



# Pulmonary Artery Intimal Sarcoma Treated as Chronic Pulmonary Thromboembolism

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## CASE

A 72-year-old woman was hospitalized for suspected chronic pulmonary thromboembolism (CPTE) with complaints of tachycardia and tachypnea; her blood pressure was stable and she had 90% oxygen saturation in room air. She reported a history of dyspnea, which started 3 years ago, along with weight loss of approximately 25 kg. Throughout this period, she was treated with oral anticoagulants, but without improvement. The axial computed tomography pulmonary angiogram (CTPA) reconstruction (Figure 1a) and coronal maximum intensity projection CTPA reconstruction (Figure 1b) scans demonstrated an enlarged pulmonary trunk with filling defects partially occupying the lumen of the right and left pulmonary arteries. The filling defects showed heterogeneous density and coarse calcifications; additionally, the right pulmonary artery filling defect had an extraluminal extension to the mediastinum. Due to the degree of consumptive involvement and the worsening of the respiratory indexes, the patient died eventually.

An autopsy was conducted, which showed that her heart was enlarged, mainly on the right side. Notably, there was also a tumor mass mimicking a thrombus, but gelatinous, adhered to the intimal layer of the pulmonary artery, causing total obstruction of the vascular lumen (Figure 1c). The histological sections showed a spindle-cell neoplasm with moderate cellularity. This mesenchymal neoplasm was characterized by the proliferation of fusiform cells, with elongated, atypical, hypertrophic, pleomorphic, and hyperchromatic nuclei, along with coarse chromatin, and some mitotic figures. The mitotic activity was found to be 10-12 mitoses/10 high-power fields (Figure 1d). Focally, atypical cells with cartilaginous differentiation, inconspicuous cytoplasm, and necrotic areas were also observed. The neoplastic cells were negative for AE1/AE3, S100, desmin, and CD34, and showed positivity for SM-actin and focally for MDM2; the Ki-67 level was approximately 40% (see Supplementary Material). The final diagnosis was made as pulmonary artery intimal sarcoma (PAIS).

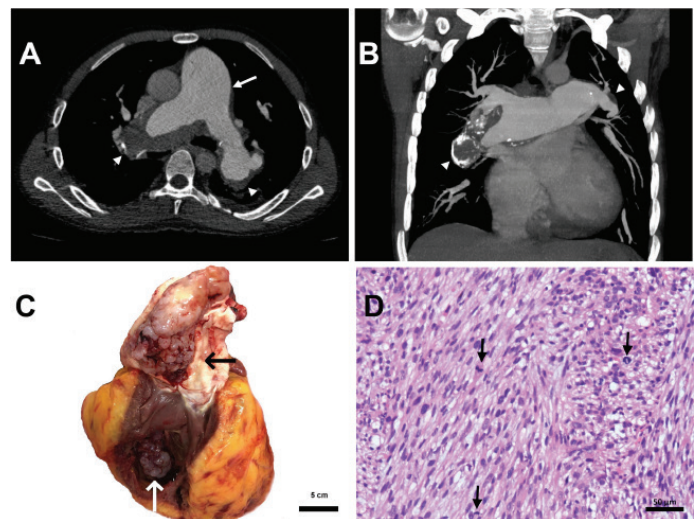


FIG. 1. (a) Axial computed tomography pulmonary angiogram (CTPA) reconstruction discovered an enlarged pulmonary trunk and filling defects partially occupying the lumen of the right and left pulmonary arteries. (b) Coronal maximum intensity projection CTPA reconstruction revealed an enlarged pulmonary trunk and filling defects partially occupying the lumen of the right and left pulmonary arteries (arrowheads). These filling defects had heterogeneous density and coarse calcifications; the right pulmonary artery filling defect showed extraluminal extension to the mediastinum. (c) On autopsy, gelatinous intraluminal masses attached to the pulmonary arterial wall (black arrow) and extending into the heart (white arrow) were seen. (d) Histopathological features revealed a spindle-cell mesenchymal neoplasm with mild atypia, moderate cellularity, cartilaginous differentiation, and hypertrophic, pleomorphic, hyperchromatic nuclei (black arrow). (Hematoxylin and eosin staining)

