Case Report: Spindle Cell Sarcoma of the Pulmonary Artery

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Abstract: Primary pulmonary artery sarcomas are rare. Moreover, they are notorious for masquerading as a chronic pulmonary embolus. There is much symptom overlap that leads to this relatively common misdiagnosis. We present such a case.

Keywords: Sarcoma, pulmonary artery, neoplasm, spindle cell.

CASE REPORT

A 45-year-old Caucasian female presented to her primary care physician with symptoms of worsening chronic cough and shortness of breath. Computed tomography angiography (CTA) of the chest was performed at an outside institution and findings were felt to be consistent with a large pulmonary embolism involving the left main pulmonary artery. She was subsequently admitted and started on anticoagulation therapy.

The patient's symptoms failed to improve, which prompted a visit to Vanderbilt University Medical Center. A ventilation-perfusion scan was ordered, which showed decreased ventilation to the entire left lung in addition to asymmetric decreased perfusion involving the same lung. An echocardiogram was then obtained and demonstrated an ill-defined mass in the main pulmonary artery with an appearance that was atypical for pulmonary arterial thrombus.

The initial CTA was then further reviewed, and the mass previously characterized as thrombus was found to have its own vascular supply. A PET-CT was obtained and demonstrated a hypermetabolic malignant tumor thrombus in the distal main and left pulmonary arteries with local hypermetabolic lymph nodes in the aortopulmonary window.

Endovascular biopsy was performed by the interventional radiology department. Pathology confirmed an intimal spindle cell sarcoma. The patient was admitted for surgical management, which ultimately resulted in left pneumonectomy.

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DISCUSSION

Sarcomatoid carcinoma of the lung is of the non-small cell variety and is rare with an estimated incidence of 0.3-1.3% of all lung cancers. Rarer still are primary sarcomatoid carcinomas involving the pulmonary artery with less than 250 cases described as of 2009 [1]. Five histological types are recognized: pleomorphic, spindle cell, giant cell, carcinosarcoma, and pulmonary blastoma. As a group, these carcinomas are poorly differentiated containing histological, cytological, and/or molecular properties of epithelial and mesenchymal cells. They are aggressive tumors with a worse prognosis and higher rate of recurrence than other non-small cell lung cancers. Median time to recurrence is roughly 11 months [3].

Spindle cell carcinoma represents approximately 0.1-0.3% of all lung tumors and 0.4% of all malignancies [2, 5]. This particular subtype affects men more than women with a ratio of approximately 4:1. Patients are typically in the 6^{th} to 8^{th} decade of life and have a smoking history [4]. Sarcomatoid carcinoma rarely involves the pulmonary parenchyma [2].

The literature reports several instances in which sarcoma of the pulmonary artery have been mistaken for chronic or recurrent pulmonary embolism. Blackmon *et al.* report that symptoms such as cough, dyspnea, hemoptysis, and chest pain are common. Clinical factors that can differentiate pulmonary embolism from sarcoma include fever, elevated erythrocyte sedimentation rate, anemia, weight loss, absence of a procoagulant state, and lack of history of deep venous thrombosis [1]. Differentiating factors on computed tomography between pulmonary embolism and sarcoma are as follows: hyperdense lesions with inhomogeneous attenuation because of hemorrhage, beaded peripheral pulmonary arteries, contiguously soft tissue-filled pulmonary artery occupying the entire

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lumen, vascular distention, distal oligemia, and extravascular spread [1], many of which were present in our patient.

Endovascular biopsy has been reported for the diagnosis of intravascular tumors and the technique is well established [6]. In this particular case, ultrasound was used to access the right common femoral vein and a sheath and guidewire were placed in the inferior vena cava (IVC). A six french pigtail catheter (Merit Medical, South Jordan, UT) was advanced to the main pulmonary artery using fluoroscopic guidance. After confirmation of the findings on CT, a seven French Raabe vascular sheath (Cook Medical, Bloomington, IN) was advanced to the edge of the tumor. Through this sheath, a seven French endomyocardial biopsy forcep was advanced (Cordis, Hialeah, FL). Multiple biopsies were performed. The biopsy fragments were placed into formalin for pathologic examination. The patient tolerated the procedure well with no procedural complications.

