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 cm²). 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...
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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 neoplasms 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.

References

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I don't feel old. I don't feel anything until noon. Then it's time for my nap.

Bob Hope (1903-2003), British-born American comedian and actor