A Great Red Herring: A Rare Case of Primary Pulmonary Artery Sarcoma

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INTRODUCTION

Primary pulmonary artery sarcoma (PPAS) is a very rare and aggressive malignant tumor which often mimics acute pulmonary embolism (PE) or chronic thromboembolic pulmonary hypertension (CTEPH), leading to delayed diagnosis and increased mortality.[1,2]

CASE

- A 45-year-old male with seizure disorder presented with nonproductive cough and progressive dyspnea on exertion.
- His workup revealed an elevated D-dimer, and a CT chest angiography (CTA) showed a large intraluminal defect in the main pulmonary artery (PA) extending to the R main concerning for PE (Figure 1). He was started on rivaroxaban.
- One month later, he still had persistent dyspnea associated with unintentional 20-pound weight loss.
- His repeat CTA showed extension of intraluminal defect to the L main concerning for progression of PE despite being on anticoagulation (Figure 2).
- His echocardiogram showed septal flattening and elevated RVSP >60 mmHg.
- He was transitioned to therapeutic enoxaparin and was referred to pulmonary hypertension center due to concern for CTEPH.
- His hypercoagulability studies were negative.
- His ventilation-perfusion (V/Q) scan showed absent perfusion of the entire R lung.
- Due to rapid progression of the thrombus despite on anticoagulation, there was a high suspicion for malignancy. His PET/CT revealed an intense uptake within the R main extending to the L main PA concerning for sarcoma (Figure 3).

IMAGES

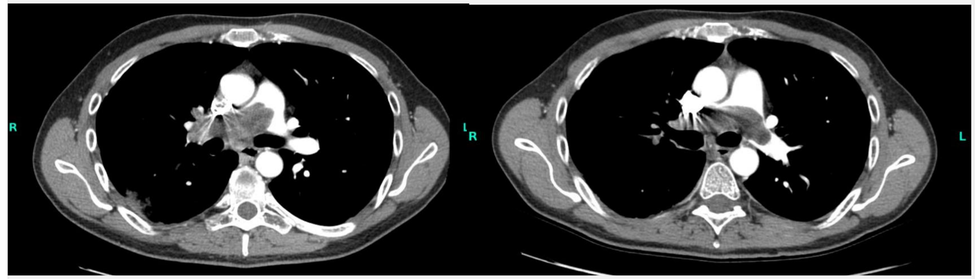

Figure 1. Initial presentation CTA showing intraluminal defect in the main PA extending to the right PA

Figure 2. After a month on therapeutic anticoagulation, CTA showed progression of intraluminal defect extending to the left PA

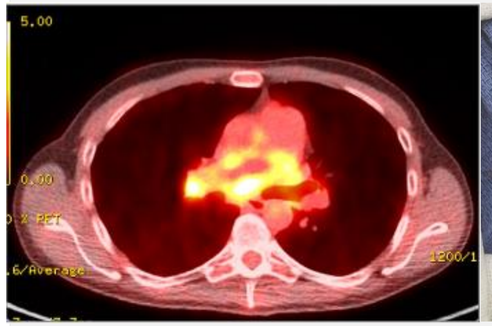

Figure 3. PET/CT showed an intense uptake within the right main PA extending to the left main concerning for sarcoma

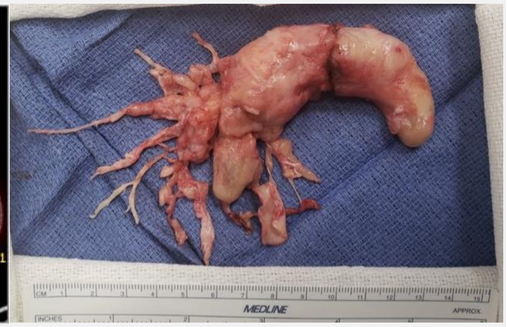

Figure 4. Tumor debulked from right pulmonary artery showing white-tan soft tissue measuring 5.2 x 5.0 x 3.9 cm in aggregate

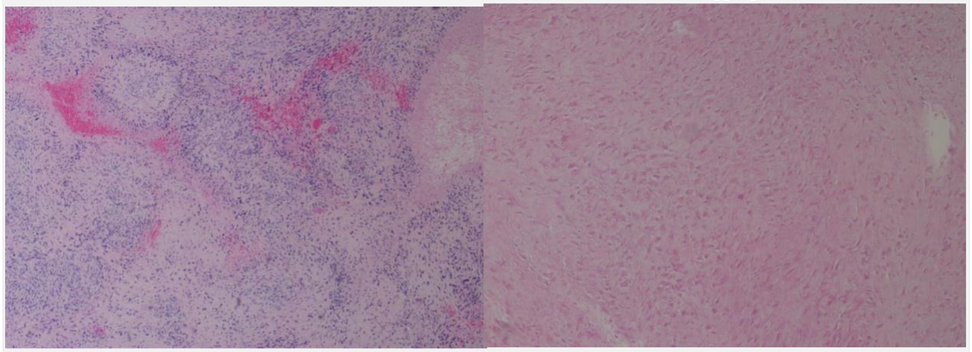

Figure 5. Histopathology of primary pulmonary artery sarcoma (intimal sarcoma) showing spindle-shaped tumor cells

CASE CONTINUED

- RHC with biopsy of the PA was non-diagnostic due to low sensitivity.
- Due to high suspicion for malignancy, he underwent pulmonary thromboendarterectomy (Figure 4).
- Histopathology revealed spindle-shaped tumor cells (Figure 5) with positive fluorescence in situ hybridization for MDM2 gene, confirming PPAS.

DISCUSSION

- **PPAS** is a rare aggressive malignancy with a median survival of 1.5 months if untreated and mimics PE or CTEPH at presentation, leading to frequent misdiagnosis.[1,2]
- A high index of suspicion for atypical presentation includes unprovoked subacute progression of PE despite adequate anticoagulation in the absence of hypercoagulable state.[3]
- Symptoms favoring PPAS include digital clubbing, weight loss, and asthenia, in addition to dyspnea, chest pain, coughing, and hemoptysis.[1,4]
- D-dimer, BNP, ESR, and CRP are also typically elevated.[1]
- Echocardiography and CTA identify intraluminal filling defect in the pulmonary outflow tract and the PA vasculature, respectively.[1,4] Magnetic resonance imaging is useful in tissue characterization, and PET also shows high metabolic activity in PPAS compared to PE.[3,5]
- Definitive tissue diagnosis is necessary.[6]
- Multimodal treatment involves neoadjuvant chemotherapy with surgical resection.[6]
- Successful surgery increases survival to 8-36 months.[7]

REFERENCES