The mainstay of treatment is surgery, as chemotherapy alone has demonstrated suboptimal results [1, 3]. However, Mainwaring *et al.* demonstrated a case of complete remission after the administration of oral Germanium Sesquioxide [4], an area for further investigation. Surgical management options include pneumonectomy, lobectomy, endarterectomy, or tumor debulking with or without pulmonary artery reconstruction. The main determinants for management include tumor location and tumor extension. Although there was no difference in median overall survival in those patients who underwent endarterectomy and those who did not, surgery provided significant symptom improvement. Adjuvant chemotherapy and radiation therapy may confer a modest survival benefit. Further studies are needed to confirm this. Palliative chemotherapy and radiation therapy are reasonable management options in patients with inoperable or metastatic disease [7].

RADIOLOGIC FINDINGS

Imaging characteristics on CTA include a large filling defect involving the main and left pulmonary arteries (Figure **1**). Coronal and sagittal reformats demonstrate vascular supply to the filling defect (Figures **2** and **3**). Three-dimensional volume rendered reformats again show the mass involving the main and left pulmonary arteries with neovascularity (Figures **4** and **5**).

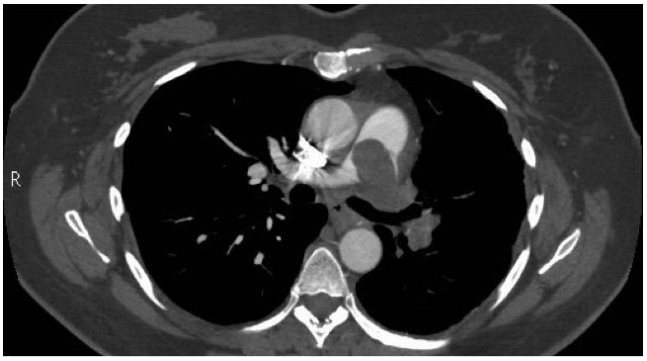

Figure 1: Contrast-enhanced CT of the chest demonstrating a large filling defect in the main pulmonary artery extending into the left pulmonary artery.

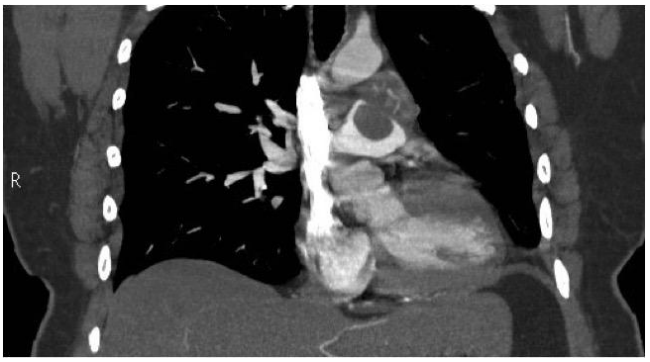

Figure 2: Coronally reformatted CTA of the chest demonstrating a large filling defect in the main and left pulmonary arteries with prominent vascularity.

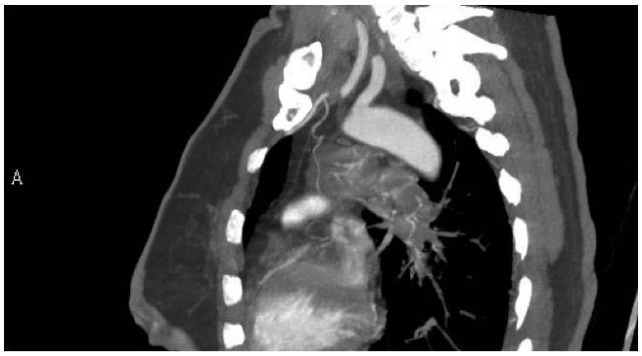

Figure 3: Sagitally reformatted CTA of the chest demonstrating a large filling defect involving the main and left pulmonary arteries with prominent vascularity.

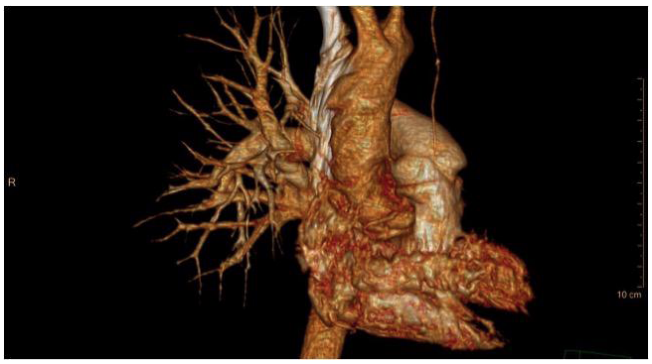

Figure 4: Three-dimensional reformatted CT of the chest demonstrating a large vascular mass involving the main and left pulmonary arteries.

