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Case Reports

Incidentally Discovered Right Atrial Mass: A Rare and Unexpected Etiology

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Abstract

Primary cardiac sarcoma is a rare type of intracardiac mass. This report describes a patient with atrial flutter who had a new right atrial mass incidentally discovered on transesophageal echocardiography. A thrombus was suspected based on radiographic appearance, but there was minimal change with anticoagulation. The mass was resected and found to be an undifferentiated pleomorphic cardiac sarcoma, an uncommon subtype within the already rare category of primary cardiac neoplasms. This report highlights the importance of considering primary malignancy and thoroughly correlating radiographic and clinical evidence during the diagnostic workup of patients with intracardiac masses.

Keywords: Intracardiac imaging techniques; cardiac tumor; echocardiography, transesophageal; magnetic resonance imaging; sarcoma

Introduction

Identification of a new intracardiac mass should prompt an astute clinician to consider a wide differential diagnosis that includes thrombus, bacterial or nonbacterial endocarditis, neoplasia, an implanted device, or an anatomic variation.[1] Primary cardiac tumors are very rare but essential to consider, given that the natural history of some tumors can be highly aggressive with a poor prognosis.[2] The patient described in this report had a newly identified right atrial (RA) mass, as seen on echocardiogram, that was initially thought to be a thrombus based; however, careful monitoring and eventual excision revealed the diagnosis of a rare and surprising neoplastic etiology.

Case Report

A 71-year-old White man presented with 4 weeks of progressive dyspnea. Electrocardiography revealed atrial fibrillation with rapid ventricular response. His medical history included melanoma with 2 excisions (7 years and 1 year before presentation), prostate adenocarcinoma (pending initiation of proton beam therapy), and 44 pack-years of tobacco use. He was afebrile, with a pulse rate of 155/min, and physical examination was notable for bilateral pulmonary rales and an irregular tachycardia. Initial laboratory studies revealed leukocytosis (14.1×10^9 cells/L), normal serum high-sensitivity troponin T, and an N-terminal pro b-type natriuretic peptide level of 2,955 pg/mL (reference range, ≤103 pg/mL). His chest x-ray showed pulmonary edema. He was treated with intravenous diuretic agents, diltiazem for rate control, and intravenous heparin.

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A transthoracic echocardiogram (TTE) revealed marked left ventricular systolic dysfunction, a left ventricular ejection fraction of 25% and severe generalized left ventricular hypokinesis consistent with tachycardia-induced cardiomyopathy. Transesophageal echocardiography (TEE) performed before electrical cardioversion showed an irregular, echogenic, 3.2 × 3-cm mass firmly attached to the lateral RA wall, with a wide base above the tricuspid annulus (Fig. 1). This mass was deemed suspicious for thrombus, malignancy, myxoma, or nonbacterial thrombotic endocarditis. Given that the patient was already sedated for the TEE and possible cardioversion, a thorough discussion was held with multiple specialists and with his wife to weigh the risks of pulmonary embolization after cardioversion against the benefits of restoring sinus rhythm and preventing cardiogenic shock in the setting of worsening symptomatic dysrhythmia and markedly depressed ejection fraction. The patient underwent cardioversion without experiencing any complications and returned to normal sinus rhythm.

Cardiac magnetic resonance (CMR) imaging was performed to assess the suspicious attachment of the mass to the atrial wall and redemonstrated the RA mass which was concerning for thrombus given the lack of dynamic early or late gadolinium enhancement and persistently low signal intensity on a long inversion recovery sequence (Fig. 2). The patient was discharged on apixaban anticoagulation for maximal simplicity and comfort. One month later, the patient underwent coronary angiography to investigate the etiology of his heart failure. This showed diffuse nonocclusive coronary artery disease, confirming the diagnosis of tachycardia-induced cardiomyopathy. On repeat TTE, the mass appeared unchanged; apixaban was changed to enoxaparin because of concern for anticoagulation failure in the setting of his known prostate cancer and the more reliable anticoagulant efficacy profile of enoxaparin in malignancy.[3]

