A Case of Primary Leiomyosarcoma of the Heart

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A 53-year-old male was hospitalized with complaints of cough, fever and backache. Two-dimensional echo-cardiography showed a pericardial echo-free space and a mass in the right atrium. Based on the MRI findings showing a pericardial mass originating from the atrial tumor, the final diagnosis of leiomyosarcoma was made by a percutaneous pericardial biopsy. Despite various therapies, the patient died after 3 wk. Because of its rareness (to date only 25 case reports), a premortem diagnosis of primary cardiac leiomyosarcoma is generally difficult. However, we feel that MRI and a subsequent biopsy is quite useful for making an early diagnosis of this disease.

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Key words: MRI, percutaneous cardiac biopsy

Introduction

Cardiac tumor is a very rare disease. In particular, primary malignant tumor of the heart is extremely rare, and thus establishment of the diagnosis while the patient is alive is difficult. For morphological diagnosis of the tumor, two-dimensional echocardiography (2DE) has been used as the first step, followed by CT scanning (including enhanced CT and dynamic CT) and cardio-angiography. Although these examinations can provide some information on the morphology of intracardiac lesions, it is very difficult to determine their properties (e.g., differentiation from thrombus and pericardial lesions). We describe here a case with primary leiomyosarcoma of the heart; for the diagnosis magnetic resonance imaging (MRI) was quite useful from a radiological point of view, and percutaneous biopsy of the tumor was possible.

Case Report

A 53-year-old male company employee had the chief complaints of backache, cough and fever. His past history and family history were not contributory. The patient began to suffer from backache and arthralgia of the left shoulder from mid-December 1988. He visited a neighborhood clinic in mid-January 1989 with the com-

plaint of cough and a fever of 37°C in addition to the above symptoms. Pleural effusion was noted on the left side of the chest X-ray film, thus the patient was admitted to our department on January 23, 1989. The medical check-up he had undergone 2 months before admission only disclosed slight enlargement of the cardiac shadow on the indirect chest X-ray film.

On admission, the patient had a body temperature of 38°C, pulse rate of 120/min and normal blood pressure. The palpebral conjunctiva was not anemic. Although weakened respiratory sounds were noted in the left thoracic region, no rale or heart murmur was heard. There was no hepato-splenomegaly or edema in the lower extremities. No cervical or axillary lymph nodes were palpable.

Laboratory examination on admission revealed anemia, markedly increased LDH, increased ESR, positive CRP, slightly decreased coagulability and an increase only in CA 125 as a tumor marker. Arterial blood gas analysis showed a decrease in oxygen concentration. The pleural effusion was bloody and exudative, negative for culture tests; cytological examination disclosed class II. Electrocardiography showed negative T waves in V₄₋₆ but was without low voltage in any lead. Chest X-ray examination revealed voluminous pleural effusion on the left side and irregular margin of the right second arch of the cardiac shadow (Fig. 1).

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Fig. 1. Chest radiograph on admission. Left pleural effusion and irregular margins of the right second arch of the cardiac shadow are shown.

A trochar® catheter was inserted into the left thorax to drain the pleural effusion. Repeated cytological examination of the effusion showed no evidence of malignant cells. 2DE revealed an echo-free space around the whole circumference of the pericardium and a mass lesion measuring about 3 cm in diameter in the right atrium. Chest CT scan showed no abnormal shadows in the lungs, but in the pericardial cavity there was an area of density slightly lower than that of the myocardium, and the structure in the right atrium was unclear (Fig. 2). On the basis of these findings, cardiac tumor was suspected, and chest MRI (Hitachi Medico MRP20, T₁-weighted image) was carried out (Fig. 3). In the pericardial cavity, tumor shadows, particularly thick on the side of the right atrium, surrounded the heart, and protruded into the right atrial cavity via the posterior wall of the right atrium. Since abdominal CT scanning and Ga scintigraphy showed no abnormalities in other organs, primary malignant tumor of the heart was suspected. On February 4, 1989, percutaneous mediastinal puncture was performed from the fourth intercostal space at the right edge of the sternum under the guidance of CT scan with reference to MR images. Malignant tumor tissues were collected. The patient's general condition worsened rapidly after admission. He complained of dyspnea with fever and tachycardia. Pleural effusion became massive around February 6, 1989. Blood pressure and urine volume decreased, which implied the occurrence of cardiac tamponade. The patient died of congestive heart failure on February 16 without receiving any radical treatment for the tumor because of rapid exacerbation.

Fig. 2. CT image of the heart showing low-density areas around the pericardium. The structure is unclear.

Fig. 3. MR image of the heart showing a polypoid lesion in the right atrium and a tumor involving the posterior wall of the right atrium.

