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Treatment of a Case of Primary Osteosarcoma of the Left Heart: A Case Report

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ABSTRACT

Intracavitary sarcoma of the heart is rare. We report a case of primary osteosarcoma of the left atrium and ventricle that was managed with surgical resection and chemotherapy, attaining more than 1-year survival.

CASE REPORT

A 46-year-old man was referred for progressive dyspnea on exertion and worsening fatigue. The physical exam on admission revealed dyspnea at rest, tachycardia, and inspiratory crackles. His chest radiography revealed bilateral pleural effusions and pulmonary congestion. A transthoracic and transesophageal echocardiogram showed an oval-shaped mass attached broadly to the posterior wall of the left atrium and left ventricle extending to the subaortic outflow tract with severe mitral valve involvement. Preoperative magnetic resonance imaging (MRI) confirmed the mass and its extension into the heart (Figure 1). A working diagnosis of primary cardiac tumor was made after neoplasm staging and skeletal survey. Coronary angiography revealed nonobstructive coronary disease and surgical resection was contemplated. Cardiopulmonary bypass with bicaval cannulation was instituted and the left atrium explored posterior to the interatrial groove. The endocardium of the left atrium was diffusely thickened and fibrotic, with multiple small conglomerations of tumor extending into the mitral leaflets and papillary muscles. There was a $3 \times 3 \times 3$ cm mass in the left atrium close to the origin of the right superior pulmonary vein. A large, white, hard mass was seen in the dome of the left atrium with extension of softer gelatinous material throughout the mitral valve annulus (Figure 2A). The left ventricular cavity exhibited a $4.5 \times 4 \times 3$ cm tumor posterolateral to the origin of the anterior papillary muscle. The tumor was sessile throughout the length of the posterior left ventricle wall to the left ventricular outflow tract. Multiple small foci of the tumor were seen on the ventricular side of the intact aortic valve. The aortotomy showed a large fungating mass just below an intact aortic valve extending from the posterior left ventricle wall, which was excised (Figure 2B). The mass was grossly removed as completely as possible. Resection was limited by the extensive involvement of the posterior ventricular wall with the tumor. The mitral valve apparatus had to be removed entirely due to tumor involvement and the valve was replaced with a St. Jude prosthesis #25 (St. Paul, MN, USA).

Permanent histological sectioning confirmed osteosarcoma of the heart. The pathological specimen under light microscopy revealed loosely arranged collagen containing scattered spindle-

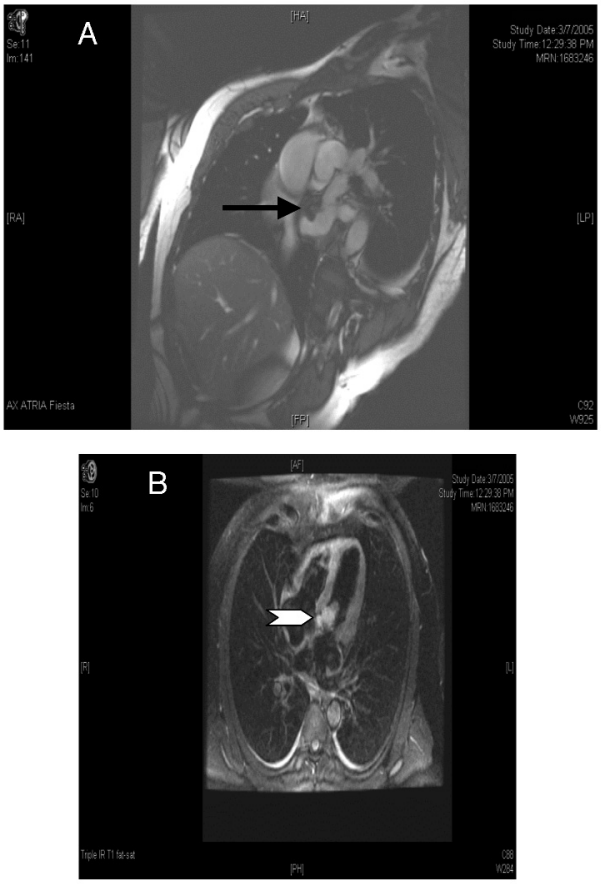

Figure 1. A and B depict T1- and T2-weighted cardiac magnetic resonance imaging, respectively. The left atrial (arrow) and left ventricular outflow tract tumor (arrow head) is revealed.