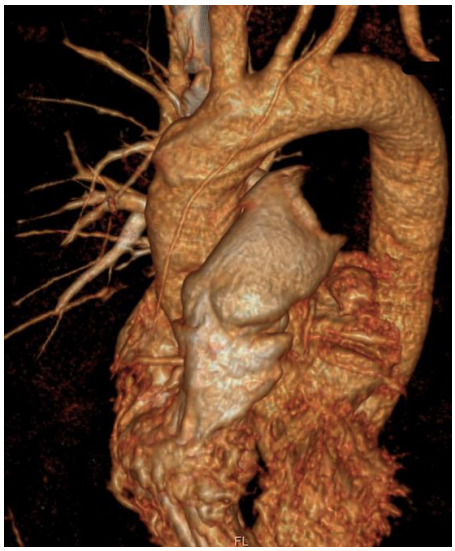

Figure 5: Three-dimensional reformatted CT of the chest demonstrating a large vascular mass involving the main and left pulmonary arteries.

PATHOLOGIC FINDINGS

Gross examination showed an 8.5 x 4.8 x 3.5 cm firm, white and tan mass adherent to the main and left pulmonary arteries that coursed distally through the interlobar and lobar arteries (Figure **6**).

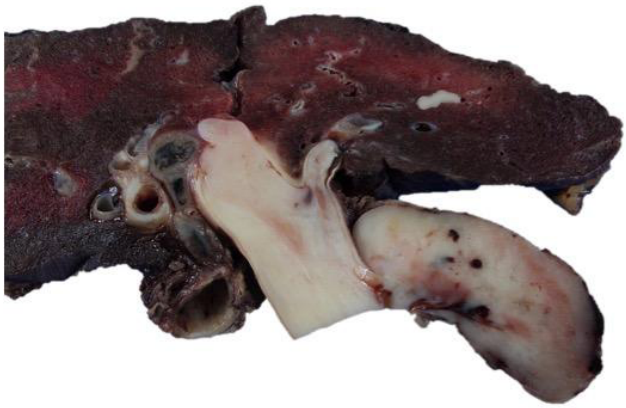

Figure 6: Tumor is present within the main pulmonary artery and extends into the lobar branches and smaller divisions (marked by *).

Microscopically, the tumor was present within and adherent to elastic arterial walls and exhibited characteristics of a sarcomatoid mass including hypercellularity, a whorling, fascicular pattern of spindle shaped cells, and rare mitoses (Figures **7-9**). Extensive immunohistochemical staining was largely negative except for a small fraction of cells that stained positive for Ki-67, a marker for cellular proliferation.

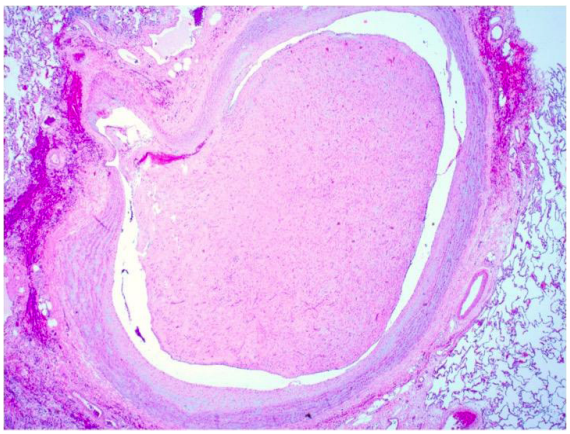

Figure 7: The tumor is located within and adherent to this branch of the pulmonary artery, while background lung appears normal (hematoxylin and eosin (H&E), 2x original magnification).

