ACCURATE DIAGNOSIS OF METASTATIC CARDIAC LEIOMYOSARCOMA WITH INFUNDIBULAR STENOSIS AND CARDIAC TAMPONADE BY TRANSSESOPHAGEAL ECHOCARDIOGRAPHY AND Gd-DTPA MAGNETIC RESONANCE IMAGING

--- Report of a Case ---

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We present a patient with metastatic cardiac leiomyosarcoma that was diagnosed antemortem by transesophageal echocardiography (TEE) and ECG-gated magnetic resonance imaging with gadolinium diethylene triaminepentaacetic acid (Gd-DTPA MRI). TEE and Gd-DTPA MRI clearly revealed the morphological characteristics of infundibular stenosis and cardiac tamponade. Our patient has survived about 30 months after cardiac surgery. Since sarcoma is usually associated with a very poor prognosis, surgical treatment should be performed as soon as possible. TEE with color Doppler imaging and Gd-DTPA MRI are very useful diagnostic methods for determining the precise anatomical characterization of cardiac tumor, and these procedures are prerequisite to precise therapy.

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ALTHOUGH the incidence of cardiac metastasis ranges from 1.5 to 20.6% (average 6%) in autopsies on patients with malignant diseases, cardiac metastasis of leiomyosarcoma is extremely rare. With regard to the antemortem diagnosis of metastatic cardiac leiomyosarcoma, only 3 cases have been reported. In this report, we present a patient with metastatic cardiac leiomyosarcoma, which was clearly characterized by transesophageal echocardiography (TEE) and ECG-gated magnetic resonance imaging with gadolinium diethylene triaminepentaacetic acid (Gd-DTPA MRI).

CASE REPORT

In 1986, a 33-year-old woman underwent extensive excision of right femoral leiomyosarcoma. In October 1990, she was admitted to Ehime University Hospital for treatment of progressive exertional dyspnea. The blood pressure was 124/72 mmHg and heart rate was 88 beats/min. Cardiac examination revealed a grade 4/6 systolic ejection murmur with thrill at the left 2nd intercostal space.

Key words:
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Fig. 1. A: Transesophageal two-dimensional echocardiogram at the level of the right ventricular outflow tract. The cardiac mass (arrow) extensively occupied the right ventricular outflow tract. B: Transesophageal color Doppler echocardiogram at the same level as in Fig. 1-A. The cardiac mass (arrow) impeded hemokinesis. Ao = ascending aorta; RA = right atrium; RVOT = right ventricular outflow tract; PE = pericardial effusion.

Fig. 2. Gd-DTPA MRI at the level of the right ventricular outflow tract. The cardiac tumor (arrow) penetrated the anterior wall of the right ventricle and protruded into the anterior pericardial space. Ao = ascending aorta; PA = main pulmonary artery.

Electrocardiography showed sinus rhythm and non-specific ST-T wave changes in right precordial leads. Chest X-ray indicated cardiac enlargement and a coin lesion 1.5 cm in diameter in the left lung. Two-dimensional transthoracic echocardiography showed a large pericardial effusion and an undefined mass on the right ventricular outflow tract.

Cytologic examination of the pericardial fluid revealed class I cells.

Transsthoracic echocardiography was insufficient for imaging the entire mass because of narrow rib spaces. Therefore, TEE was performed using an Aloka SSD 870 imaging system with a 5 MHz transducer. TEE revealed that a 2.5 cm × 1.5 cm mass, which was mobile and attached at the anterior wall of the right ventricular outflow tract, produced severe infundibular stenosis (Fig. 1-A). Color-Doppler TEE clearly displayed a narrow flow area (shown in red in Fig. 1-B), and pulsed-Doppler TEE indicated that the pressure gradient of the infundibular stenosis was more than 20 mmHg. However, TEE could not detect a difference in tissue characterization between the mass and normal myocardium.

T1-weighted spin-echo MRI (1.5-T) was obtained after the administration of Gd-DTPA (0.1 mmol/kg) with an echo time (TE) of 12 msec, and a repetition time (TR) equal to the RR interval of the ECG. Gd-DTPA MRI clearly enhanced the homogeneous cardiac mass which penetrated the right ventricular anterior wall and protruded into the anterior pericardial space (Fig. 2).

