Leiomyosarcoma of the pulmonary artery diagnosed by endobronchial ultrasound-guided transbronchial needle aspiration

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ABSTRACT
Leiomyosarcoma of the pulmonary vasculature is an extremely rare condition that has not been previously diagnosed by endobronchial ultrasound (EBUS) bronchoscopy. We present the case of a 43-year-old white male with a history of leiomyosarcoma who was diagnosed with pulmonary embolism 2 years ago. As the filling defects on follow-up chest computed tomography continued to worsen despite anticoagulation, EBUS-guided transbronchial needle aspiration (EBUS-TBNA) of the right pulmonary artery lesion was safely and successfully performed. Cytopathological examination revealed the “thrombus” to be metastatic leiomyosarcoma. In experienced hands, and carefully selected cases, EBUS-TBNA seems to be a safe and effective in diagnosing thoracic endovascular lesions.

Key words: Biopsy, endobronchial ultrasound, leiomyosarcoma, pulmonary artery

INTRODUCTION
Metastatic tumors of the pulmonary vasculature are uncommon. Only a few cases of pulmonary vascular tumors diagnosed using endobronchial ultrasound-guided transbronchial needle aspiration (EBUS-TBNA) have been reported in the literature.¹⁻⁴ They frequently present as filling defects in the pulmonary vasculature on computed tomography (CT) making it challenging to distinguish between vascular tumor and thrombotic emboli.

Although fluorodeoxyglucose-positive emission tomography (PET) scan may reportedly help differentiate malignant growth from benign emboli,⁵²⁶ PET scans are not routinely performed for this purpose. Increased uptake on the PET scan increases the malignant suspicion of vascular tumors but in most cases, cytopathologic confirmation remains essential to confirm the diagnosis. EBUS-TBNA of mediastinal and hilar lymph nodes has proven value in diagnosis and staging for lung cancer.⁷ In addition, EBUS-TBNA may provide an opportunity to identify and sample endovascular abnormalities in the mediastinal and hilar area. We report a case of metastatic leiomyosarcoma diagnosed with EBUS-TBNA.

CASE REPORT
A 43-year-old white male was diagnosed with leiomyosarcoma of proximal left thigh involving femoral vessels 2 years back. He underwent preoperative radiation followed by surgical resection with clear margins. No postoperative chemotherapy was indicated. In the preoperative period, he was found to have pulmonary embolism involving right pulmonary artery...
for which he was started on anticoagulation and an inferior vena cava filter was placed prior to surgery. Despite being on anticoagulation postoperatively, serial surveillance imaging continued to show persistent “thrombus.” Eventually, an interval increase in size of the filling defect in right main pulmonary artery extending to the right lower lobe pulmonary artery was noted on contrast-enhanced chest CT [Figure 1]. At this point, intravascular metastatic tumor was suspected and the patient was referred to our interventional pulmonary service for diagnostic evaluation. The patient did not have any other imaging studies such as PET scan or magnetic resonance imaging of the chest. EBUS (BF-UC160F-OL3; Olympus, Tokyo, Japan) procedure was performed under general anesthesia. With the ultrasound and color flow Doppler, an endovascular hyper-echoic lesion was identified in right pulmonary artery [Figure 2]. Only one endovascular lesion was identified, which extended to the right lower lobe branch of the pulmonary artery. Because of the nature of the lesion’s extension in the pulmonary artery, it was not possible to accurately size it but it was estimated to be around 20 mm in the largest diameter. EBUS-TBNA of this lesion using the 22 Gauge EBUS needle (Olympus, NA-201SX-4022) was performed under general anesthesia with laryngeal mask airway using propofol intravenous drip and fentanyl intravenous intermittent injections. A core biopsy was obtained using the 22 Gauge EBUS needle and a total of five passes were performed during this procedure. Rapid-on-site evaluation of fixed slides was bloody, and no malignant cells were identified. A cellblock block was sent using the Roswell Park Memorial Institute solution. The procedure was performed without any complications and cytopathological and immunohistochemical evaluation confirmed the diagnosis of metastatic leiomyosarcoma [Figure 3]. The patient was started on gemcitabine plus docetaxel combination chemotherapy.

