there was no necrosis and mitosis was 3/10. Differentiations were excluded based on immunohistochemistry staining: Miyogenin (-) and SOX (-). The final diagnosis was fibrosarcoma (Figure 4). A left lower sleeve lobectomy was performed to achieve tumor negative margins after the diagnosis was confirmed. A pathological examination of the lobectomy material revealed no residual tumor. The chest X-ray following surgery is shown in Figure 5. After 4 months of surgery, a PET/CT scan showed no FDG uptake in any part of the body. A fiberoptic bronchoscopy was applied 1 month after the PET/CT scan, and no endobronchial lesion was observed in the left upper lobe or lingula. The patient's status was stable at postoperative 24 months. No chemo/radiotherapy was considered necessary after surgery.

DISCUSSION

Fibrosarcoma is a malignant neoplasm of mesenchymal origin, and is a very rare malignancy that can occur anywhere in the body and in any age group. Primary pulmonary sarcoma occurs in 0.1% of all primary pulmonary malignant neoplasms (2). Intrathoracic fibrosarcomas tend to present as endobronchial masses in the main or lobar bronchi in children and young adults, while in adult lungs they tend to occur as solitary or multiple nodules or masses (3). Although intrapulmonary fibrosarcomas often produce no symptoms, especially when small, endobronchial lesions typically manifest with cough, hemoptysis or chest pain. Our patient presented only with hemoptysis.

In radiological analyses, fibrosarcomas in the chest wall and heart usually manifest as masses that are often heterogeneous in attenuation and signal intensity on CT and MR images (4). In the lung, fibrosarcomas manifest as well-marginated smooth or lobular nodules, or as masses on CT (3). Endobronchial tumors can manifest as atelectasis or post-obstructive pneumonitis. Our patient's chest radiographic finding was similar to those of previously reported cases, involving atelectasis and no pulmonary/pleural mass.

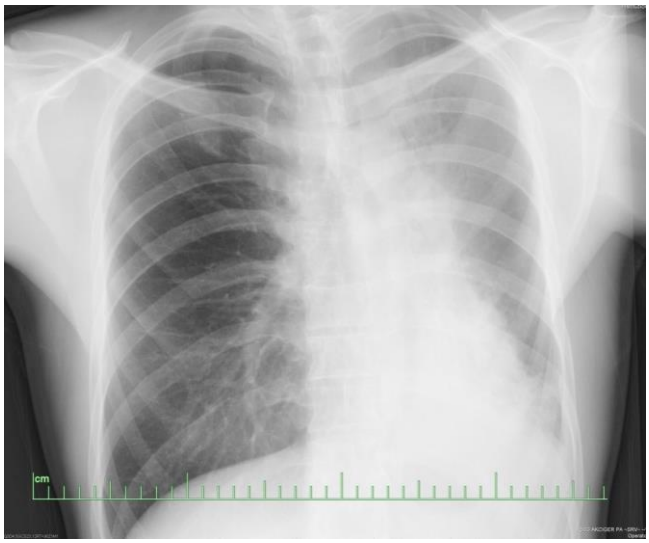

Figure 1: *Chest radiography revealed volume reduction in the left lung, and a mediastinal shift toward to the left side*

