Case Report

Left Atrial Rhabdomyosarcoma Mimicking Mitral Valve Stenosis

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A rare case of left atrial rhabdomyosarcoma is presented in a patient with symptoms of congestive heart failure mimicking a hemodynamically obstructive mitral stenosis and secondary pulmonary hypertension. Although the diagnosis of a cardiac neoplasm is often difficult, it should be suspected in any patient with idiopathic heart failure refractory to conventional therapy, or with systemic or pulmonary emboli without an obvious source. The possibility of a "tumor plop" should always be considered during auscultation of a diastolic click. If clinically suspected, echocardiography will usually establish the diagnosis and allow follow-up for recurrences. If the tumor is benign, cardiac surgery will be curative and, if malignant, chemotherapy or radiotherapy should be considered.

MOST CARDIAC neoplasms are discovered during autopsy.[1] Primary rhabdomyosarcoma is an extremely rare neoplasm among patients who die with cardiac tumors.

This report describes a case of hemodynamically obstructive rhabdomyosarcoma of the left atrium in an elderly woman. Of interest is the patient's initial clinical presentation and recurrence of her tumor 3 months later. The main clinical feature was a recent onset of congestive heart failure refractory to conventional therapy.

CASE REPORT

History

An 82-year-old woman was hospitalized for evaluation of increasing shortness of breath. One month earlier, she had been admitted to another hospital with dyspnea and syncope. The patient was discharged after therapy with digoxin and diuretics; however, she was later brought to our emergency room because of recurring dyspnea, syncope and paroxysmal nocturnal dyspnea. She denied a history of angina, myocardial infarction or congenital heart disease.

Physical Examination

The patient was in mild respiratory distress with a respiratory rate of 18/minute, temperature of 37°C and pulse of 72 beats per minute and regular. Blood pressure was 120/72 mm Hg. Neck veins were distended; the chest had scattered rales bilaterally. There was a distinct early diastolic click and a Grade 2/6 diastolic decrescendo murmur at the apex that was variable with postural changes. A Grade 2/6 systolic ejection

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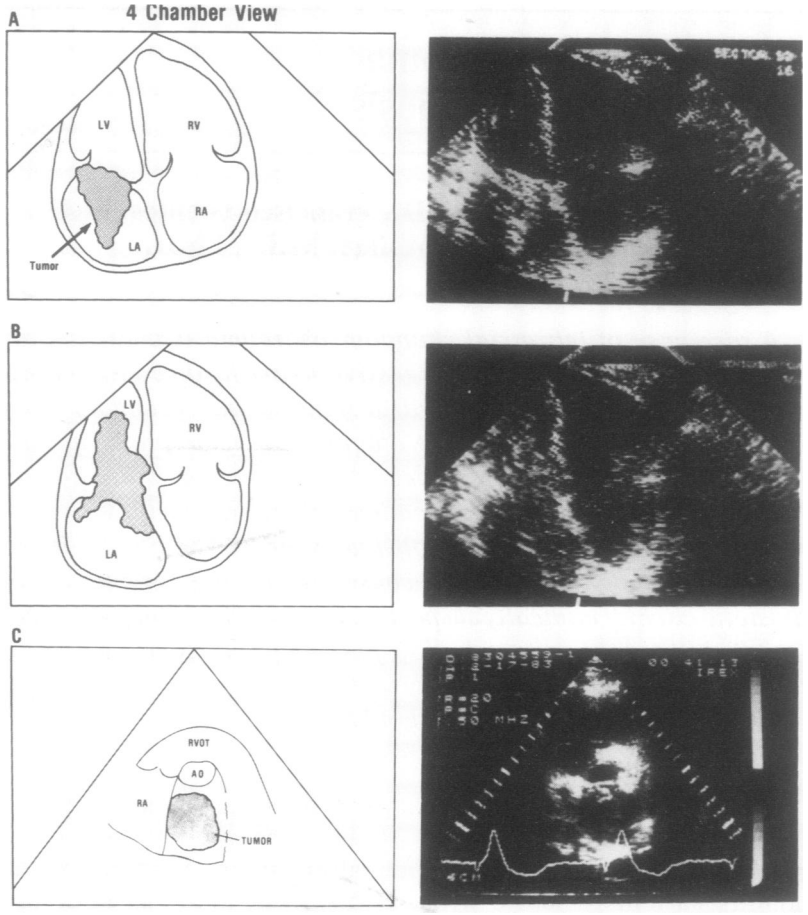

