Turkish Journal of Thoracic and Cardiovascular Surgery 2023;31(4):577-580

Case Report / Olgu Sunumu

A rare phenomenon, primary pulmonary smooth muscle tumor and its management

Nadir bir fenomen, primer pulmoner düz kas tümörü ve tedavisi

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ABSTRACT

Smooth muscle tumors of uncertain malignant potential, the borderline tumors arising from the smooth muscle cells, usually grow slowly and do not fulfill the diagnostic criteria of leiomyosarcoma and its variants, but may behave in a malignant manner. A 15-year-old female patient with an endobronchial mass in the left main bronchus on thoracic computed tomography underwent thoracotomy and tracheobronchoplasty with a wide and safe margin. Histopathological evaluation revealed a smooth muscle tumor of uncertain malignant potential. There were no complications related to the operation during the hospital stay. At six months of surgery, there were no symptoms or signs suggesting any recurrence in her follow-up. In conclusion, In conclusion, pulmonary leiomyomas may rarely present as an endobronchial mass and may mimic asthma by causing respiratory symptoms developing as attacks due to displacement of the mass within the lumen.

Keywords: Endobronchial mass, pulmonary leiomyoma, pulmonary leiomyosarcoma, smooth muscle tumor.

ÖZ

Düz kas hücrelerinden kaynaklanan borderline tümörler olan malign potansiyeli belirsiz düz kas tümörleri, genellikle yavaş büyürler ve leiomiyosarkom ve varyantlarının tanı kriterlerini karşılamazlar; ancak malign bir şekilde davranabilirler. Toraks bilgisayarlı tomografide sol ana bronşta endobronşiyal kitle saptanan 15 yaşındaki kadın hastaya geniş ve güvenli sınırla torakotomi ve trakeobronkoplasti uygulandı. Histopatolojik değerlendirmede malign potansiyeli belirsiz düz kas tümörü görüldü. Hastanede kaldığı süre boyunca ameliyat ile ilgili herhangi bir komplikasyon gelişmedi. Ameliyatın altıncı ayında, takibinde nüksü düşündüren herhangi bir belirti veya bulgu yoktu. Sonuç olarak, spulmoner leiomyomlar nadiren endobronşiyal kitle şeklinde presente olabilirler ve kitlenin lümen içinde yer değiştirmesine bağlı olarak ataklar şeklinde gelişen solunumsal semptomlara neden olup astımı taklit edebilirler.

Anahtar sözcükler: Endobronşiyal kitle, pulmoner leiomiyom, pulmoner leiomiyosarkom, düz kas tümörü.

The most well-known smooth muscle tumors are uterine leiomyomas/leiomyosarcomas, but they may occur in any tissue consisting of the smooth muscle cells.[1] Pulmonary smooth muscle tumors are mostly seen as a metastasis from the uterus to the lung, but they rarely also present as primary tumors of the lung. More commonly, they arise in immunocompromised patients or patients with acquired immunodeficiency syndrome or after lung transplantation in association with Epstein-Bar virus, but they may also arise in immunocompetent patients without any predisposing factor.[2] As they usually originate from the airway smooth muscle cells, they present with signs and symptoms of airway obstruction. The ones originating from the pulmonary vessels may mimic pulmonary thromboembolism. Additionally, pleural, mediastinal, and intraparenchymal localizations even with no symptom, are also possible.[3] Based on the characteristics of mitotic index, cytological atypia, and coagulative necrosis, they are classified as

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Doi: 10.5606/tgkdc.dergisi.2023.21893

Received: March 31, 2021
Accepted: September 05, 2021
Published online: October 19, 2023

Cite this article as: Dirol H, Başaran AE, Hicran Özbudak İ, Erdoğan A, Bingöl A. A rare phenomenon, primary pulmonary smooth muscle tumor and its management. Turk Gogus Kalp Dama 2023;31(4):577-580. doi: 10.5606/tgkdc.dergisi.2023.21893

