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REVIEW ARTICLE

Primary pulmonary leiomyoma

Leiomioma pulmonar primario

Ricardo Adolfo Zapata-González[1], Camilo Andrés Díaz-Quintero[2], Alejandro Cardona-Palacio[3], Victoria Murillo-Echeverri[4]

[1] Physician, specialist in General Surgery and Chest Surgery, Clínica CardioVID, Medellín, Colombia.
[2] Physician, resident of General Surgery, Universidad Pontificia Bolivariana, Medellín, Colombia.
[3] Physician, resident of Pathology, Universidad de Antioquia, Medellín, Colombia.
[4] Physician, specialist in Pathology, Hospital Pablo Tobón Uribe, Medellín, Colombia.

Abstract

Primary lung leiomyoma is a rare benign tumor of mesodermal origin, that accounts for approximately 2% of all benign lung tumors. Since it was first described by Forkel in 1910, at least 150 cases have been reported in the literature. A review of the subject is made regarding the case of an oligosymptomatic postmenopausal woman, with the finding of a solid lung lesion, associated with a vascular malformation, in whom the initial biopsy reported a spindle cell tumor, for which she was taken for a right upper lung lobectomy and whose definitive pathological diagnosis was primary pulmonary leiomyoma. This case is presented due to its low incidence and the little literature on this type of benign tumor lesions.

Keywords: leiomyoma; lung; lung neoplasms; thoracoscopy; thoracic surgery, video-assisted.

Resumen

El leiomioma primario de pulmón es un tumor benigno raro, de origen mesodérmico, que representa aproximadamente el 2 % de todos los tumores benignos de pulmón. Desde que fue descrito por primera vez por Forkel en 1910, se han reportado al menos 150 casos en la literatura. Se hace una revisión del tema a propósito del caso de una mujer posmenopáusica oligosintomática, con hallazgo de una lesión pulmonar sólida, asociada a una malformación vascular, en quien la biopsia inicial reportó un tumor fusocelular, por lo que fue llevada a lobectomía superior del pulmón derecho, y cuyo diagnóstico patológico definitivo fue leiomioma pulmonar primario. Este caso se presenta por su baja incidencia y la poca literatura sobre este tipo de lesiones tumorales benignas.

Palabras claves: leiomioma; pulmón; neoplasias pulmonares; toracoscopía; cirugía torácica asistida por video.

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Correspondence: Camilo Andrés Díaz Quintero, Clínica Cardio VID, Calle 78B # 75-21, Medellín, Colombia. Phone: 3005149932
Email: camilodzq@gmail.com
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Introduction

Primary lung leiomyoma is a rare benign tumor of mesodermal origin, representing about 2% of all benign lung tumors[1]. It develops from smooth muscle fibers of the tracheobronchial tree, blood vessels or heterotopic embryonic muscle islets in the lung and could be parenchymal or tracheobronchial[2]. Since it was first described by Forkel in 1910[3], have been reported at least 150 cases in the literature.

The importance of its knowledge lies in the low incidence, which leads to confusion in the diagnosis on multiple occasions, with treatments that may be insufficient. Classifications and immunohistochemical characteristics still on debate; for the above, and in order to contribute to the knowledge of pathology, a case presentation is made including a review of the literature with the current evidence on clinical, imaging, pathological and surgical characteristics.

Clinical case

Female patient of 64-year-old, with medical history of arterial hypertension, left bundle branch block, non-insulin diabetes mellitus and dyslipidemia with adequate control, who consulted with a clinical picture consisting of nonspecific and intermittent thoracic pain, in right hemithorax, long-standing and without specific management, that exacerbated after a fall from his own height, associated with non-productive cough. A chest computed tomography (CT) scan with contrast is performed where a nodule in the right upper lobe measured 30 x 26 mm was observed, with well-defined edges and soft tissue attenuation, without contrast medium uptake; additionally, abnormal drainage of the right superior pulmonary vein was found (Figures 1 and 2).

PET-CT studies were performed with little increase in metabolism, no increase in size compared to previous studies, and right hilar lymphadenopathy with slight increase in metabolism.

Fine needle CT-guided biopsy was performed that reported a lymphocytic inflammatory infiltrate, with presence of spindle-shaped cell proliferation, without atypia, which are arranged forming a solid pattern in the middle of hyalinized stroma downright. The immunohistochemistry report described smooth muscle actin positive in neoplastic cells, desmin and CD34; negative for myogenin, cytokeratins and S100. The description was consistent with a spindle cell injury of smooth muscle.

Given these findings, the case was discussed at a multidisciplinary committee, considering lung injury of sarcomatous lineage as a diagnosis, possibly low-grade, therefore an upper right total lobectomy and mediastinal lymphadenectomy by single-port thoracoscopy was proposed as treatment, which was carried out without complications, adequate postoperative evolution and early discharge.