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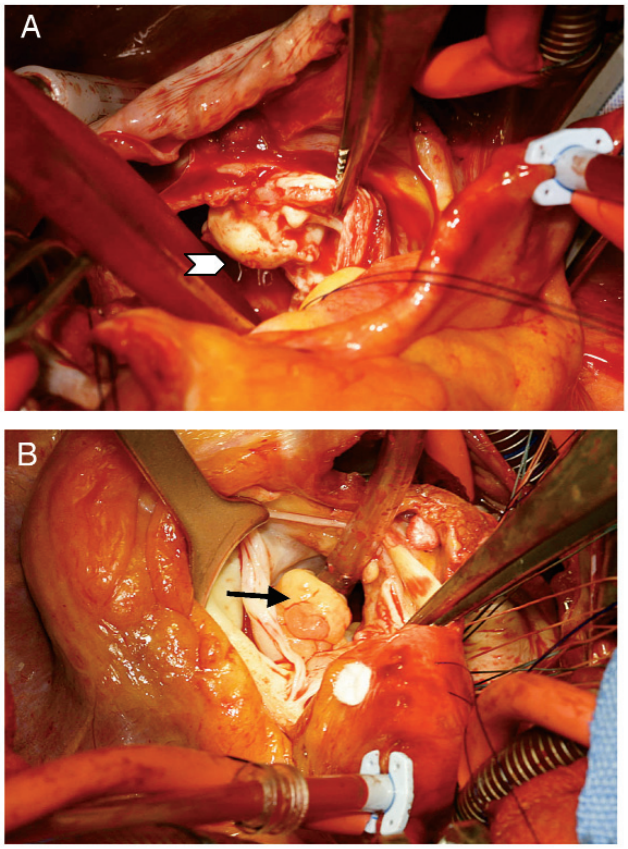

Figure 2. A, A large calcified mass in the posterior wall of the left atrium (arrow head). B, Subaortic presence of osteosarcoma tumor after retracting via transverse aortotomy (arrow).

type cells poorly differentiated in nature (Figure 3). The initial transesophageal echocardiography at 6 weeks after surgery revealed no evidence of intracardiac tumor, but repeat echocardiography at about 3 months showed evidence of tumor recurring in the left ventricular outflow tract. After thorough multidisciplinary discussions, the patient was treated with anthracycline-based chemotherapy. Multiple repeat transesophageal echocardiographies revealed the mass to be stable, and at 16 months the patient reported to be feeling well and remained fully employed.

DISCUSSION

Primary osteosarcomas of the heart are exceptionally rare tumors that comprise 3% to 9% of all primary sarcomas of the heart [Vander Salm 2000; Sarjeant 2003]. They usually arise in the left atrium; however, occasionally they can arise from the left ventricle. Osteosarcomas in general are bulky, sessile, masses 4 to 10 cm in diameter and are usually attached to the wall of the left atrium. Destruction of the valve and conduction tissue has been reported. Current theory postulates that these tumors arise from fibrous tissue that has undergone metaplasia with de-differentiation into malignant bony and cartilaginous elements [Sarjeant 2003]. Patients present usually with heart failure symptoms due to valve dysfunction or conduction defects. Complete surgical excision is the main therapy for these tumors. In the absence of extracardiac metastasis, complete cardiac excision with orthotopic cardiac transplantation has been attempted [Goldstein 1995]. However, the idea of transplantation in the face of this aggressive tumor and in light of the required immunosuppression and the scarcity of the available organs does not seem to be an appealing option.

