

Pulmonary artery sarcoma mimicking a pulmonary embolism

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Abstract

Sarcomas involving the lung are a rare occurrence, often a result of metastatic disease from primary malignancies involving the skin, liver, breast or heart. Primary pulmonary artery sarcomas are rarer still, with limited cases reported world-wide and consequently data regarding treatment modalities are sparse and largely experimental. These tumors are often mistaken for a pulmonary embolism and seemingly supported by radiological findings. Patients will often present without symptom resolution despite therapeutic anticoagulation. The following case illustrates how a soft tissue sarcoma of the pulmonary artery can mimic a pulmonary embolism, thus, resulting in both a diagnostic and therapeutic dilemma. A positron emission tomography scan was an invaluable tool in this case, showing increased radiotracer uptake and placing neoplasm at the top of the differential diagnosis. This ultimately led to a biopsy that was vimentin positive, cytokeratin negative and CD117 negative, thus consistent with soft tissue sarcoma.

Key words: Positron emission tomography, pulmonary embolism, sarcoma

Introduction

Soft tissue sarcomas involving the pulmonary artery are extremely rare and often go unrecognized. They may imitate the presentation of a pulmonary embolism, both clinically and radiologically, making the diagnosis difficult and potentially delaying treatment. The following case helps demonstrate how the utilization of a positron emission tomography (PET) scan allowed for our patient to be diagnosed and thus begin treatment for his underlying neoplastic process.

Case History

A 56-year-old gentleman presented to an outside Emergency Department with exertional shortness of breath. His past medical history included hepatitis C and hypothyroidism. A computed tomography (CT) scan revealed a saddle pulmonary embolism but no other abnormalities. He was anticoagulated with heparin and later discharged on warfarin. Thereafter, he remained well for approximately six weeks, at which time he again developed severe exertional dyspnea. A repeat CT scan

ordered by his primary physician, showed extension of the clot, accompanied this time by the appearance of lung nodules [Figure 1]. He was recommended discontinuation of warfarin and to be evaluated in hematology/oncology to rule out a neoplastic process.

When seen at our institution, his only complaint was ongoing exertional shortness of breath, with no associated hemoptysis, chest pain, cough, night sweats or loss of consciousness. His review of systems was otherwise non-contributory. On account of the suspicion of a possible neoplastic process, a PET scan was obtained, which revealed increased radiotracer uptake over the area of the presumed pulmonary thrombus, as well as other areas of the right lung, including a lower lobe sub pleural nodule [Figure 2].

During his outpatient evaluation he developed two syncopal episodes, with associated exertional dyspnea and presented to our Emergency Department. His electrocardiogram showed ST elevations in leads V3-V5 along with a small elevation in the troponin I of 0.31 (normal is less than 0.20), indicative of

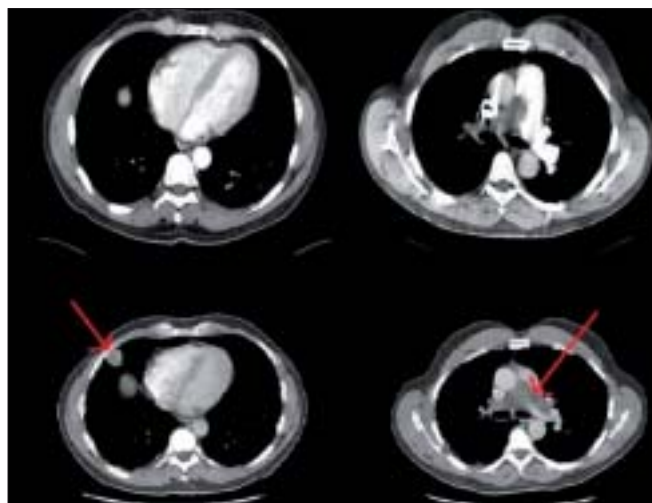


Figure 1: Top: Initial CT scan with the presumed pulmonary embolism. Bottom: CT scan, 1 month later, showing the appearance of a new lung nodule and extension of the previously suspected saddle pulmonary embolism

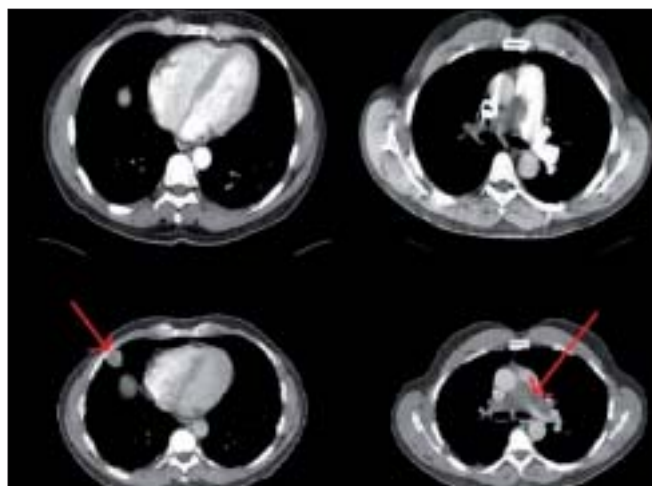


Figure 2: PET scan revealing hypermetabolism of the lung nodule and the previously presumed pulmonary embolism

