

Primary Cardiac Osteosarcoma

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ABSTRACT

A 52-year-old woman, who was suffering from aplastic anemia, presented with the clinical features of severe heart failure. The transthoracic echocardiogram showed a heterogeneous, huge mass on the base of the posterior mitral valve. We guessed that the mass would be a benign neoplasm and probably myxoma, and we decided upon surgical resection. After tumor resection, an unexpected result of the histopathology was a high grade osteosarcoma. The other studies that were done after we had the diagnosis could not reveal any evidence of metastatic malignancy. (Korean Circulation J 2006;36:764-766)

KEY WORDS : Cardiac neoplasms ; Osteosarcoma.

Introduction

Primary cardiac neoplasms are rare tumors. About 25% of primary cardiac neoplasms are considered as malignancy, and the majority of these malignancies are angiosarcoma and undifferentiated sarcoma. Cardiac osteosarcomas account for only 3-9% of all cardiac sarcomas,¹⁾ and few cases of cardiac osteosarcoma have been reported. We report here on an extremely rare case that proved to be a primary cardiac osteosarcoma.

Case

A 52-year-old woman, who had been receiving treatment for 5 years due to aplastic anemia, was referred to our cardiologic department because of her aggravated dyspnea on exertion for several weeks. At the time of admission, her vital signs were relatively stable and the physical examination was unremarkable exclusive for soft crackle sounds on both lower lung fields. The laboratory findings were pancytopenia (white blood cells: $2,740 \times 10^3/\mu\text{L}$, hemoglobin: 3.7 gm/dl, platelets: $84,000 \times 10^3/\mu\text{L}$), serum iron: 139 $\mu\text{g/dL}$, serum ferritin: 11336 ng/mL and serum TIBC: 192 $\mu\text{g/dL}$. The cardiac enzy-

mes and serum electrolytes were normal. Chest radiography demonstrated cardiomegaly and pulmonary congestion. A transthoracic echocardiogram (TTE) showed a small sized echogenic mass in the left atrium (LA), a dilated left ventricular dimension (left ventricular end diastolic dimension: 58 mm), a decreased left ventricular systolic function (left ventricular ejection fraction: 34%) and significant mitral regurgitation (Fig. 1A). The transesophageal echocardiogram demonstrated about a 1 cm sized heterogeneous mass on the base of the posterior mitral valve, which extended to the mid portion of it (Fig. 1B). At that time, our diagnosis was nonbacterial, thrombotic endocarditis and dilated cardiomyopathy due to hemochromatosis. After conventional therapy for congestive heart failure and anticoagulation, she became much better and was discharged.

Alas, she arrived at the emergency department about 4 months later because of severe dyspnea and orthopnea. The physical examination revealed an apical diastolic murmur and coarse crackle sounds on the mid to lower lung fields. On TTE, the mass shown by the previous study had rapidly increased to about 5×4 cm in size with broad base, and this mass occupied most of the left atrial chamber. Further, the echogenicity of the mass was more inhomogeneous. It had relatively low echogenicity within the center and it was attached to the base of the posterior mitral valve (Fig. 2). We guessed that the mass would be a benign neoplasm and probably myxoma, and we decided upon surgical resection. During the operation, we saw the palpable mass in the LA and it had invaded the base of the posterior mitral leaflet with extension into

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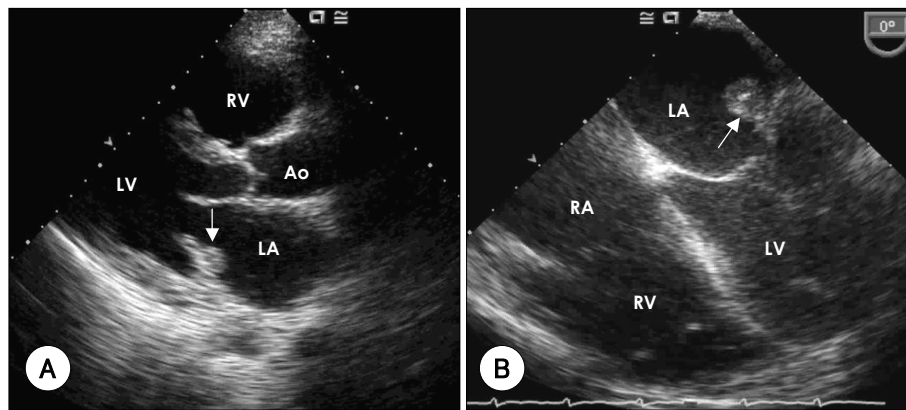


