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Left Atrial Undifferentiated Pleomorphic Sarcoma Causing Mitral Valve Obstruction

Eric H. Yang, MD; Victor Gabrielian, MD; Ping Ji, MD; Bassam Omari, MD;
Joseph L. Thomas, MD; Jina Chung, MD

A 61-year-old Latino woman with no past medical history presented to the emergency department with 2 weeks of gradual onset of dyspnea on exertion, cough, and palpitations. She denied any recent illnesses, fevers, or unintentional weight loss. She was not taking any medications or using tobacco, alcohol, or illicit drugs.

On physical examination, she had a temperature of 100.6°F, a heart rate of 122 bpm, respiratory rate of 25 breaths per minute, blood pressure of 155/92 mm Hg, and oxygen saturation of 92% on bilevel positive airway pressure. Her physical examination was notable for jugular venous distension, a loud S₁ and normal S₂, a grade 2/6 middiastolic murmur heard loudest at the apex, crackles throughout her posterior lung fields, and trace bilateral lower-extremity edema.

Her ECG showed sinus rhythm with no significant ST-T abnormalities (Figure 1). A portable chest radiograph showed

mild enlargement of the cardiac silhouette, bilateral air space opacities, and small bilateral pleural effusions (Figure 2). Computed tomography of the chest with intravenous contrast was done to evaluate for pulmonary embolism; it revealed a mass with lobulated margins within the left atrium (Figure 3) protruding into the left ventricular cavity and infiltrating the left atrial wall.

An emergent transthoracic echocardiogram showed normal left and right ventricular systolic function. A large mass in the left atrium was seen prolapsing into the left ventricle during diastole, causing functional mitral stenosis, and was attached to the posterior wall of the left atrium. The patient was admitted for intravenous diuresis and was evaluated by the cardiothoracic surgery service for tumor excision.

A preoperative coronary angiogram demonstrated extensive tumor blush with filling from the left atrial recurrent

