

A Case of Primary Cardiac Rhabdomyosarcoma with Surgical Removal and Mitral Valve Repair

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ABSTRACT

Primary cardiac tumors are rare. Nearly 25% of primary cardiac tumors are malignant, with rhabdomyosarcoma being the second most common primary sarcoma. Symptoms are variable, and the clinical presentation depends on the location and propagation of the tumor. Transthoracic and transesophageal echocardiography are preliminary tests in diagnosing the disease. Echocardiographic findings should be supported by other imaging methods. In appropriate cases, surgery combined with chemotherapy and radiotherapy is suggested. We present a case of primary cardiac rhabdomyosarcoma with surgical removal and mitral valve repair.

INTRODUCTION

Rhabdomyosarcomas constitute 21% of the primary neoplasms of the heart and are the second most common primary neoplasm in adults [Vander Salm 2000]. These tumors usually arise from the ventricular [Schmaltz 1981] and atrial [Sokullu 2008] walls. Cardiac rhabdomyosarcomas are often asymptomatic until disease is advanced, and even then they produce nonspecific symptoms (eg, dyspnea, chest pain, congestive heart failure, arrhythmias) that inevitably require surgical resection. We report a case of left atrial rhabdomyosarcoma localized on the mitral valve that led to hemodynamic obstruction.

CASE REPORT

A 38-year-old man was admitted to the hospital because of increasing dyspnea and weakness. A physical examination revealed an extremely loud first sound with a late-systolic murmur and a rumbling mid-diastolic murmur. His heart rate was 92 beats/min, and the rhythm was regular. The patient's blood pressure was 120/70 mm Hg. A chest radiograph

indicated the presence of increased pulmonary vascularity. The results of routine blood tests were normal. Transthoracic and transesophageal echocardiography evaluations revealed a large mass attached to the posterior mitral valve leaflet. The mass protruded into the left ventricle, causing transient obstruction between the cardiac chambers (Figure 1). Furthermore, a continuous wave Doppler examination showed the presence of a diastolic transmitral flow curve typical of severe mitral stenosis, with a maximal diastolic flow velocity of 2 m/s and a mean diastolic pressure gradient of 10 mm Hg. Echocardiographically, these observations were consistent with cardiac myxoma. Coronary angiography revealed a patent vasculature.

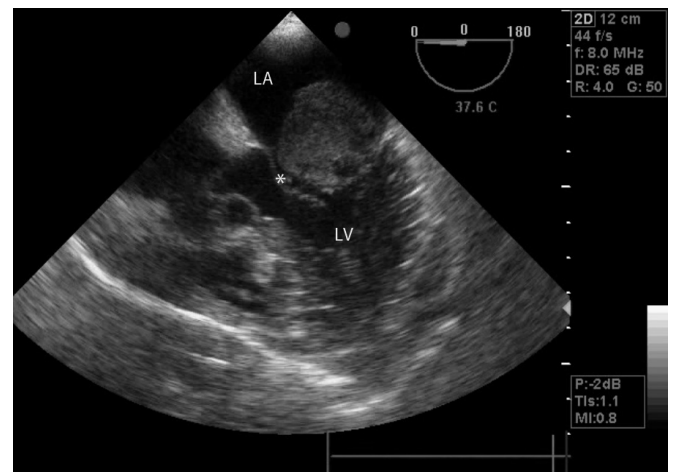


Figure 1. Transesophageal echocardiographic view during diastole showing a large mass prolapsed into the left ventricle and completely obstructing the mitral valve. LA indicates left atrium; LV, left ventricle; *, anterior mitral valve leaflet.

Operative Technique

A median sternotomy was performed, and the pericardium was opened. Ascending aorta and bicaval cannulation were completed. Cardiopulmonary bypass was established. Antegrade and retrograde cold blood cardioplegia were used for myocardial protection. The left atrium was opened. A solid cardiac mass (3 × 4 cm) was detected on the atrial side of the posterior mitral valve leaflet

Received January 4, 2013; accepted May 29, 2013.

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(Figure 2). The tumor and its large peduncle were excised along with part of the posterior mitral valve cusp. The posterior leaflet was reconstructed with a pericardial patch, followed by implantation of a 29-mm flexible annuloplasty ring. After tumor removal and mitral valve repair, an echocardiographic examination showed a normal pattern of diastolic flow through the mitral valve.

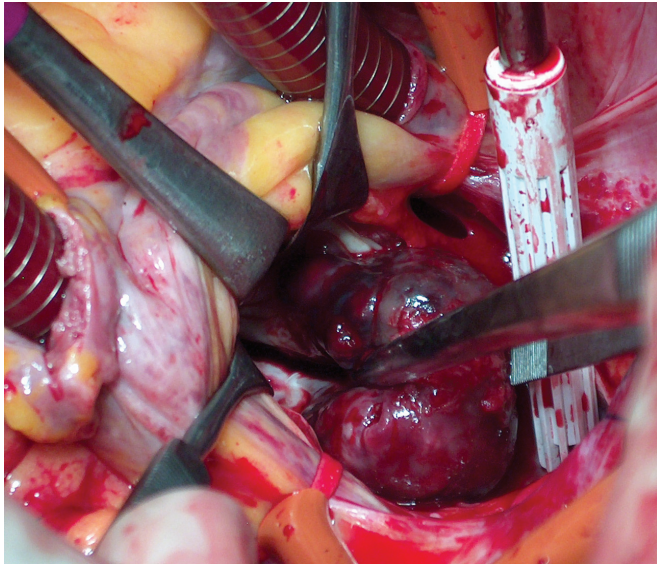


Figure 2. Intraoperative view of the tumor.

