MALIGNANT FIBROUS HISTIOCYTOMA OF THE HEART

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A case of cardiac sarcoma was reported. The tumor, which arose from the posterior wall of the left atrium, was demonstrated by echocardiography, contrast-enhanced computed tomography and direct observation during surgery. The tumor was successfully excised under emergency operation and was diagnosed as malignant fibrous histiocytoma after histologic examination. The patient had been free of symptoms for several months, but died of rapidly progressive congestive heart failure 6 months after surgery.

CASE REPORT

A 61-year-old woman was admitted to the Ishikawa Prefectural Central Hospital on December 2, 1980 with chief complaints of intermittent precordial discomfort and transient atrial fibrillation since November 27, 1980 and an episode of unconsciousness on November 30. She also complained of progressive fatigability and exertional dyspnea.

On admission, her pulse rate was 98/min and regular. Her blood pressure was 50/20 mmHg. Auscultation revealed a grade 2/6 low pitched mid-diastolic murmur at the apical region and moist rales in both lung fields. There was no evidence of hepatosplenomegaly, ascites or peripheral edema.

Her electrocardiograms showed sinus tachycardia, left atrial load and non-specific ST-T changes. Chest X-rays showed cardiomegaly with a cardiothoracic ratio of 59%, severe pulmonary congestion and a calcified abnormal shadow in her cardiac silhouette. Laboratory studies showed a hemoglobin content of 14.2%, a white blood cell count of 14500/mm³, an erythrocyte sedimentation rate of 72 mm (one hour), C-reactive protein of 4+, glutamic oxalacetic transaminase of 137u (normal: 0~38), glutamic pyruvic transaminase of 50 u (normal: 4~35), lactate dehydrogenase of 589 u (normal: 80~300) and creatine phosphokinase of 88 u (normal: 5~50).

M-mode echocardiography suggested the presence of a left atrial tumor: abnormal echoes behind the anterior mitral valve leaflet during ventricular diastole (Fig. 1-a). Cross-sectional echocardiography also showed a pendular motion of the tumor between the left atrium and the left ventricle from ventricular systole through diastole (Fig. 1-b).

Contrast-enhanced computed tomography was done at heart level 12 sec after the initiation of the bolus injection, by hand, of 30 ml of diatrizoate (Angiografin®) into the femoral vein to estimate the size and location of the tumor.

Keyword:
- Sarcoma
- Malignant fibrous histiocytoma
- Cardiac tumor
- Computed tomography

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Fig. 1. Echocardiogram before surgery. (a): An M-mode echocardiogram shows abnormal echoes below the anterior mitral valve during ventricular diastole. (b): Cross-sectional echocardiograms show a tumor from the left atrium to the left ventricle, which is attached to the posterior wall of the left atrium and slips into the ventricular cavity during ventricular diastole.

(Fig. 2). A calcified abnormal mass with a radiolucent halo was 2.5 cm in diameter and was located at the left posterolateral wall of the enlarged left atrium.

Hemodynamic measurements were done using a Swan-Ganz catheter. The pressure of the right atrium was 7 mmHg, the right ventricle 53/4, the pulmonary artery 57/21 and the pulmonary capillary wedge pressure 25. The cardiac index was 2.77 L/min/m². Left heart cineangiograms by injecting a contrast medium into the main pulmonary artery showed a large filling defect in the left atrium which prolapsed into the left ventricle through the mitral orifice during ventricular diastole.

Emergency operation was decided and left atriotomy was carried out under cardiopulmonary bypass. A lobulated, soft, yellowish-white tumor, weighing 50 g, occupied most of the left atrial chamber. The tumor was attached to the posterior wall of the left atrium by a partially calcified thick stalk. The tumor was excised from its attachments by blunt dissection and diagnosed histologically as a malignant fibrous histiocytoma (Fig. 3-a, b): the micrograph showed a storiform pattern of spindle cells, atypical, bizarre mitotic figures and multinucleated giant cells. PAS stains showed no glycogen in any type of tumor cells. No muscle type cross-striations were identified on hematoxylin and eosin or PTAH stains.

