A 33-year-old man noted increasing dyspnea over a period of 6 weeks. Initial evaluation with transthoracic echocardiography demonstrated severe right ventricular dilation and systolic contractile dysfunction along with moderately severe tricuspid valve regurgitation on color Doppler with an estimated right ventricular systolic pressure of 108 mm Hg based on continuous wave Doppler assessment of the tricuspid regurgitant jet (Figure 1 and online-only Data Supplement Movies I and II). Subsequent evaluation for the cause of pulmonary hypertension included contrast-enhanced computed tomography of the chest that demonstrated a large homogeneous filling defect within the main pulmonary artery extending into the proximal right and left branches (Figure 2). Bilateral lower-extremity venous duplex scan was negative for acute deep venous thrombosis. Intravenous thrombolytics were subsequently administered without change in size of the pulmonary artery filling defect on follow-up chest computed tomography performed the next day. Cardiac magnetic resonance imaging demonstrated a mildly expansile, heterogeneous, partially enhancing mass (Figure 3 and online-only Data Supplement Movie III). The patient underwent surgical resection of a large, fleshy tumor within the main pulmonary artery. The tumor was attached to the main pulmonary artery and had invaded through the wall. The pulmonary valve was replaced with a homograft. Histological examination revealed a high-grade intimal sarcoma with osteosarcomatous differentiation (Figure 4). The patient was discharged from the hospital 1 week after surgery with plans to follow-up with oncology as an outpatient.

Disclosures

None.

Figure 1. A, Two-dimensional transesophageal echocardiography reveals an echogenic mass present in the right ventricular outflow tract and proximal main pulmonary artery. B, Continuous-wave Doppler echocardiography depicts severely elevated right ventricular systolic pressure.
Figure 2. Computed tomographic chest post contrast reveals a large mildly expansile mass in the main pulmonary artery that extends into the right and left pulmonary arteries. Mag indicates magnification.

Figure 3. Magnetic resonance imaging of the chest. A, Steady-state free precession sagittal image reveals a large intermediate-signal mass (arrow) at the right ventricular outflow tract that invades the pulmonic valve. B, T2-weighted spin-echo image demonstrates homogeneous increased signal of the mass. C, T1-weighted spin-echo precontrast image depicts heterogeneous enhancement of the mass. D, Postgadolinium phase-sensitive inversion recovery image showing large mass with absence of delayed hyperenhancement.

Figure 4. A, Low-magnification microphotograph of the pulmonary artery mass shows invasion through the pulmonary artery wall (arrowheads) and destruction of the pulmonary valve (arrows; Movat pentachrome, ×15). B, Histologically, the tumor is composed of malignant spindle cells arranged in fascicles (hematoxylin-eosin, ×400). C, The tumor cells produce lace-like osteoid in focal areas (hematoxylin-eosin, ×400).
Pulmonary Artery Intimal Sarcoma Masquerading as Pulmonary Embolism
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