Fig. 1. Photograph of the transthoracic echocardiogram showing the parasternal long axis view of a 1 cm sized, round shaped, echogenic mass in the left atrium (arrow) (A). Transesophageal echocardiogram showing the heterogeneous mass on the base of the posterior mitral valve (arrow) (B). LA: left atrium, LV: left ventricle, RA: right atrium, RV: right ventricle, Ao: aorta.

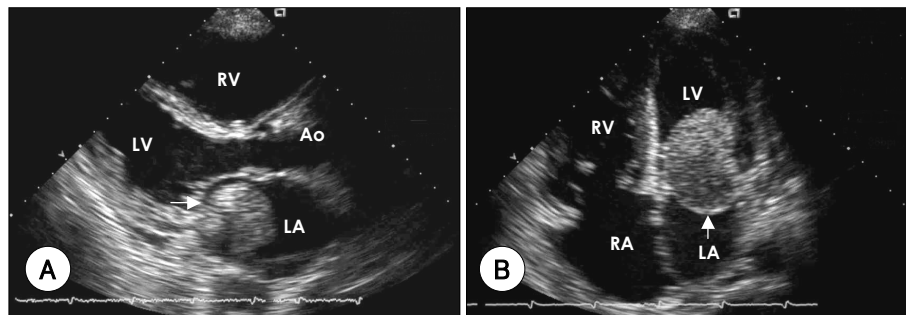


Fig. 2. Photograph of the follow up transthoracic echocardiogram showing the parasternal long axis view of the 5 × 4 cm sized mass (arrow). It had relatively low echogenicity within the center and it was attached to the base of the posterior mitral valve (A). The apical four chamber view showed the mass (arrow) occupying most of the left atrial chamber and protruding into the left ventricle during diastole (B). LA: left atrium, LV: left ventricle, RA: right atrium, RV: right ventricle, Ao: aorta.

the posterior wall of the LA. After resection, it seemed to be a myxoma in appearance (Fig. 3), but the histopathology was that of a high grade osteosarcoma composed of polygonal to spindle cells, patchy areas of necrosis and calcification with cartilage and osteoid formation (Fig. 4).²⁾ The bone marrow biopsy and a 131-Iodine whole body bone scan along with the other studies we conducted revealed there was no evidence of metastatic malignancy. We finally came to a conclusion that the mass was a rapid growing, primary cardiac osteosarcoma.

Discussion

Primary cardiac neoplasms are infrequent tumors, with metastases to the heart being about 20-40 times more common than primary tumors. An autopsy series reported a prevalence of 0.1% to 0.3% for primary cardiac tumors.¹⁾ About 25% of primary cardiac tumors are considered as malignancy; the majority (95%) are sarcomas, primarily angiosarcomas and undifferentiated sarcomas. Cardiac osteosarcoma accounts for only 3-9% of all cardiac sarcomas. They may be predominantly osteoblastic or they may show chondroblastic or fibroblastic differentiation.³⁾ Unlike metastatic osteosarcoma, primary cardiac

osteosarcomas occur predominantly in the LA and they frequently involve the pulmonary veins.⁴⁾⁵⁾ Therefore, they are usually accompanied by the signs and symptoms of congestive heart failure. Histologically, they contain malignant bone-producing cells. The heterogeneous tissue contains spindle cells, mature osteoid with acellular lacunae and calcification. Calcification is often noted in the gross surgical specimens. Although cardiac neoplasms can arise from any cardiac structure, most exhibit a predilection for a specific chamber. Mainly because of their left atrial location, primary cardiac osteosarcomas are often confused upon radiology with left atrial myxomas. Echocardiography has been the modality of choice for making the diagnosis of intracardiac disease.⁶⁾ It allows real-time imaging, and it can show tumor mobility and those features that are typically seen for atrial myxomas and less often for sarcoma.

