The Heart Surgery Forum #2007-1176
11 (2), 2008 [Epub April 2008]
doi: 10.1532/HSF98.20071176

Online address: http://cardenjennings.metapress.com

Primary Cardiac Rhabdomyosarcoma of the Right Atrium: Case Report

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ABSTRACT

Rhabdomyosarcoma is a rare malignant tumor of the heart that accounts for 20% of all primary malignant neoplasms of the heart. Symptoms vary in accordance with the location of the mass; unfortunately, by the time the patient becomes symptomatic, the tumor has already metastasized to other organs. Diagnosis is frequently obtained via transthoracic or transesophageal echocardiography and nuclear magnetic resonance imaging. Surgery is indicated for malignant cardiac neoplasms to relieve cardiac symptoms and to prolong patient survival. Subsequent postoperative chemotherapy or radiotherapy is necessary, and the long-term prognosis is poor. We present a case of a primary cardiac rhabdomyosarcoma that arose from the lateral wall of the right atrium and required implantation of a permanent cardiac pacemaker after surgery.

INTRODUCTION

Primary cardiac tumors are rare. They occur at an incidence of 0.001% to 0.28%, and 25% of such tumors are malignant [Holley 1995]. Rhabdomyosarcoma accounts for almost 20% of all primary malignant neoplasms of the heart. It is usually localized in the ventricular walls, the interventricular septum [Schmaltz 1981], or the right atrium [Chan 1985]. Symptoms vary according to the location of the mass. In adults, the disease sometimes mimics atrioventricular valve stenosis if the tumor has arisen from the atrial walls. It is also associated with a poor prognosis, owing to diagnostic delay, therapeutic difficulty, and the disease's high metastatic potential. We report a case of right atrial rhabdomyosarcoma that presented with dyspnea and chest pain.

CASE REPORT

A 50-year-old man was admitted to our hospital because of a 15-day history of exertional dyspnea and chest pain. A physical examination, routine blood tests, and a chest radiographic evaluation were performed, and the results were all nonspecific. The patient's blood pressure was 110/70 mm Hg, and his heart rate was 88 beats/min. A transthoracic echocardiography evaluation revealed an echogenic mass of 3.2 × 2.4 cm at the lateral wall of the right atrium. Computed tomography and nuclear magnetic resonance imaging examinations revealed the origin of the mass to be in the lateral wall of the right atrium (Figure 1). A positron emission tomography (PET) scan revealed the malignant potential of the mass, but there was no evidence of metastasis to lymph nodes or other organs. A coronary angiographic examination was performed because of the patient's history of chest pain, and the results showed isolated noncritical stenosis of the circumflex coronary artery. The patient was prepared for surgery.

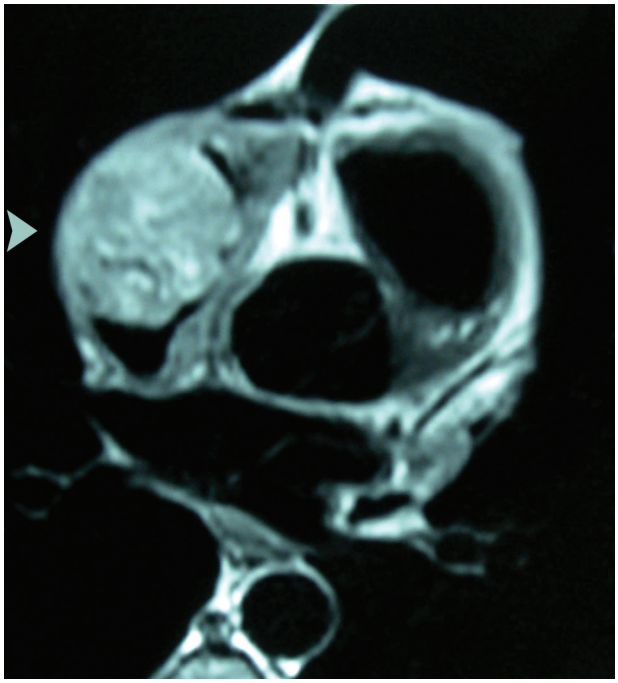

Figure 1. Preoperative nuclear magnetic resonance image of the right atrial mass (arrowhead).

Received August 16, 2007; accepted December 20, 2007.

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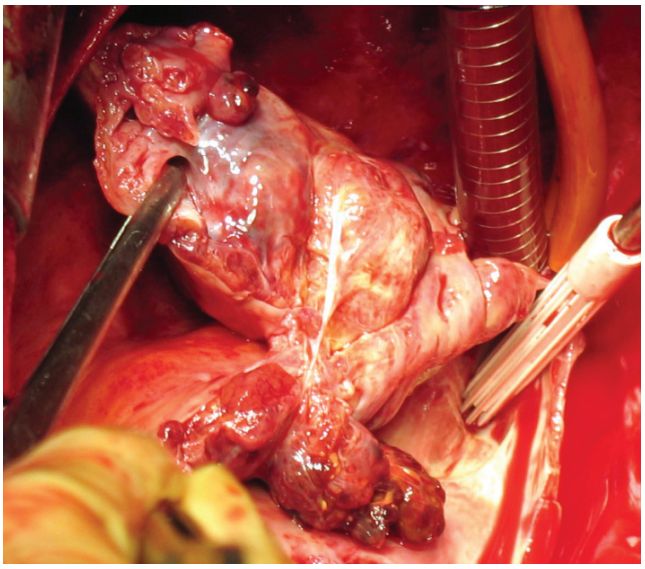

Figure 2. Intraoperative view of the tumor.