Fig. 1 Panels A and B: two-dimensional preoperative apical 4-chamber view showing a large, left atrial mass. In diastole, the left atrial mass protrudes into the left ventricular chamber. Panel C: the short axis basal view 3 months after complete removal of the atrial tumor again showing a large left atrial mass. LV = left ventricle; LA = left atrium; RV = right ventricle; RA = right atrium; Ao = aorta; RVOT = right ventricular outflow tract.

murmur was heard at the left lower sternal border. There was mild pedal edema.

Laboratory Results

Hematocrit was 39%; an electrocardiogram showed normal sinus rhythm, and the chest X ray illustrated an enlarged cardiac silhouette, upper vascular redistribution, and right pleural effusion. Two-dimensional echocardiography revealed a large pedunculated mass attached to the left atrial chamber, protruding into the left ventricular cavity (Fig. 1A and B). A radionuclide ventriculogram demonstrated a left ventricular ejection fraction of 56% and a mobile left ventricular filling defect originating in the left atrium.

Cardiac Catheterization

The mean right atrial pressure was 14 mm Hg and right ventricular pressure was 82/40 mm Hg with a mean of 50 mm Hg. The mean pulmonary capillary wedge pressure was 35 mm Hg. The cardiac index was 1.6 L/min/m^2. The systemic vascular resistance was 2232 dynes·sec·cm^{-5} with a pulmonary vascular resistance of 571 dynes·sec·cm^{-5}. The mean mitral transvalvular gradient was 28 mm Hg. The coronary arteries were normal.

Therapy

In view of severe mitral valve obstruction by the atrial mass, surgical removal of the

tumor was recommended. At operation, the patient was discovered to have a polypoid irregular 8 × 7 × 3.5 cm tumor mass based posteriorly between the mitral valve and the pulmonary veins and extending across the mitral valve orifice. The tumor, shown in Figure 2, was removed by clean dissection from the atrial wall; the mitral valve appeared normal without evidence of tumor invasion. The operating surgeon thought the tumor had the appearance of an atrial myxoma, and was unwilling to replace the posterior wall of the left atrium with possible obstruction of the pulmonary veins or damage to the mitral valve. On examination of the posterior atrial wall where the tumor was growing, there was a slightly roughened appearance; however, it did not appear permeated by tumor. There was no evidence of intramyocardial or pericardial metastases. Histology revealed typical characteristics of rhabdomyosarcoma.

The patient tolerated her surgery well, but the postoperative course was complicated by pneumonia, atrial and ventricular arrhythmias and small bowel obstruction caused by adhesions. She underwent a second operation to relieve her bowel obstruction and did well. There was no histological evidence of tumor on any pathological bowel specimen. After 2 months of hospitalization, the patient was discharged on a regimen of digoxin and procainamide. Because of her age, the absence of obvious sites of metastases, and reluctance by the family, chemotherapy was not given.

