Primary Cardiac Angiosarcoma Presenting with Pericardial Effusion and Cardiogenic Shock — Two Cases Report

Ying-Chieh Wang, Hin-Wing Law, Ai-Hsian Li and Shu-Hsun Chu

Primary tumors of the heart occur with an incidence rate of 0.001% to 0.28%. Sarcomas comprise the largest proportion of primary malignant cardiac tumors. Angiosarcoma is the most common malignant tumor of the heart. This tumor is found most often in the right atrium and frequently extends to the pericardium, vena cava, or tricuspid valve, causing tamponade and/or heart inflow obstruction. We present 2 cases of angiosarcoma with cardiogenic shock due to massive pericardial effusion and obstruction to blood flow in the tricuspid valve area. After having resection of the original tumors, 1 patient needed to have pacemaker implantation due to sinus node dysfunction with junctional bradycardia, while the other patient had recurrent cardiac tumor and lung metastases. Despite diagnosis by noninvasive imaging procedures and aggressive early surgical intervention, survival in both cases was less than few months. Thus, optimal therapy is unclear.

Key Words: Angiosarcoma • Cardiogenic shock • Pericardial effusion

INTRODUCTION

Angiosarcoma is the most common primary malignant tumor of the heart. It commonly occurs in middle-aged men. The clinical pictures are often non-specific, which is related to the location of the tumor and involved area of the heart, with or without metastasis. Pericardial effusion is a common manifestation. There are many non-invasive methods (echocardiography, magnetic resonance imaging, computed tomography) to diagnose cardiac angiosarcoma. But, in most cases, the tumor has already extended widely at the time of presentation, with pericardial, vena cava, and valvular involvement rendering most cases inoperable and rapidly fatal.

The treatment protocols for cardiac angiosarcomas include combination of surgery and radiation and/or chemotherapy. We report 2 cases of right atrial angiosarcoma with the presence of massive pericardial effusion and cardiogenic shock.

CASE 1

Prior to his admission, this 37-year-old man had had chest pain and shortness of breath for 1 week. On July 8, 1999, he experienced sudden loss of consciousness while on the way to the lavatory. He was sent to our emergency room and then admitted to our cardiovascular ward. On physical examination, the patient was acutely ill-looking, with blood pressure of 90/60 mmHg and pulse rate of 112 beats per minute. The jugular vein was engorged. Breath sounds in both lung fields were clear and without crackles. Irregular heart beats were noted on auscultation.

Twelve-lead surface electrocardiography (ECG) showed atrial fibrillation, complete right bundle block and general low voltage. Cardiac enzyme markers were within normal limits. A chest X-ray showed marked enlargement of the cardiac silhouette. Chest computed tomography (CT) suggested pericardial effusion. Transthoracic echocardiography (TTE) (Figure 1 A,B) revealed a huge...
tumor mass on the right atrium (RA) which protruded to RV occasionally. Large amount of pericardial effusion with congested inferior vena cava and hepatic vein was also found. Cardiac tamponade was considered. Disturbed blood flow from right atrium to right ventricle in the tricuspid orifice was shown. The patient’s blood pressure dropped and consciousness changed soon.

He was transferred to another cardiovascular center for emergency operation.

According to their operation report, right atrial tumor invaded the tricuspid valvular annulus, SA node and free wall. There was active bleeding from RA free wall. Unfortunately, it was not possible to totally remove the tumor and bovine pericardium was used for reconstruction of RA. The histopathological finding confirmed a diagnosis of angiosarcoma. The whole-body bone scan was negative on July 15, 1999. A pacemaker was implanted on July 19, 2002 for sinus node dysfunction with junctional bradycardia. Chemotherapy was not done. The patient died 3 months after being discharged.

CASE 2

A 62-year-old male was diagnosed as constrictive pericarditis, ruling out of tuberculous pericarditis with pericardial effusion on November 16, 2001 in another hospital. Pericardium biopsy showed chronic inflammation on November 17, 2001.

He had received 4 anti-tuberculosis drug treatments for 2 months. During the course of medication there, 2-dimensional echocardiogram showed a RA mass (size 5.3 × 3.3 cm) on November 23, 2001. However, 2 months later, he suffered from bilateral leg edema, decreased urine amount and orthopnea. There was no fever nor chills. Transesophageal echocardiogram (TEE) found the mass had become larger (size 7.26 × 5.75 cm) and located in RA roof, protruding into the superior vena cava on January 21, 2002. Large amount of pericardial effusion with signs of cardiac tamponade were found.