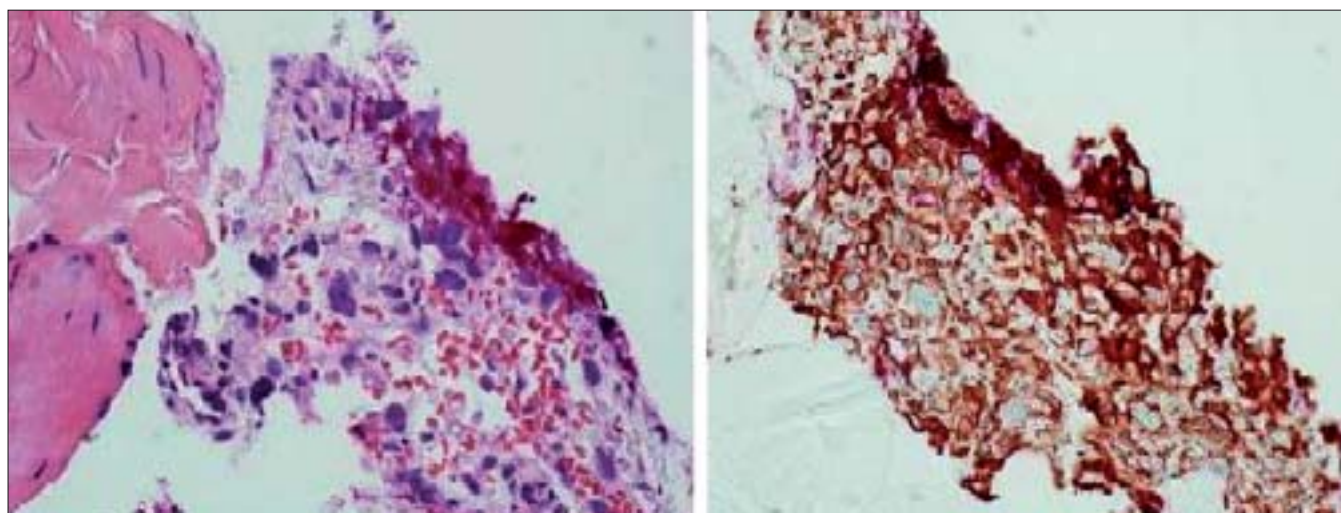


Figure 3: Left: Tissue fragment with malignant spindle cells displaying enlarged, angulated hyperchromatic nuclei. Right: Vimentin stain positive (Cytokeratin stain negative)

cardiac damage. He was transferred to the cardiac intensive care unit where he was empirically started on aspirin, metoprolol, lisinopril and atorvastatin. His echocardiogram revealed an ejection fraction of 55-65%, aortic root dilatation, right ventricle dilatation, mild pulmonic regurgitation, normal right atrium, systolic pulmonary artery pressure of 33 mmHg and an echogenic mass in the pulmonary artery, with mild tricuspid regurgitation. He was taken to the catheterization lab and was found to have 30-31% stenosis in the left main coronary artery, consistent with non-ischemic cardiomyopathy.

Multidisciplinary team approach was initiated; a cardiothoracic approach was deemed to be too dangerous and so was an embolectomy. After much consideration, the patient underwent a CT guided biopsy of the right lower lobe nodule revealing a malignant tumor, consistent with a soft tissue sarcoma. The sarcoma was classified as high grade; however the exact subtype could not be determined. On immunohistochemistry, the tissue was vimentin positive, cytokeratin negative and CD117 negative, being thus consistent with a soft tissue sarcoma [Figure 3]. Based on the site of initial involvement, a primary pulmonary artery sarcoma was suspected.

Based on the anatomical position and extent of his disease, surgery was not considered a realistic option and he was admitted for systemic chemotherapy using Mesna, Adriamycin, Ifosfamide and Dacarbazine (MAID).^[1] He seemed to be responding after four cycles of therapy, as demonstrated on repeat imaging studies. However, following additional two cycles, he was found to have progressed. Following this, he was treated with single agent Gemcitabine, but continued to show evidence of progression. At this time, twelve

months following his initial diagnosis, he enrolled in a clinical trial using ET 743.

Discussion

Sarcomas involving the lung are a rare occurrence, often a result of metastatic disease from primary malignancies involving the skin, liver, breast or heart. Primary pulmonary artery sarcomas are rarer still, with limited cases reported world wide and consequently data regarding treatment modalities are very sparse and largely experimental. They were initially described in 1923 by Mandelstamm. These tumors run an indolent course and often do not come to attention until there are symptoms of right outflow obstruction, including hemoptysis, dyspnea, cough, chest pain, swelling of the lower extremities and loss of consciousness. They are often mistaken for a pulmonary embolism and seemingly supported by radiological findings.^[2-7] These findings include a ventilation perfusion mismatch in the area of the pulmonary artery obstruction. CT scanning can also yield an imagery disguise and thus what appears to be a mass in the pulmonary artery is often mistaken for a pulmonary embolism. Thus, patients will often present without symptom resolution despite therapeutic anticoagulation. A repeat CT scan often shows an enlarging mass, with accompanying hilar masses. A PET scan can be an invaluable tool in this setting, showing increased radiotracer uptake in the mass.^[7-10]

Tumor embolization from the pulmonary artery is a commonly recognized phenomenon, presenting as distal metastases or worse still, as compromised blood flow to distal organs and structures. The prognosis is poor, with patients dying within months to a year of presentation. Surgery has been the accepted treatment modality of choice, although emerging data show that these tumors are radiosensitive. Interleukin 2 and Paclitaxel have also been noted to be beneficial in tumor regression and survival.^[7,11] Most of the data and evidence are largely experimental and due to case scarcity these studies are

confined to isolated cases, without reproducible large scale trials.^[12-14]

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