Hypervascular Metastatic Cardiac Tumors: An Unknown Cause of Mitral Valve Prolapse

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A 40-yr-old woman with alveolar soft-part sarcoma and multiple hypervascular cardiac tumors involving both left and right ventricles is reported. Papillary muscle dysfunction and mitral valve prolapse with mitral regurgitation were caused by the largest tumor in the left ventricle.

(Internal Medicine 31: 78–81, 1992)

Key words: alveolar soft-part sarcoma, echocardiography, coronary arteriography

Introduction

Cardiac metastases are not an uncommon finding in autopsies of patients who have died from malignant neoplasms. The origin of most metastatic cardiac tumors is melanoma, leukemia and carcinoma (1). Alveolar soft-part sarcoma is a rare, slow-growing neoplasm (2). The tumor tends to metastasize to the lung and the brain. However, only one case of cardiac metastasis diagnosed during life has been reported by Stark et al (3).

We report a patient with alveolar soft-part sarcoma. She was found to have multiple hypervascular cardiac tumors involving both left and right ventricles. She also had mitral valve prolapse caused by a left ventricular tumor. Papillary muscle dysfunction due to neoplasm is not to be a cause of mitral valve prolapse.

Case Report

The patient, a 40-yr-old woman, was referred to our department for evaluation of a heart murmur in August 1986. She had a past history of alveolar soft-part sarcoma in her left thigh. The tumor was resected and irradiation was applied in May 1973. Then, she was found to have multiple lung tumors, which were resected in July 1984. The histological study confirmed the diagnosis of alveolar soft-part sarcoma. In 1985, she had paresis of the left leg and multiple brain tumors were found. The largest brain tumor was resected and she received cranial irradiation in December 1985. Small tumors in the lungs and the brain still remained.

On admission to our department (August 1986), blood pressure was 130/82mmHg and pulse rate was 80 beats/min with regular rhythm. The jugular venous pressure was not elevated. A grade 3/6 systolic regurgitant murmur was heard at the apex of the heart. Respiratory sound was normal. Slight left hemiparesis was present.

Laboratory data were within normal limits. Twelve-lead electrocardiogram showed sinus rhythm with T-wave inversion in the inferior leads. The chest X-ray film showed small coin-like lesions in both lung fields. Heart size and lung vasculature were normal.

Two-dimensional echocardiography revealed a large mass in the left ventricular cavity attached to the inferior wall appearing to take the place of the posterior papillary muscle (Fig. 1A). The posterior mitral valve leaflet prolapsed to the left atrium in late systole with mitral regurgitation (Fig. 1B).

Cardiac catheterization revealed normal intracardiac pressures and cardiac output. Coronary arteriography showed multiple hypervascular tumors (Fig. 2A–C). The largest tumor on echocardiography was predominantly supplied by the posterior descending artery. There were many other tumors in the left ventricle. A small tumor was also found in the right ventricle (Fig. 2C).

Because she had no symptoms associated with the heart disease and radical operation could not be per-
formed, we decided to observe her under medical care without surgical intervention. She had neither symptoms nor signs until her sudden death at her home in December 1986. Autopsy could not be performed.

Discussion

Cardiac metastasis is relatively common. Autopsy studies show heart involvement in about 20% of the patients with fatal malignant neoplasms (1). Most cardiac metastases originate in carcinomas, melanomas, leukemias or lymphomas (4).

Alveolar soft-part sarcoma is a soft tissue sarcoma, which was first defined and named by Christopherson et al in 1952 (5). The tumor is an uncommon neoplasm and usually presents as a slow-growing painless mass without functional impairment. It has a characteristic histologic pattern of granular tumor cells arranged in alveolar nests surrounded by thin-walled vascular sinusoids. The tumor, resected from the present patient’s lung in 1984, showed such features (Fig. 3). Although some other tumors, including renal cell carcinoma, paraganglioma and granular cell tumor, often resemble alveolar soft-part sarcoma, the clinical features can be useful for differential diagnosis (2, 5). The tumor occurs principally in patients between 15 and 35 yr of age. The occurrence in female patients is greater than in males. In adults, it occurs predominantly in the lower extremities, especially the anterior portion of the thigh. The tumor tends to metastasize in the lung and brain. Its histogenesis is still unknown though four theories have been proposed regarding the nature of alveolar soft-part sarcoma; angio-reninoma, a form of granular cell myoblastoma, paraganglioma or muscle tumor (6).

As diagnostic procedures improve, cardiac tumors can be found more often during life, especially by echocardiography (7). However, on alveolar soft-part sarcoma, only one case of cardiac metastasis can be found in the literature, that reported by Stark et al. That patient had a right ventricular metastasis of alveolar soft-part sarcoma, which was successfully resected (3). We believe that the present patient was possibly the second case of cardiac metastasis. Although we could not perform a histologic study on the cardiac tumors, her clinical history suggested that they were alveolar soft-part sarcoma. The coronary arteriogram supported this diagnosis because it is known
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Fig. 2. Coronary arteriogram showing multiple hypervascular tumors (A–C). The largest tumor was predominantly supplied by the posterior descending artery (A; arrows). A small tumor in the right ventricle was also found (C; arrows).

Fig. 3. Photomicrograph (×100) of the lung tumor resected from the patient showing the characteristic features of alveolar soft-part sarcoma.

that arteriograms of such tumors show hypervascularity (8, 9). There were multiple discrete tumors in both left and right ventricles of her heart. Their distribution implied that they might have metastasized through coronary arteries.

The present case had mitral valve prolapse with mitral regurgitation. Her largest cardiac tumor involved the posterior papillary muscle. Therefore, we consider that the tumor might have caused papillary muscle dysfunction and mitral valve prolapse. Metastatic tumors sometimes affect cardiac valves in various ways, including direct invasion, compression, or release of humoral factors (10). However, mitral valve prolapse due to papillary muscle dysfunction caused by tumor invasion is not known as a mechanism of valvular involvement. Then, the present case presents a possible additional manner of valvular dysfunction caused by the tumor, which then leads to mitral valve prolapse.

In the present case the cardiac tumors were first found by echocardiography and mitral valve prolapse was also diagnosed. Echocardiography has been recognized as a valuable diagnostic method for cardiac tumors (7). However, it should be noted that not all tumors shown on coronary arteriography are detectable by echocardiography.

Acknowledgments: The authors thank Dr. Kazuaki Yamada of the Tachikawa National Hospital, for the proposition of photomicrograph of the lung tumor resected from the patient.

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