Operative Technique

A median sternotomy was performed, and the pericardium was distended. We detected a large cardiac mass (4 × 5 × 9 cm) on the right atrium that infiltrated the sinus node. Moderate systemic hypothermia (32°C) and antegrade and retrograde cold blood cardioplegia were used for myocardial protection. We completely excised the mass, which was solid, encapsulated, and irregular (Figure 2), along with a large amount of atrial tissue and the sinus node. We then reconstructed the right atrium with a polyethylene terephthalate fiber (Dacron) graft into its anatomical shape and placed temporary cardiac-pacing leads. The cardiopulmonary bypass time was 77 minutes, and the aortic cross-clamp time was 52 minutes. We easily weaned the patient from cardiopulmonary bypass and transferred him to the intensive care unit without significant inotropic support. The patient was extubated 9 hours after the surgery. He was discharged from the intensive care unit on the first postoperative day with temporary cardiac pacing.

A microscopy analysis revealed fusiform tumor cells with pleomorphic nuclei (Figure 3). Positive results in monoclonal antibody tests for desmin, vimentin, and muscle actin confirmed the histologic diagnosis of rhabdomyosarcoma.

A permanent cardiac pacemaker was implanted in the patient on the 18th postoperative day, and he was discharged from the hospital on the 21st postoperative day for subsequent oncologic treatment.

DISCUSSION

Primary malignant tumors of the heart are rare. Twenty-five percent of cardiac neoplasms have malignant potential [Holley 1995]. These tumors primarily arise from the ventricular walls and deteriorate valvular function [Schmaltz 1981]. In adults, they sometimes arise from the atrial walls [Chan 1985]. Angiosarcomas generally originate from the right side of the heart, usually from the right atrium. In our patient, the tumor was located on the right atrial wall. The diagnosis is frequently confused with pericardial inflammatory disease because of the pericardial involvement [McFadden 1997].

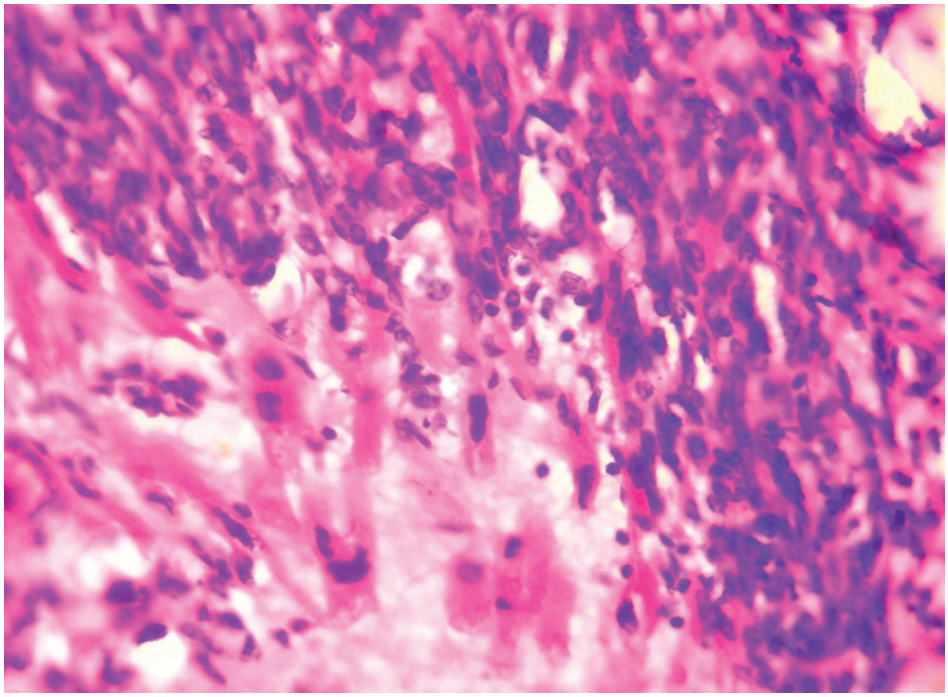

Figure 3. Microscopical view of the malignant tumor cells, which have a fusiform nucleus and an acidophilic cytoplasm.

Like other sarcomas, rhabdomyosarcoma is very aggressive. Patients often present with cardiac failure. By the time the cardiac symptoms have become evident, the metastatic lesions are usually diffuse. The prognosis is very poor, and patients usually survive less than 1 year, despite excision of the primary tumor and subsequent radiation and chemotherapy [Castorino 2000].

Transthoracic and transesophageal echocardiography can be used to determine the location, size, shape, attachment site, and mobility of the tumor [Reeder 1991; Centofanty 1999]. Computed tomography and magnetic resonance imaging are more useful for defining the nature of the mass [Castorino 2000]. Coronary angiography can exclude coexisting coronary artery disease. We used all of the diagnostic imaging techniques in our patient. In addition, the PET scan revealed no evidence of any metastasis and provided information about the survival prospects for the patient. Microscopy and immunomorphologic evaluations enabled the diagnosis of malignant rhabdomyosarcoma.

Surgery is indicated for malignant cardiac neoplasms to relieve symptoms of acute heart failure and to prolong the survival of the patient [Chitwood 1988; Murphy 1990]. In the presented case, we completely removed the tumor, anatomically reconstructed the right atrium, and established the cardiac rhythm with a permanent cardiac pacemaker. Oncologic treatment has been initiated.

In conclusion, rhabdomyosarcoma is an aggressive malignant cardiac tumor for which surgery is indicated to relieve cardiac symptoms and to prolong survival. Echocardiography and nuclear magnetic resonance imaging are beneficial for diagnosis. A PET scan can be performed to detect potential metastasis, and subsequent oncologic treatment is necessary. Finally, permanent cardiac pacing is indicated if the sinus node has been infiltrated by the tumor and has to be excised during the surgery.

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