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*Case Reports*

# Rhabdomyosarcoma Involving the Left Atrium and the Mitral Valve

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## Abstract

Rhabdomyosarcoma represents almost 20% of all primary malignant neoplasms of the heart. In the majority of cases, the tumor arises from the ventricular wall, although these tumors can also arise from the atrial walls and mimic atrioventricular valve stenosis. This report presents a case of a patient diagnosed with an atrial mass that was detected by transthoracic echocardiography and confirmed by histopathology. The mass was successfully resected during an uneventful surgical intervention, but the patient died 6 months after the procedure.

**Keywords:** Rhabdomyosarcoma; heart atria; mitral valve; heart neoplasms; case reports

## Case Report

### Presentation, Medical History, and Physical Examination

A 61-year-old woman presented to the emergency department with a 2-month history of progressive shortness of breath on exertion, orthopnea, and intermittent chest discomfort. Two days before presentation, her symptoms had worsened substantially, prompting her to seek medical advice. She had an unremarkable medical and surgical history.

On examination, the patient did not have dyspnea at rest. Her oxygen saturation was 80%, blood pressure was 130/80 mm Hg, and heart rate was 98/min. Cardiac auscultation revealed a low-pitched diastolic murmur. Respiratory examination was unremarkable, with no signs of pulmonary edema, pleural effusion, or wheezing. Laboratory results were within normal ranges.

An electrocardiogram indicated a normal sinus rhythm, but chest x-ray showed signs of atrial enlargement resembling mitral valve pathology and evidence of bilateral pulmonary congestion. Transthoracic echocardiography was performed for further evaluation and showed severe, noncalcified mitral valve stenosis, with a mean pressure gradient of 14 mm Hg. In addition, a large left atrial mass measuring 4.5 × 3.5 cm was noticed, attached to the superior wall of the left atrium and accompanied by an atrial thrombus (Fig. 1). Given the patient's history of chest pain, coronary angiography was performed, revealing isolated mild proximal calcification of the left anterior descending coronary artery.

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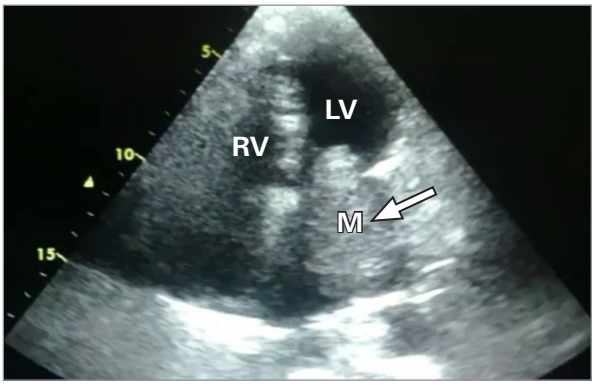


**Fig. 1** Transthoracic echocardiography shows a large left atrial mass (arrow) measuring 4.5×3.5 cm and attached to the superior wall of the left atrium. The image also shows the right ventricle and left ventricle.

*Supplemental motion image available for* Figure 1

LV, left ventricle; M, mass; RV, right ventricle.

## Abbreviation

RT, radiation therapy

## Key Points

- Acute worsening of chronic symptoms such as exertional shortness of breath and orthopnea should prompt evaluation for structural heart or vascular abnormalities, such as a mass or thrombus.
- Transthoracic echocardiography is essential in evaluating mitral stenosis and identifying underlying causes, such as atrial masses or thrombi, which may alter management.
- Surgical excision of cardiac masses enables definitive diagnosis through histopathology because rare conditions such as rhabdomyosarcoma can mimic more common etiologies of mitral valve stenosis.

## Supplementary Materials

For supplemental materials, please see the online version of this paper.

## Differential Diagnosis

The differential diagnosis of a left atrial mass includes myxoma, thrombus, and malignant tumors. Myxomas are the most common primary cardiac tumors; they are typically mobile and attached to the interatrial septum, whereas thrombi are associated with atrial fibrillation and usually found in the left atrial appendage. Malignant tumors, such as sarcomas, show rapid growth and tissue invasion. Echocardiography helps distinguish these entities based on mobility, attachment, and vascularity.