**DISCUSSION**

Historically, most cases of pulmonary artery tumors were diagnosed with surgery\(^9\,\text{–}\,\text{12}\) or during postmortem examination.\(^{13,14}\) Successful diagnosis of primary pulmonary artery leiomyosarcoma with catheterization and biopsy has also been reported.\(^{15}\) In the last few years, several cases of primary\(^{16,17}\) and metastatic\(^{1–6}\) pulmonary artery tumors diagnosed by EBUS-TBNA have been reported without any procedure-related complications. Although pulmonary artery hematoma has been described from EBUS-TBNA, the pulmonary

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**Figure 1.** (a) Chest computed tomography (CT) image showing right pulmonary artery filling defect (arrow) diagnosed as pulmonary thrombotic emboli 2 years ago. (b) Chest CT 2 years later showing enlarged right pulmonary artery filling extending to the right lower arterial branch consistent with vascular tumor (arrow).

**Figure 2.** (a) Endobronchial ultrasound (EBUS) image of the right hilar area showing a vascular tumor of the right lower lobe pulmonary artery (arrow). (b) Color Doppler showing the blood flow around the intravascular tumor. (c) EBUS image of pulmonary artery tumor (arrows). (d) EBUS-guided transbronchial needle aspiration (arrow) of the right pulmonary artery tumor. The color Doppler was used to ensure sampling the tumor area with the least blood flow.

**Figure 3.** (a) This is the tissue core biopsy of the right pulmonary mass lesion showing cells with abundant eosinophilic cytoplasm and spindled nuclei (H and E, ×200 original magnification). (b) These cells cytologically shows fine granular chromatin without prominent nucleoli (Papanicolaou stain, ×600 original magnification). (c) Immunohistochemical stains for smooth muscle actin shows strong immunoreactivity in these cells (SMA, ×400 original magnification). (d) Immunohistochemical stains for desmin also shows strong positivity in these malignant cells (Desmin, ×200 original magnification).
artery was inadvertently punctured in that case during a routine biopsy of mediastinal lymph node. In addition, there have been reports of using EBUS-guided trans-pulmonary vascular lymph-node sampling without any complications. Here, we present the first case of metastatic leiomyosarcoma of pulmonary artery diagnosed with EBUS-TBNA. The case reported is a clear example of the challenges in diagnosis of pulmonary artery tumors due to their endovascular location and the potential risk of bleeding. The use of color flow Doppler during EBUS-TBNA of pulmonary vascular tumor is essential to avoid area of higher blood flow and to perform the biopsy in the tumoral area of least blood flow. Given the potential higher risk of bleeding, anticoagulation or clopidogrel therapy should probably be discontinued prior to this procedure. Clinically, the importance of high index of suspicion is also demonstrated as the patient was initially diagnosed with pulmonary embolism. Misdiagnosing patients with pulmonary artery tumor emboli as thrombotic emboli can result in delaying effective tumor therapy with chemotherapy or surgery. The diagnosis of pulmonary embolism in patients with malignant diseases can be challenging especially in ill patients where the Wells score is already high. Our patient was asymptomatic, and the chest imaging was performed as part of surveillance and staging of his sarcoma. Intravascular metastasis was suspected as an alternate diagnosis after the presumed failure of anticoagulation therapy and increased of the pulmonary artery lesion size. EBUS-TBNA is a safe procedure and serious complications are extremely rare. Until now, EBUS-TBNA of the pulmonary vascular lesions did not report any serious complications.

**CONCLUSION**

The role of EBUS-TBNA in diagnosing mediastinal pathology continues to expand. EBUS has been successful in identifying many vascular conditions such as pulmonary vascular thromboembolism. EBUS-TBNA seems to be a safe, feasible, and minimally invasive tool for the diagnosis of vascular tumors such as metastatic pulmonary artery leiomyosarcoma.

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