Malignant Fibrous Histiocytoma of the Heart: Case Report and Review of 46 Cases in the Literature

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Abstract

A rare case of cardiac malignant fibrous histiocytoma (MFH) is reported. A 55-year-old woman complained of palpitation due to atrial fibrillation. Echocardiography, magnetic resonance imaging, and angiography demonstrated a tumor arising from the posterior wall of the left atrium. At surgery, the tumor was almost entirely resected and histologically defined as MFH. Neither chemotherapy nor irradiation was administered. Echocardiography revealed a local recurrence two months after the surgery and the patient died of advanced cachexy and heart failure 2 years later. The details of this case are presented with a review of the literature.

Key words: sarcoma, malignancy, left atrium

Introduction

We report a case of cardiac malignant fibrous histiocytoma (MFH) and review 46 other reported cases in order to clarify the nature of this rare heart disease (1–48). A primary heart tumor is a rare disease, the incidence of which has been reported in only 0.0017% of autopsies (49). More than sixty percent of primary heart tumors are benign and most are curable by surgery; the remaining cases are malignant (50). Regarding benign tumors, 40% are myxomas, 20% are rhabdomyomas, and the remaining 40% is composed of a variety of mesodermal tumors (51). Most malignant tumors are sarcomas, one-third of which are angiosarcomas (50). MFH is the most common soft-tissue sarcoma in adults (52). However, MFH has been rarely reported as a primary tumor occurring in the heart.

Case Report

The patient was 55 years old upon diagnosis and a previously healthy Japanese woman. She had a one-month history of palpitation and shortness of breath on exertion. An electrocardiogram obtained by her family doctor showed transient atrial fibrillation and she was subsequently transferred to our hospital. On admission, her pulse rate was 72 regular beats/min and blood pressure was 120/72 mmHg. There were no abnormal findings in the chest on auscultation. The abdomen was soft and flat and no evidence of hepatosplenomegaly, ascites, or peripheral edema was revealed. Superficial lymph nodes were not palpable by careful palpation and no tumorous lesions were detected in the legs or arms. Neurological examination did not show any abnormalities.

Laboratory studies disclosed the following values. The erythrocyte sedimentation rate was 7 and 20 mm per 1 and 2 hours, respectively. Hemoglobin concentration was 14.2 g/dl; the white blood cell count was 5,700/mm³; the platelet count was 21.3 × 10⁴/mm³; the C-reactive protein level was within the normal range; the aspartate aminotransferase level was 12 U (normal: <40 U); the alanine aminotransferase level was 9 U (normal: <30 U); and lactic dehydrogenase level was 417 U (normal: 100–400 U). Tissue polypeptide antigen was momentarily elevated during her hospitalization; other common tumor markers were not particularly abnormal.

The chest X-ray showed a cardio-thoracic ratio of 0.47 and mild enlargement of the left atrium. Neither pulmonary congestion nor a space occupying lesion was observed in the lung. The electrocardiogram upon admission showed regular sinus rhythm. Transient atrial fibrillation was also detected during the hospitalization, which was controllable with oral administration of disopyramide. The chest X-ray showed a cardio-thoracic ratio of 0.47 and mild enlargement of the left atrium. Neither pulmonary congestion nor a space occupying lesion was observed in the lung.

The electrocardiogram upon admission showed regular sinus rhythm. Transient atrial fibrillation was also detected during the hospitalization, which was controllable with oral administration of disopyramide. Two-dimensional echocardiography revealed a pedunculated tumor in the posterior wall of the left atrium (Fig. 1). M-mode scanning showed neither left ventricular dysfunction nor a protruding tumorous echo into the left ventricle during diastole. A broad-based tumor attached to the
MFH of the Heart

Figure 1. Transthoracic echocardiography revealed a left atrial mass (T). LA: left atrium, RA: right atrium, Ao: aorta.

Figure 2. Magnetic resonance images of the chest revealed a cardiac mass (T) at the posterior portion of the left atrium.

Figure 3. Gross (A) and microscopic (B, HE stain, ×200, Bar=100 μm) appearance of a heart tumor, diagnosed as malignant fibrous histiocytoma. The area of the neoplasm discloses a storiform pattern of growth, malignant spindle cell stroma, focal myxoid changes, and multinucleated giant cells.

posterior wall of the left atrium was also identified on magnetic resonance images (Fig. 2). Angiography showed a tumor stain with the feeding artery arising from the atrioventricular node artery of the right coronary artery. A feeding artery also originated from the atrial branch of the left coronary artery. No tumor was found in the other cardiac chambers and no metastasis was noted outside of the heart.

Upon cardiac surgery, the left atrium was opened and a sessile multinodular tumor was found to be attached to the posterior and lateral walls of the atrium. All parts of the visible tumor were excised, together with surrounding normal tissues. The resected tumor (5×4×3 cm) was yellowish-white and weighed 30 g (Fig. 3A). The micrograph showed a storiform pattern of spindle-shaped cells, bizarre mitotic figures, and multinucleated giant cells (Fig. 3B). Periodic acid-Schiff staining failed to demonstrate glycogen accumulation in the cytoplasm. With silver impregnation, individual tumor cells were enmeshed with reticulin fibers. Immunohistochemically, the tumor cells stained positive for both α-antichymotrypsin and α-antitrypsin. Staining for cytokeratin, S-100 protein, and myoglobin were completely negative. Accordingly, the tumor was finally diagnosed as ordinary MFH, storiform variant.