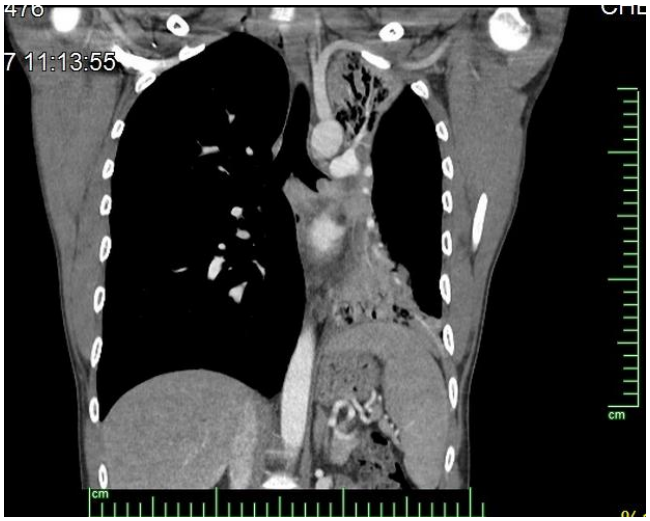

Figure 2: *A CT scan of the chest showed an endobronchial polipoid lesion in the left main bronchus, and atelectasis on the left upper lobe apicoposterior segment, lingular segment and lower lobe subsegments*

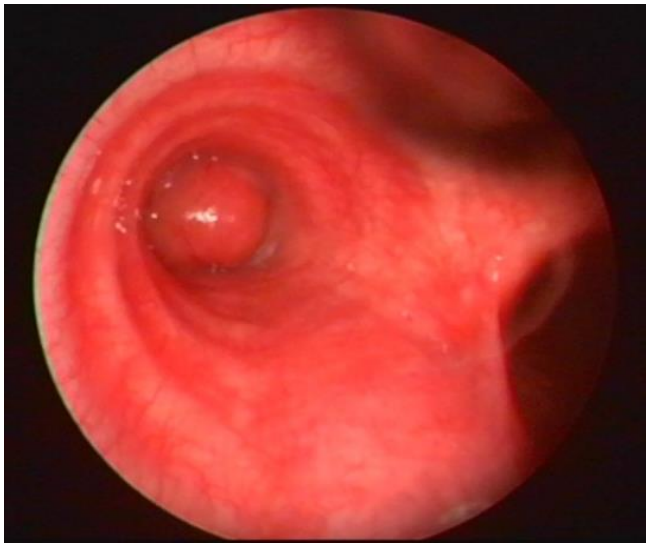

Figure 3: *A polipoid lesion was observed in the main left bronchus, and mucosal hypervascularity was also present*

The histological immunohistochemical evaluation and electron microscopy of tumor cells is critical for tumor diagnosis. Immunohistochemistry is helpful when differentiating fibrosarcoma from other similar tumors in the chest (fibrous mesotheliomas, malignant fibrous tumors of the pleura, and other sarcomas, such as myxofibrosarcoma, synovial sarcoma or nerve sheath sarcoma). Tumors are usually highly cellular, consisting of spindle cells with a fusiform nucleus, arranged in a herringbone or broad fascicular pattern (5). The diagnosis of our patient was confirmed through an immunohistochemical evaluation of the resected endobronchial mass.

