Primary Cardiac Leiomyosarcoma Growing Rapidly and Causing Right Ventricular Outflow Obstruction


Leiomyosarcomas are extremely rare primary cardiac tumors. We report a rapidly growing primary leiomyosarcoma of the right ventricle, which obstructed the right ventricular outflow tract within one month after symptom onset in a 68-year-old man. Two-dimensional echocardiography was useful in diagnosing the extent and progression of the tumor. The tumor was surgically resected on an emergency basis, and the right ventricle and pulmonary artery were successfully reconstructed. Recurrence of the tumor on the right ventricle was observed, and the patient was overcome by sudden dyspnea and died three months after surgery.

Key words: transesophageal echocardiography, transthoracic echocardiography

Introduction

Primary tumors of the heart are very rare; about 30% of them are malignant tumors and the prognosis is very poor. Most of the tumors are sarcomas. Primary leiomyosarcoma of the heart is extremely rare with only 37 cases previously reported including 6 cases in Japan (1–6).

Case Report

A 68-year-old man had dyspnea on effort and general fatigue. His past medical history was not remarkable. He was free of cardiovascular or other symptoms until he developed dyspnea while playing golf on October 15, 1994. The severity of the symptoms increased rapidly, and he was admitted to our department on November 1, 1994.

On admission, he looked unwell. His temperature was 36°C, radial pulse was regular at 70 beats/min, and blood pressure was 130/100 mmHg. The palpebral conjunctiva was not anemic. The bilateral jugular veins were distended. On auscultation, the first and second heart sounds were normal in intensity, and the third and fourth heart sounds were not audible. A grade 3 high-pitched mid-systolic murmur was heard along the left sternal border and was loudest at the meso-apex. The breathing sounds were normal. The abdomen was soft and flat, and the liver was palpable 1 finger breadth beneath the right costal margin. There was slight edema of the face and the lower extremities.

The chest X-ray film disclosed mild cardiomegaly with a cardiothoracic ratio of 0.5. The electrocardiography on admission revealed right ventricular hypertrophy with right QRS axis deviation (Fig. 1). Transthoracic two-dimensional echocardiography revealed a large mass extending from the right ventricular anterior free wall to the right ventricular outflow tract and the pulmonary artery, with enlargement of the right ventricle and oppression of the ventricular septum toward the left ventricle. Doppler echocardiographic measurement demonstrated a maximum instantaneous pressure gradient of 80 mmHg.
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Figure 1. Electrocardiograms obtained during a health checkup on March 23 and on admission, November 8. Note the marked changes in the QRS complex, which indicate a right ventricular hypertrophy pattern, on admission.

mmHg across the right ventricular outflow tract. Transesophageal echocardiography disclosed invasion by the tumor from the right ventricle to the main pulmonary arterial trunk (Fig. 2).

Right heart catheterization showed a mean right atrial pressure of 8 mmHg, right ventricular pressure of 73/15 and pulmonary artery pressure of 17/7 mmHg. The pressure gradient between the right ventricular apical portion and the outflow tract was 33 mmHg. The coronary arteriogram revealed feeding arteries of the tumor arising from the right ventricular branch. Right ventriculography showed a large filling defect in the right ventricular outflow tract (Fig. 3).

On November 13, the severity of the dyspnea increased progressively. The chest X-ray film disclosed an increased cardiothoracic ratio, to 0.66, and decreased pulmonary arterial shadow, especially in the right lung field (Fig. 4). Two-dimensional echocardiography revealed an increase in the tumor size. Acute progressive obstruction of the pulmonary artery was suspected and an emergency operation was performed. Surgery revealed that the tumor invaded the outer layer of the right ventricular anterior wall, and the right ventricular outflow tract was almost obstructed. The right ventricular wall, the main pulmonary arterial trunk and the both pulmonary arteries were resected as completely as possible (Fig. 5) and reconstructed using a porcine pericardium and Dacron patch. Neither chemotherapy nor radiation therapy was administered postoperatively in accordance with the expressed wishes of the patient and his family. Pathological analysis of the resected specimen revealed that the nuclei of the tumor cells varied widely in size, shape, and chromaticity (Fig. 6). The patient was discharged one and a half months after surgery.