Four months after his initial presentation, TTE and TEE (Fig. 3) demonstrated markedly improved systolic function (ejection fraction, 55%), but the mass appeared unchanged. Because he had been taking anticoagulation therapy for 4 months, a neoplastic etiology (RA myxoma in particular) appeared to be more likely than a throm-

Abbreviations and Acronyms

CMR	cardiac magnetic resonance
HMB45	human melanoma black
MART-1	melanoma antigen recognized by T cells 1
PET-CT	positron emission tomography-computed tomography
RA	right atrial
SOX10	SRY-box transcription factor 10
TEE	transesophageal echocardiogram
TTE	transthoracic echocardiogram

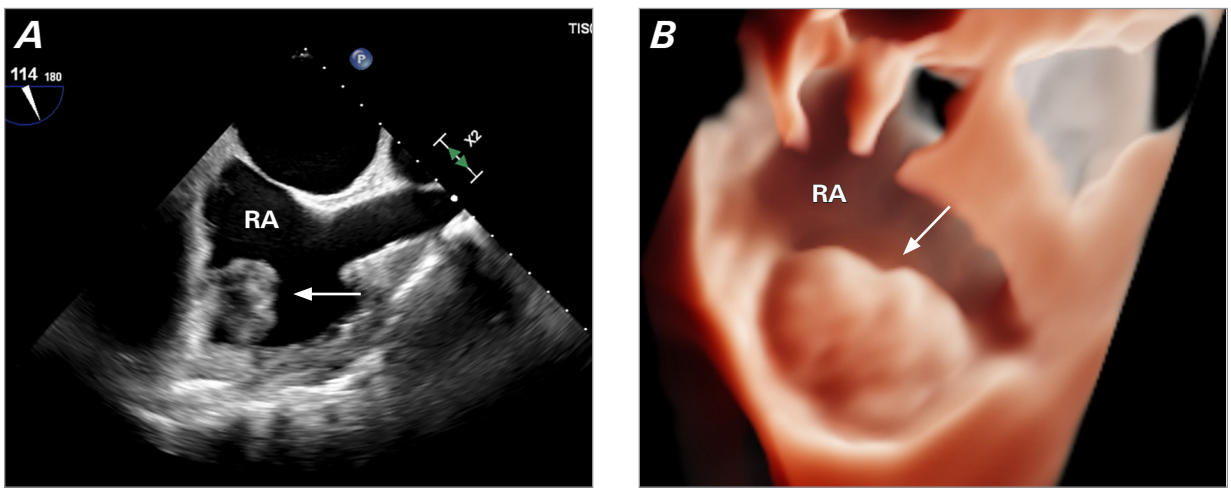

Fig. 1 A) Two-dimensional and **B**) 3-dimensional transesophageal echocardiogram performed before cardioversion at initial presentation reveals an irregular 3.2 × 3-cm echogenic mass (arrow) attached to the lateral right atrial wall just above the tricuspid annulus.

Supplemental motion images are available for Figure 1A and Figure 1B.

RA, right atrium.

bus at this point. After consultation with cardiovascular surgery, a plan was made to perform median sternotomy, right atriotomy, mass excision, a modified Maze procedure, and left atrial appendage ligation because of continuing paroxysmal atrial fibrillation and flutter.