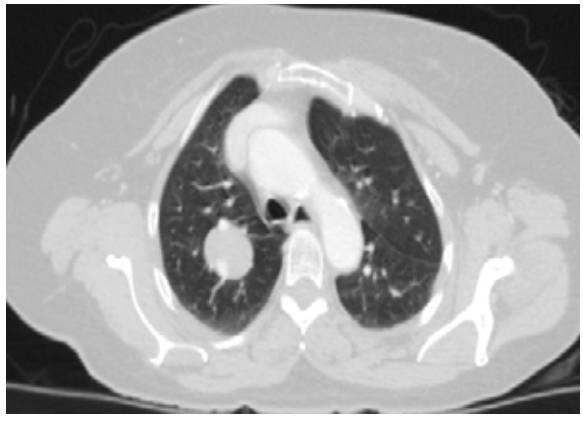

Figure 1. Chest computed tomography where a nodule in the right upper lobe is noticed.

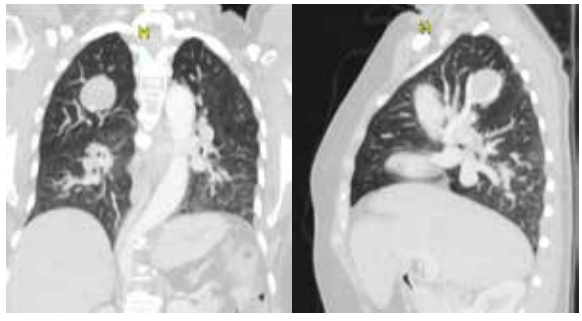

Figure 2. Chest tomography showing a nodule in the right upper lobe and abnormal drainage of the right superior vein.

The final specimen pathology reported a circumscribed mass of 3 cm in diameter, firm, cream colored, that enucleates easily when trying to cut, with negative surgical margins (Figure 3). Immunohistochemical studies were positive for desmin, CD34, H-caldesmon, estrogen and progesterone receptors, and negative for cytokeratins, Bcl2 and Stat 6 (Figure 4). The lymph nodes were negative for malignancy (Figure 5).

Findings are consistent with primary lung leiomyoma based on negative gynecological history for primary neoplasms in genital tract, as well as previous ultrasound where no lesion was appreciated. Up to date, the patient shows an adequate evolution, without complications.

Literature review and discussion

Epidemiology

Primary pulmonary leiomyoma is a rare entity, most cases are eported secondary to lung metastatic lesions, with a primary usually localized at the uterine level. Leiomyomas can also be found in the gastrointestinal tract, but it is exceptionally rare for the tumor to develop in the pulmonary system[4].

Leiomyomas represent 2% of all benign tumors in the pulmonary system, can be located anywhere in the trachea (16%), bronchi (33%) and pulmonary parenchyma (51%)[5]. Its presentation has been described in young, middle-aged women, with an average age of 35 years. In children, immunodeficiency and concomitant infection with Epstein-Barr virus have been implicated as a predisposing factor for proliferation of benign and malignant tumors of the smooth muscle[6].

Our case was a female patient in her seventh decade of life, no previous symptoms or gyn-obstetric history, in whom additionally abnormal pulmonary venous drainage was found.

Clinical presentation

The clinical presentation varies by location and size of the lesion. In cases when bronchi are affected, the symptoms are related with the degree of obstruction of the affected bronchus and the state of the lung parenchyma. The tracheal

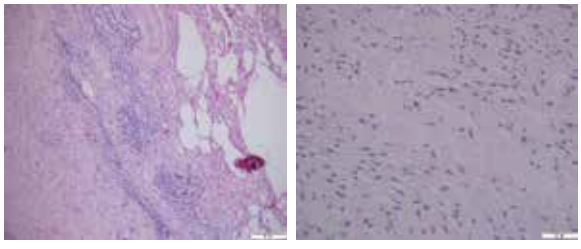

Figure 3. Lobectomy: lung parenchyma adjacent to the leiomyoma. From left to right, hematoxylin and eosin 4x and 20x.

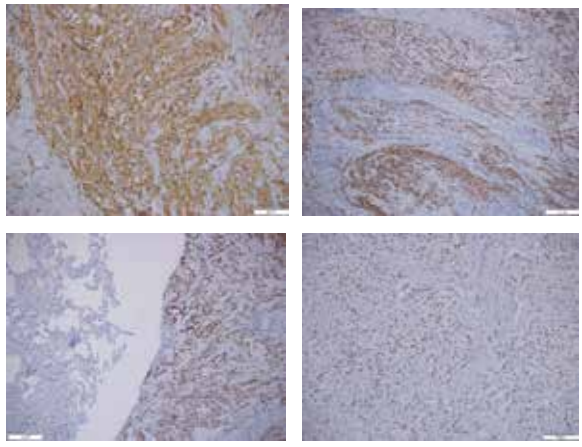

Figure 4. Lobectomy: adjacent lung parenchyma leiomyoma. From left to right, positivity for CD34 and H-caldesmon, desmin, and progesterone.