## Technique

The patient was transferred to the operating room, where a median sternotomy was performed under general anesthesia and cardiopulmonary bypass was initiated. Moderate systemic hypothermia (30 °C) and cold crystalloid cardioplegia were used for myocardial protection. A left atriotomy revealed a large, firm mass measuring 5 × 5 × 2 cm and adherent to the superior wall of the left atrium (Fig. 2A). The mass partially obstructed the right pulmonary veins and substantially reduced the mitral valve area. It extended from the posterior superior wall of the left atrium to the anterior leaflet of the mitral valve, with a broad base approximately 5 cm in diameter. The mass exhibited a firm consistency, with scattered friable areas.

Complete excision was performed without difficulty, preserving the left atrial wall. The mass was removed along with the left atrial appendage, and 3 foci were identified on the anterior leaflet of the mitral valve (Fig. 2B). Given the presence of scattered foci on the anterior leaflet, mitral valve excision and replacement were deemed necessary. A St Jude Medical 29 mm mechanical heart valve was implanted using interrupted pledgeted sutures. The cardiopulmonary bypass and aortic cross-clamp times were 104 minutes and 76 minutes, respectively. The patient was successfully weaned from cardiopulmonary bypass and transferred to the intensive care unit in stable condition with minimal inotropic support.

## Outcome and Follow-Up

Histopathologic examination of the mass revealed cells displaying large pleomorphic nuclei with abundant eosinophilic cytoplasm, resembling myoblasts (Fig. 3A). Frequent mitotic figures, including atypical forms with foci of necrosis, were noted. These findings confirmed the diagnosis of rhabdomyosarcoma with embryonic differentiation (Fig. 3B). The margins of the excised mass were friable, however, and the pathologist could not easily determine the extent of the tumor. Immunohistochemical staining was positive for myogenin and desmin, further supporting the diagnosis (Fig. 3C). Moreover, histopathologic findings of the

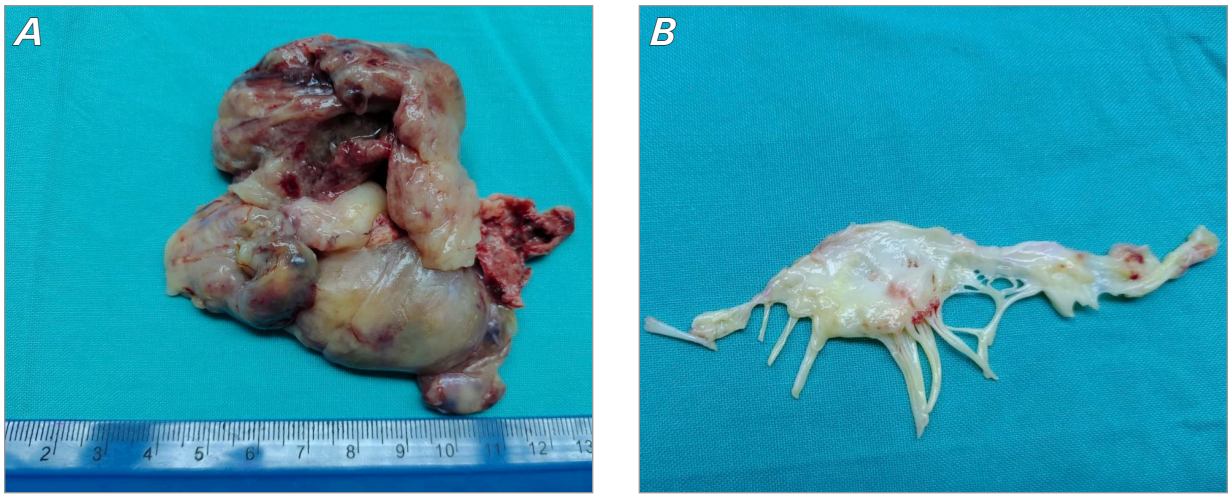


**Fig. 2** (**A**) Gross examination of the resected mass measuring approximately 5 × 5 × 2 cm. (**B**) Anterior leaflet of the mitral valve, with 3 spots for histopathologic examination.