Her postoperative course was uneventful except for transient episodes of supraventricular tachycardia. No radiation or anti-cancer drugs were given. Five months after surgery the patient was rehospitalized because of heart failure, pleural effusion and pericardial effusion. Contrast-enhanced computed tomography demon-
Fig. 2. Computed tomography scan obtained after a bolus intravenous injection of a contrast medium. A calcified abnormal tumor with radiolucent halo is visualized in the left atrium. RV = right ventricle, RA = right atrium, LV = left ventricle, LA = left atrium

Fig. 3-a. Storiform pattern of spindle cells is found in the excised tumor. x100

Strated a large mass in the right ventricle (Fig. 4). She died of heart failure 6 months after surgery.

Autopsy was performed 2 hours after death. The heart weighed 600g. A tumor arising from the anterior wall of the right ventricle and occupying most of the right ventricle was found (Fig. 5). No tumors were found in other cardiac chambers and no metastasis was noted outside the heart. The histological picture was identical to that of the tumor excised from the left atrium.

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DISCUSSION

Arrhythmias and intractable and rapidly progressive heart failure are common features of malignant primary cardiac tumors1-4. The interval between the onset of the symptoms and death varies from a few weeks to 2 years, but most often it is only few weeks. Therefore, accurate diagnosis during life is difficult. In recent years echocardiography has been used as the preferred noninvasive method for the identification of cardiac tumors6,7. Cross-sectional
echocardiography shows the location, attachment and the size of the tumor with particular clarity. Myxomas account for more than half of all primary cardiac tumors and they are located in the atria in 95%, usually in the left atrium. We considered this tumor a left atrial myxoma at first, but cross-sectional echocardiography and computed tomography revealed that it arose from the posterior wall of the left atrium rather than from the interatrial septum. Left atrial myxomas usually arise from the left atrial septum at the level of the fossa ovalis. Contrast-enhanced computed tomography, which was obtained following a bolus injection of intravascular contrast medium, has been shown to be also useful in detecting lesions in the heart.

An emergency operation was performed because of the occurrence of ball-valve blockade phenomenon, syncope and hypotension. Histological examination of the resected material showed the findings of a malignant fibrous histiocytoma, one of the cardiac sarcomas. About 25% of all primary cardiac tumors are malignant and sarcoma is the most common. The types of sarcomas of the heart reported previously include hemangioendothelial sarcomas, rhabdomyosarcomas, angiosarcomas, fibrosarcomas, fibromyxosarcomas, myxosarcomas and lymphosarcomas. In 1978 Shah et al. have reported the first case of malignant fibrous histiocytoma of the heart and in 1980 Hamada et al. have reported the second case, in which the tumor arose from the interatrial septum of the left atrium and from the posterior wall of the left atrium. The present case is the third case reported. Sarcomas of the heart usually occur in the right side of the heart but these 3 cases of malignant fibrous histiocytoma originated in the left atrium. Weiss et al. reviewed the clinicopathological findings in 200 cases of malignant fibrous histiocytoma by follow-up studies. This tumor typically involves the deep fascia or skeletal muscle of the extremities or retroperitoneum. Metastasis occurs most frequently to the lung (80%) and lymphnodes (32%). Tumors, which are small and superficially located, or have a prominent inflammatory component, metastasize less frequently than larger and more deeply located tumors. In our case, the tumor with the typical histologic features of malignant fibrous histiocytoma, such as a storiform pattern of histiocytic cells, cellular atypism, bizarre neoplastic cells and multinucleated giant cells, was at first noted only in the left atrium on echocardiograms, on computed tomograms and during surgery. The tumor in the right ventricle was not thought to be a metastasis, but was considered one of the multiple cardiac lesions, because it arose from the anterior wall of the right ventricle while the tumor of the left.
atrium arose from the posterior wall. No tumor was found in the lungs and no recurrence was noted at the operation area in the left atrium. Soule et al.\textsuperscript{21} have reported that recurrence of tumor after local excision occurred in 73\% of the patients with malignant fibrohistiocytic tumors, and that regional lymphnode metastases, lung metastases and widespread metastases were found.

No therapy has been effective, although radiation and chemotherapy have been tried with minimal success. All patients in one series\textsuperscript{20} died within 2 years from the onset of symptoms.

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