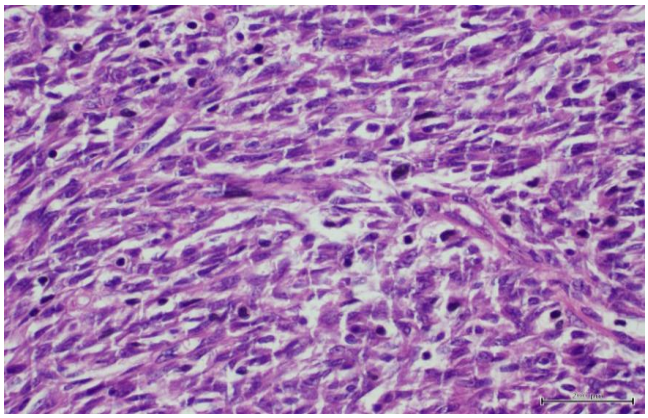

Figure 4: *Microscopy of fibrosarcomatoeus cells (H&E – 20x10)*

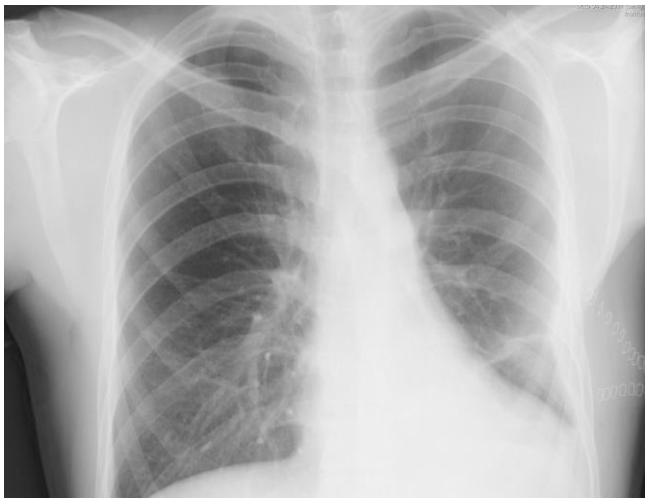

Figure 5: *A chest X-ray after surgery revealed no atelectasis*

Endobronchial lesions are usually treated with local excision, and long-term survival is common. The treatment of chest wall and mediastinal lesions is resection, with preoperative chemotherapy used to improve resectability. Postoperative radiation therapy is applied if the surgical margins are positive or if a complete resection is not possible (6). There are several reports in literature highlighting the effective role of adjuvant chemotherapy after tumor resection for the treatment of infantile fibrosarcoma. The Mayo Clinic reported 40 patients with infantile fibrosarcoma who were treated with only surgery and radiotherapy (7). Our patient's endobronchial lesion was treated with local excision, cryotherapy and APC. Following diagnosis, the patient underwent a left lower sleeve lobectomy, and no adjuvant treatment options were recommended.

The response to therapy is often poor, with larger masses tending to recur locally, whereas smaller lesions have a high likelihood of metastasis (3). Our case had no hematogenous or lymphatic metastasis. Prognosis is correlated with tumor size, histologic grade, mitotic count and the location of the tumor (endobronchial vs. intra-

parenchymal) (3,8). In one case series, all patients with a primary fibrosarcoma of the lungs larger than 5 cm in diameter eventually died of the tumor (9). Primary pulmonary sarcomas are generally associated with worse overall survival than with soft tissue sarcomas of the extremities (10). An aggressive approach to treatment is advocated, with radiotherapy and chemotherapy preferred for unresectable cases. Endobronchial tumors are usually detected earlier and have a better prognosis. Intraparenchymal tumors often behave in a highly malignant fashion, with death occurring within 2.5 years. Tumors with eight mitotic figures per 10 high-power fields behave in an aggressive fashion, while others remain stationary or tend to grow slowly (11). Our patient is still alive, although the ultimate prognosis remains unknown.

In conclusion, primary pulmonary fibrosarcomas with polypoid endobronchial growth are rare. Although interventional bronchoscopy can rapidly and efficiently remove endobronchial tumors, surgical excision is the preferred treatment option in such patients.

CONFLICTS OF INTEREST

None declared.

AUTHOR CONTRIBUTIONS

Concept - D.T., M.A.O., L.C., M.A.B.; Planning and Design - D.T., M.A.O., L.C., M.A.B.; Supervision - D.T., M.A.O., L.C., M.A.B.; Funding -; Materials - D.T.; Data Collection and/or Processing - D.T.; Analysis and/or Interpretation - L.C.; Literature Review - L.C.; Writing - M.A.O.; Critical Review - M.A.B.

YAZAR KATKILARI

Fikir - D.T., M.A.O., L.C., M.A.B.; Tasarım ve Dizayn - D.T., M.A.O., L.C., M.A.B.; Denetleme - D.T., M.A.O., L.C., M.A.B.; Kaynaklar -; Malzemeler - D.T.; Veri Toplama ve/veya İşleme - D.T.; Analiz ve/veya Yorum - L.C.; Literatür Taraması - L.C.; Yazıyı Yazan - M.A.O.; Eleştirel İnceleme - M.A.B.

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