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PAIS is a rare neoplasm with approximately 250 cases reported so far the literature.<sup>1</sup> Since the disease is often misdiagnosed as CPTE, the actual incidence of PAIS is still unknown.<sup>2</sup> The gold standard method for a noninvasive diagnosis is CTPA, the most frequent findings of, which are vascular distension due to tumor growth, continuity with periarterial tissues, arterial lumen filling, extravascular involvement, nonhomogeneous hyperdense hemorrhagic lesions, and involvement of peripheral pulmonary arteries.<sup>3</sup> Features including expansion of affected vessels, extraluminal extension, and contrast enhancement allow differentiation between PAIS and CPTE. However, owing to low clinical suspicion, the diagnosis is often made by postmortem examination.<sup>4</sup>

**Informed Consent:** Written informed consent was obtained from a proxy for the patient described in the manuscript.

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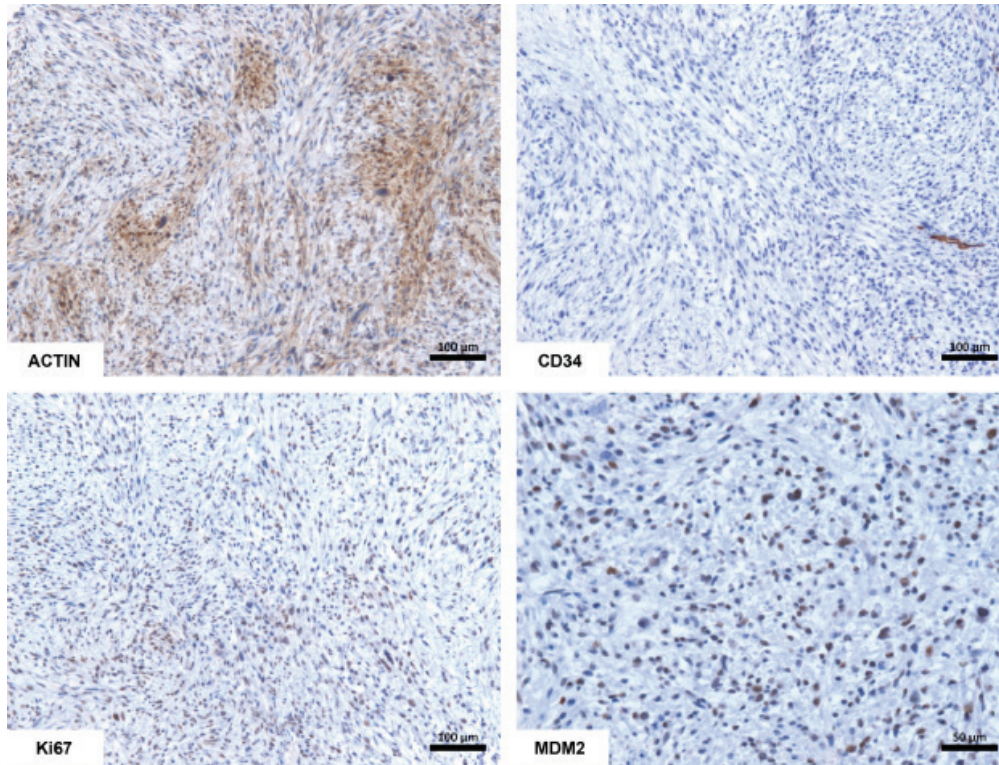
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**Supplementary Figure 1:** <http://xxxxxxxxxxx.pdf>

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Supplementary Figure 1. Immunohistochemical panel: neoplastic cells were negative for AE1/AE3, S100, desmin, and CD34, and showed positivity for SM-actin and focally for MDM2; Ki-67 level was approximately 40%.