During surgery under cardiopulmonary bypass, oblique right atriotomy revealed a large RA mass. The mass was attached to the RA wall, just above the tricuspid valve and atrioventricular groove. There was no true stalk; rather, the mass originated from a large-based area on the atrium wall. The atriotomy was extended toward the mass, and a 3 × 3-cm area of the RA wall was resected to remove the mass and "stalk" en bloc (Fig. 4). All margins were grossly negative visually. The resection abutted the atrioventricular groove but did not disrupt the right coronary artery. An ablation probe was used

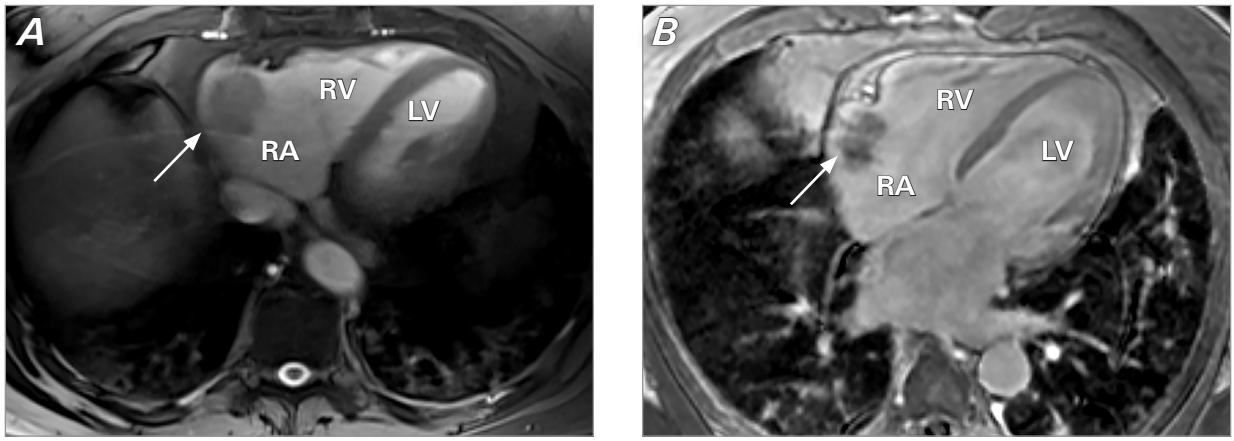

Fig. 2 A) After discovery of the right atrial mass, cardiac magnetic resonance imaging with steady-state free precession protocol demonstrated the T1 hypointense lobulated mass measuring 4.1 × 2.7 cm (arrow) in the lower right atrium, attached to the anterior wall just above the tricuspid annulus and in the region of the Chiari network. **B**) The T2 slightly hyperintense lesion (arrow) was mobile and did not display late gadolinium enhancement and maintains low signal intensity on long inversion recovery sequences.

LGE TI, late gadolinium enhancement inversion time; LV, left ventricle; RA, right atrium; RV, right ventricle.

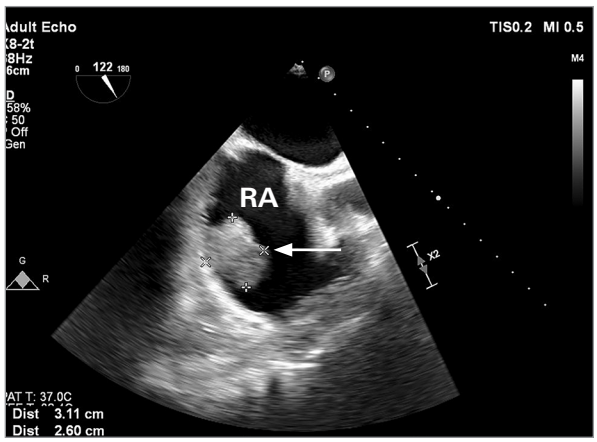

Fig. 3 Transesophageal echocardiography after 4 months of anticoagulation shows the RA mass (arrow) with minimal change in size.

Supplemental motion image is available for Figure 3.

RA, right atrium.

