



Left-Sided Angiosarcoma of Heart: A Rare Case

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Abstract

A 22-year-old man presented with exertional dyspnea commencing one month prior to his admission. Echocardiography revealed a non-homogenous mass, and the pathology examination of the pericardial biopsy was compatible with angiosarcoma.

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Introduction

Primary tumors of the heart are extremely rare, with a prevalence rate of around 0.01% in collective autopsy studies.¹ Angiosarcomas with the inclusion of Kaposi sarcomas account for 30% of primary cardiac sarcomas.² In this article, we describe a case of a malignant angiosarcoma.

Case Report

A 22-year-old man presented with dyspnea functional class II-III. Aside from a history of progressive exertional dyspnea commencing one month previously, the patient had no other symptoms. Because of massive pleural effusion, a chest tube had been inserted and pleural and pericardial biopsy obtained. The pericardial biopsy showed fibrinoid pericarditis, and the cytology of his pleural effusion revealed no malignant cells. He was subsequently referred to our center for further evaluations. On presentation, the patient's vital signs were stable, but there was a decreased lung sound especially in the left lung and the laboratory

studies were all normal. Transthoracic and transesophageal echocardiographic examinations were conducted, which demonstrated a large left atrium mass (5×6 cm) with the involvement of the interatrial septum, roof, and the lateral side of the right atrium with extra cardiac extension and pleuropericardial effusion (Figures 1, 2, and 3). A thoracic surgeon was consulted, and biopsy was taken via thoracoscopy so as to define the nature of the mass, which was determined to be malignant secondary to the pleuropericardial effusion and the extension of the mass. Thoracoscopy revealed multiple small nodules on the pericardium, and biopsy was taken. The result of the pathological examination was compatible with angiosarcoma, which was confirmed by immunohistochemistry staining. Chemotherapy was commenced at the discretion of our hematologist, but an acute sudden dyspnea in the first session of chemotherapy led to the patient's death.

Discussion

Primary tumors of the heart are extremely rare, with a prevalence rate of around 0.01% in collective autopsy

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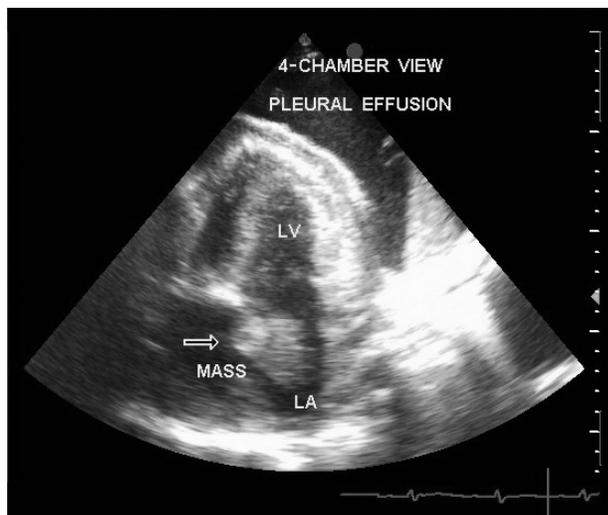


Figure 1. Left atrial mass (arrow) with pericardial effusion (4-chamber view)
LV, Left ventricle; LA, Left atrium

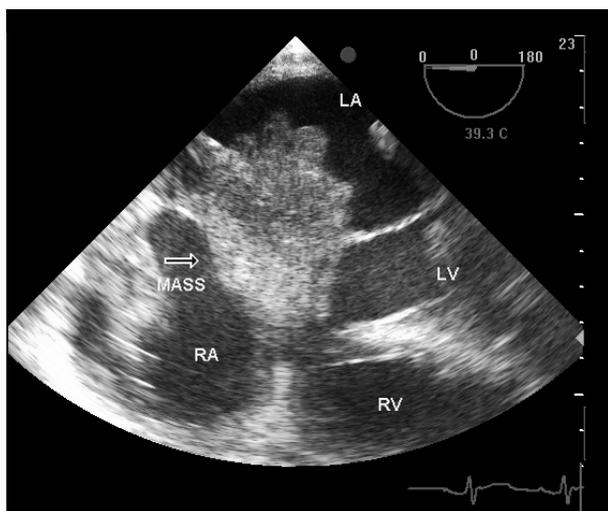


Figure 2. Transesophageal echocardiographic view of the mass (arrow) with involvement of interatrial septum
RA, Right atrium; LV, Left ventricle; RV, Right ventricle; LA, Left atrium

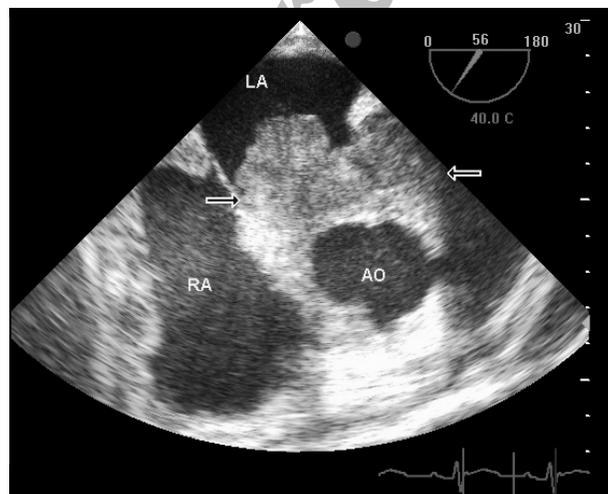


Figure 3. Transesophageal echocardiographic view of the mass (arrows)
AO, Aorta; RA, Right atrium; LA, Left atrium

studies. The majority of primary cardiac tumors are benign. Myxomas are the most common primary cardiac tumors, while angiosarcomas are the commonest primary malignant tumors.¹ Angiosarcomas with the inclusion of Kaposi sarcomas account for 30% of primary cardiac sarcomas.² There is a 3:1 male-to-female ratio amongst patients with angiosarcomas.³ Patients usually present with right-sided heart failure or tamponade as well as systemic signs such as fever and weight loss.³ This case of angiosarcoma of the heart is presented herein because of the extreme rarity of its location. Not only did our patient have extensive cardiac involvement but his other organs were involved as well and the tumor was not primarily from the right side of the heart. Unfortunately, the progression of the disease after diagnosis was extremely rapid and the patient died following the first course of chemotherapy.

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