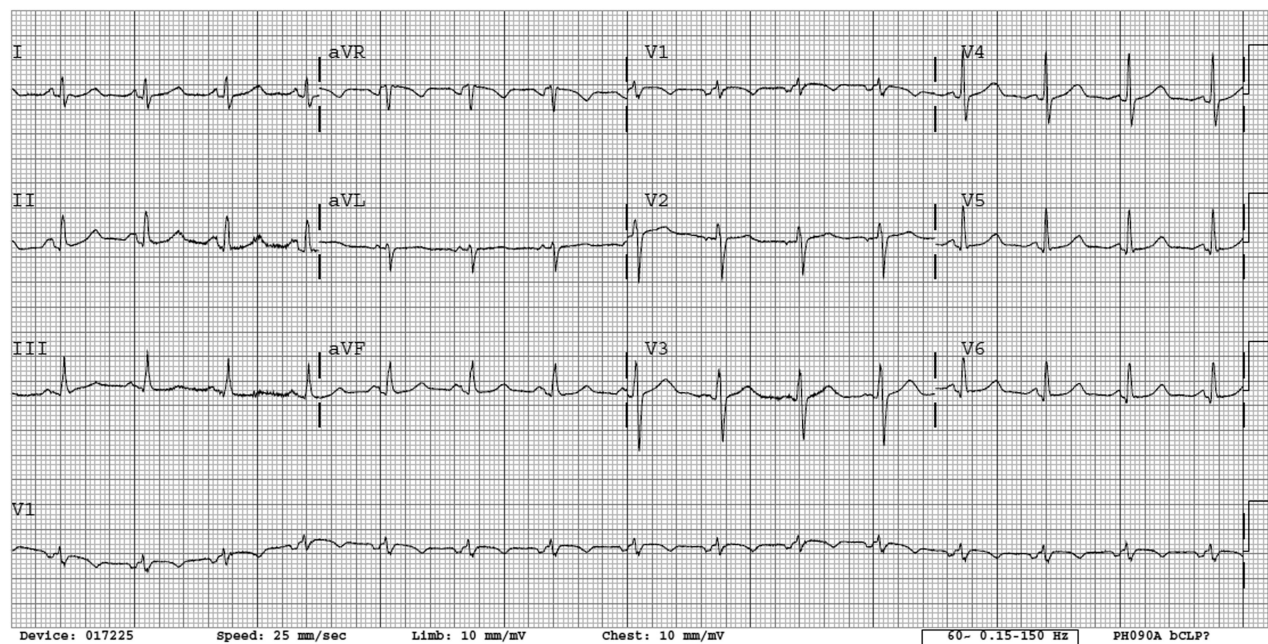


Figure 1. Twelve-lead ECG demonstrating normal sinus rhythm with no significant ST-T segment abnormalities.

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The online-only Data Supplement is available with this article at <http://circ.ahajournals.org/cgi/content/full/CIRCULATIONAHA.110.974345/DC1>. Correspondence to Eric H. Yang, MD, Division of Cardiology, Department of Medicine, Harbor-UCLA Medical Center, 1000 W Carson St, Box 405, Torrance, CA 90509. E-mail Datsunian@gmail.com

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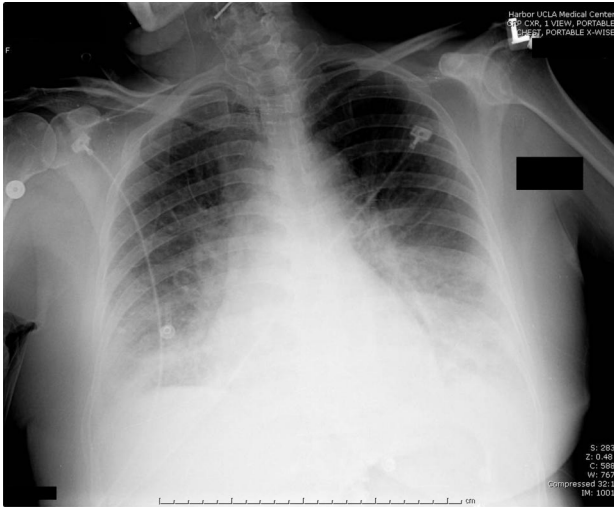


Figure 2. Portable chest radiograph showing mild enlargement of the cardiac silhouette, bilateral air space opacities, and small bilateral pleural effusions.

branch from the circumflex artery and from the sinoatrial artery and posterior left ventricular branch of the right coronary artery feeding the suspected tumor (Figure 4 and Movies I and II in the online-only Data Supplement).

The intraoperative transesophageal echocardiogram confirmed the transthoracic echocardiographic findings of a large 5-cm soft tissue density in the left atrium with a mobile, pedunculated component prolapsing into the left ventricle during diastole (Figure 5 and Movie III in the online-only Data Supplement). The mass caused a functional mitral stenosis with a valve area calculated as 1.1 cm² and a mean gradient of 7 mm Hg. The left atrium was accessed by first opening the right atrium and then the interatrial septum. The



Figure 3. A 64-slice non-ECG-gated computed tomogram of the chest with intravenous contrast. A, Oblique multiplanar view of the tumor. A large filling defect suspicious for a tumor within the left atrium (LA) with irregular margins is seen (arrow) with a maximum diameter of ≈6.7 cm. The filling defect infiltrates the posterior left atrial wall and pericardium (*). The axial, coronal, and sagittal views centered about * are seen in B, C, and D, respectively. A portion of this tumor is prolapsing into the left ventricular cavity. Bilateral moderate pleural effusions and bilateral upper lobe consolidation were present. RA indicates right atrium; PA, pulmonary artery; and T, tumor.

