Primary Cardiac Pleomorphic Sarcoma: An Aggressive Tumor

Georgios P. Georghiou MD[1], Yaron Shapira MD[2], Ana Tobar MD[3], Bernardo A. Vidne MD[1] and Gideon Sahar MD[1]

Departments of [1]Cardiothoracic Surgery and [2]Cardiology, and [3]Institute of Pathology, Rabin Medical Center (Beilinson Campus), Petah Tiqva, Israel
Affiliated to Sackler Faculty of Medicine, Tel Aviv University, Ramat Aviv, Israel

Key words: echocardiography, heart pathology, mitral valve replacement, sarcoma

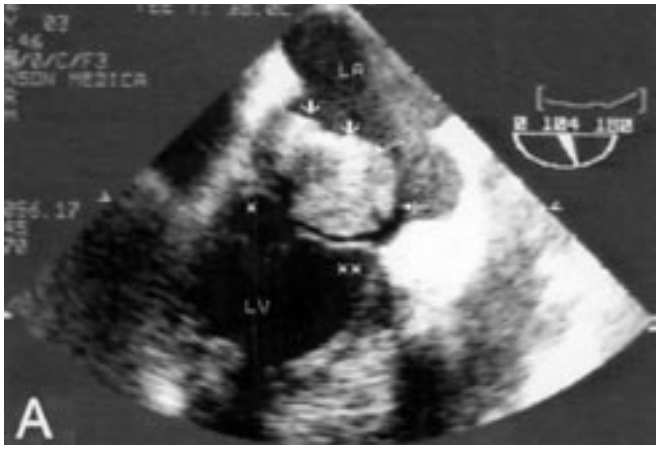
IMAJ 2005;7:470–471

Soft tissue sarcoma is the most common malignant neoplasm of the heart, pericardium, and great vessels [1]. Primary cardiac sarcomas have an incidence at autopsy of only 0.0001% [2]. They are the second most common type of primary cardiac tumor, after myxoma [3]. Symptoms are usually the result of tumor invasion or anatomic obstruction of the blood flow, necessitating resection to prevent early death [3]. Historically, the long-term survival of patients with primary cardiac sarcoma has been poor, especially when complete surgical excision is unfeasible [1]. In these instances, median survival is generally 10 months or less [2]. There are no proven benefits of chemotherapy and radiotherapy [1]. A more aggressive approach with transplantation in highly selected patients with primary cardiac sarcomas proved disappointing [4].

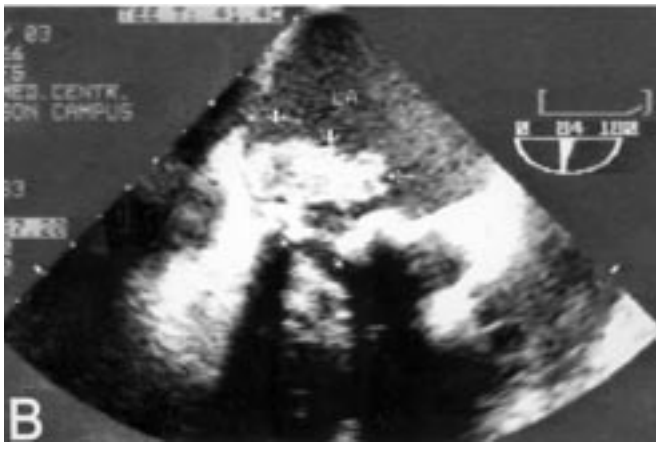
Patient Description

A 65 year old previously healthy woman presented at another hospital with a dull substernal chest pain of 3 weeks duration accompanied by progressive shortness of breath. The initial physical examination was unremarkable except for a grade 3/6 diastolic murmur at the left sternal border. Transesophageal echocardiography [Figure A] revealed a 3 x 5 cm mass attached to the posterior left atrial wall just above the posterior mitral valve leaflet insertion. The mass prolapsed into the left ventricle in diastole causing increased gradients through the mitral valve (peak 14 mmHg, mean 7 mmHg) with reduced mitral valve area (1.4 cm^2). The patient was referred for urgent removal of the mass.

Intraoperatively, the tumor was found to be fixed in the left atrium, just above the posterior mitral valve leaflet insertion. Partial thickness resection of the left atrial wall with the endocardium was performed, and a 5.5 x 4.0 x 2.0 cm yellowish-pink mass was excised, including the mitral valve because of the mass-induced deformity of the posterior valve leaflet. Pathologic examination revealed a malignant pleomorphic sarcoma. The margins were close but clear of the tumor. Microscopically, the tumor was composed of pleomorphic cells, most of them multinucleated, with tails. Some had no



[A]. Transesophageal echocardiography. Left atrial mass measuring 3 x 5 cm (arrows) attached to the left atrial wall just above the posterior mitral valve leaflet insertion. * = anterior mitral leaflet, ** = posterior mitral valve leaflet. LA = left atrium, LV = left ventricle



[B]. Transesophageal echocardiography, 3-chamber view. Note echogenic, mobile mass measuring 3.5 x 1.3 cm (arrows) in the left atrium, attached to the mitral valve prosthesis (asterisk).

striations. The cells expressed high mitotic activity, including atypical mitosis. There was extensive necrosis and a rich vascular stroma. Immunohistochemical stains for vimentin, desmin and actin were positive. Staining for keratin, S100, CD30, CD68 and FVIII was negative. Staging workup, completed postoperatively, was negative for metastasis.

The oncologist considered the patient unsuitable for adjuvant therapy. Two months later, a 3.5 x 1.3 cm oscillating echodensity was noted in the left atrium, attached to the posterior left atrial wall and partially obstructing the mitral valve prosthesis, consistent with tumor recurrence [Figure B]. The patient died one month later.

Comment

This case is unique because serial cardiac imaging provided insight into the aggressive behavior of primary cardiac sarcoma. Local tumor recurrence was noted already 2 months after debulking. Histologic study confirmed the tumor's rapid growth. However, this was impossible to predict accurately, because the growth and necrosis occurred simultaneously within the same mass.

As in most cases of cardiac tumor, the diagnosis of sarcoma was not made preoperatively. The decision to operate was based on the need to relieve the left ventricular inflow obstruction, prevent embolism, and obtain tissue for diagnosis. At surgery, the mitral valve was replaced because of the mass-induced deformity of the posterior valve leaflet.

This is an unusual tumor, and we found only one similar description in the literature [5]. In that case, the pleomorphic sarcoma of the mitral valve extended into the myocardium. There, too, the mitral valve was replaced during surgery. The patient died of recurrent cardiac neoplasm and multiple metastases 30 months after diagnosis [5].

Debulking surgery is the mainstay of treatment for all primary cardiac malignancies, although success is limited by tumor spread, metastasis at the time of diagnosis, and early recurrence. The role of adjuvant chemotherapy following surgery to prevent the development of metastatic disease has not been established. Further research is needed to develop improved methodologies that will prevent local recurrence.

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Correspondence: Dr. G.P. Georghiou, Dept. of Cardiothoracic Surgery, Rabin Medical Center (Beilinson Campus), Petah Tiqva 49100, Israel.
Phone: (972-3) 937-6701
Fax: (972-3) 924-0762
email: georgios@clalit.org.il