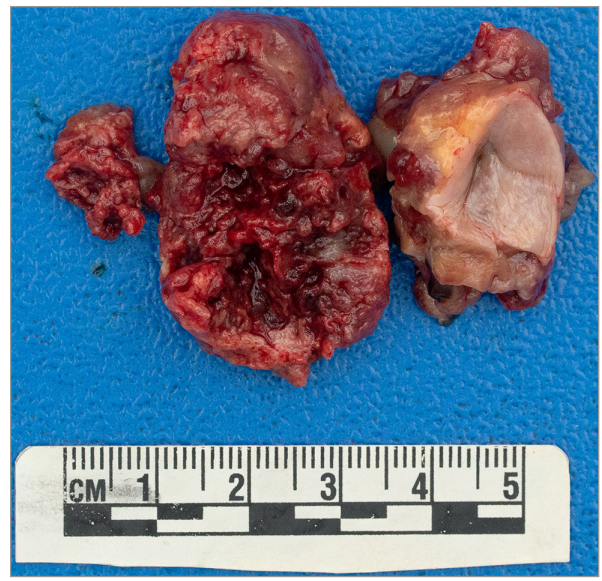

Fig. 4 Right atrial mass after resection.

to create ablation lines up to the RA appendage, up to the superior vena cava, down to the inferior vena cava, and down to the tricuspid annulus. A bovine pericardial patch was used to reconstruct the right atrium. With the heart still arrested, the heart was lifted and the left atrial appendage identified and ligated. The right and left pulmonary veins were isolated using 4 burn lines in each location. This completed the patient's modified Maze procedure, and the surgery concluded without complications.

On pathologic examination, the tumor was an irregular, lobulated, malignant, spindled, and epithelioid neoplasm with marked nuclear pleomorphism and inconspicuous mitotic activity (Fig. 5). Large areas of

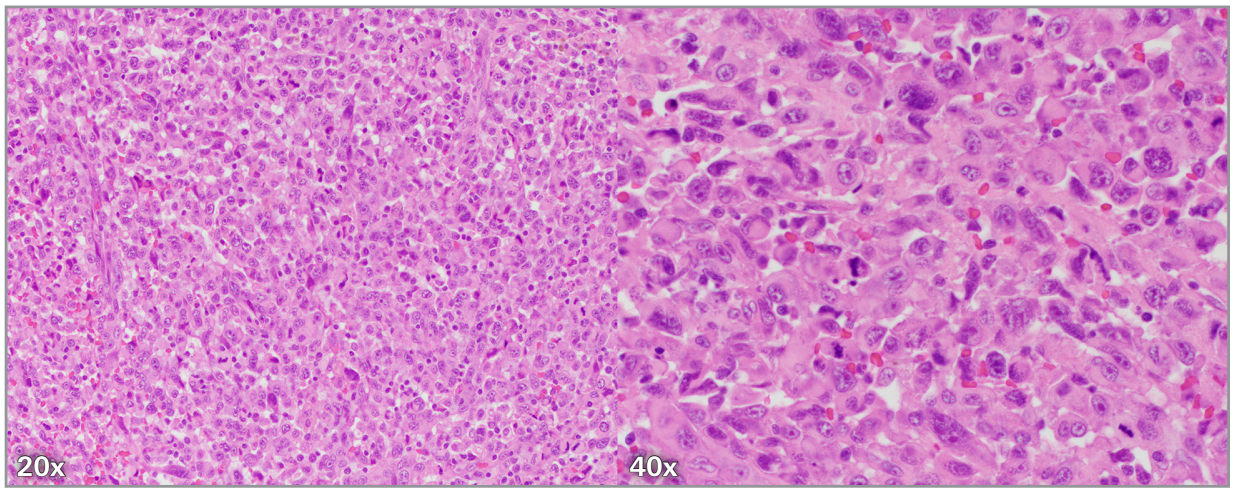

Fig. 5 Hematoxylin and eosin staining of the tumor; magnification, ×20 (left) and ×40 (right).

