

A Primary Cardiac Sarcoma Preoperatively Presented as a Benign Left Atrial Myxoma

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Primary cardiac sarcomas are extremely rare. We report a case of a primary cardiac sarcoma with myxoid change, which originally presented as a benign cardiac myxoma on a two-dimensional echocardiogram. On operating, the mass was found to extend into the posterior left atrial wall, the left pulmonary vein, and the mitral valve. The patient underwent wide resection of the left atrium, a mitral valve replacement and a left pneumonectomy. The histological diagnosis was of an undifferentiated primary cardiac sarcoma. The patient had postoperative chemotherapy. The patient expired 11 months after surgery due to a recurrence of the cardiac sarcoma. Although most tumors that develop in the left atrium are benign myxomas, we should make a preoperative differential diagnosis.

Key Words: Cardiac sarcoma, benign myxoma, echocardiography

INTRODUCTION

The prevalence of primary tumors of the heart is extremely rare.¹ Since the advent of echocardiography, the clinical experiences of cardiac neoplasms have become more frequent. Of the primary cardiac tumors, a myxoma is the most common lesion.¹ Usually, the presence of a benign cardiac myxoma is considered when a mobile mass is found in the left atrium.

We present a case of a primary cardiac sarcoma,

which was preoperatively diagnosed as a benign myxoma in the left atrium. We also discuss the differential points between a benign myxoma and cardiac sarcoma in the left atrium.

CASE REPORT

A 33-year-old female patient was admitted to our hospital because of dyspnea on exertion and orthopnea of 10 days duration. She had experienced fever, weight loss, and malaise for 1 month prior to her admission. On physical examination, the patient appeared well, and her vital signs were unremarkable. Both breath sounds were decreased. The heart rate was regular, and there was a grade II diastolic murmur in the mitral area. Laboratory studies, on admission, were normal. Chest X-rays confirmed the clinical impression of pulmonary edema. A two-dimensional echocardiogram showed a 6 × 5 cm-sized, mobile mass adhered to the posterolateral left atrial wall. During diastole, the mass elongated into the mitral valve orifice, with a semisolid consistency (Fig. 1). A preoperative diagnosis of a benign left atrial myxoma was made.

On operating, a 6 × 5 cm sized semisolid left atrial mass was found, attached with a broad stalk to the posterior left atrial wall. The globular shape and gelatinous consistency of the mass were reminiscent of a myxoma, but the attachment to the posterior wall, with a broad base, was unusual. The mass extended into the left superior pulmonary vein, obstructing its lumen. Another 1 × 0.5 cm sized mass was found on the anterior

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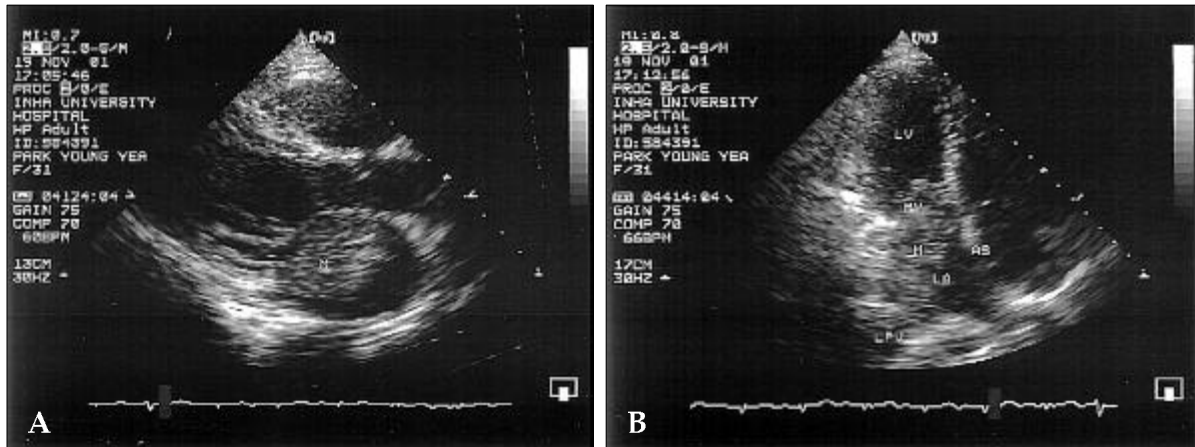


Fig. 1. (A) Two-dimensional echocardiogram, parasternal long-axis view. A mass occupying the left atrium simulates a left atrial myxoma. (B) Two-dimensional echocardiogram, apical four chamber view. The left atrial mass prolapsing into the mitral valve orifice (MV). Note, the mass is adhered, not to the septum, but to the lateral wall, and extends to the pulmonary vein side (AS=atrial septum, LA=left atrium, LV=left ventricle, M=mass, MV=mitral valve, LPV=left pulmonary vein).

mitral leaflet. A frozen pathological examination suggested a myxosarcoma. We extended the incision into the left superior pulmonary vein, and confirmed tumor invasion to the secondary division of the pulmonary vein. The posterior left atrium, involving the pulmonary vein, was widely resected. The left lung could not be saved because of tumor invasion up to the hilar vessels. After the resection of a portion of the anterior mitral leaflet, we tried to repair it, but the defect was too large to repair. Finally, the patient underwent a mitral valve replacement and a left pneumonectomy via a median sternotomy. The left atrium could be closed primarily. The bypass, and aorta cross-clamping (ACC) times were 270 and 240 minutes, respectively. Permanent histological examination revealed a primary cardiac sarcoma, with tumor invasion into the myocardium, and a focal myxoid change (Fig. 2). Immunohistochemical stains for vimentin, cytokeratin, desmin and smooth muscle actin (SMA), were performed. There was a positive result with the vimentin, and the cytokeratin, desmin and SMA were focally positive. The tumor was classified as an undifferentiated, high grade (Grade III/III) sarcoma. The postoperative course was uneventful (Fig. 3). Postoperative chemotherapy (adriamycin and dacarbazine) was instituted. The patient complained of dyspnea on exertion 6 months postoperatively. A computed tomography of the heart revealed a recurrence of

