Long-Term Survival With Heart Transplantation for Fibrosarcoma of the Heart

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Primary sarcoma of the heart is a rare disease that has an ominous prognosis with either medical or surgical therapy. We report a case of a 25-year-old woman with sarcoma of the heart who received a transplant and is clinically well after 7 years. We believe that transplantation must be considered in this kind of pathology for selected cases.

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Primary tumors of the heart are rare clinical entities and represent 0.001% to 0.03% of all cardiac surgical cases [1–3]; of these, 25% are malignant tumors, and among them, sarcomas are the most prevalent. Sarcomas of the heart are known to be highly invasive, and when the diagnosis is made, about 80% are already disseminated. Average survival at 9 months is 10% [2].

The best therapeutic options for these patients are still being discussed. Total resection is not possible for most patients, and the local recurrence rate is very high. Transplantation has certainly been an option for selected isolated cases, but long-term results and precise indications are still not clear [1].

Our patient was a 25-year-old woman who was diagnosed with Hodgkin lymphoma at age 16 years. She was treated with chemotherapy and mediastinal radiotherapy and experienced total remission. As sequela, she had some degree of lung fibrosis and hypothyroidism.

In August 2001, the patient was unexpectedly admitted to the hospital with complaints and signs of right-sided heart failure. An echocardiogram showed a tumor mass filling the right ventricular cavity, and a computed tomography scan and cardiac magnetic resonance imaging confirmed those findings and excluded any other locations. Without other diagnosis and given her severe New York Heart Association functional class IV condition, she underwent an open heart operation.

A biopsy specimen was obtained from the tumor mass inside the right ventricle, and the pathology report identified sarcoma. Considering the ominous result, partial debulking of the tumor was done, trying to maintain the cavity-filling function of the ventricle without putting the patient at risk with a too-aggressive resection, namely at

the septal area where we could create a ventricular septal defect.

The patient's postoperative period was without complications. She was referred for chemotherapy and underwent a perfusion of ifosfamide (1500 mg/m²) for 3 days.

One month later, she was readmitted with gross right-sided heart failure, and the echocardiogram showed extensive recurrence of the tumor mass, with protrusion of the tumor through the tricuspid valve into the right atrium (Fig 1). At this point, there seemed to be no other medical or surgical options owing to the type of the recurrent tumor involving the heart. Computed tomography scans and liver and bone perfusion scans were able to exclude any metastasis or invasion at distant locations, and cardiac transplantation came as a last option.

The patient underwent cardiac transplantation in December 2001 that was performed with a biatrial technique. The tumor invaded the ventricular wall, the papillary muscles, and the cords of the tricuspid valve, but not the anulus, the right atrium, or the epicardium. This was confirmed by the microscopic examination.

She experienced no major events in the postoperative period. The patient is receiving routine triple-drug immunosuppression with azathioprine (25 mg daily), cyclosporine (75 mg twice daily), and prednisolone (7.5 mg daily). Follow-up extends to 7 years now. The patient is at New York Heart Association functional class I, is very active, and has had no episodes of rejection and no evidence of recurrence. Echocardiograms reveal normal function, and control computed tomography and liver and bone scans results have been entirely normal. On no occasion and in accumulation with the standard immunosuppression, was any type of extra chemotherapy given to this patient.

Comment

Optimal therapy for sarcomas of the heart has not been established yet; however, wide resections with reasonable safety margins would seem appropriate [1, 2, 4, 5] if

Fig 1. Intraoperative photograph shows the right ventricle (Rv), the tumor (Tu), and the right atrium (Ra).

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metastases at distant locations in the body [5] are not found by computed tomography or perfusion scans. Transplantation might be an alternative, but due to immunosuppression, there is always the risk of tumor recurrence or de novo malignancies related to chronic use of immunosuppressant agents; for example, lymphoma has an incidence of 2.3% to 13% in transplant recipients [1, 5, 6].

Another issue is long-term survival after transplantation for cardiac malignancies. In a series of 21 patients with nonresectable cardiac tumors for whom transplantation was offered, mean survival was 12 months, and only 7 patients survived a mean of 29 months free of recurrence [5]. Despite these poor results, transplantation remains the only option for selected patients with nonresectable tumors but localized disease [5]. Adjuvant chemotherapy or radiotherapy, or both, do not seem to improve survival and are not useful [1, 5] and were not used in our patient.

In favor of transplantation for our patient, we had a nonresectable, recurring tumor in a young patient and the disease was strictly localized to the heart. Against transplantation was the history of previous lymphoma (eventually predisposing to de novo lymphoma induced by malignancy immunosuppression) and the poor results usually obtained with this treatment for this type of patients, although case series are very limited. Mean survival for patients with cardiac sarcoma has been established between 9 and 11 months [1]. Our patient has been monitored for 7 years until now, without evidence of recurrence of any type and without cardiac rejection, which certainly is an unusually good result.

Cardiac transplantation must be considered a valid therapeutic option for selected patients with nonresectable cardiac sarcomas without evidence of distant metastatic disease. Intermediate-term or even long-time survival can sometimes be achieved.

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Extended Cardiac Resection for Obstructing Pseudotumor Due to Ormond Disease

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A 60-year-old man presented with symptoms from an intracardiac mass. His medical history included retroperitoneal fibrosis (Ormond disease). Magnetic resonance imaging revealed an obstructing bilobular mass in the right atrium, located at the caval junction and extending intramurally into the atria, septum, and right ventricle. En bloc resection of the right atrium, interatrial septum, dome of the left atrium, vena cava, anterior tricuspid annulus, right coronary artery, and partial right ventriculectomy was completed with right ventricular repair, tricuspid valve replacement, and left and right atrial replacement with bovine pericardium. This lesion was a myofibroblastic tumor with the same histologic features as his retroperitoneal fibrosis.

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Primary cardiac tumors present a rare and complex challenge in cardiac reconstructive surgery. With an incidence of 0.0017% to 0.19%, these tumors account for less than 0.2% of all cardiac operations [1]. The categories of primary malignant cardiac tumors include histiocytomas, undifferentiated sarcomas, leiomyosarcomas, malignant osteosarcomas, and myxoid sarcomas. Benign tumors, which account for approximately 80% of primary cardiac tumors [1], include myofibroblastic tumors (pseudotumors), paragangliomas, and myxomas [2]. Myxomas account for nearly 80% of these benign tumors [1, 3]. This report details the treatment of a massively obstructive, benign cardiac tumor that caused superior vena cava syndrome, respiratory compromise, and functional tricuspid stenosis.

The patient was a 60-year-old man who presented with shortness of breath, bilateral pleural effusions, anasarca, and chronic renal failure; echocardiography revealed an intracardiac mass. His medical history was notable for asbestosis, industrial solvent exposure, a meningioma that was treated with radiotherapy, and retroperitoneal fibrosis (Ormond disease) requiring repeated ureteral stenting. Cardiac magnetic resonance imaging revealed a

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