After the surgery, no radiation or anti-cancer drugs were administered. Except for bradycardia, the patient’s postoperative course was uneventful. The bradycardia was controlled with orally administered isoproterenol. Two months after the operation, echocardiography revealed a small irregular mass (ap-
proximately 1×1 cm large) in the left atrium. That mass gradually grew to 3×3 cm within a few months. Direct metastatic invasions into the chest wall and skin developed and the patient died of advanced cachexy and heart failure 2 years later. Postmortem examination was not performed in this patient.

**Discussion**

To understand the nature of cardiac MFH, we reviewed 46 cases in the literature in addition to the present case. The first reported case of cardiac MFH was published by Shah et al in 1978 (1). Some reports in the literature discussed the same patient from different points of view; thus we counted each independent patient as one case (1–48). Cited reports were limited to the English literature at least in the abstract.

**Age and gender**

The analysis of a total of 47 cases revealed that the youngest patient was 14 and the oldest was 77 years old. Twenty-nine patients (62%) were female and 18 were male (38%). The mean age of the female patients was 50.1 and that of the males was 42.1; the mean age of all cardiac MFH patients was 47.1. Cardiac MFH occurs with relatively equal frequency in elderly and young adults. Among the reported cases, patients younger than 50 years old accounted for 57% of this disease.

**Localization**

It has been reported that cardiac sarcomas occur more frequently in the right side of the heart, especially in the right atrium (53). However, cardiac MFH is most frequently located in the left atrium. Among the 47 reported cases, 38 tumors (81%) were located in the left atrium and usually attached to the posterior wall, as was the present case. In the remaining cases, the tumor was located in the pericardium (3 cases) (4, 13, 46), right ventricle and/or pulmonary valve (3 cases) (10, 30, 42), right atrium (1 case) (48), left ventricle (1 case; a multicentric tumor was also found in the left atrium) (47), or at the site of an atrial septal defect (1 case) (43). Cardiac MFH tends to metastasize in addition to local recurrence after surgery. Twenty cases in the literature indicated metastatic lesions. Most commonly organs involved in metastasis were brain (9 cases), and subsequently, lung (5 cases), bone (5 cases), and adrenal glands (4 cases); these results suggest hematogenous metastasis.

**Clinical feature**

In 43 cases of cardiac MFH, various symptoms were indicated; four reports did not provide information on symptoms. The most frequent symptom was difficulty in breathing. This symptom was seen in 32 cases (74%). Other chest symptoms such as palpitation or chest discomfort were observed in 17 cases (40%). The early symptoms of the present case also consisted of palpitation and shortness of breath. If an MFH tumor obstructs the inflow into the left side of the heart, the clinical signs resemble those of mitral stenosis. Upon auscultation of such cases, classic "tumor plop" as well as a variety of murmurs are audible (50). Some patients complained of hemiparesis, dysarthria, unconsciousness, headache, defects in the visual field, epigastralgia, or abdominal discomfort. It appears that tumor size, tumor locality, and/or presence of metastasis influence the symptoms and clinical findings. Taken together, the symptoms of cardiac MFH are nonspecific.

Echocardiography is now the imaging modality of choice for the noninvasive assessment of heart tumors. Computed tomography scans and magnetic resonance images are also useful (54). In the present case, the detected cardiac mass attached to the posterior wall of the left atrium was considered to be a cardiac sarcoma in terms of the findings of echocardiography and MRI. A definite diagnosis was made from the histological findings after surgery and the possibility of thrombus or benign tumors was finally ruled out. Out of 36 cases in the literature which were histologically examined in detail, 28 cases including the present case were storiform variant of ordinary MFH. The other type of MFH such as pleomorphic variant of ordinary MFH (3 cases), myxoid MFH (4 cases), and giant cell MFH (1 case) were also reported in the heart. Thus, the histological features of cardiac MFH are not different from those of extra cardiac MFH which were well described by Weiss and Enzinger (52).

**Treatment and prognosis**

Most patients with cardiac MFH undergo surgical resection. In spite of the high rates of local recurrence and metastasis, surgery is considered to be beneficial in order to obtain histological information and to ameliorate symptoms (55). In the present case, a definite histological diagnosis of MFH was necessary for the choice of therapeutic modalities and for the estimation of prognosis. No specific complications were reported in the surgical treatment of cardiac MFH. In our case, the bradyarrhythmia which occurred in the postoperative course was transient. Thus, we assumed that it was not associated with organic influence of the surgery or the involvement of MFH. However, we had no chance to examine the cardiac conducting system at autopsy.

It has been reported that the potential use of adjuvant chemotherapy and/or radiation therapy remains controversial (55, 56). For example, four patients treated with chemotherapy and/or radiation without surgery died within a year after diagnosis (7, 13, 24, 36). Thus, we did not add therapeutic regimens other than the surgery, which was based on informed consent.

Survival of cardiac MFH ranged from less than 1 month to 6 years after diagnosis. The mean survival period among the total 47 cases was about one year. One patient survived for 6 years and 5 additional surgeries were performed (38). A second operation was not indicated in our patient because of the tumor dissemination into the chest wall. It has been reported that heart transplantation offers good results for cardiac malignancies (45, 57), however the indication is considered to be strictly limited due to the highly metastatic potential.

In summary, we presented a rare case of cardiac MFH and reviewed 46 additional cases in order to clarify the nature of this rare disease. Development of an optimal therapy is desired in order to improve the prognosis and to enhance the quality of
life in patients with cardiac MFH.

References