Since medical treatment did not prevent clinical deterioration due to cardiac tamponade and right ventricular outflow stenosis, the patient underwent cardiac surgery on November 5, 1990. After the hemorrhagic fluid (400 ml) was removed from the pericardial sac, a 2.0 cm × 3.3 cm solid tumor and a 4.2 cm × 4.7 cm cystic tumor were found near the anterior wall of the right ventricular outflow tract (Fig. 3-A). The solid tumor penetrated the myocardium and protruded into, and outside of, the myocardium. These operative findings corresponded to those in TEE and MRI. A bloody cyst was connected to the solid tumor, but did not involve the myocardium. Slight bleeding from the cyst was also observed. These tumors and the left lung tumor were excised simultaneously. The solid tumors were pathologically identified as the metastatic leio
coma that had been removed 4 years previously (Fig. 3-B). The postoperative course is uneventful and the patient has survived about 30 months after the cardiac surgery.

**DISCUSSION**

Rosenblatt and Featherston\(^5\) were the first to report a case of metastatic cardiac leiomyosarcoma. Our report is the first case in which the morphological characteristics of metastatic cardiac leiomyosarcoma were accurately diagnosed by TEE and Gd-DTPA MRI methods. Cardiac metastasis of the tumor affects the right side of the heart more frequently than the left.\(^6\) Cardiac tumors can usually be examined more clearly by TEE than by transthoracic echocardiography. TEE is especially useful for identifying the tumor attachment point and precise morphological details, such as cysts? In our study, however, even TEE could not determine the exact distribution of the metabolic tumor or qualitatively characterize the tumor. In contrast, Gd-DTPA MRI could differentiate between intramural tumor and normal myocardium.

In our case, TEE clearly displayed the intracardiac morphology of metastatic cardiac leiomyosarcoma, but did not show the protruding bloody cystic tumor in the anterior pericardial space. This limitation of TEE might be based on the characteristics of the 5 MHz transducer, which has an inherently weak ability to resolve distinct images. Therefore, this TEE method is limited for differentiating intramural tumors from normal myocardium and for identifying cardiac extension of the tumor to adjacent structures.

MRI, on the other hand, can determine the more extensive vascular, intracardiac and extracardiac nature of cardiac tumors, provides important additional anatomic information that can not be obtained by echocardiography, as reported by Freedberg et al. \(^5\) However, according to previous MRI studies,\(^7-10\) T1-weighted MRI can not differentiate between intramural tumors and normal myocardium, and T2-weighted MRI degrades the image of the heart due to prolonged TE. In our study, Gd-DTPA MRI was able to differentiate tumors from normal myocardium, as reported by Funari et al.\(^10\) Gd-DTPA, as a paramagnetic contrast agent, results in T1 shortening and enhancement of the lesion on T1-weighted images. Gd-DTPA usually accumulates in higher concentrations in tissues which contain augmented vascularization or a substantial proportion of interstitial space and are thus preferentially enhances.\(^11\) This contrast agent can also provide information about the internal architecture, i.e., nonvascularized, necrotic or cystic areas, of a tumor.\(^9\) In our case, only Gd-DTPA MRI could clearly differentiate intramural tumor from normal myocardium. As compared with the operative findings, the tumor that was homogeneously enhanced by Gd-DTPA MRI seemed to correspond to the solid part of the tumor. Enhancement of the metastatic leiomyosarcoma with Gd-DTPA in our case is believed to be due to neovascularization\(^10,11\) or distinctive characteristics of the leiomyosarcoma. These findings represent a significant advantage over TEE in the assessment of intramural tumors. However, with regard to the mobile tumor in the outflow tract, TEE provided a superior image compared to MRI. Therefore, the combined use of TEE and Gd-DTPA MRI compensated each other and provided a great deal of information about the cardiac tumors.

In conclusion, TEE and MRI are useful for determining the precise anatomical characteristics of cardiac tumors that are not adequately defined by usual examination procedures.

**REFERENCES**

5. ROSENBLETT WH, FEATHERSTON WP: Metastatic leiomyosarcoma of the heart. Am J Cardiol 1960; 6: 672–675
7. MUGGE A, DANIEL WG, HAVERICH A, LIBHTLEN PR: Diagnosis of noninfective cardiac mass lesions by two-dimensional echocardiography: Comparison of the transthoracic and transesophageal approaches. *Circulation* 1991; 83: 70—78
8. FREEDBERG RS, KRONZON I, RUMANCIK WM, LIEBESTKM D: The contribution of magnetic resonance imaging to the evaluation of intracardiac tumors diagnosed by echocardiography.

*Circulation* 1988; 77: 96—103