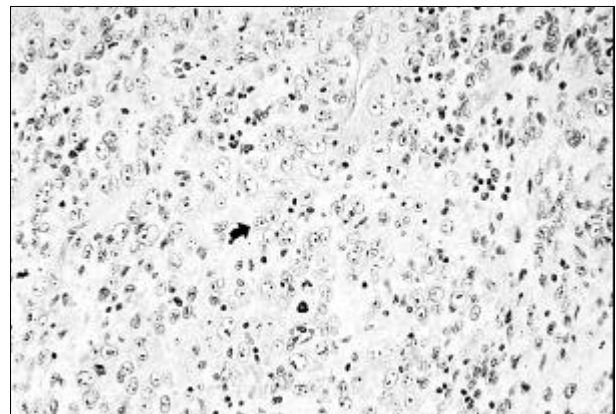


Fig. 2. Photomicrograph of the undifferentiated cardiac sarcoma, with myxoid change. Small round neoplastic cells (arrow) are observed focally in the myxoid background (original magnification $\times 200$).

a sarcoma at the posterior wall of the left atrium. The patient was treated with mediastinal radiotherapy, but refused further surgical management. The patient expired 11 months after surgery.

DISCUSSION

An endocardial myxoma, the most common form of primary cardiac tumors, is generally considered to represent a benign growth, with an excellent prognosis.¹ Clinically, patients with an



Fig. 3. Postoperative chest PA reveals the status of the mitral valve replacement and left pneumonectomy.

endocardial myxoma may manifest one or more of the classic triad of symptoms: hemodynamic obstruction, embolism and constitutional effects.¹ Patients with a primary cardiac sarcoma present similar symptoms as in a left atrial myxoma.¹⁻³ Patients with a left atrial tumor often present with dyspnea, orthopnea, paroxysmal nocturnal dyspnea, with or without hemoptysis as in mitral stenosis, and systemic embolization.

Echocardiography can provide diagnostic confirmation. The differential diagnoses of intracavitary left atrial echoes include: vegetation, thrombi and primary or secondary cardiac tumors. It is not difficult to distinguish non-neoplastic intracardiac masses from benign atrial myxomas by their echocardiographic appearances.⁴ As illustrated in our patient, however, a sarcoma with a myxoid change can closely mimic the clinical and echocardiographic appearance of left atrial myxomas. The identification of features that distinguish a cardiac sarcoma from a benign myxoma may be difficult.

From our case, and other cases in the literature,^{4,5} we can suggest several findings of left atrial sarcomas for their differential diagnosis from a benign myxoma; 1) non-septal origin of the mass, 2) extension into the pulmonary vein, 3) multiple masses, 4) a broad attachment on the left

atrial wall, and 5) semisolid consistency. Of these, the septal or non-septal origin of the mass is most important. It has been reported that only 10% of left atrial myxomas originate from sites in the atrium other than the septum, including the posterior and anterior walls, and the atrial appendages.⁴ In our case, a two-dimensional echocardiogram showed a mass adhered to the left posteriolateral atrial wall, rather than to the septum, from a retrospective view (Fig. 1B).

Of the primary cardiac tumors, 72% were benign and 28% were malignant, from the autopsies of 75 cases, from the Armed Forces Institute of Pathology.⁶ They classified 75 primary sarcomas of the heart as; angiosarcomas (26 cases), undifferentiated sarcomas (18 cases), osteosarcomas (9 cases), fibrosarcomas (6 cases), malignant histiocytomas (6 cases), leiomyosarcomas (4 cases), myxosarcomas (3 cases), synovial sarcomas (2 cases) and a neurofibrosarcoma (1 case). Our case was identified as an undifferentiated sarcoma, with myxoid change, from its histological and immunohistochemical staining. The myxoid change made the mass mobile, and thus, it simulated a left atrial myxoma.

Compared with extracardiac sarcomas, the prognosis of a cardiac sarcoma is relatively poor, as complete resection of the tumor is difficult due to the proximity of the tumor to vital structures. Previous reports on patients undergoing surgery for cardiac sarcomas have shown that the initial operation is commonly successful, as in our case, but that few patients with cardiac sarcomas survive more than 1 year.³⁻⁶ A cardiac transplantation can offer another hope for patients with malignant cardiac tumors.^{5,7}

In conclusion, the preoperative confirmation of a primary cardiac sarcoma is relatively easy if the clinician has a high degree of suspicion. Although, two-dimensional echocardiography has become the diagnostic test of choice for detecting cardiac tumors,¹⁻⁵ magnetic resonance imaging (MRI) or computed tomography (CT) is needed with indeterminate echocardiographic findings. Cardiac malignancies are difficult to treat with any modalities (operation, chemotherapy, radiation, or transplantation). A complete resection is most important for the patient's prognosis, and accurate preoperative information on the tumor invasion is

essential for the successful operation.

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