On January 24, 2002, the patient was transferred to our hospital for operation. Physical examination on the day of operation displayed the blood pressure at 91/51 mmHg and heart rate at 110 beats per minute. A cardiac tumor (size 7 × 8 × 5 cm) was excised. Pathological finding showed a picture of angiosarcoma. There was no bony metastasis in whole-body bone scan examination. He was discharged on January 31, 2002. Magnetic resonance imaging (MRI) study on March 21, 2002 revealed no residual tumor found in cardiac wall or cardiac cavity with homogenous enhanced signal intensity across cardiac wall. Unfortunately, the follow-up MRI on June 13, 2002, showed a recurrent cardiac tumor filling the main chamber of RA and part of tumor protruding into RV. Lung perfusion study and pulmonary function test showed lung metastases. Chemotherapy was started on June 26, 2002. Transthoracic echocardiography on July 3, 2002 (Figure 2) showed massive pericardial effusion with a larger tumor mass on RA, protruding into RV in diastole. Febrile neutropenia pneumonia was found during the course of treatment. The patient’s general condition became worse. He expired on

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**Figure 1.** Transthoracic echocardiography of case 1. (A) parasternal long-axis view showing a tumor mass (TM) located in the dilated right atrium (RA) and large amount of pericardial effusion (PE). (B) apical 4-chamber view revealed a huge tumor mass in RA with obstruction of blood flow to right ventricle (RV).
July 20, 2002.

DISCUSSION

Cardiac angiosarcoma is a very rare malignant tumor of the heart, which is found most often in the right atrium. It occurs in males more often than in females in a ratio of 2 to 3:1, in the age group of 20-50 years. The diagnosis of cardiac tumor is complicated by the fact that most cardiac tumors cause a variety of nonspecific clinical manifestations similar to those of many other common cardiovascular and systemic diseases. The clinical presentation is related to the myocardial and regional involvement, with or without the presence of metastasis. The patient’s initial signs and symptoms are often suggestive of pericarditis and pericardial effusion. If the tumor extends into the pericardial sac or atrial chambers, it often leads to hemopericardium, cardiac tamponade, or obstruction to blood flow. Electrocardiographic changes (such as elevated ST segment, T-wave inversion, low QRS voltages, right bundle branch block, atrial fibrillation, or electrical alternans) occur frequently, and may signal intra-myocardial spread. Other nonspecific symptoms, such as dyspnea, fever, malaise, or weight loss, also may be present. The associated nonspecific clinical and physical findings often delay the diagnosis, with resultant poor outcome.

The 2 cases here were admitted to our ward with the chief complaint being shortness of breath and some non-specific symptoms of cardiac diseases. The differential diagnosis of case 1 in emergency room included chest pain, cardiogenic shock, rule out myocarditis, and pericardial effusion. Case 2 was initially diagnosed as tuberculosis pericarditis with pericardial effusion. Cardiac tumor was found later in echocardiographic examination. Primary cardiac angiosarcomas are rare tumors and their clinical features are usually non-specific, which makes early diagnosis difficult. Most of them are found postmortem.

Imaging procedures, such as 2-dimensional echocardiography, CT and MRI, may allow recognition of malignant primary cardiac tumors. Transthoracic echocardiogram is relatively insensitive in detecting cardiac tumors. Transesophageal echocardiogram, CT and MRI are the newer, more sensitive and non-invasive modalities. CT can assess the location, the extent of tumors and evaluate for possible metastases. Echocardiogram adds information about the mobility of tumors and permits assessment of their hemodynamic significance. Therefore, it is not necessary to use invasive procedures to make the diagnosis of cardiac tumor if we use the combination of CT and echocardiography examination.

The indication of emergency operation of our 2 cases was the hemodynamic deterioration due to massive pericardial effusion and tumor mass obstruction in the right heart. During operation in case 1, it was not possible to completely excise the tumor due to extensive involvement. Recurrent tumor in RA was found in the follow-up MRI on June 13, 2002. Both patients died later in spite of using/or not using chemotherapy. Operation is not an effective treatment for cardiac angiosarcoma because of the large area of cardiac tissue involved or if metastases is found during diagnosis. But it can allow for differential diagnosis between a malignant cardiac tumor and a curable benign tumor through surgery procedure. Despite the absence of randomized trials proving efficacy, surgical excision is the mainstay of treatment. Further critical treatment modality decisions depend on the histological findings. Local and distant recurrence are both common. Most patients have distant metastases at time of clinical presentation. Prognosis of this rapid progression and early metastasizing of tumor is poor, with a median survival time of 3-6 months. Survivals of 1 to 3 years have been reported following partial resection, chemotherapy and radiation therapy. The prognosis of cardiac angiosarcoma remains poor. Cardiac transplantation
may have a role in the future for patients with localized disease.

REFERENCES