Primary Cardiac Malignant Fibrous Histiocytoma in the Right Ventricular Infundibulum Treated With a Cavo-Pulmonary Shunt and Coronary Embolization

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A 51-year-old woman presented with progressive right ventricular infundibular wall thickening and outflow obstruction. She had had an aorto-coronary bypass for left main coronary artery disease 1 year after radiation therapy for left mammary cancer. Enhanced computed tomography showed a mass in the right ventricular free wall with no connection to the mediastinum; the tumor extended into the main pulmonary artery, but there was no other evidence of a primary or metastatic tumor. A biopsy specimen was obtained and based on the microscopic and immuno-histochemical findings (vimentin and Kp-1 positive) the diagnosis was primary cardiac malignant fibrous histiocytoma, which is very rare. A cavo-pulmonary artery connection lessened her symptoms, but embolization of the coronary artery to try and to reduce the mass had minimal effect. Four months after the tumor was diagnosed she died of extended pulmonary artery obstruction. (Jpn Circ J 2000; 64: 982–984)

Key Words: Cardiac tumor; Cavo-pulmonary artery connection; Malignant fibrous histiocytoma

Malignant fibrous histiocytoma (MFH) is a common soft tissue sarcoma in adults that mainly is treated by orthopedic surgeons. MFH of cardiac origin is very rare1–4 and right-sided cardiac MFH even more so. We report a patient with MFH in the right ventricular infundibulum who had a history of radiation therapy for breast cancer and who had had a coronary bypass operation 10 years earlier.

Case Report

A 51-year-old woman who had a 10-month history of exertional dyspnea was admitted to hospital. She had had mammary cancer and received radiation to the left chest wall and mediastinum 11 years before. One year later she experienced severe angina pectoris and coronary angiography revealed significant stenosis (90% reduction of lumen diameter) of the left main trunk. Subsequently she underwent a coronary bypass operation: left internal mammary artery graft to left descending artery and a saphenous vein graft to a posterolateral branch. Five years ago, oliguria, edema, ascites and exertional dyspnea developed; treatment with diuretics promptly lessened her symptoms.

Ten months before admission she complained of nocturnal dyspnea and chest discomfort. Auscultation revealed a new ejection murmur in the second left sternal border; an echocardiogram showed thickening of the right ventricular free wall and stenosis of the right ventricular outflow tract (RVOT). Right heart catheterization showed a pressure gradient of 22 mmHg between the RVOT and main pulmonary artery. Enhanced computed tomography (CT) showed that the thickening of the right ventricular free wall did not have a connection to the mediastinum. Gallium-67 radioisotope scanning provided no further information about the systemic extension. Five months later she experienced abdominal distention, epigastric discomfort, anorexia, and nausea. The exertional dyspnea increased, and 3 months later she could not take a bath. She was admitted to hospital in March 1999.

The physical examination revealed hypotension (BP 96/60 mmHg) and tachycardia (102 beats/min), neck vein distension, generalized mild edema, and ascites. A chest roentgenogram showed bilateral small pleural effusions. The routine laboratory examination showed only mild liver dysfunction. Electrocardiography showed complete right bundle branch block with right ventricular hypertrophy (Fig 1).

An echocardiogram showed advanced obstruction of the RVOT by an amorphous mass that extended into the main pulmonary artery. Enhanced CT pinpointed the location of the right ventricular free wall thickening as the anterior and outflow portion of the right ventricle, and CT showed extension of the mass into the main pulmonary trunk (Fig 2).

The biopsy specimen obtained from the mass in the RVOT comprised spindle-shaped polymorphic cells with highly atypical nuclei proliferating in a dense array (Fig 3). Immunohistochemically these cells were positive for vimentin (interstitial cell marker) and Kp-1 (histiocyte cell marker). Other immunohistochemical cell markers (endothelial (Factor VIII), epithelial (CK22, EMA), lymphatic (LCA), myogenic (Desmin, HHF35, alpha SMA), and neurogenic (S 100) cells) were all negative. The tumor...
Fig. 1. Electrocardiogram of August 1998 showing vertical axis and T wave inversion in the anterior chest leads. Electrocardiogram of March 1999 showing right axis deviation and complete right bundle branch block with right ventricular hypertrophy.

Fig. 2. Enhanced CT scans showing the right ventricular outflow mass and an intra-pulmonary artery mass. Three dimensional CT (above right) also shows severe outflow obstruction.

Coronary arterial embolization was attempted in April to prevent further tumor proliferation. Two platinum coils were inserted in the proximal portion of the right coronary artery to obstruct the feeding arteries that were originating from there (Fig. 4). Ventricular function was not impaired because the right coronary artery only supplied a small area of the left ventricle. Low-grade chest pain, fever and an elevation of creatine kinase (to 1,848 IU/L; normal, 165 IU/L) were observed, and reduction of the mass was minimal.

Four months after TCPA she died of sustained hypotension resulting from tumor invasion of the left main pulmonary artery and coagulation abnormality. Autopsy was not performed.
Discussion

Primary sarcomas of the heart are rare, and MFH makes up only 2–3% of all cardiac tumors. A left atrium origin for primary cardiac MFH is relatively frequent, but right-sided MFH is considered to be extremely rare. The diagnosis and selection of treatment for this rare malignant tumor in the ventricular wall are difficult.

The present patient had MFH in the infundibulum of the right ventricle. It was very difficult to diagnose her newly developed infundibular stenosis as the early phase of this rare primary cardiac tumor using only pathological diagnostic modalities. From first suspicion to definite diagnosis took 7 months, and the patient died 4 months after the tumor was correctly diagnosed. In all but one report, right-sided MFH occupied the infundibulum of the right ventricle; although metastatic MFH to the right ventricle has been reported. The patient reported here showed no evidence of an extracardiac primary site or distant metastasis during the clinical course of her illness.

After the tumor was diagnosed, palliative shunt surgery remarkably lessened her symptoms and did prolong her life. The reported treatment of cardiac MFH consists of maximal surgical resection followed by conventional fractionated radiation therapy and chemotherapy as the second-line salvage therapy. In the present case, the graphical findings of tumor extension into the interventricular septum ruled out surgical resection, so embolization of the coronary artery was attempted to reduce the mass, but had minimal effect. Cardiac transplantation has been recently reported as an alternative.

The patient had received high-dose radiation therapy for mammary cancer 11 years earlier and it is possible that the left main coronary stenosis and rare cardiac sarcoma resulted from this. Such a possibility should be kept in mind when selecting treatment for thoracic cancer and the use of late bracing coronary brachytherapy be considered.

References


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