Pulmonary Artery Sarcoma: Not Every Filling Defect is a Pulmonary Thromboembolism

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A 76-year-old man with a past medical history of hypertension only presented to the hospital with dry nonproductive cough, pleuritic chest pain, and dyspnea. Computed tomographic (CT) angiography scan revealed a 'filling defect' in the left main pulmonary artery (Fig. 1*A*). Unfractionated heparin was started and bridged to Coumadin. No deep venous thrombus was revealed on the upper and lower extremities duplex venous scans. The patient was discharged home with the diagnosis of pulmonary thromboembolism (PTE). After discharge, the patient's symptoms kept worsening with additional anorexia and weight loss despite the ongoing anticoagulation treatment. Due to the worsening dyspnea and a weight loss of 20 pounds over the next 12 months, a follow-up CT-chest angiogram was performed which revealed a large 16 × 10 × 8 cm mass in the left main pulmonary artery which seemed to have originated from the filling defect seen on the initial CT-scan (Figs. 1*B–D*). An ultrasound-guided biopsy of the mass was consistent with the diagnosis of pulmonary artery sarcoma (PAS) (Fig. 2). Due to the extensive disease, only the palliative chemotherapy was offered, which the patient declined.

Described first by Mandelstamm[1] in 1923, PAS is a rare and often lethal tumor which is usually diagnosed only during surgery or autopsy. They are thought to arise from the mesenchymal cells of the intima of pulmonary artery

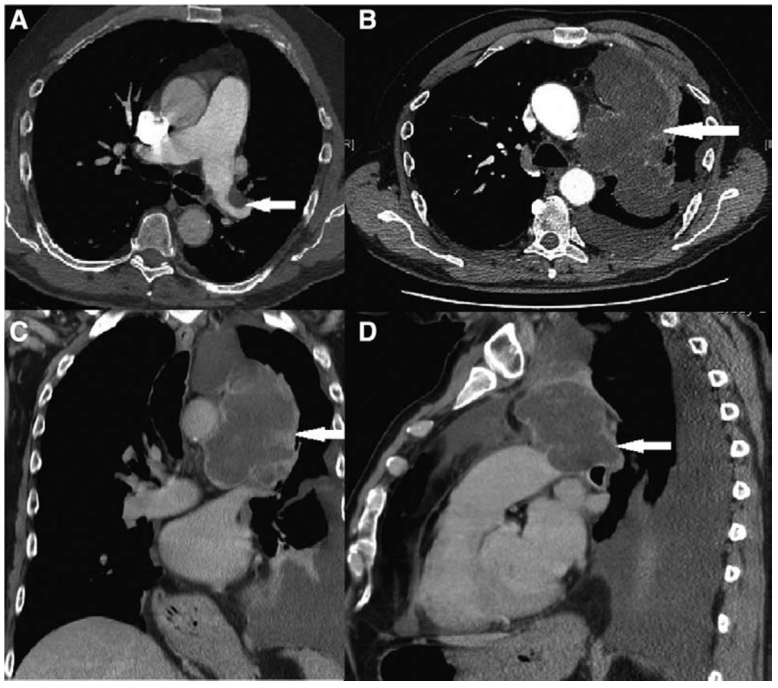

FIGURE 1. *A*, Computed tomographic scan (CT)-chest (with contrast) showing a filling defect in the left main pulmonary artery (arrow), (*B*), CT scan-chest (with contrast) a year later showing large left mediastinal mass (arrow), (*C*), CT scan-chest (with contrast), coronal reconstruction showing large mediastinal tumor (arrow), (*D*), CT scan chest (with contrast), sagittal reconstruction showing the same tumor originating from the pulmonary artery (arrow).

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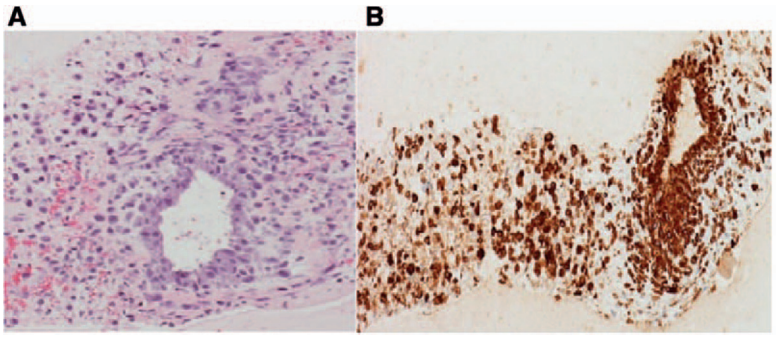

FIGURE 2. *A,* Histopathological examination (hematoxylin-eosin stain) demonstrates spindle cells with pleomorphic nuclei in a myxoid and collagenized background. *B,* Immunohistochemical analysis showing diffuse positivity of tumor cells to vimentin stain, a marker of tumors of mesenchymal origin including sarcomas.

trunk or the bulbous cordis.[2] PAS is often misdiagnosed as PTE, as both of the diseases initially appear as intraluminal 'filling defects' in the pulmonary artery system on contrast enhanced CT scans. The differentiation between these two disease entities is very important to avoid misdiagnosis of a potentially fatal malignancy and unnecessary anticoagulation therapy. An absence of predisposing factors for PTE and the progressive worsening dyspnea in the patients with an intraluminal "filling defect" despite anticoagulation therapy are factors suggestive of PAS. Recently, it has been reported that, positron emission tomographic scan may help to make a differential diagnosis between PAS and PTE.[3] Surgical resection in conjunction with neoadjuvant chemotherapy remains the primary treatment, if detected early in the course of the disease.[4]

Clinicians should maintain high clinical suspicion of PAS in patients with a filling defect in the pulmonary artery without any underlying risk factors for PTE and worsening of the symptoms despite anticoagulation therapy.

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