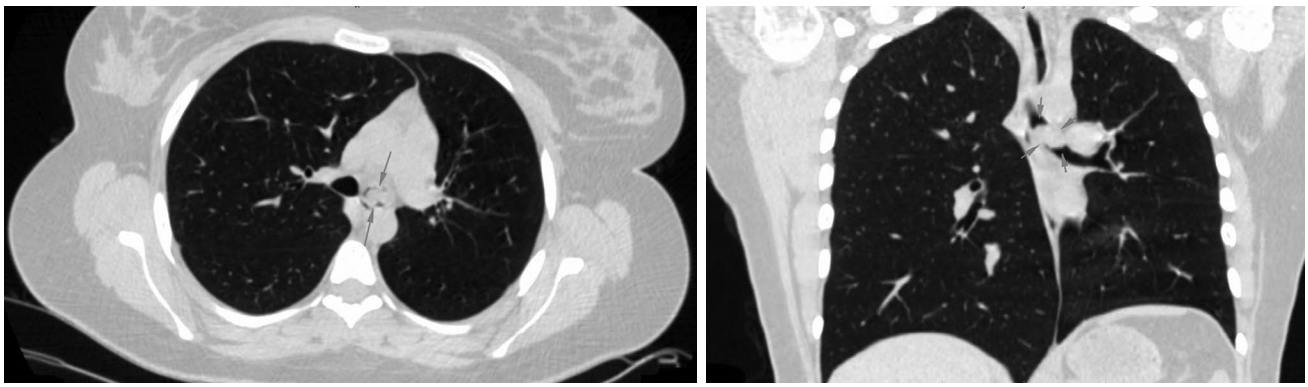

Figure 1. Endobronchial mass in the left main bronchus in coronal and axial plane.

leiomyoma, leiomyosarcoma, and smooth muscle tumor with uncertain malignant potential (STUMP). The STUMPs are distinctly different from leiomyosarcoma histologically, and more similar to leiomyoma but clinically may behave in a malignant manner, similar to a low-grade leiomyosarcoma. What is known about the management of STUMPs consists of information obtained from case series, and uncertainties about classification, treatment, and follow-up are challenging the clinicians.

In this article, we report a case of STUMP and its management in the light of literature data.

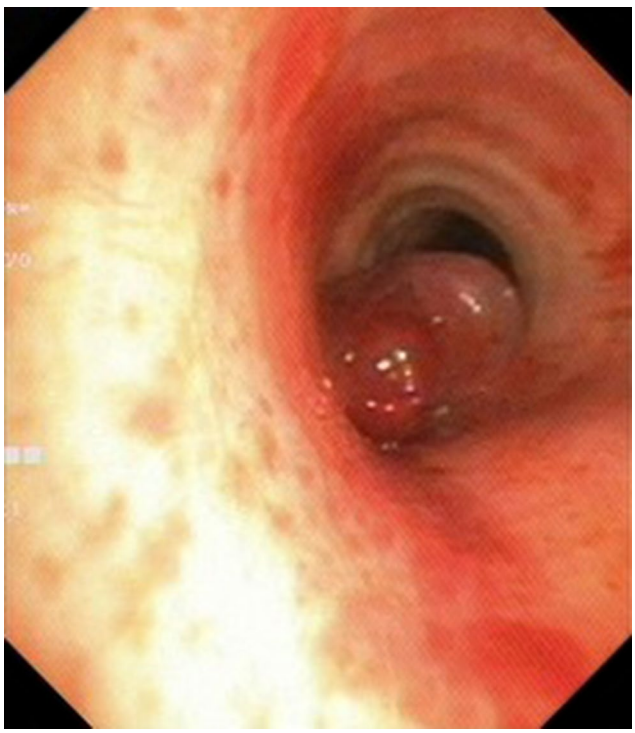

Figure 2. Bronchoscopic appearance of the endobronchial mass.

CASE REPORT

A 15-year-old female patient presented to our outpatient clinic with the complaint of wheezing and dyspnea for a few months. Dyspnea was not related to exercise and sometimes preceded with wheezing or increased, when she lied down. She was diagnosed with asthma and was started on bronchodilators, but she was not benefitted from this treatment and recently her symptoms increased and the night before the day she administered, she was taken to emergency room due to a sudden onset dyspnea while sleeping. She was a passive smoker and denied using illicit drugs. She had no comorbidity and no feature in her medical history either. Her brother had allergic rhinitis and her family history was unremarkable.