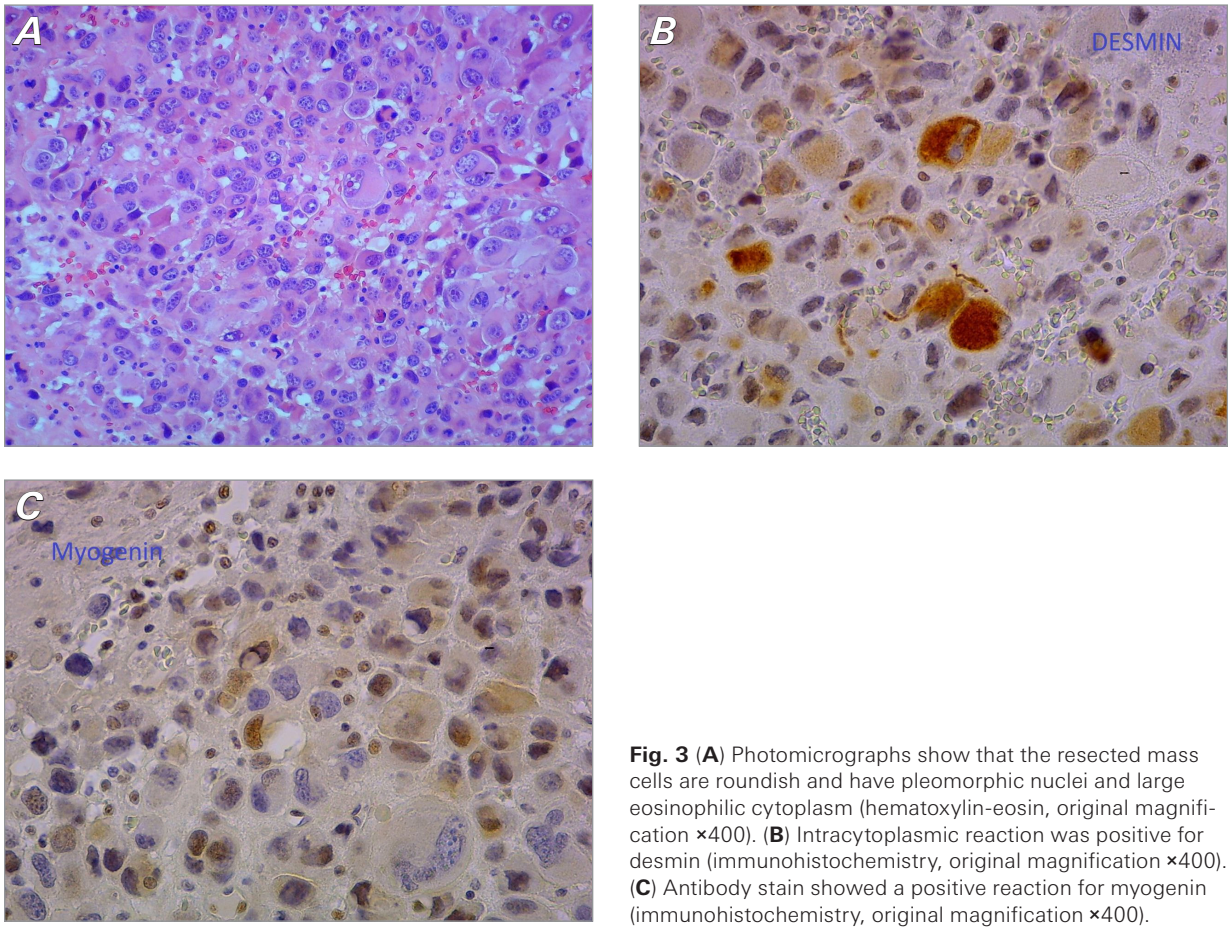


**Fig. 3** (**A**) Photomicrographs show that the resected mass cells are roundish and have pleomorphic nuclei and large eosinophilic cytoplasm (hematoxylin-eosin, original magnification ×400). (**B**) Intracytoplasmic reaction was positive for desmin (immunohistochemistry, original magnification ×400). (**C**) Antibody stain showed a positive reaction for myogenin (immunohistochemistry, original magnification ×400).

lesion forming along the mitral valve showed foci of necrosis with few atypical cells.

Postoperatively, the patient developed atrial fibrillation, which was successfully managed with amiodarone. She had an otherwise uneventful recovery and was discharged home in stable condition. A postoperative positron emission tomography scan revealed no evidence of metastasis. The patient was recommended for chemotherapy and radiation therapy (RT), but she refused any such treatment. She died 6 months after the surgical resection because of progressive heart failure and acute pulmonary edema.