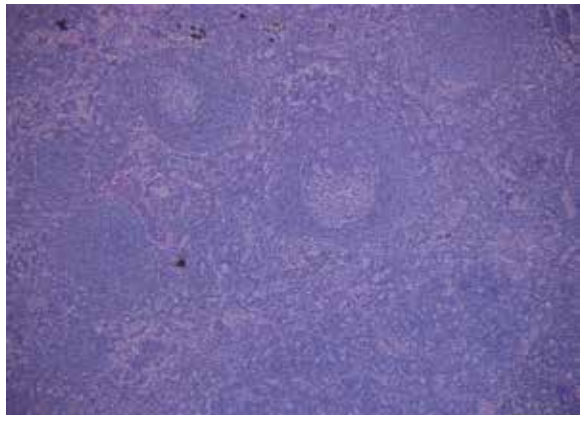

Figure 5. Lymph node station 4 R. Hematoxylin and eosin 10x.

leiomyoma occurs predominantly in the membranous portion of the lower third of the trachea. Produces wheezing and even dyspnea, permanent or intermittent, which can be confused with bronchial asthma.

Symptoms of pulmonary location included cough, wheezing, chest pain, hemoptysis, fever and recurrent pneumonia with risk of bronchiectasis or destruction of parenchyma as the final stage. The parenchymal location is asymptomatic up to 90% of cases, being frequently an incidental finding on a chest x-ray, where usually is evidenced as a single rounded mass, or in an autopsy[7].

As a relevant symptom our patient had non-specific, intermittent chest pain, long-standing, but it was finally an incidental finding after trauma.

Diagnosis

The diagnosis is usually by imaging. The chest x-ray shows a round opacity, homogeneous, well defined. Computed tomography confirms the finding of a round or ovoid lung lesion, well defined, rarely calcified, with or without atelectasis or post-obstructive pneumonia [8].

The work up is complemented with bronchoscopy. The final diagnosis is histological and can be obtained from bronchial biopsies by bronchoscopy, in the central forms of the disease, or by transparietal biopsies guided by CT in peripheral parenchymal forms[9].

Differential diagnosis

The differential diagnosis must be distinguished from low-grade leiomyosarcoma and benign leiomyomatosis metastatic disease, which is a rare lesion with very few reported cases in our setting[10]. The lesion was described the first time by Steiner in 1939[11] who reported a patient who had died from corpulmonale and had a benign uterine fibroid with metastasis in the lung; Steiner used the term "metastatic leiomyoma" to describe it[12]. The metastatic leiomyomatosis can metastasize at a distance, located in the lung, skin, bone, mediastinum, lymph nodes, muscle, heart and retroperitoneum[13]. Usually are multiple and bilateral non-infiltrative lesions, whose growth ceases with the onset of menopause and have a good response to hormonal treatment[14]. The diagnosis is based on the coexistence histology of a benign proliferation spindle cell in addition to a distant lesion of similar characteristics[15].

Macroscopically leiomyomas are solid masses, white to pearl gray when cut it. The microscopic characteristics are smooth muscle cells, with a swirling pattern with bundles of smooth muscle, arranged in longitudinal or transverse manner, eosinophilic cytoplasm with discrete, oval nuclei, cigarette-shaped; characteristically have low cellularity, absence of mitotic count, lack of cytological atypia and absence of necrosis. The neoplastic cells are strong and diffusely positive for smooth muscle actin, confirming the smooth muscle origin of this benign tumor.

Immunohistochemistry is negative for CD117, the c-kit proto-oncogene, ruling out an extra gastrointestinal stromal tumor, and cells are S100 negative, excluding essentially a tumor of neural origin. The metastatic pulmonary leiomyomatosis typically expresses estrogen and progestin receptors, which favors its possible uterine origin[16]. In our case gynecological and abdominal tracts were thoroughly reviewed, without finding neoplasms compatible with leiomyomas.

Treatment

The treatment of choice for lung leiomyoma is a surgical resection and the technique depends on the location and size of the lesion. Tracheobronchial lesions of small sizes can be resected by bronchiectomy, segmental resection, laser endoscopic or electrocautery, especially in high-risk surgical patients[17]. However, an extensive parenchymal resection may be required in case of destruction of the parenchyma. In a series of 76 cases of operated bronchopulmonary leiomyomas, 13 were treated by pneumonectomy, 30 by lobectomy, two by segmentectomy, 12 by segmental resection and 19 by bronchiectomy[18].

There are currently no guidelines for treating primary lung leiomyoma. The treatment strategy depends on the location of the injury in the airway and size of the lesion. In our case, the lesion was 3 cm in diameter, located in the right upper lobe, in the presence of an abnormal venous drainage of the upper lobe, with an initial presumptive diagnosis where a low-grade sarcomatous lineage could not be rule out, for which a resection was performed by lobar resection with mediastinal emptying by video-thoracoscopy technique through one port, with curative intention.

Conclusion

Pulmonary leiomyoma is an entity with benign characteristics, very rare, whose symptoms may go unnoticed by long time and as an incidental finding. Its treatment is based on surgical resection, which can be conservative or not, depending on the affected site and the associated findings. The definitive diagnosis can only be obtained once the lesion is resected. After resection, they are associated with an excellent prognosis, as in the case presented, where until now there is no report of relapses.

Compliance with ethical standards

Informed consent: The written informed consent was obtained from the patient, appropriate for the publication of this case report and the accompanying images.

Conflict of Interest: No declared conflict of interest on the part of the authors.

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