On her examination, she was overweight. Her vital signs were normal and oxygen saturation was

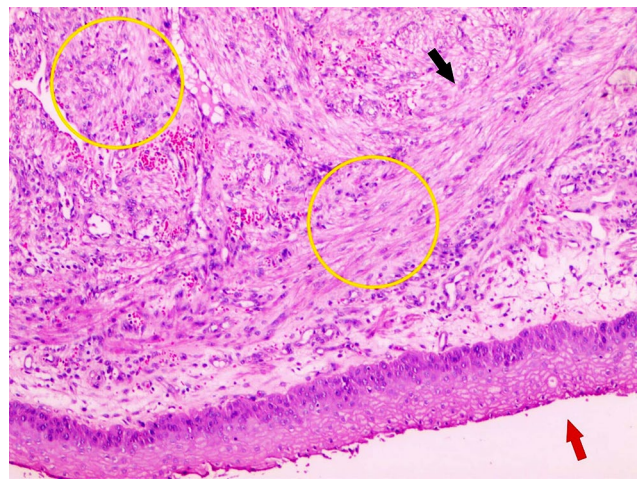

Figure 3. In tumor sections, a well-circumscribed mesenchymal tumor, consisting of intersecting short fascicles, adjacent to the bronchial epithelium was seen. Tumor cells are elongated bland cells with blunt-ended nuclei (H&E, ×100).

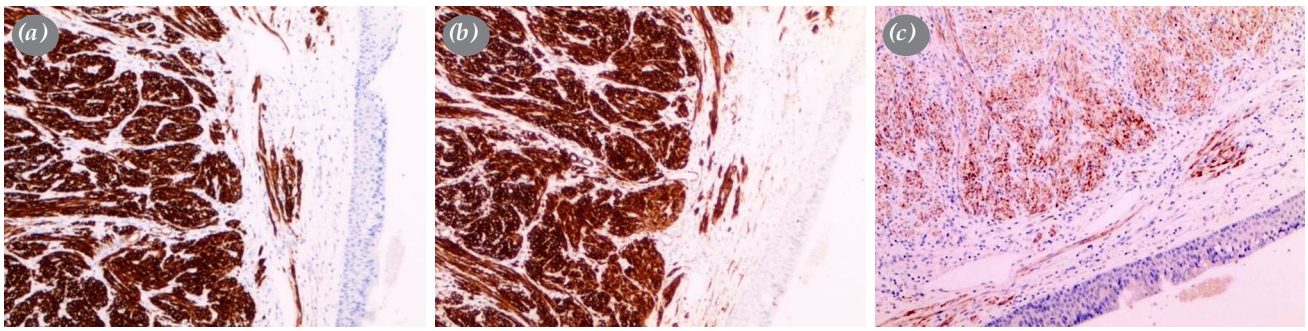

Figure 4. (a) Tumor cells are diffusely immunopositive with Caldesmon and **(b)** muscle-specific actin, **(c)** focal and mild staining was observed with Desmin (×100).

98% at room air. There was a localized rhonchus over the left hemithorax. She had no abdominopelvic mass or tenderness with palpation. Initial laboratory investigations revealed normal biochemical blood tests, a volume loss, and a mild increase in radiopacity of left hemithorax with slightly elevated left hemidiaphragm on chest X-ray. Her pulmonary function test showed an expiratory plateau in the flow-volume loop. Subsequent thoracic computed tomography (CT) revealed a polyploid endobronchial lesion in the proximal left main bronchus (Figure 1). Then, we performed fiberoptic bronchoscopy and observed a fragile, mobile, polypoid endobronchial lesion obliterating the left main bronchus inlet, and we obtained forceps biopsies from that lesion (Figure 2). However, biopsies were not sufficient for the diagnosis.