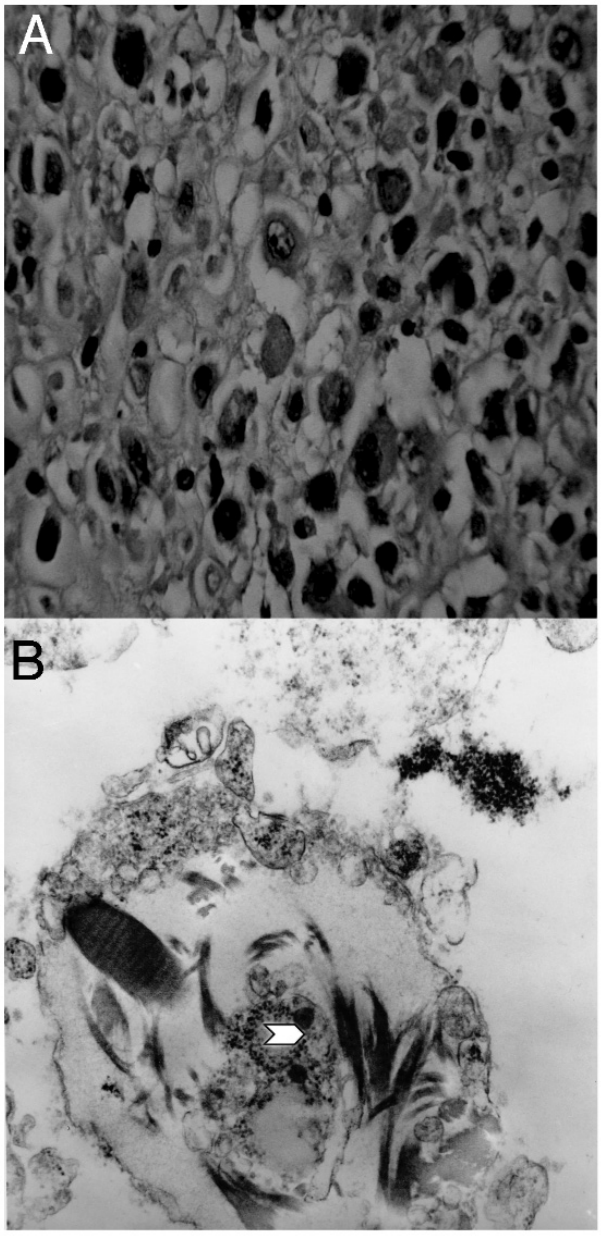

Figure 3. Hematoxylin and eosin staining, medium power with atypical malignant cells. There is a presence of amorphous extracellular material, consistent with osteoid deposition (A). In the electron micrograph (B), the adjacent intercellular space shows a low electron-dense matrix enclosing haphazardly arranged collagen and elastic fibers. Additionally, hydroxyapatite deposits (arrow head) are seen.

Complete excision of the tumor offers the highest chance for cure. The aim of surgery is to remove the tumor with minimal cardiac functional loss. Lateral and in-depth negative surgical margins are usually impossible with extensive intracardiac involvement, as in our case. Full-thickness excision of ventricular sarcomas has been described and is limited by small residual cavity size and coronary anatomy. Endocardial patch ventriculoplasty is sometimes needed for partial thickness excision [Putnam 1991; Calderon 1996].

The prognosis after surgical resection of malignant cardiac tumors is poor. Patients with osteosarcoma live for a mean of 3 months to 1 year [Vander Salm 2000; Sarjeant 2003]. The therapeutic role of chemotherapy and radiotherapy remains a work in progress. Systemic chemotherapy is mostly beneficial in patients with systemic metastases or incompletely resected or unresectable tumors [Sarjeant 2003]. However, patients with localized disease have been considered for neoadjuvant chemotherapy to downstage the tumor prior to complete resection.

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