Rapid Progression of Primary Cardiac Leiomyosarcoma with Obstruction of the Left Ventricular Outflow Tract and Mitral Stenosis

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Abstract

We report a 73-year-old woman with primary cardiac leiomyosarcoma in the left atrium and ventricle. The tumor progressed very rapidly for 2 months after initial clinical evaluation. Obstruction of the left ventricular outflow tract and mitral stenosis were induced by the tumor. Urgent surgical resection was performed because she had cardiogenic shock due to paroxysmal atrial fibrillation. We could not resect the tumor completely because of severe invasion. She refused postoperative chemotherapy and radiotherapy, and died suddenly at home 89 days after surgery. To our knowledge, this is the first observation of mitral stenosis concomitant with obstruction of the left ventricular outflow tract in a patient with primary cardiac leiomyosarcoma.

Key words: cardiac leiomyosarcoma, obstruction of the left ventricular outflow tract, mitral stenosis

Case Report

A 73-year-old Japanese woman was admitted to our hospital because of dyspnea induced by physical exertion. Her local physician had pointed out mitral stenosis 2 months before (Fig. 1A). There was no known personal nor family history of cardiac disease.

On admission, her height was 146 cm and body weight 46 kg (body mass index 21.6). She had lost 6 kg of body weight during the 2 months before admission. Cardiac auscultation showed loud S1, and grade III/VI systolic murmurs at the apex. There was no swelling of superficial lymph nodes.

Transthoracic echocardiography showed a very large mass in the posterior mitral leaflet (Fig. 1B, C), which induced obstruction of the left ventricular outflow tract and mitral stenosis. The mitral valve area estimated by the pressure half time method was 0.85 cm² (Fig. 1D) and the pressure gradient in the left ventricular outflow tract was 53.6 mmHg (Fig. 1E). Transesophageal echocardiography and cardiac computed tomography (Fig. 2) demonstrated multiple separate masses in the left atrium. These masses were immobile. Whole body computed tomography revealed no metastases and no primary lesions.

On hospital day 3, she had cardiogenic shock immediately after paroxysmal atrial fibrillation. Therefore, urgent surgical resection was performed. The tumor occupied most of the left atrial cavity and adhered to the wall. The tumor, which was solid, was carefully dissected and resected. We confirmed that the tumor extended into the mitral valves and interventricular septum. We excised and removed as much of the tumor as possible, but no attempt was made to replace the mitral valve with a mechanical valve, because invasion of the tumor was very severe. Therefore, we could not resect...
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Figure 1. (A): Parasternal long axis view and M-mode echocardiogram of the mitral valve on initial clinical evaluation (two months before admission). There was mild thickening of the posterior mitral leaflet with reduced diastolic (EF) slope. Tumor was not apparent. (B–E): Echocardiograms on admission (two months after initial clinical evaluation). Parasternal long axis and apical views on admission showed a very large and immobile mass in the posterior mitral leaflet (arrows). The mitral valve area was 0.85 cm$^2$ by Doppler pressure half-time methods. Pressure gradient in the left ventricular outflow tract by Doppler flow velocity was 53.6 mmHg. LV, left ventricle; LA, left atrium.

She died suddenly at home 89 days after surgery.

**Discussion**

About 25% of cardiac tumors are malignant, mostly represented by sarcomas. Primary cardiac leiomyosarcoma is the rarest among them (2) and constitutes less than 0.25% of all cardiac tumors (3). The mean age of patients with leiomyosarcoma is 45 years (range, 6 weeks to 77 years) and the incidence is twice as common in females (4, 5).
Primary Cardiac Leiomyosarcoma

Figure 2. Contrast-enhanced computed tomograms on admission showed occupation of the left atrium (A, B), and invasion into the atrial septum (C) and the left ventricle (D) by multiple separate tumors (arrows).

The present patient had the multiple separate masses in the left atrium and ventricle. The left atrium is the typical site of origin for most cardiac sarcomas (76%), followed by the right atrium (16%). Eight percent are found in the ventricles (6). Half of the leiomyosarcoma are located in the left atrium (4). Leiomyosarcoma tends to invade the pulmonary veins and the mitral valve as observed in the present patient (7). Our patient had a large mass in the left atrium and the left ventricle which obstructed transmural and left ventricular outflow blood flow. Although these hemodynamic conditions may be thought to be typical in cardiac leiomyosarcomas, there is no report showing obstruction of the left ventricular outflow tract and mitral stenosis in a patient with leiomyosarcoma.

Transthoracic echocardiography showed a very large mass mimicking benign myxoma. The macroscopic appearance of deformable myxomas is gelatinous, papillary and friable, whereas the nondeformable tumors are smooth, firm and nonfriable. On the other hand, cardiac sarcomas have a wide range of cell differentiation (8). Cardiac sarcomas often form intracavitary left atrial lesions with partial myxomatous change. These similarities make discrimination of benign myxoma from malignant sarcoma impossible by only the echocardiographic appearance (9). Although the echocardiographic appearances of cardiac tumors do not indicate the specific tumor type, immobile tumors as in this patient may indicate malignancy. In addition, contrast-enhanced computed tomograms demonstrating multiple separate masses may indicate a malignant tumor in this patient.

We observed very rapid growth of the tumor. The average duration from symptom onset until final diagnosis is shorter for malignant cardiac sarcoma than benign myxoma. This period usually is 15 months in benign myxomas, but 4.7 months in malignant sarcomas (10).

Surgical resection is essentially palliative. The role of adjuvant radiotherapy and/or chemotherapy is still controversial (11, 12). Therefore, the prognosis for these tumors is very poor and most patients die within 1 year of diagnosis (3, 6). The mean survival after surgery and adjuvant therapies is 6.8 months (6). Aggressive and complete surgical resection seems to offer the best hope for palliation, longer survival and effectiveness of adjuvant therapies (5, 11). Heart transplantation may be an option for malignant cardiac tumors with extensive local disease (12, 13). However, the results of heart transplantation for treatment of malignant primary heart tumors are not convincing (6).
Figure 3. Macroscopic appearance of the resected tumors showed a white/brown color with areas of hemorrhage and necrosis (Top). Microscopy disclosed spindle-shaped cells with a high rate of mitosis and pleomorphic nuclei indicating malignancy (HE stain, ×100) (Middle). Tumor cells exhibited immunoreactivity for α-smooth muscle actin. Positive cells were brown (×100) (Bottom).

References