JACC: CASE REPORTS VOL. 2, NO. 12, 2020

MINI-FOCUS ISSUE: CLINICAL CARDIOLOGY

INTERMEDIATE

IMAGING VIGNETTE: CLINICAL VIGNETTE

What Looks Like a Clot But Is Not a Clot?

Cardiac Leiomyosarcoma Mimicking Pulmonary Embolism

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ABSTRACT

Primary cardiac tumors in the right ventricular outflow tract are often misdiagnosed as pulmonary embolism due to rarity and inadequate imaging characterization. Multimodality imaging offers advantages and facilitates subsequent diagnostics and management. We present a case of a woman with suspected submassive pulmonary embolism who was found to have pleomorphic leiomyosarcoma. (**Level of Difficulty: Intermediate.**) (J Am Coll Cardiol Case Rep 2020;2:1966–8) © 2020 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 woman in her 70s with a history of 2 primary lung cancers and nonhuman immunodeficiency virus-related cutaneous Kaposi's sarcoma presented with increasing dyspnea and was transferred for advanced management of a large filling defect in the main pulmonary artery. Vital signs at the time of transfer were notable for a heart rate of 82 beats/min, blood pressure 118/84 mm Hg, and oxygen saturation at rest on room air was 98%. Examination was notable for regular rhythm with a soft systolic ejection murmur at the cardiac base, jugular venous pulsation was noted at the ear while the patient was seated at 90 degrees, lungs were clear bilaterally, and bilateral pitting edema was present to the mid-shins.

She had 2 primary lung cancers for which she underwent a resection without adjuvant therapy for an adenocarcinoma in the 1980s and a right lower lobectomy for adenocarcinoma with adjuvant chemotherapy in 2015. She has never smoked and was considered to be in remission before admission. She also had a history of Kaposi's sarcoma of her right posterior lower leg which had been excised.

A repeat chest tomography (CT) showed a lobulated lesion in the right ventricular outflow tract (RVOT) (**Figure 1A**) extending into the proximal main pulmonary artery (PA) with right ventricular (RV) and right atrial enlargement. Positron-emission tomography performed 2 months prior for cancer surveillance showed a fludeoxyglucose-avid soft tissue lesion in the main PA corresponding to the mass seen on the CT scan (**Figure 1B**). A transthoracic echocardiogram showed a severely dilated RV with moderate-to-severe systolic dysfunction and RV free wall hypokinesis with sparing of the RV apex. There was also evidence of mild-to-moderate supravalvular pulmonic stenosis. Cardiac magnetic resonance imaging showed a 30 × 25-mm lobulated mass with late gadolinium enhancement in the RVOT and PA (**Figure 1C**, Video 1) consistent with a tumor. Given possible intercurrent thrombus, she received systemic parenteral anticoagulation. She underwent surgical resection of RV and PA mass with reconstruction of RVOT and proximal main PA along with a

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The authors attest they are in compliance with human studies committees and animal welfare regulations of the authors' institutions and Food and Drug Administration guidelines, including patient consent where appropriate. For more information, visit the *JACC: Case Reports* author instructions page.

Manuscript received June 22, 2020; accepted August 21, 2020.

ISSN 2666-0849 https://doi.org/10.1016/j.jaccas.2020.08.018

pulmonic valve replacement. Pathology of the mass showed high-grade pleomorphic leiomyosarcoma involving the endocardium (Figures 1D and 1E).

This patient presented with acute cardiopulmonary symptoms and was found to have a primary cardiac sarcoma. Acute thrombus was initially suspected; however, gross pathology ultimately showed no thrombus present (Figure 1F). Primary cardiac malignancies are rare with an incidence rate of ~0.02% based on autopsy studies (1). Conventional CT imaging is often not sensitive enough to distinguish tumor from thrombus, therefore multimodal imaging is helpful to better characterize RVOT lesions. Cardiac magnetic resonance imaging is particularly useful in distinguishing tumor from thrombus and malignant tumors from benign (2,3). Positron-emission tomography can be useful in further delineating intracardiac masses as thrombi tend to not be fludeoxyglucose avid. This patient did well in the post-operative period and was discharged to rehabilitation. This case highlights the utility of multimodal imaging in characterizing intracardiac masses and in guiding therapeutic and further diagnostic strategies.