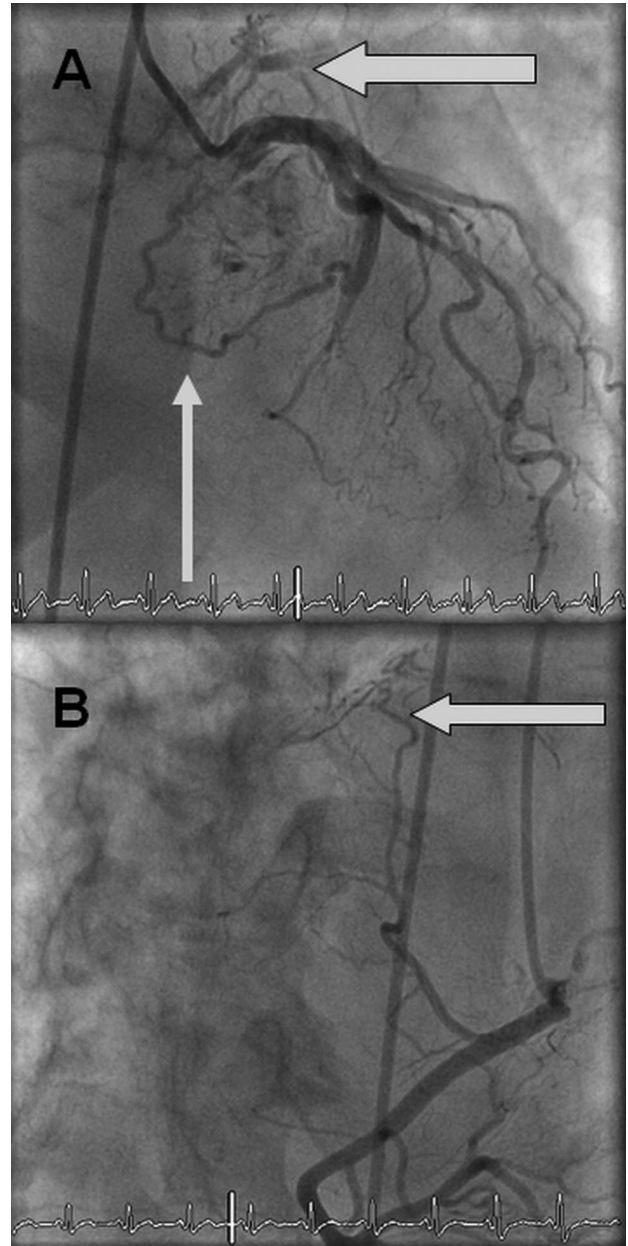


Figure 4. Coronary angiography of the left and right coronary arteries. A, Right anterior oblique view of the left coronary artery, where the left atrial recurrent branch off the circumflex provides perfusion to the tumor. B, Right anterior oblique view of the right coronary artery, with collaterals branching from the sinoatrial artery and posterior left ventricular branch to the tumor.

left atrial mass was adherent to the posterior wall of the left atrium and extended to the orifice of the left atrial appendage but spared the mitral leaflets (Figure 6). A pedunculated portion protruding through the mitral valve was excised (Figure 7); however, the rest of the mass could not be excised because it was infiltrating through the posterior wall to the visceral pericardium and into the atrioventricular groove (Figure 8). The pathological specimen later revealed a pleomorphic undifferentiated high-grade sarcoma (Figure 9).

