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IMAGING VIGNETTE

CLINICAL VIGNETTE

Pulmonary Artery Intimal Sarcoma Mimicking Right Ventricular Outflow Tract Thrombus

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ABSTRACT

A 72-year-old man presented with breathlessness and a systolic murmur. Extensive diagnostic work-up revealed a rare pulmonary artery intimal sarcoma mimicking a right ventricular outflow tract thrombus and impacting a cardiac pacemaker lead. Surgical resection, pathology confirmation, and management strategies are discussed, highlighting the challenges of treating this rare malignancy. (J Am Coll Cardiol Case Rep 2023;28:102101) © 2023 The Authors. Published by Elsevier on behalf of the American College of Cardiology Foundation. This is an open access article under the CC BY-NC-ND license (http://creativecommons.org/licenses/by-nc-nd/4.0/).

A 72-year-old man presented with progressive shortness of breath, diaphoresis, headache, and a newly detected systolic murmur. Symptoms had been worsening over time. On examination, vital signs were stable, but the presence of a systolic murmur raised concerns about the patient's cardiac health. The medical history was significant for essential hypertension, prostate cancer, obstructive sleep apnea, and a permanent pacemaker implanted for complete heart block. The patient was a current smoker, reported occasional alcohol consumption, and denied any illicit drug use. Differential diagnosis was right ventricular outflow tract (RVOT) thrombus, primary or secondary cardiac malignancy, and cardiac vegetation.

A transthoracic echocardiogram showed an elevated velocity (3.3 m/s) and gradient (45 mm Hg) across the pulmonic valve with restricted valve opening caused by a large, irregular, echo-dense mass in the main pulmonary artery. A subsequent transesophageal echocardiogram confirmed moderate pulmonary valve stenosis and revealed a 2.9 × 2.6 cm echo-dense mass attached to the pulmonic valve leaflet in the pulmonary artery and invading the tissue planes, causing partial RVOT obstruction. Computed tomography revealed a large thrombus at the pulmonic valve and narrowing of the proximal main pulmonary artery (**Figure 1A**). Cardiac magnetic resonance imaging showed a 3.5-cm perfusion-enhanced mass in the RVOT with characteristics suggestive of an invasive process, possibly angiosarcoma or metastatic disease (**Figures 1B to 1E**).

The patient was started on broad-spectrum antibiotics because of the possibility of endocarditis. The electrophysiology service was consulted about pacemaker lead extraction, deeming it unnecessary because the pacemaker was functioning normally and blood cultures remained negative. Coronary angiography revealed no significant coronary artery disease. In the operating room, a large mass invading the septum and pulmonary