Computed tomography (CT) and Magnetic Resonance Imaging (MRI) provide better soft-tissue contrast and visualization of extracardiac structures than echocardiography. CT can depict calcification and fat, and it may allow tissue diagnosis of some masses such as lipomas. CT may show dense calcifications within a low-attenuation mass. However, calcification may also be

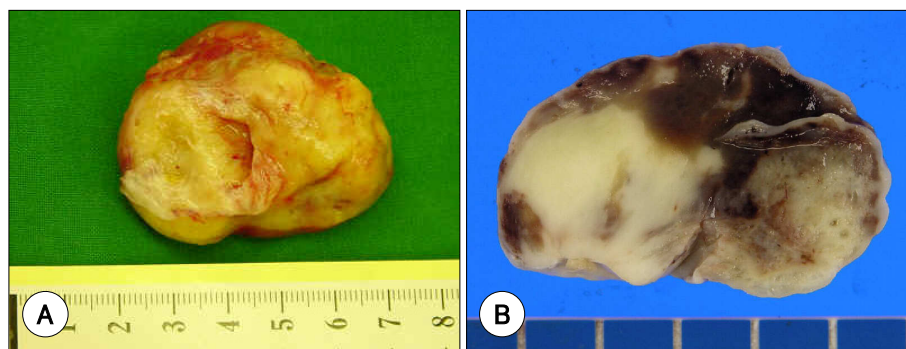


Fig. 3. The left atrium mass measured 31.5 g in weight and $5.1 \times 4.2 \times 3.0$ cm in size. The outer surface was pale yellow to grayish brown with a slightly lobulated contour (A). On sectioning, the cut surface was homogeneously pale, rubbery to firm and it showed a somewhat myxomatous appearance and focal hemorrhage (arrows) (B).

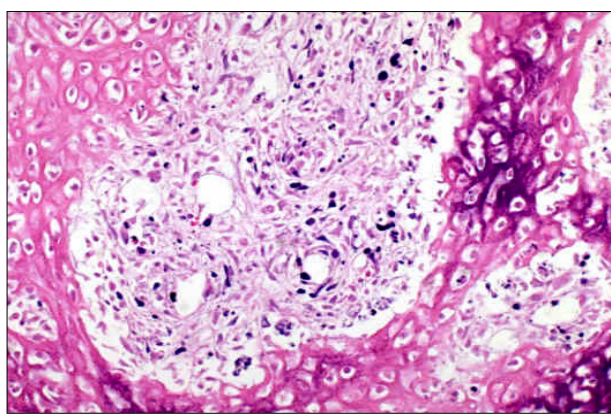


Fig. 4. The histopathology showed a high grade osteosarcoma composed of polygonal to spindle cells, patchy areas of necrosis and calcification with cartilage and osteoid formation.

minimal and it can in may be mistaken in the early stages for benign, dystrophic calcification.³⁾ The suggestive image features of osteosarcomas are a broad base of attachment, an aggressive growth pattern such as extension into the pulmonary veins, invasion of the atrial septum or infiltrative growth along the epicardium, and a tumor location within the left atrium. Not only are

they unusual, but they are also often asymptomatic until they become large, and even then they produce non-specific symptoms. Complete excision has been associated with increased survival. Therefore, an early diagnosis may permit more complete debulking and improve the outcome. However, patient survival is poor for this malady. Despite resection, chemotherapy and even cardiac transplantation, these patients generally succumb to their tumor burden or metastases.

REFERENCES

- 1) Lurito KJ, Martin T, Cordes T. *Right atrial primary cardiac osteosarcoma. Pediatr Cardiol* 2002;23:462-5.
- 2) Yamagishi M, Bando K, Furuichi S, Ishibashi-Ueda H, Yutani C, Miyatake K. *Primary cardiac osteosarcoma in right ventricular outflow tract. Circulation* 2000;101:2220-1.
- 3) Araoz PA, Eklund HE, Welch TJ, Breen JF. *CT and MR imaging of primary cardiac malignancies. Radiographics* 1999;19:1421-34.
- 4) Burke AP, Virmani R. *Osteosarcomas of the heart. Am J Surg Pathol* 1991;15:289-95.
- 5) Burke AP, Cowan D, Virmani R. *Primary sarcomas of the heart. Cancer* 1992;69:387-95.
- 6) Chang BI, Lee SJ, Sung HD, et al. *A case of primary malignant mesenchymoma of the heart. Korean Circ J* 2003;33:625-8.