Primary Cardiac Angiosarcoma of the Right Atrium Undiagnosed by Transvenous Endocardial Tumor Biopsy

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A 50-year-old man was admitted with acute pericarditis. Echocardiography demonstrated a large mass on the right atrial free wall along with a pericardial effusion. We performed transvenous biopsy of this mass under transesophageal echocardiographic guidance. Though the biotome obtained the mass, the pathological findings were of organized thrombus. Two weeks later, a new precordial mass appeared around the left third rib and was suspected to be a metastasis. Incisional biopsy of this mass gave the diagnosis of angiosarcoma.

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Introduction

Primary cardiac tumors are rare. Among these tumors, less than 30% are malignant and one-third of the malignant lesions are angiosarcomas (1). It is difficult to diagnose primary cardiac angiosarcoma and the diagnosis is almost always at autopsy except when open biopsy can be done (2–4). In this case, we attempted diagnosis by transvenous biopsy under transesophageal echocardiographic guidance.

Case Report

A 50-year-old man had no medical problems until April 1997, when he visited his local hospital complaining of shortness of breath and orthopnea. At that time, his temperature was 38°C, C-reactive protein (CRP) was 13.9 mg/dl, erythrocyte sedimentation rate (ESR) was 93 mm/h, cardiothoracic ratio (CTR) was 75% on the chest X-ray film, and a pericardial effusion was found by echocardiography. He was admitted to this hospital with a diagnosis of acute pericarditis. Although his pericarditis improved, transthoracic echocardiography (TTE) demonstrated a large mass on the free wall of the right atrium with an irregular surface and poor movement. These findings suggested that the mass was malignant. So he was admitted to our hospital for examination of this tumor.

On physical examination, no abnormalities were found and lymphadenopathy was not detected. Laboratory tests showed no abnormalities except for an elevated ESR (79 mm/h). The chest X-ray film showed a CTR of 46%. The electrocardiogram showed normal sinus rhythm and was within the normal limits including the ST-T changes. Transesophageal echocardiography (TEE) demonstrated a large mass (4.6×4.3 cm) on the free wall of the right atrium extending from just proximal to the superior vena cava down to the neighborhood of the tricuspid valve (Fig. 1). Hemodynamic data were normal. Computed tomography (CT), magnetic resonance imaging (MRI), and Ga citrate scan did not show evidence of metastasis. Then we performed cardiac catheterization. On inferior vena cavography, a large right atrial tumor was detected. The feeding arteries of this tumor originated from both the right coronary artery and the right internal mammary artery. We tried transvenous biopsy under TTE guidance, but could only get normal right atrial myocardial and fibrous tissue. We then tried transvenous biopsy under TEE guidance. TEE clearly visualized the biotome tip in contact with the tumor surface, and we obtained four specimens. The tumor was very hard and could be clearly observed to vibrate when in contact with the biotome tip. But the specimen obtained was mainly fibrous tissue which was found to organized thrombi by light microscopy. There were no findings suggestive of a malignant tumor. We discussed this case with surgeons and decided to try total tumorectomy. However, a new skin mass was found on his left third rib two weeks later. CT and MRI (Fig. 2) clearly revealed irregular signals in the mass after contrast enhancement. These findings...
were similar to those of the right atrial tumor. We then performed incisional biopsy of the skin tumor. On hematoxylin-eosin stained sections, spindle-shaped tumor cells were seen composing small vascular structures with fibrous areas. The tumor showed strong mitotic activity. Most of these cells were positive for both CD31 and factor VIII-related antigen (Fig. 3). Therefore, we finally diagnosed this tumor as angiosarcoma.

The patient developed lung and brain metastases, and then he died of hemorrhage from the lung metastasis about 8 months after diagnosis. At autopsy, the right atrium and pericardium were involved by a huge mass and the tumor invaded the chest wall. However, the right atrial endocardium was covered with thick thrombus and we did not detect any tumor in the endocardium microscopically. Histologic examination showed that tumor did not invade the endocardial side and the part of the endocardium covered with fibrous tissue (Fig. 4).

**Discussion**

Cardiac angiosarcoma is located in the right atrium in more than 70% of cases. Invasion of the vena cava, tricuspid valve, and pericardium is common, while involvement of the interatrial septum or the pulmonary artery is rare (2, 4).

The clinical presentation is nonspecific. Initial signs are often caused by pericarditis or the obstruction of the inferior vena cava, superior vena cava, or tricuspid valve. On echocardiography, cardiac angiosarcoma is often lobulated, focally necrotic, or hemorrhagic. Findings are the same with CT and MRI, but these characteristics are not specific to cardiac angiosarcoma. Thus, the final diagnosis must depend on tumor biopsy (2, 4).

A biopsy diagnosis of angiosarcoma was established ante-mortem in 50% of 46 cases in a 1986 review (2). Hermann et al reported that the mean survival time of patients who only had biopsy was 9 months, while that for patients having surgical excision was 10 months (including eight survivors for 12 to 36 months). Chemotherapy and irradiation were reported not to improve survival (4). Recently, interleukin-2 (IL-2) therapy was reported to be effective for angiosarcoma of the skin, but its value is not known for cardiac angiosarcoma. Total resection of the tumor may improve the prognosis in a few selected cases of cardiac angiosarcoma. Though there are some case reports that transvenous biopsy under TEE guidance was a useful diagnostic procedure (5–7), other authors have stated that the tissues obtained from transvenous biopsy show only false-negative findings. Transvenous biopsy is less invasive than open biopsy and surgical resection, but it often false-negative in cardiac angiosarcoma. The reason for this is not known. In our patient, the surface of the right atrial endocardium was irregular on imaging and endocardial continuity with the tumor was unclear. Thus, we thought that tumor cells could probably be obtained by endocardial biopsy. However, autopsy findings showed that the tumor originated from the outer portion of the right atrium or the pericardium, and no tumor cells were detected in the endocardium which was covered with thick thrombus. Hattori et al reported that six out of the 31 angiosarcomas diagnosed from 1945 to 1988 in Japan originated from the pericardium and autopsy showed that the endocardium was intact (8). Based on these findings, we probably could not diagnose the tumor by endocardial biopsy because it originated from the outer portion of the right atrium or the pericardium and the endocardium was covered with organized thrombus. Thus, endocardial biopsy has limitations for diagnosis in cases such as the present case. When a malignant tumor is suspected from imaging findings but transvenous endocardial biopsy shows no malignancy, incisional biopsy or total tumorectomy might be important diagnostic procedures. In addition, total tumorectomy might be
**Primary Cardiac Angiosarcoma**

**Figure 2.** T1-weighted magnetic resonance imaging (MRI) demonstrated a large low intensity mass (arrow) in the right atrium. The mass invaded the superior vena cava (SVC) and tricuspid valve. The dilatation of SVC was not found.

**Figure 3.** Histologic features of the skin tumor obtained by incisional biopsy. A) HE stain, ×20, B) HE stain, ×400. Spindle-shaped tumor cells form small vascular structures with fibrous areas. The tumor shows high mitotic activity (arrows). C) anti-CD-31, ×400. Most of the cells are positive for CD31, an endothelial marker, by immunohistochemistry (brownish staining).
Figure 4. Autopsy findings: The right atrial tumor and subendocardial myocardium are respectively shown in panel A (HE stain, x20) and panel B (Masson’s trichrome stain, x20). The tumor tissue (*) is seen on the outer aspect. The atrial myocardium (arrows) is covered with organized thrombi (Th). C (HE stain, x100); Endocardial view of panel A.

useful to improve the prognosis of selected patients with cardiac angiosarcoma in whom curability is expected.

References