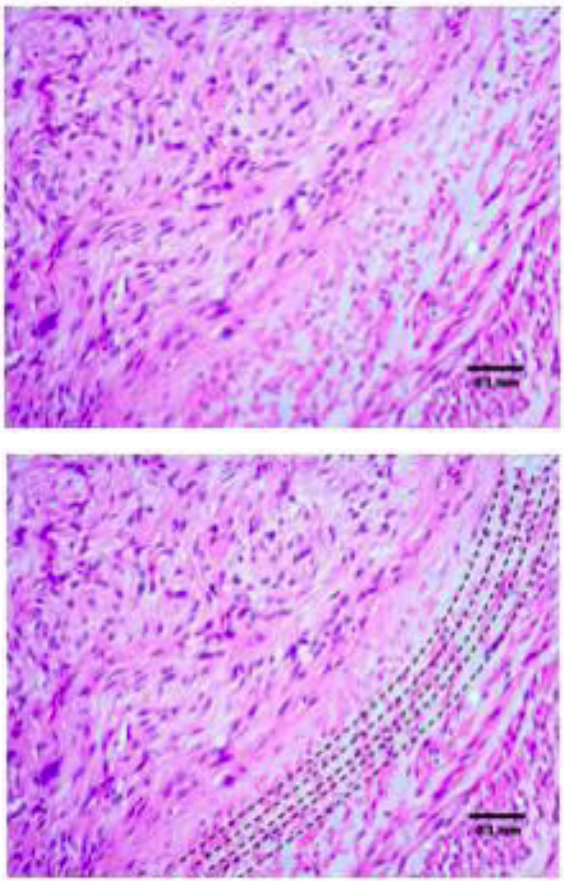

Figure 8: The tumor arises from the intimal lining overlying the elastic lamina (dotted lines on bottom) with cells arranged in waving fascicles (H&E, 20x original magnification).

CONCLUSION

The literature demonstrates a handful of examples of pulmonary artery sarcomas mistaken for chronic pulmonary embolus. This case report is intended to expound upon the existing literature regarding the clinical presentation of pulmonary artery sarcoma,

treatment of this rare, but clinically important entity, and highlight areas of further investigation.

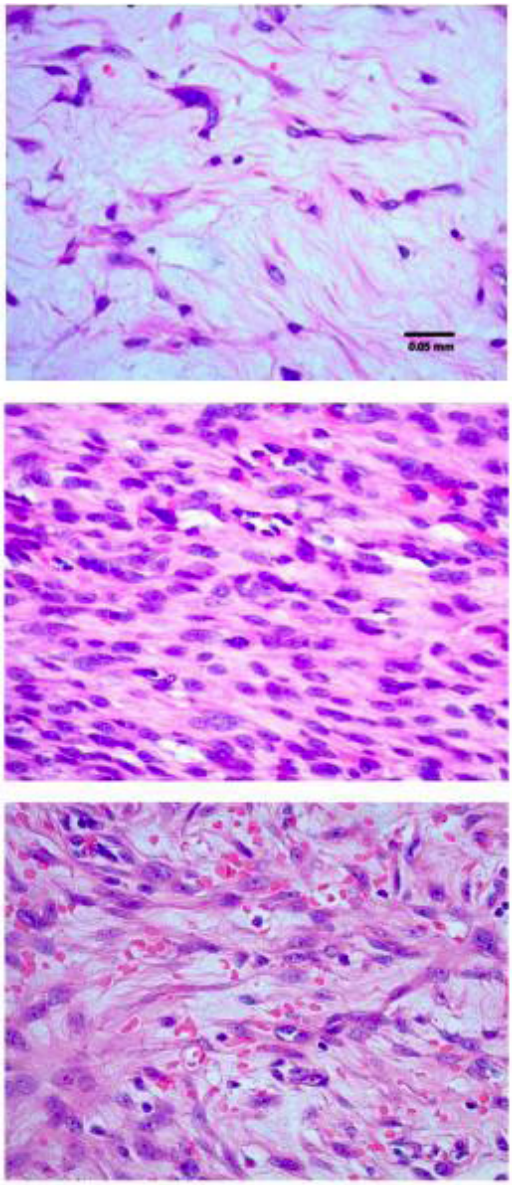

Figure 9: Histologic Patterns: Tumor cells are spindle to epithelioid in shape with oval nuclei and frequent prominent nucleoli. Throughout the tumor, multiple patterns are seen, including loose 'tissue culture' like (top), hypercelullar areas (middle) and areas of red blood cell extravasation (bottom), (H&E, 40x original magnification).

CONFLICTS OF INTEREST

None.

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