On January 25, 1995, two months after surgery, two-dimensional echocardiography disclosed a recurrence of the tumor on the right ventricle and the chest X-ray film revealed metastasis of the tumor in the left lung. On February 28, the patient was overcome by a sudden dyspnea during a trip to another city. He was admitted to a hospital and treated for pulmonary embolism caused by the right ventricular tumor, however died on March
Figure 2. The long-axis-view transthoracic echocardiogram (A) shows a large mass in the right ventricle (marked by arrows), and the horizontal-view transesophageal echocardiogram (B) shows the intrusion of the mass into the main trunk of the pulmonary artery.

Figure 3. Right ventriculograms obtained in the postero-anterior view (P-A view) and left-lateral view (L-L view). The arrows indicate a large filling defect in the right ventricular outflow tract.

1, 108 days after surgery.

Postmortem examination performed 2 hours after the patient’s death revealed recurrence of the tumor in the right ventricle (Fig. 7) and a tumor embolus in the right main pulmonary artery. Histological examination revealed the tumor cells to be of spindle, often bizarre, shape, with hyperchromatic or vesicular nuclei with prominent nucleoli. The cytoplasm of some of these cells was stained red by Masson stain. Immunohistochemical examination revealed that the tumor cells were positive for vimentin, but negative for desmin, actin, myoglobin, keratin, and S-100 protein. Z-bands were not noted and a considerable number of myogenic, intermediate size filaments were noticed in the tumor cells by electron microscopic examination (Fig. 8). These findings suggested a leiomyogenic malignant tumor, and because of the absence of a primary lesion in other organs, primary leiomyosarcoma of the heart was diagnosed.
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Figure 4. Chest X-rays obtained on admission (A) and before emergency surgery (B). On the latter, the cardiac silhouette is enlarged and the pulmonary vascular shadow is decreased especially in the right lung.

Figure 5. Specimens of the resected tumor. Note the invasion from the right ventricular outflow tract to the pulmonary arteries and valves (RV: right ventricle, rt and lt PA: right and left pulmonary arteries).

Figure 6. Photomicrograph of the tumor stained by HE stain (A; ×200, B; ×400). The tumor consisted of various types of pleomorphic tumor cells, which have a spindle shape with hyperchromatic or vesicular nuclei with prominent nucleoli.
Discussion

Cardiac malignant tumor is a rare disease. Most of these tumors are sarcomas, comprised of angiosarcoma, rhabdomyosarcoma, fibrosarcoma, and liposarcoma. Primary cardiac leiomyosarcoma is very rare. Takamizawa et al (1) reviewed 25 cases in 1992, and 12 additional cases have been reported. In Japan, only 6 cases have been reported (1–6), of which the tumor was located in the right atrium and ventricle, in the right atrium and in the right ventricle in one patient each, and in the left atrium in 3 patients.

Previously, the majority of the patients were not diagnosed during life, but advances in echocardiography, computed tomography, and magnetic resonance imaging have made it possible to diagnose this disease in living patients. The most useful diagnostic method for intracardiac tumor is two-dimensional echocardiography. Protrusion of the tumor into the cardiac cavity and involvement of the myocardium, valves, and pericardium are easily and repeatedly detected by echocardiography. Therefore, the progression and recurrence of the tumor can be observed. In the present case, transesophageal echocardiography clearly disclosed the invasion of the tumor into the main pulmonary arterial trunk.

Clinically, the patients most often present dyspnea, followed by cough, chest pain, palpitation, malaise, and body weight loss. As in the present case, the progression of this tumor is very rapid. Because it originates from the heart, the tumor is not easily resected completely. Patients with primary cardiac leiomyosarcoma thus have a poor prognosis. Most of them die within one year of diagnosis. Combined chemotherapy and/or radiation therapy after the excision of the tumor is somewhat effective, and some patients have survived more than two years after surgery with radiation therapy and/or chemotherapy (1, 4, 7–9). One report describes an infant with an incompletely resected cardiac leiomyosarcoma who was treated

![Figure 7. At autopsy the heart showed a recurrent tumor in the right ventricle (marked by arrow) and in the right atrium.](image)

![Figure 8. Electronmicrograph of the tumor at autopsy (x27,000). A considerable number of myogenic, intermediate size filaments is noticed in the tumor cells, with dense bodies in some areas.](image)
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postoperatively with ifosfamide and etoposide and was still disease-free 5 years following the completion of the therapy (10). The treatments for cardiac leiomyosarcoma should be evaluated, although this is difficult due to the rarity of this disease.

References