Unusual Presentation of Cardiac Angiosarcoma Mimicking Left Ventricular Myxoma

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Introduction

Diagnosis of cardiac tumor is a challenging issue for the echocardiographer most of the time. Patients with obviously large intracardiac masses (several centimeters in diameter) can be referred to cardiac surgery without doubt. On the other hand, it is difficult to refer for surgery patients with small intracardiac masses, since they usually do not interfere with cardiac function. In these cases, misdiagnosis will result in unnecessary surgery, thus clinicians frequently prefer to follow up the patient. However, follow-up by periodical echocardiographic examination may allow malignant tumors to metastasize.

Several features of the tumors are described in previous studies. Myxomas usually occur as nodular or polypoid masses with a stalk. However, the appearance of a tumor may not give us an idea about the malignant nature of the tumor. In such a case, atypical location of the mass may be a clue for surgical resection.

Case Report

A 73-years-old man with long standing hypertension and ventricular arrhythmias of right bundle branch block (RBBB) morphology was referred to the department of cardiology because of heart failure symptoms. Transthoracic echocardiography revealed dilatation of all cardiac chambers, diffuse hypokinesia of left ventricle, mitral regurgitation (moderate severity) and tricuspid regurgitation (moderate severity). In addition, a small mass of 6 mm in diameter on left ventricular side of the interventricular septum was also observed (Figure 1). Its globular shape with a short peduncle (Figure 2), and absence of infiltration or polypoid appearance at first suggested the diagnosis of left ventricular myxoma (1). Transesophageal echocardiography did not reveal any additional abnormality. The clinical history was not remarkable for any embolic event. Thorax computed tomography (CT) was performed in order to search for the possible presence of a thoracic extracardiac tumor (or metastasis). However, thorax CT did not reveal any abnormality. Cardiac magnetic resonance imaging was not performed since transthoracic echocardiography clearly showed a single mass with a peduncle rather than multiple cardiac masses.

The tumor was suspected to cause the ventricular arrhythmias of RBBB morphology. Because of this possible link and the bizarre location of the mass, the patient underwent cardiac surgery for removal of the tumor. Pathological examination was compatible with angiosarcoma (Figure 3). The patient did not receive chemotherapy or radiotherapy. Transthoracic echocardiography, which was performed 8 months after the operation did not show any tumor mass in the left ventricle. Left ventricular wall motion abnormalities and chamber size dilation did not resolve after the surgery. Ventricular arrhythmias of RBBB morphology also persisted after the operation.

Discussion

Primary cardiac tumors are very rare findings. Only 2 % of cardiac tumors are primary. About 25% of them

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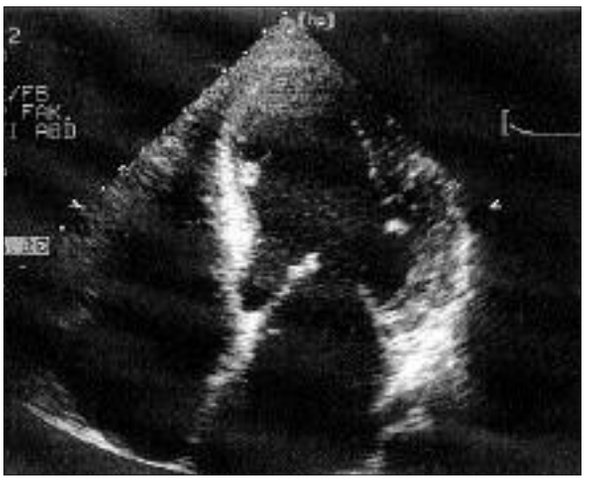

Figure 1: Apical-4-chamber view demonstrates the small tumor (6 mm in diameter) on left ventricular side of the interventricular septum. LA indicates left atrium; LV, left ventricle; and RV, right ventricle.

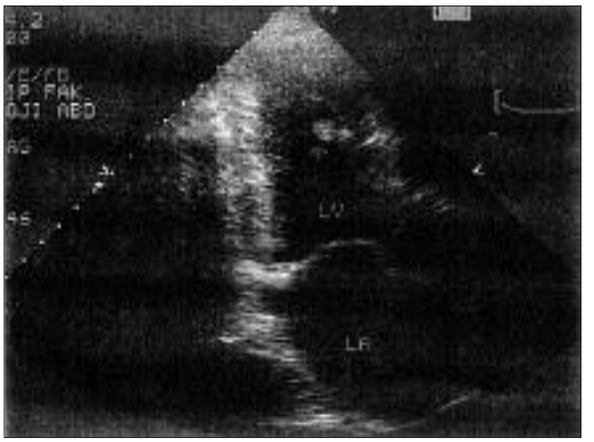

Figure 2: Apical long-axis view clearly demonstrates the nodular tumor (arrow) and its stalk. Tumor removal with limited endocardioectomy may allow complete resection. LA indicates left atrium; and LV, left ventricle.

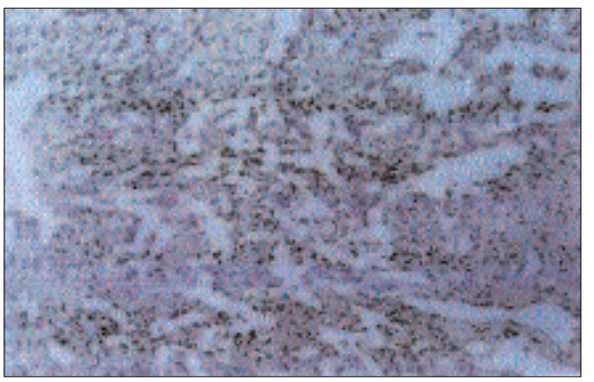

Figure 3. Pathological examination was compatible with well-differentiated cardiac angiosarcoma composed of irregular vascular channels.

are malignant and angiosarcoma is the most common (2). Cardiac angiosarcomas have several foremost features including a striking predilection for right atrium, infiltrative or polypoid appearance and a relatively large tumor size at the time of initial diagnosis (3). Approximately 80% of them are reported to originate in the right atrium as large mural masses (4). Early detection of angiosarcoma during life is very difficult. In most cases, this highly malignant tumor is diagnosed at autopsy.

If reliable echocardiographic criteria for the diagnosis of a malignant cardiac tumor were available, then it would be very easy for the clinician to refer patient for surgical removal of the mass. Indeed, several studies attempted to define the echocardiographic features of cardiac tumors (5,6). In our patient, the origination of tumor from interventricular septum without mural location or distortion of the shape of the interventricular septum, and the presence of a smooth surface with a stalk were not suggestive of angiosarcoma. Atypical presentation of angiosarcoma in our case clearly shows that echocardiographic appearance is not a reliable finding to estimate the malignant nature of a cardiac tumor. In patients with suspected intracardiac malignancies, atypical location of the mass rather than its size or appearance may be a more important clue for the clinician. Our case signifies the importance of early surgery for removal of small cardiac masses at bizarre locations.

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