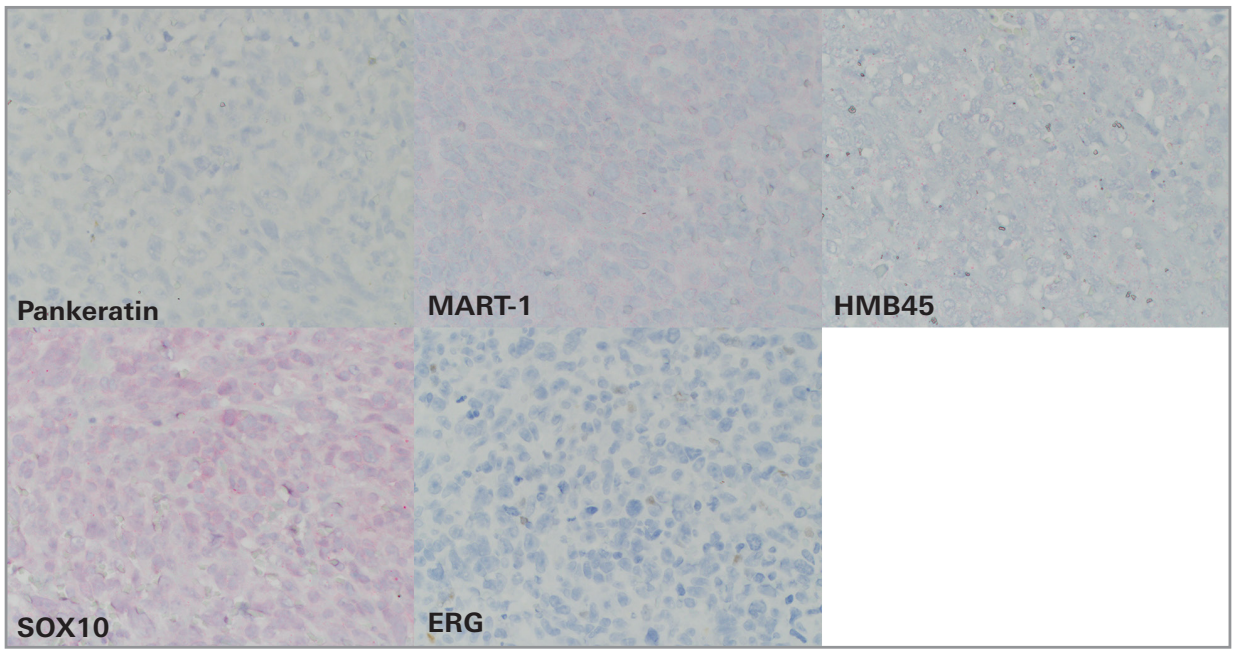

Fig. 6 Immunohistochemical staining of the tumor is negative for pankeratin, MART-1, HMB45, SOX10, and ERG; magnification, ×40.

HMB45, human melanoma black; MART-1, melanoma antigen recognized by T cells 1; SOX10, SRY-box transcription factor 10.

tumor cell necrosis were present. Immunohistochemical staining was negative for pankeratin, melanoma antigen recognized by T cells 1 (MART-1), human melanoma black (HMB45), SRY-box transcription factor 10 (SOX10), and ERG (a transcription factor) (Fig. 6). These immunohistochemistry results excluded the diagnoses of metastatic melanoma, metastatic carcinoma, and angiosarcoma. In addition, *MDM2* fluorescence in situ hybridization was negative for amplification, essentially excluding intimal sarcoma. The final pathologic diagnosis was undifferentiated pleomorphic primary cardiac sarcoma.

Positron emission tomography-computed tomography (PET-CT) and brain magnetic resonance imaging did not reveal any evidence of metastasis. The patient underwent adjuvant chemotherapy with 4 cycles of gemcitabine and docetaxel over 4 months. Follow-up whole-body PET-CT and CMR imaging shortly after completion of chemotherapy showed no evidence of recurrence. Future follow-up plans include imaging every 3 to 4 months for 1 year.