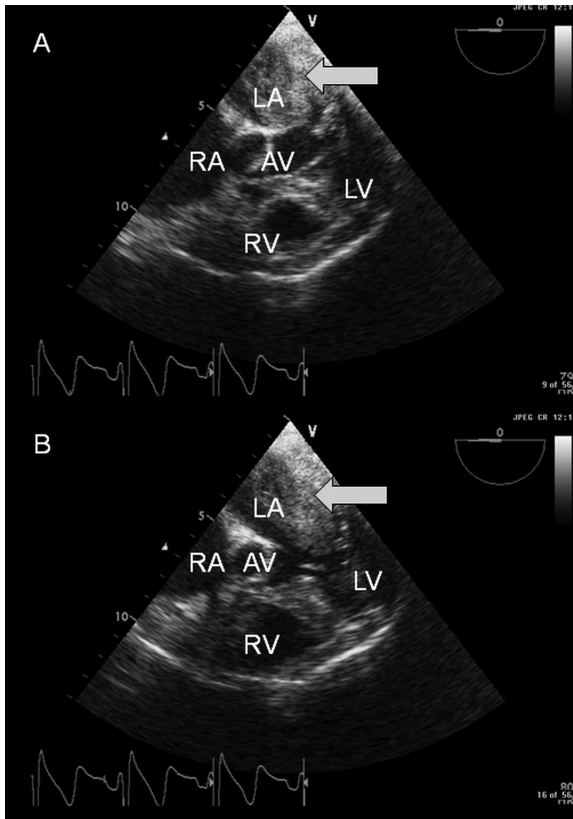


Figure 5. Transesophageal echocardiogram, 5-chamber view, showing a large soft tissue density (arrow) in the left atrium (LA) attached to posterior, superior, and anterolateral left atrial walls. The mass measures ≈ 5 cm in thickness and extends to the posterior mitral annulus and spares the mitral leaflets as seen during systole (A). A mobile, pedunculated component of the left atrial mass, measuring $>4 \times 2$ cm, prolapses into the left ventricle (LV) during diastole (B). Extension of the mass was also seen in the orifice of the left atrial appendage. A portion infiltrates the pericardium. RA indicates right atrium; AV, aortic valve; and RV, right ventricle.

The patient's postoperative course was complicated by several days of hypotension and accelerated junctional rhythm requiring inotropic support with dopamine, which gradually resolved with return of the patient's native sinus

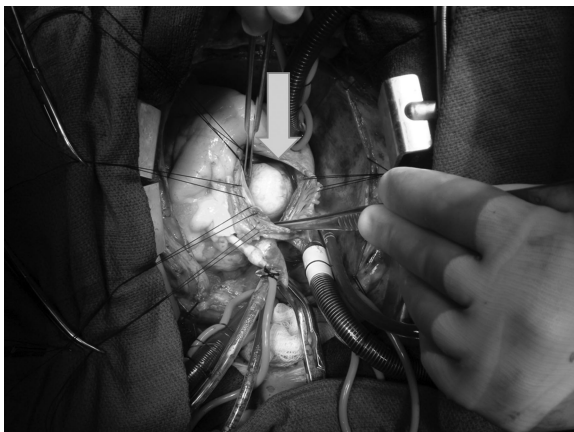


Figure 6. Exposure of the left atrial mass (arrow) by opening the right atrium and interatrial septum.

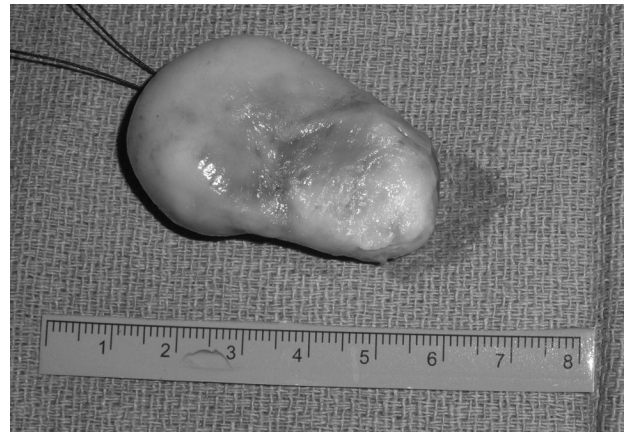


Figure 7. Gross specimen of the partially resected tumor that was obstructing the mitral valve.

rhythm. The patient was discharged with recommendation for follow-up at the Oncology and Cardiology Clinic with serial transthoracic echocardiogram surveillance and for evaluation of chemotherapy/radiation therapy options. She is doing well 2 months after discharge and is receiving outpatient chemotherapy with doxorubicin and cyclophosphamide.

Primary cardiac tumors are rare, with an incidence of $\approx 0.0017\%$ to 0.019% , with 25% of these tumors being malignant.¹ The undifferentiated sarcomas are very rare, are known to primarily occupy the left atrium, and can be mistaken for a myxoma,¹ causing pulmonary congestion. Sarcomas can also cause symptoms of dyspnea, syncope, chest pain, fever, malaise, and weight loss due to valvular inflow obstruction, arrhythmias, pericardial involvement and effusion, and tumor emboli. This case illustrates many of these features as a new presentation.

