A 27-year-old man complained of frequent chest pain and recurrent syncope for 3 months. He was in mild respiratory distress with a respiratory rate of 20/min and a heart rate of 94 bpm, regular in rhythm. His blood pressure was 120/70 mmHg. Cardiac auscultation revealed a distinct early diastolic click followed by a grade 2/5 diastolic decrescendo murmur at the apex that was variable in character with postural changes. A grade 2/6 systolic ejection murmur was heard at the left lower sternal border.

Transthoracic echocardiography revealed a large pedunculated mass within the left atrial chamber, protruding into the left ventricular cavity and obstructing mitral inflow (Figure 1). Computed tomography of the chest showed a large mass in the dilated left atrium.

In view of severe mitral valve obstruction by the atrial mass, surgical removal of the tumor was recommended. During operation, an irregular tumor mass measuring 5×6×6 cm was found attached to the roof of the left atrium, with adhesions to the left atrial posterior and lateral walls. Pericardial effusion was also noted. The tumor occupied almost the entire left atrium.

Histological examination of the excised tumor mass and confirming the normal structure of the atrial wall to which the tumor was attached appeared to be slightly roughened, but no tumor penetration of the atrium wall was evident. There was also no evidence of intramyocardial or pericardial spread of the tumor mass. Histological examination of the excised tumor mass revealed features typical of a rhabdomyosarcoma (Figure 2B).

Predictive echocardiography showed normal ventricular function and no significant dysfunction of the cardiac valves, and the left atrium appeared to be clean without residual tumor. The patient was symptom free and was discharged 10 days after the operation.

The patient did well after discharge and returned for a follow-up 38 days after the operation. At this time, echocardiography revealed a recurrent small echodense mass attached to the atrial septum and another larger mass in the left atrial appendage (Figure 3A and 3B). These 2 masses had progressively enlarged on the echocardiography performed 54 days after the operation (Figure 3C and 3D), as demonstrated also by computed tomography (Figure 4 and Movies III and IV in the online-only Data Supplement).

Primary malignant cardiac rhabdomyosarcoma is a rare neoplasm that has been reported in all age groups and occurs with similar frequency in both male and female subjects.1 The usual location of this tumor in order of frequency is right atrium, left atrium, and both ventricles.2 Rhabdomyosarcomas usually originate intramurally, sometimes protruding into cardiac chambers, causing obstruction.3 The mass in our patient originated in the left atrium and protruded through the mitral valve, causing mechanical obstruction to the left ventricular inflow, clinically mimicking mitral stenosis with a low-pitched diastolic murmur, manifested clinically as recurrent syncope that was indicative of intermittent cardiac and brain ischemia.

Echocardiography is useful in revealing the location of the tumor mass and confirming the normal structure of the mitral valve. Tumor invasion to the right side of the heart can mimic tricuspid or pulmonary valve diseases with clinical presentation of right heart failure. Tumor metastases are usually found in the lung, liver, thoracic lymph nodes, and pancreas. Occasionally, cardiac rhabdomyosarcoma can present with cerebral embolism.4

Before surgery, differentiation between benign and malignant tumors is important but sometimes difficult. The most common benign cardiac tumor is the atrial myxoma, which may be accompanied by systemic embolization mimicking the metastasis of malignant tumors. In our case, atrial myxoma was suspected initially. However, atrial myxoma is typically more mobile because of its attachment to the atrial septum through a stalk compared with a malignant tumor that may have a more broad-based adhesion to the chamber walls, which may be at multiple sites. Moreover, the highly vascular nature demonstrated by color Doppler flows within the tumor mass is more suggestive of a vascularized malignant tumor than a myxoma. This case illustrates the role of echocardiography in differentiating a malignant tumor mass from a benign atrial tumor before surgery. Echocardiographic contrast perfusion imaging aids in the differentiation of cardiac masses.
Compared with the adjacent myocardium, malignant and vascular tumors are hyperenhanced by contrast.5

Disclosures

None.

References


Figure 1. A, Echocardiography (parasternal long-axis view) showed a huge mass occupying most of the left atrium (LA), with blood flow within the tumor on color Doppler examination. B, Modified 4-chamber view showed the mass protruding into the left ventricle (LV) during diastole. RA indicates right atrium; and RV, right ventricle.

Figure 2. Left, Gross appearance of the excised tumor showing hemorrhagic and necrotic areas. Right, Histology revealed roundish tumor cells with pleomorphic nuclei and large eosinophilic cytoplasm that are typical of rhabdomyosarcoma.
Figure 3. **A**, On postoperative day 38, a parasternal long-axis view of echocardiography revealed a small mass attached to the atrial septum. **B**, A modified parasternal short-axis view showed another mass in the left atrial (LA) appendage (LAA). **C**, Fifty-four days after surgery, a parasternal long-axis view showed an enlarging irregular echodense mass attached to the atrial septum. **D**, The mass in the left atrial appendage also significantly increased in size. AO indicates aorta; LV, left ventricle; and RV, right ventricle.

Figure 4. Several soft tissue masses in the left atrium (LA) were demonstrated by computed tomography 60 days after the operation, which suggested tumor recurrence. **A**, Computed tomographic 4-chamber view showed 3 masses in the left atrium. **B**, Computed tomographic 2-chamber view indicated 2 tumors in the left atrium. **C**, Computed tomographic short-axis view showed several masses in the left atrium. **D**, Computed tomographic reconstructed image showed the rhabdomyosarcoma in the left atrium close to the anterior wall. AA indicates ascending aorta; LAD, left anterior descending coronary artery; LCX, left circumflex artery; LV, left ventricle; PA, pulmonary artery; RA, right atrium; and RV, right ventricle.
Left Atrial Rhabdomyosarcoma
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**Movie Legend**

**Movie 1.** Echocardiography (parasternal long-axis view) showed a huge mass occupying most of the left atrium, with blood flow within the tumor on color-Doppler examination. Best viewed with Windows Media Player.

**Movie 2.** Modified 4-chamber view showed the mass blocking the mitral inflow partially. Best viewed with Windows Media Player.

**Movie 3.** The parasternal long axis view showed that there is a small echodensity mass attached to atrial septum 38 days post operation. Best viewed with Windows Media Player.

**Movie 4.** A modified parasternal short-axis view showed the tumor mass in left atrial appendage significantly increased in size 54 days after surgery. Best viewed with Windows Media Player.