Primary pulmonary leiomyosarcoma with invasion of the pulmonary vein—A case report

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INTRODUCTION: Primary pulmonary leiomyosarcomas are a subset of the rare sarcomatous lung neoplasms, found to be less than 0.5% of the organ’s primary malignant tumors (Attanoos et al., 1996). PRESENTATION OF CASE: We describe here a case of a 69-year-old woman who presented with heart palpitations, incidentally found to have a large lung mass abutting the left inferior pulmonary veins. Challenging tissue diagnosis led to invasive alternatives; attempted full neoplastic resection and pneumonectomy.

DISCUSSION: The mainstay of treatment for these tumors is complete surgical resection. Chemotherapy and radiation can be helpful adjuncts as well. CONCLUSION: This case presents a unique invasion pattern of a primary leiomyosarcoma of the lung, our diagnostic process, and surgical intervention.

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1. Introduction

Primary pulmonary leiomyosarcoma is a tumor rarely encountered by thoracic surgeons. We report a case of a left pneumonectomy performed for a large left lower lobe mass that was diagnostically challenging preoperatively, however postoperatively, final pathology demonstrated a high-grade leiomyosarcoma.

2. Presentation of case

A 69-year-old woman presented to thoracic surgery clinic for evaluation of a newly diagnosed lung mass discovered after presentation for heart palpitations. Initial imaging, including a chest CT and PET, revealed an isolated left hilar mass measuring 5.5 × 3.9 × 6.1 cm (Fig. 1A & B) which was intensely hypermetabolic without uptake elsewhere (Fig. 2). Tissue diagnosis was attempted via transbronchial biopsy, however, this was nondiagnostic, at which point she presented to our clinic for further recommendations.

While primary malignancy was the most obvious concern, we felt a second attempt at attaining conclusive tissue diagnosis was reasonable. A percutaneous CT guided needle biopsy of the mass was arranged as well as further staging with a brain MRI. These tests were to be followed by mediastinoscopy and possible lower lobectomy/pneumonectomy for the presumed primary lung cancer.

Percutaneous needle biopsy was attempted, however pathology showed no definitive diagnosis or evidence of malignancy. MRI of the brain revealed a solitary microhemorrhagic, enhancing intravascular lesion within the medial right cerebellar hemisphere measuring 7.5 × 10.4 mm, presumed to be metastasis. Plans for resection were aborted and she was taken to the operating room for repeat bronchoscopy with biopsies as well as mediastinoscopy. Bronchoscopy was again nondiagnostic and mediastinoscopy showed no evidence of mediastinal involvement.

Over the next few weeks the patient underwent radiation to the brain lesion, which left her with an undiagnosed, left lower lobe mass. She was scheduled to undergo chemotherapy for presumed non-small cell lung cancer, establishing a definitive diagnosis was still prioritized in her treatment plan. After discussions with the patient, her family, and her oncology team it was decided to pursue further biopsies and possible surgical resection for definitive diagnosis.

She was taken to the operating room and a one-centimeter incision was made in the midaxillary line at the eighth intercostal space, and through this, a thoracoscope was inserted and the left hemithorax was inspected. There was no evidence of pleural disease. Several core needle biopsies were taken and sent for frozen section in attempts to establish a definitive diagnosis before proceeding with resection. Pathology on the biopsy returned as inflammatory cells without evidence of malignancy.

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Fig. 1. (A) Transverse section of chest computed tomography in lung window showing a large left lower lobe mass. (B) Transverse section of chest computed tomography in a mediastinal window demonstrating the close involvement of the mass with the inferior pulmonary veins.

Fig. 2. Transverse section of a fused chest computed tomography PET scan demonstrating the intensely hypermetabolic central left lower lung mass.

Fig. 3. Spindle shaped tumor cells arranged in ill-defined fascicles and nodules. A central focus of necrosis is present (arrow).

A lateral thoracotomy was therefore performed and the mass further inspected. It extended into the hilum and surrounded the inferior pulmonary vein. Further biopsies were taken from the base of the mass and again sent for frozen section and a diagnosis of carcinoma was made.

The mass was resectable, but given its location and extensive hilar involvement a pneumonectomy rather than merely a lobectomy was warranted to ensure complete resection. Due to the involvement with the inferior pulmonary vein, this section was taken last. The pulmonary artery and bronchus were freed from surrounding structures and divided. Attention was then turned toward the veins. Due to the medial extent of the tumor the pericardium was opened. The veins were divided by placing a stapler proximal to the confluence of the superior and inferior veins on the left atrium and proximal to visible disease. Any more proximal and cardiopulmonary bypass with left atrial reconstruction would have been necessary. The pericardial defect was then closed using a bovine pericardial patch and a mediastinal lymphadenectomy was performed.

The patient had an uneventful postoperative course and was discharged home on postoperative day four.

Final pathology demonstrated a high-grade leiomyosarcoma measuring 7.0 × 6.5 × 5.2 cm (Fig. 3). By immunohistochemistry, the neoplastic cells showed moderate to strong cytoplasmic expression of calponin and smooth muscle actin (SMA), and focal, weak expression of muscle specific actin (MSA), Desmin and CD30. Overall, the immunoprofile and morphology were deemed most consistent with a poorly differentiated malignant mesenchymal neoplasm with smooth muscle differentiation, a high-grade leiomyosarcoma. Bronchial and pulmonary artery margins were negative for malignancy, however, tumor was found to microscopically invade the pulmonary venous wall, extending into atrial
endomyocardium (Fig. 4). Five of sixteen peribronchial lymph nodes were positive for malignancy. The final pathologic stage was determined to be T4 N1. The patient was referred to our oncology colleagues who recommended adjuvant radiation and chemotherapy.

3. Discussion

Primary pulmonary leiomyosarcoma is a tumor rarely encountered by thoracic surgeons. Sarcomas in the lung are typically due to metastatic disease, with primary lung sarcomas being exceedingly rare and reported to be less than 0.5% of all malignant pulmonary tumors [1]. The most frequent of these are leiomyosarcomas [2]. Nath et al. recently reviewed the literature and reported approximately 127 case reports of primary pulmonary leiomyosarcoma [3].

Leiomyosarcomas can arise within the pulmonary artery or pulmonary vein or from the smooth muscle within the lung parenchyma as in this case [4]. These tumors can be difficult to diagnose, however a diagnosis is crucial, even if major surgery is required to obtain it. They are much more common in adults, although they have been reported in children. They are also more common in males with a 2.5:1 male to female ratio [4]. Patients with primary pulmonary leiomyosarcoma can be asymptomatic or present with the same array of symptoms as any other lung tumor patient. Because there is no distinguishing clinical finding, histopathological examination is considered the gold standard for diagnosis.

The mainstay of treatment for these tumors is complete surgical resection. Chemotherapy and radiation can be helpful adjuncts as well [4]. In this case, based on final pathology, the patient was advised to undergo adjuvant chemotherapy and radiation therapy.

4. Conclusion

In conclusion, we report a case of a left pneumonectomy performed for a large left lower lobe lung mass involving the hilum. Final pathology demonstrated a primary pulmonary leiomyosarcoma with involvement into the inferior pulmonary veins.

Conflict of interest
None of the authors have any conflicts of interest.

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Consent
No identifying information or pictures were used.

Author contribution
Colwell—preparation of manuscript. Gasparri—preparation and editing of manuscript. Alghahim—editing of manuscript. Rao—pathology slides.

Guarantor
Elizabeth Colwell MD.

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