Postmortem examination was carried out 2 h and 40 min after the patient's death. The tumor had invaded the whole circumference of the epicardium, involving the ascending aorta and superior vena cava. On the pleural side, there was scattered invasion centering on the lower lungs, showing continuous invasion into some areas of the pulmonary parenchyma. Lymph-nodes at the tracheal bifurcation and the bilateral pulmonary hilum were swollen, measuring 1 cm in diameter. A cross-section of the right atrium showed a tumor (measuring 2.5 cm) involving the entire layer of the posterior wall, suggesting a primary lesion. In the right atrial cavity, a polypoid mass measuring 3.5 cm in diameter was found (Fig. 4). Histological examination showed medullary tumor tissue mainly consisting of spindle-shaped cells showing atypia, polymorphism and degenerative necrosis with some proportion of giant cells. These cells had many hyaline droplets and/or vacuoles in the cytoplasm. Structures resembling those of smooth muscle tissue were also noted (Fig. 5). These structures were stained

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Fig. 4. Autopsy specimen of the heart with the right atrium and ventricle exposed. A tumor of the right atrial wall with an external nodule on the epicardial surface is seen.

red with Masson stain but did not stain with PTAH stain. The structures did not show positivity for Alcian blue stain or digestion with hyaluronidase, both of which are characteristic of mesothelioma. Immunohistochemical examination revealed that the tumor tissue was strongly

Fig. 5. Highly pleomorphic and atypical area showing some spindle-shaped cells.

positive for muscle actin or desmin which are specific for myofibrils, and also positive for vimentin, as were mesenchymal cells. However, the tumor tissue was negative for EMA, cytokeratin and factor 8, ruling out epithelial or vascular endothelial origin. These findings and the absence of any possible primary lesion in other organs led to the definitive diagnosis of primary leiomyosarcoma of the heart.

Table 1. Primary Leiomyosarcomas of the Heart

Reported case	Author	Year	Localization	Pericardial invasion	Diagnostic procedure
1	Weir (1)	1940	RA	⊕	auto
2	Friedman (2)	1944	LV	⊕	auto
3	Whorton (18)	1948	RA-LA		auto
4	Kennedy (3)	1967	RV	⊕	Xp→op
5	Shimada (4)	1967	RA-RV	⊕	auto
6	Sande (19)	1970	LA		auto
7	Hardin (20)	1974	LA		auto
8	Bearman (21)	1974	RV		auto
9	Burnett (22)	1975	RV		auto
10	Birman (9)	1976	LA		angio→op
11	Sasaki (23)	1976	LA		auto
12	Karasawa (24)	1981	LA		auto
13	Donovan (10)	1982	LA		auto
14	Donovan	1982	LA		angio→op
15	Reece (11)	1984	LA		op
16	Fine (7)	1985	RA		angio→op
17	Segesser (12)	1986	RA		UCG→CT→op
18	Klima (13)	1986	LA		op
19	Chaloupka (14)	1986	RA-LA		UCG→CT→op
20	Okabayashi (8)	1986	RV		UCG→CT→angio→op
21	Talley (15)	1986	RV		angio→op
22	Miki (5)	1987	LA	⊕	UCG→CT→angio→necro
23	Faught (6)	1988	RA	⊕	angio→op
24	Watanabe (16)	1989	RA		CT→MRI→auto
25	Lund (17)	1989	(details unknown)		
26	Takamizawa	1990	RA	⊕	UCG→MRI→bio

RA: right atrium, auto: autopsy, UCG: 2D echography, RV: right ventricle, Xp: chest X-ray, CT: CT scanning, LA: left atrium, op: operation, necro: necropsy, LV: left ventricle, angio: angiography, bio: biopsy

Discussion

Cardiac tumor is a rare disease, and is usually a result of metastasis from other organs. Of all primary cardiac tumors, 75% are benign lesions, consisting mainly of myxoma and rhabdomyoma in the order of frequency. Malignant tumors are very rare, most of them being sarcoma mainly comprising angiosarcoma, rhabdomyosarcoma, and fibrosarcoma. To our knowledge, only a total of 25 cases of primary cardiac leiomyosarcoma have been reported in the literature, including 5 Japanese cases (1–24) (Table 1). Six cases with epicardial involvement have been reported to date but it should be emphasized that these only showed partial invasion (1–6), where as the tumor in the present case grew in the cardiac cavity showing invasion into the whole circumference of the epicardium. The pattern of tumor growth was so unlike those previously reported that it was clinically suspected to be of pericardial origin.

In general, the prognosis of primary cardiac leiomyosarcoma is poor. Although some cases of prolonged survival after surgery and radiation therapy have been reported (7, 8), most cases were not operable or resulted in death soon after surgery. Surgical treatment is impossible in cases with involvement of the whole circumference of the heart, as in the present case. Here, because the progression of the disease was very rapid, neither irradiation or chemotherapy could be initiated in the present case. This suggests that early recognition of the tumor is necessary in order for sufficient treatment.

Premortem diagnosis was made in as few as 11 of the 25 cases previously reported in the literature; in all 11 cases, it was achieved through surgically resected tissue specimens (3, 6–15). In contrast, in the present case, MRI provided distinct tumor shadows which were not detectable by standard 2DE or CT scanning, and the definitive diagnosis was obtained by percutaneous mediastinal biopsy. Chaloupka et al noted the usefulness of CT scanning in their description 2 cases of cardiac tumor, but their investigation was limited to a comparison with echo-cardiography (14). The efficiency of MRI in the diagnosis of cardiac tumor has been reported by Watanabe et al (16) and Lund et al (17). They stated that this procedure is superior to any other currently available diagnostic methods in terms of the precision of the image, and unnecessary of the use of contrast media. In fact, MRI was much more useful than CT scanning in differentiating the lesion in the present case. Although the histological diagnosis was only made by biopsy, MRI provided an important clue to the clinical diagnosis of cardiac tumor, with images the same as those of intramyocardial or epicardial tumor, excluding thrombus or myxoma. We emphasize the usefulness of MRI especially at an early stage of the disease for the determination of the appropriate therapeutic regimen.

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