Discussion

Primary cardiac tumors are exceptionally rare, with an incidence of only 0.02% to 0.06%.[4,5] Among primary benign neoplasms, papillary fibroelastomas and myxomas are the most common.[6] Papillary fibroelastomas mostly occur on valvular surfaces, with left-sided valves being more common. Myxomas are smooth masses that can occur in any cardiac chamber but are most frequently found in the interatrial septum. Although it may be challenging to differentiate myxomas from thrombi on echocardiography, CMR imaging is quite helpful given the significant T2 hyperintensity seen in myxomas because of their high water content.

Primary malignant tumors are even more uncommon. Sarcoma is the most common primary cardiac malignancy, with angiosarcoma being the most common subtype (21%-40%). Undifferentiated pleomorphic sarcoma accounts for only 3% to 7% of primary cardiac sarcomas.[7] In most patients with malignant masses such as cardiac sarcoma, the echocardiographic features are easier to recognize than they were in the patient featured in this report, given their invasive nature (which leads to frequent crossing of tissue planes) and their association with pericardial effusions. Cardiac sarcomas are associated with high mortality and an overall poor prognosis given the local impairment of blood flow and the propensity to recurrence and metastasis.

This report reinforces that a comprehensive differential diagnosis is essential for patients with an intracardiac mass. The clinician must consider thrombus; bacterial or nonbacterial endocarditis; metastatic, primary, or benign intracardiac tumors; implanted devices; or anatomic variations. The differential can be narrowed based on the mass location, as different cardiac masses are more likely to manifest in specific chambers. For example, angiosarcomas are more likely to arise in the right atrium and undifferentiated sarcomas in the left atrium (although the tumor in this report did not follow this rule).[8] Metastatic melanoma is also an important consideration for the patient in this report, given his history of recurrent melanoma, although it was ruled out on immunohistochemical examination. Because the patient had a mild leukocytosis that resolved without intervention, and because he lacked other infectious stigmata, there was minimal concern for infection. He was given anticoagulation from the time of presentation for his arrythmia, which simultaneously served as empiric treatment for a potential thrombus. After detecting the mass, CMR imaging was quickly pursued; this modality identifies thrombus not solely by morphology, as in echocardiography, but by tissue characteristics such as gadolinium-based contrast uptake. Thus, CMR imaging is highly specific (99%) for thrombus.[9] Because of the lack of contrast enhancement and persistent low signal on a long inversion recovery sequence, CMR imaging was most suggestive of thrombus, and this patient received continued anticoagulation and observation.

Notably, the final pathologic diagnosis in this patient differed from the imaging characteristics of his mass, emphasizing the need for careful follow-up. An intracardiac thrombus typically resolves after 6 months of anticoagulation. Because this patient experienced little observable change after 4 months, neoplasia quickly became the suspected etiology. Despite the incongruence between imaging and pathology in this patient, CMR imaging can typically detect distinct morphologic features and tissue components unique to primary cardiac sarcomas. One possible explanation for the thrombotic appearance of this tumor on CMR imaging is that, although angiosarcoma is characterized by a high first pass and late gadolinium enhancement because of its extensive vascularity, an undifferentiated sarcoma (as in this report) can display more variability on first pass and late gadolinium enhancement, pos-

sibly confounding the CMR imaging results.[10] Given the lack of contrast enhancement, this tumor may have been minimally vascularized; this would have increased the difficulty of identification on contrast imaging but prevented more aggressive tumor growth and dissemination, which could explain the overall static disease course over 4 months.

This report describes an unusual presentation in a patient with undifferentiated pleomorphic primary cardiac sarcoma discovered incidentally via echocardiography. Although the mass was initially thought to be a thrombus by appearance on TEE and CMR imaging, it failed to resolve with anticoagulation and was eventually resected, leading to pathologic identification of this rare primary cardiac malignancy. This case demonstrates the importance of keeping primary cardiac malignancy in the differential diagnosis for patients with intracardiac masses, especially when those masses are refractory to anticoagulation. Timely resection and consultation with oncology is crucial to guide adjuvant care, given the high metastatic potential of these neoplasms.

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