Survival depends on the extent of successful resection of sarcomas because of their high potential for recurrence and the presence of metastatic disease. Mean survival after excision has been documented to be ≈ 9 to 10 months; a

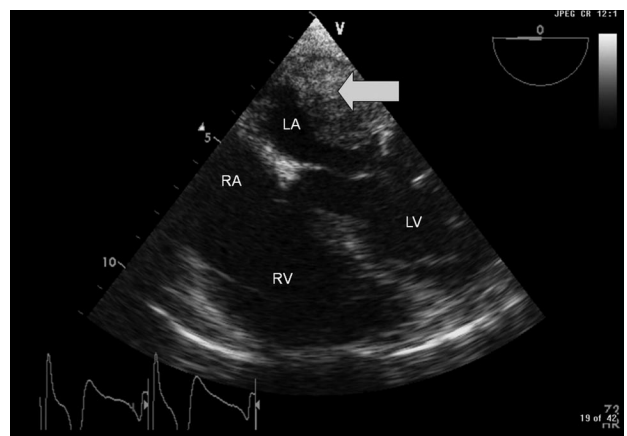


Figure 8. Transesophageal echocardiogram, 4-chamber view, after surgical excision of suspected tumor (arrow). There is still a residual burden of the density but with no functional obstruction of the mitral valve. LA indicates left atrium; RA, right atrium; LV, left ventricle; and RV, right ventricle.

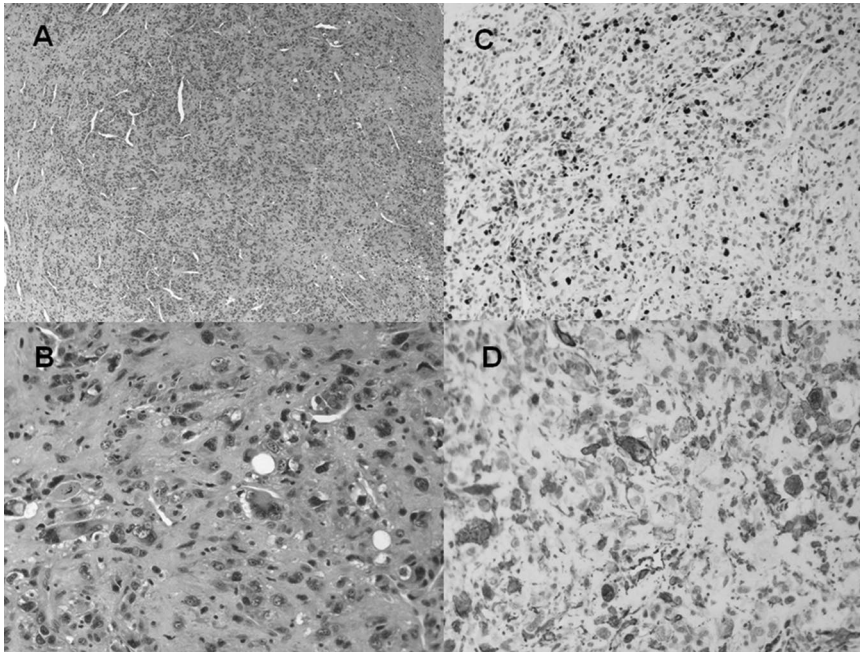


Figure 9. Histopathological analysis of tumor with findings consistent with high-grade undifferentiated pleomorphic sarcoma. A, Low magnification of hematoxylin and eosin stain showing hypercellular lesion. B, High magnification of hematoxylin and eosin stain showing the nuclei of tumor cells with hyperchromatic and marked pleomorphism. C, MIB-1 stain for proliferative index is 25% to 50%. D, Positive caldesmon stain in the large pleomorphic tumor cells.

significant predictor of long-term mortality is New York Heart Association Class III and IV symptoms.² The benefit of adjuvant chemotherapy and radiotherapy is unknown given the limited data and the lack of randomized trials; complete surgical section has been the only factor that influences survival.^{1,3}

Disclosures

None.

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