## Discussion

Primary heart tumors account for only 0.01% to 0.3% of autopsy findings, making the heart an extremely rare location for primary neoplasms.[1] Among these neoplasms, only 25% are malignant.[1,2] Rhabdomyosarcoma, a rare malignant cardiac tumor originating from mesenchymal soft tissue, represents less than 20% of all malignant primary cardiac tumors. After angiosarcoma, rhabdomyosarcoma is considered the second-most common malignant cardiac tumor.[3-6] Like other types of sarcomas, primary cardiac rhabdomyosarcoma is a highly aggressive tumor, with a rapidly deteriorating clinical course and poor prognosis. The most common site for primary cardiac rhabdomyosarcoma is the ventricles. In contrast, this patient's tumor was located in the left atrium and attached to the mitral valve, a particularly rare site, with only 12 cases documented in the literature.

Clinically, patients with a primary cardiac mass, including rhabdomyosarcoma, usually present with symptoms of heart failure and blood flow obstruction, such as syncope, dyspnea on exertion, chest discomfort, easy fatiguability, or (in severe cases) cardiogenic shock.[7] Symptom severity is primarily influenced by the tumor's size and location.[7,8] Moreover, patients can experience complications from tumor-related thromboembolic events, such as cerebrovascular, pulmonary, or peripheral vascular embolisms.[7,8] Other complications related to malignancy infiltration, such as cardiac tamponade, arrhythmia, and restrictive cardiomyopathy, can develop.[9-12] This patient presented with heart failure symptoms, including dyspnea and chest discomfort, without any other complications. Chaturvedi et al[3] reported a similar case of a patient with dyspnea. Further evaluation by echocardiography showed the presence of an intra-atrial mass attached to the mitral valve and the interatrial septum.

In alignment with this case, imaging modalities such as echocardiography, computed tomography, and magnetic resonance imaging are important for identifying and characterizing intracardiac tumors. These modalities help determine the tumor's size, location, and relationship to surrounding structures, guiding management planning.[11] Histopathologic examination of the surgical biopsy specimen, however, is essential for definitive diagnosis and staging, which will determine the appropriate management plan.

Rhabdomyosarcoma is typically treated with a combination of surgery, chemotherapy, and RT.[3,11,12] Although complete surgical excision is essential for restoring blood flow and alleviating symptoms, chemotherapy plays a central role in the management of rhabdomyosarcoma due to its high metastatic potential by the time symptoms appear. The standard chemotherapy regimen for rhabdomyosarcoma is vincristine, dactinomycin, and cyclophosphamide, and RT is useful for local control of residual tumor after surgery.[13,14]

In the present case, the patient underwent complete surgical resection of the tumor without postoperative chemotherapy or RT, which the patient refused. Despite the significantly poor prognosis of this tumor type, surgical removal is essential for symptom relief and prevention of further complications. As in this case, all previous cases of left atrial rhabdomyosarcoma were treated initially with surgical resection, whether complete or partial. The operation was not easy to perform, however, given the patient's clinical condition, the tumor's size and firm attachment to the cardiac structure, and the involvement of valvular leaflets.[9]

Although combined treatment modalities have led to improved outcomes for rhabdomyosarcoma in common sites, such as the head, neck, and genitourinary tract, primary cardiac rhabdomyosarcoma remains associated with poor outcomes. Most patients survive less than a year, regardless of treatment approach.[3,12,15] Chaturvedi et al[3] reported an 8-month survival after successful resection of a left atrial mass. Kimura et al[12] described a case of right atrial rhabdomyosarcoma treated with incomplete surgical resection followed by RT, although the patient died within 2 months of surgery. This poor prognosis is related to functional and structural cardiac loss following surgical resection of the tumor with free margins, which is not present in surgical resection of rhabdomyosarcoma in other body sites.

Notably, rhabdomyosarcoma outcomes are worse in adults than in pediatric patients. Because rhabdomyosarcoma primarily occurs in children, most treatment protocols are derived from pediatric studies, highlighting the need for further research into the disease's behavior and management in adults.[16]

## Article Information

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