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ABBREVIATIONS AND ACRONYMS

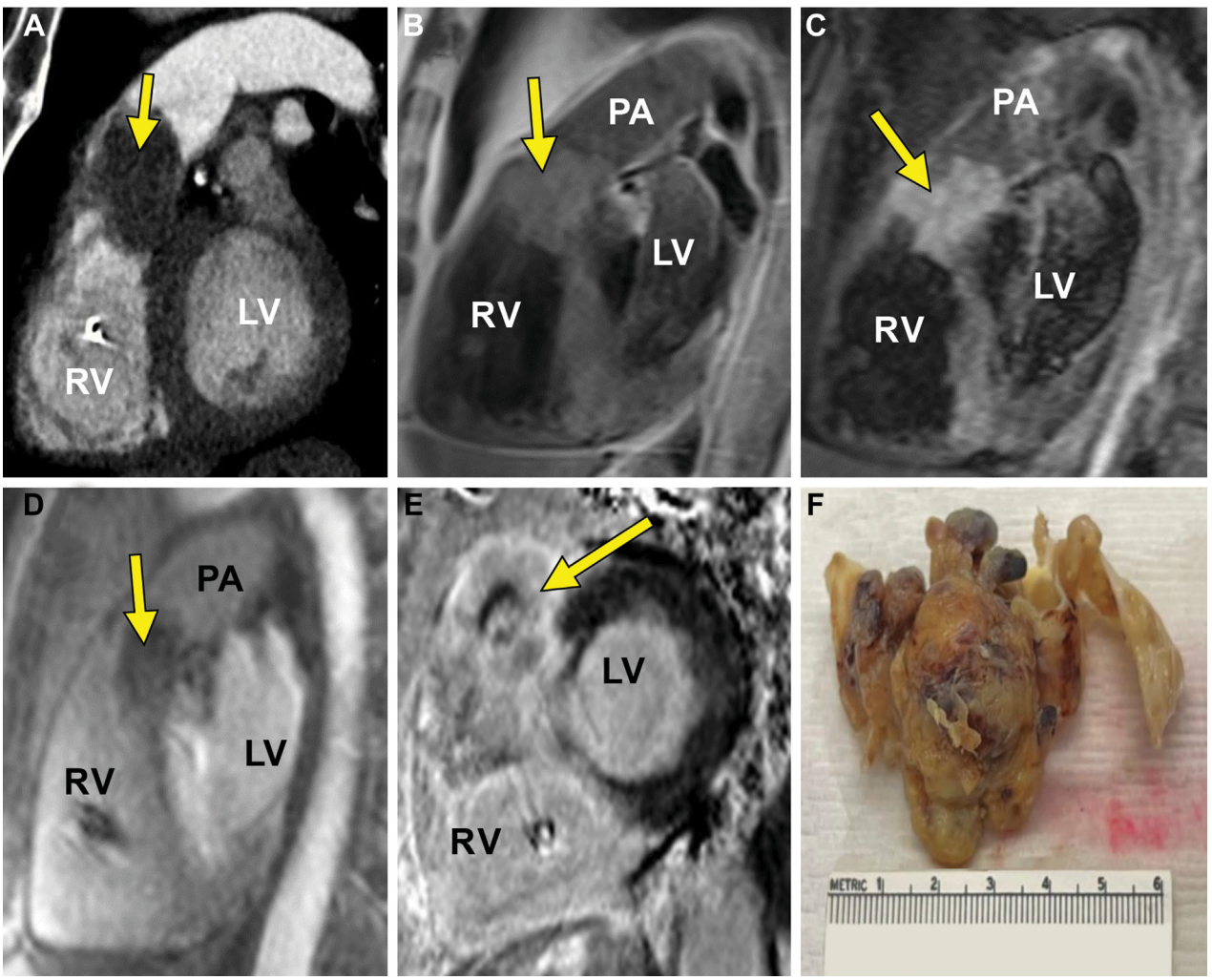
RVOT = right ventricular outflow tract

valve was resected, and a bioprosthetic aortic valve was placed in the pulmonary position (Figure 1F). A small tear in the innominate vein was repaired, but no mass was found attached to the pacemaker leads. Postoperative pathology confirmed the presence of a malignant epithelioid and spindle cell neoplasm, favoring pulmonary artery intimal sarcoma.

Postoperative follow-up revealed pleural effusions, pulmonary nodules, and an adrenal gland nodule. Because of the risk of cardiotoxicity, doxorubicin-based chemotherapy was not pursued. Gene sequencing identified an actionable allelic variant, and the patient was started on pembrolizumab. One month after discharge, the patient experienced a cardiac arrest and did not survive.

Pulmonary artery intimal sarcoma is a rare, highly aggressive malignancy. Early diagnosis can be challenging, but multimodality imaging is crucial. Surgical resection is the primary treatment; adjuvant chemotherapy and radiation therapy may extend survival in some cases. Gene sequencing can guide selection of chemotherapeutic agents.[1-3]

FIGURE 1 Imaging



(A) Initial computed tomography pulmonary embolism protocol shows a large filling defect in the right ventricular (RV) outflow tract (arrow). Cardiac magnetic resonance imaging shows (B) an isointense RV outflow tract lesion (arrow) on T_1-weighted sequence and (C) a hyperintense lesion (arrow) on T_2-weighted sequence with fat saturation. (D) First pass perfusion demonstrates gadolinium uptake suggestive of vascularization (arrow). (E) Partial late gadolinium enhancement (arrow) is noted. (F) Gross specimen of the surgically resected mass. LV = left ventricle; PA = pulmonary artery.

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KEY WORDS mass, pacemaker lead, right ventricular outflow tract