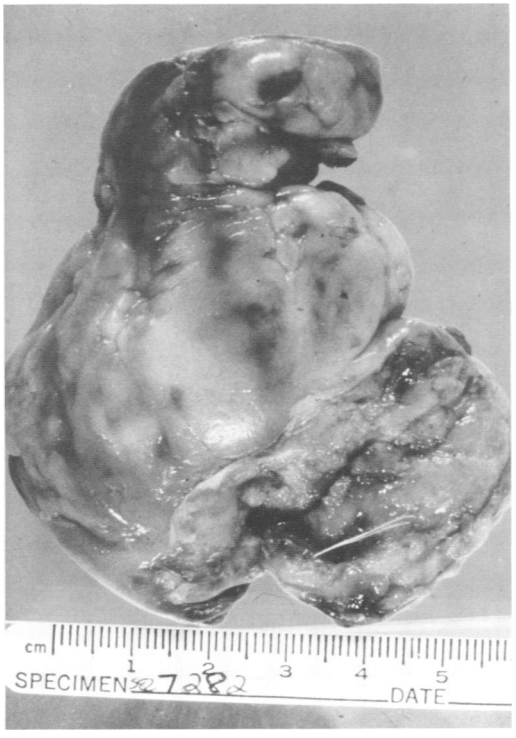

Fig. 2 Photograph of the specimen excised at the first operation. The border attached to the posterior atrial wall is at the bottom of the photograph.

Follow-up

The patient did well for about 1 month and was readmitted to our intensive care unit after being found in an unresponsive condition at home. A repeat two-dimensional echocardiogram revealed a large recurrent left atrial mass (Fig. 1C). The patient expired soon after a second cardiac arrest. At autopsy, a tumor mass completely filling the left atrium was discovered. Histology revealed the typical pathological findings consistent with rhabdomyosarcoma.

DISCUSSION

Primary malignant cardiac neoplasms are almost invariably sarcomas.[2] Rhabdomyosarcoma is a rare neoplasm that has been reported in all age groups and occurs with similar frequency in both males and females.[3] The usual location of this tumor in order of frequency is right atrium, left atrium and both ventricles.[1]

Rhabdomyosarcomas usually develop intramurally, sometimes protruding into the cardiac chamber, causing obstruction. The mass in our patient was mobile, causing partial mechanical obstruction of the mitral valve. Patients usually present with symptoms suggestive of a malignancy, that is, anemia, anorexia, malaise, weight loss, fever, or cachexia; however, our patient presented with symptoms indicative of congestive heart failure.

Tumor involvement of the left heart mimicking mitral valve stenosis is a rare presentation, and the "tumor plop" may be confused with an "opening snap" of the mitral valve. Echocardiography is useful

in revealing normal motion and thickness of the mitral leaflets.

Tumor invasion of the right side of the heart can mimic tricuspid or pulmonic valvular disease with clinical evidence of right heart failure. In 1975, O'Reilly reported a case of a myocardial rhabdomyosarcoma with pulmonary outflow obstruction and rapid onset of right heart failure.[4]

Patients with primary cardiac neoplasms may also present with complex arrhythmias, conduction disturbances or pericarditis with hemopericardium. The electrocardiogram usually shows nonspecific changes. Chest X ray will often reveal no abnormalities, although an abnormal bulging of a cardiac border may be indicative of a neoplasm. Systemic emboli have been reported in several patients with left-sided cardiac neoplasms.

There is an increased incidence of rhabdomyosarcoma in patients with neurofibromatosis,[5] but, as far as we know, only a single case of cardiac rhabdomyosarcoma with cerebral emboli has been reported in the literature.[6] Another case report described a right ventricular rhabdomyosarcoma associated with polyarthritis.[7] The association of cardiac neoplasm and autoimmune abnormalities has not been clearly defined. Tumor metastases are usually found in the lung, liver, thoracic lymph nodes and pancreas.

With the availability of two-dimensional echocardiography, the ability to diagnose intracavitary masses or tumors has improved. In the past two decades, echocardiography has allowed the physician to observe large intramural or intracavitary masses throughout the cardiac cycle and long-term follow-up of surgically excised cardiac neoplasms. Newer echocardiographic techniques utilizing tissue signature may provide information as to whether a cardiac mass is cystic, solid, or encapsulated.

Cardiac catheterization and angiography provide hemodynamic information as well as outlining of the tumor, especially if the neoplasm involves the left side of the heart. The angiogram may also reveal tumor vascularity.

The recommended therapy of rhabodomyosarcoma is surgery followed by chemotherapy and radiation. The results of radiotherapy have been disappointing. Despite chemotherapy, the response in the majority of patients is poor, with most patients dying within 1 year of diagnosis.

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