The mass was frozen and sectioned. Examination of frozen sections stained with hematoxylin and eosin revealed a myxoma. The patient was easily weaned from cardiopulmonary bypass and had an uncomplicated postoperative course. He was discharged from the hospital on the sixth postoperative day.

Although the preliminary diagnosis based on the observations of frozen sections was myxoma, the final diagnosis rendered was rhabdomyosarcoma. Immunohistochemistry tests revealed diffuse cytoplasmic positivity for desmin (Figure 3). The patient was referred to the oncology department.

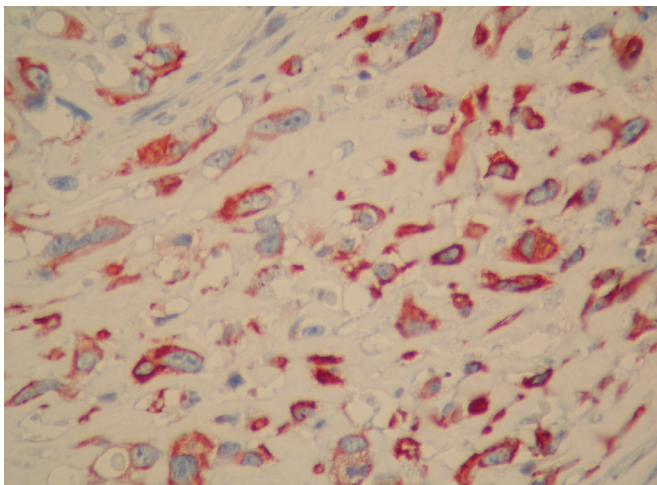


Figure 3. Microscopical view of the malignant tumor cells indicating diffuse positivity for desmin in the cytoplasm (immunoperoxidase $\times 400$).

Follow-up

After the final diagnosis, a positron emission tomography scan detected no metastasis. At the sixth postoperative month, the patient was asymptomatic and tolerated chemotherapy well. Cardiac magnetic resonance imaging and positron emission tomography scans were repeated. The survey indicated promising results; however, at postoperative month 14, metastatic brain lesions were detected after the patient reported neurologic problems. At the same time, a hemodynamically insignificant local recurrence was detected. The cardiac lesion was followed, and the patient underwent neurosurgery for the brain metastasis. Subsequently, combined chemotherapy and radiotherapy were administered. Three months later, the cardiac mass that had originated on the mitral valve grew rapidly and produced hemodynamic instability. The patient underwent reoperation at a different center. Three months later the patient died.

DISCUSSION

Cardiac neoplasms can be primary or secondary. Primary cardiac tumors are rare, with an incidence ranging from 0.0017% to 0.28% in autopsy series. Approximately 75% of primary cardiac tumors are benign [McAllister 1979]. Sarcomas are the largest group of primary malignant cardiac neoplasms. The most frequent neoplasm seems to be angiosarcoma (35%-40%), followed by rhabdomyosarcoma (20%-25%) [Miralles 1991].

In rhabdomyosarcoma cases, the ventricular septum and the ventricular wall are frequently involved [Schmaltz 1981]. The tumor usually has an intramural development, sometimes entering cardiac chambers, expanding polypoidally, and causing disturbances in atrioventricular flow [Villasenor 1985]. In our patient, findings of the physical examination suggested a valve pathology. Involvement with the atrioventricular valve played a major role in early recognition and diagnosis of the tumor.

Transthoracic and transesophageal echocardiography can diagnose the presence of a mass, but these techniques are frequently insufficient for identifying the nature of a tumor [Castorino 2000]. A malignant tumor can be misdiagnosed as a myxoma [Corradi 2012]. Cardiac magnetic resonance imaging and computed tomography are useful techniques for defining the nature of an intracardiac mass. The operative strategy should be determined after assessing the results of such imaging studies [Shanmugam 2006]. The objective therapy for benign valve tumors is tumor resection and valve preservation. For a malignant atrioventricular valve tumor, however, complete resection of the valve and the subvalvular apparatus is advised [Huang 2003]. Therefore, valve replacement should have been preferred to protection in our case. The former might have offered better prospects for survival.

In conclusion, cardiac rhabdomyosarcoma is often an aggressive disease. Clinical signs and symptoms depend on the location of the tumor and its propagation. Approaching these lesions on the assumption that the probable diagnosis is a myxoma is not ideal and could compromise the extent and type of resection, which can influence the outcome. Wide surgical resection may reduce the chances of local recurrence.

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