The patient underwent operation. She was intubated with a single-lumen tube under general anesthesia. For an isolated bronchial sleeve, we entered the thoracic cage through the fifth intercostal space with a standard left posterolateral thoracotomy incision and placed a 2/0 polypropylene suture in the left main bronchus and a sling suture to the right main bronchus. We cut the left main bronchus 0.5 cm below the carina level and the upper part of the left lower lobe and upper lobe carina and performed end-to-end anastomosis with a 3/0 polypropylene. Using two lateral traction sutures, we achieved a better orientation control and created a tension-free anastomosis while bringing the bronchial cut ends together. We completed the operation by placing a chest tube in the thorax. Histopathological evaluation revealed a pulmonary STUMP (Figures 3 and 4). The patient has still no symptom at six months of the operation.

DISCUSSION

Primary pulmonary smooth muscle tumors are fairly rare neoplasms originating from the smooth muscle cells of the airway or vessels in the lung. Based on the histopathological and clinical features, they are classified as leiomyoma and leiomyosarcoma but besides these, there are borderline tumors of the smooth muscle cells called STUMPS that do not fulfill the diagnostic criteria of leiomyosarcoma and its variants, but may behave in a malignant manner.

Primary pulmonary leiomyosarcomas comprise less than 0.5% of primary pulmonary malignancies. They are mostly localized in the upper lobes of the lung as a round-shaped, well-demarcated mass. Symptoms are non-specific and depend mainly on the localization and the size. Positron emission tomography (PET)/CT is useful for the evaluation of distant metastasis or for the differentiation, if it is primary or metastatic. On PET/CT, lymph nodes are commonly not hypermetabolic, as metastasis occurs through the bloodstream, not lymphatics. The PET/CT has even been suggested to be even useful in respect to the grading of the tumor, as the metabolic activity correlates well with necrosis and mitotic activity. Since these tumors are resistant to chemotherapy, the most optimal treatment option is surgery with a wide-safety-margin resection. Neoadjuvant chemotherapy with doxorubicin and ifosfamide may give chance for surgery in cases with unresectable leiomyosarcoma.[4] Palliative surgery and radiotherapy may contribute to survival in relapses.

Pulmonary leiomyomas usually present as asymptomatic solitary pulmonary nodules. Endobronchial leiomyomas may cause cough, hemoptysis, and shortness of breath. It may mimic a carcinoid tumor radiologically.[5] The diagnosis can be made by a histopathological evaluation. The history of uterine surgery and the presence of estrogen/progesterone receptors are useful for the differentiation of benign metastasizing leiomyoma,

which occurs mostly in premenopausal women with a myomectomy or hysterectomy. Radical resection is the preferred treatment option, but bronchoscopic debulking of an endobronchial leiomyoma is also possible.

In general, STUMPs are characterized by slow growth and long-term patient survival compared to leiomyosarcomas. Despite their slow growth, they can invade adjacent structures. No metastasis has been reported so far.[3] Most of the information about these borderline tumors have been obtained from uterine STUMPs. Tumor embolization is not proper due to collateral vessels supplying the tumor. Neither chemotherapy nor radiation therapy is effective; radical surgical resection with a wide and safe margin seems to be the only treatment option.[6]

In conclusion, primary pulmonary smooth muscle tumors should be kept in mind in patients with primary pulmonary smooth muscle tumors should be kept in mind in patients with positional dyspnea that occurs as attacks due to displacement of the polypoid lesion in the airway. Samples obtained by bronchoscopy may not be enough and, for both the diagnosis and treatment, radical surgical resection with a safe and wide margin seems to be the most optimal option.

Patient Consent for Publication: A written informed consent was obtained from the parent of the patient.

Data Sharing Statement: The data that support the findings of this study are available from the corresponding author upon reasonable request.

Author Contributions: Concept: H.D., A.E.B.; Design: H.D., A.E.B., İ.H.Ö., A.E., A.B.; Processing: H.D., A.E.B., İ.H.Ö.; Analysis and/or Interpretation: H.D., A.E.B., A.E.; Literature search: H.D., A.E.B., A.B.; Writing: H.D., A.E.B.

Conflict of Interest: The authors declared no conflicts of interest with respect to the authorship and/or publication of this article.

Funding: The authors received no financial support for the research and/or authorship of this article.

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