ABBREVIATIONS AND ACRONYMS

CT = computed tomography

PA = pulmonary artery

RVOT = right ventricular outflow tract

RV = right ventricle

AUTHOR RELATIONSHIP WITH INDUSTRY

Dr. Vaduganathan has received the KL2/Catalyst Medical Research Investigator Training award from Harvard Catalyst (NIH/NCATS Award UL 1TR002541); serves on advisory boards for Amgen, AstraZeneca, Bayer AG, Baxter Healthcare, Boehringer Ingelheim, Cytokinetics, and Relypsa; and participates on clinical endpoint committees for studies sponsored by Novartis and the NIH. All other authors have reported that they have no relationships relevant to the contents of this paper to disclose.

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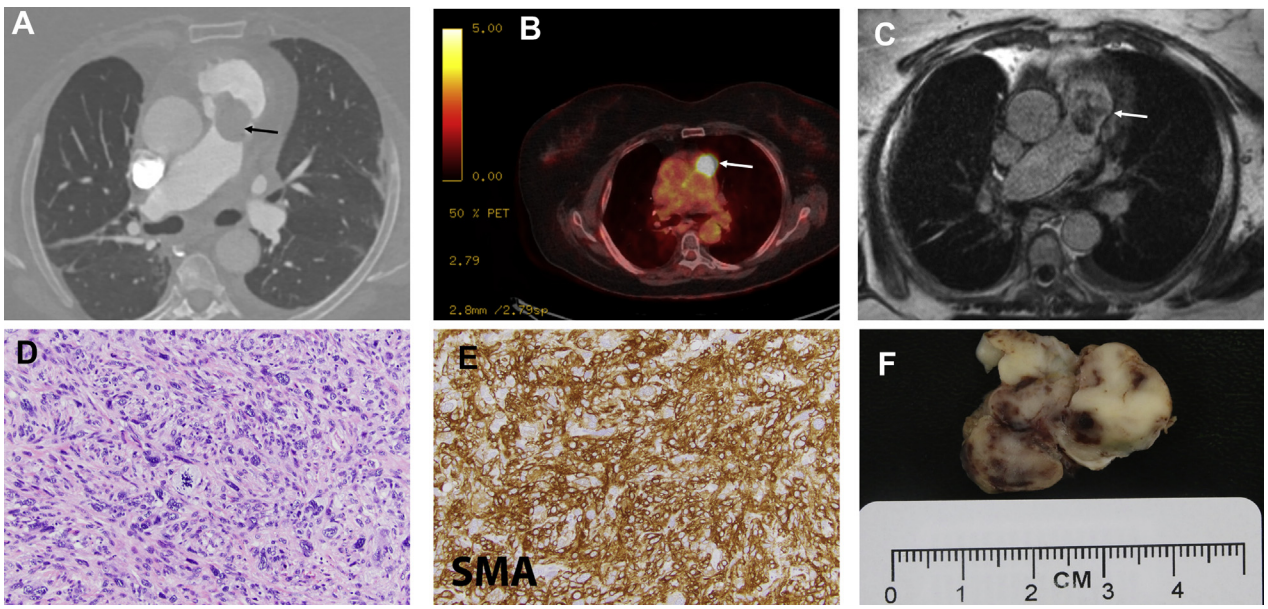
FIGURE 1 Multimodal Imaging and Pathology of Right Ventricular Outflow Tract Mass



(A) Computed tomography pulmonary angiography showing lobulated lesion in the right ventricular outflow tract crossing the pulmonic valve and extending into the proximal main pulmonary artery **(black arrow). (B)** Positron-emission tomography (PET)–computed tomography showing fludeoxyglucose-avid mass along the main pulmonary artery **(white arrow). (C)** Late gadolinium enhancement imaging showing heterogeneous uptake of gadolinium in the mass consistent with tumor. **(D)** This leiomyosarcoma diffusely infiltrates the myocardium and is composed of fascicles of spindle cells with eosinophilic cytoplasm and elongated, blunt-ended nuclei (hematoxylin and eosin stain). **(E)** The tumor cells show diffuse expression of smooth muscle actin (SMA) and desmin multifocally (not shown). Tumor cells were negative for vascular endothelial markers CD31 and ERG (not shown). **(F)** Gross pathology of excised leiomyosarcoma showing no associated thrombus but with necrosis and hemorrhage within the tumor.

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KEY WORDS cardiac magnetic resonance, echocardiography, right ventricle

APPENDIX For a supplemental video, please see the online version of this paper.