LONG-TERM SURVIVAL AFTER CARDIAC AUTOTRANSPLANTATION FOR SARCOMA WITH SUCCESSFUL LATE REDO CARDIAC AUTOTRANSPLANTATION AND RIGHT PNEUMONECTOMY FOR RECURRENT LEFT ATRIAL SARCOMA

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INTRODUCTION

Primary cardiac malignancy is rare and represents a therapeutic challenge. At the Methodist DeBakey Heart Center we have developed the cardiac autotransplantation procedure - consisting of cardiac explantation, ex vivo tumor resection with cardiac reconstruction, and cardiac reimplantation - for left atrial and intracavitary left ventricular tumors. Here we report a case of successful cardiac autotransplantation for left atrial sarcoma, and successful redo autotransplantation and right pneumonectomy when the malignancy recurred five years after the initial procedure.

CASE REPORT

Our patient, a 34-year-old male, presented in 1996 with acute onset of shortness of breath at an outside institution. Evaluation revealed a large left atrial mass. Resection was undertaken with the presumed diagnosis of left atrial myxoma. At the rime of surgery, a large sessile mass with pathological features of sarcoma was encountered. Resection with positive margins was achieved, and the patient underwent five months of chemotherapy. In 1999, an echocardiogram revealed a recurrence. The patient was referred to M.D. Anderson Cancer Center and subsequently ro the Methodist DeBakey Heart Center for possible resection. Resection was undertaken using rhe technique of cardiac explanracion for complete tumor visualization, complete radical resection of the tumor and accurate cardiac reconstruction. At the time of surgery, a large sessile mass involving che entire back wall of the left atrium was found (Figure 1). This exposure allowed radical excision of the rumor mass (Figure 2) and accurate reconscrucrion of the left atrium with bovine pericardium (Figure 3). An additional implant was removed with a full thickness resection to the anterior left atrial wall just above the mitral valve (Figure 4). The heart was reimplanted and the post-operative course was uneventful. The patient underwent post-operative chemotherapy of ifosfamide and adriamycin, which resulted in renal failure requiring dialysis. Follow-up consisted of echocardiogram every six months, all of which were negative until June 2004. Echocardiogram

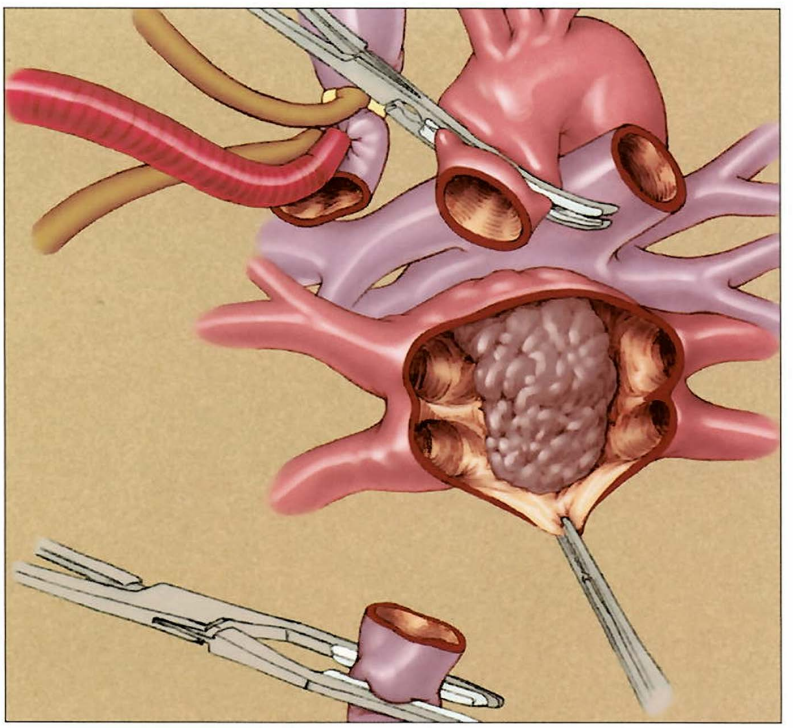

Figure 1. Tumor located in the patient's left atrium

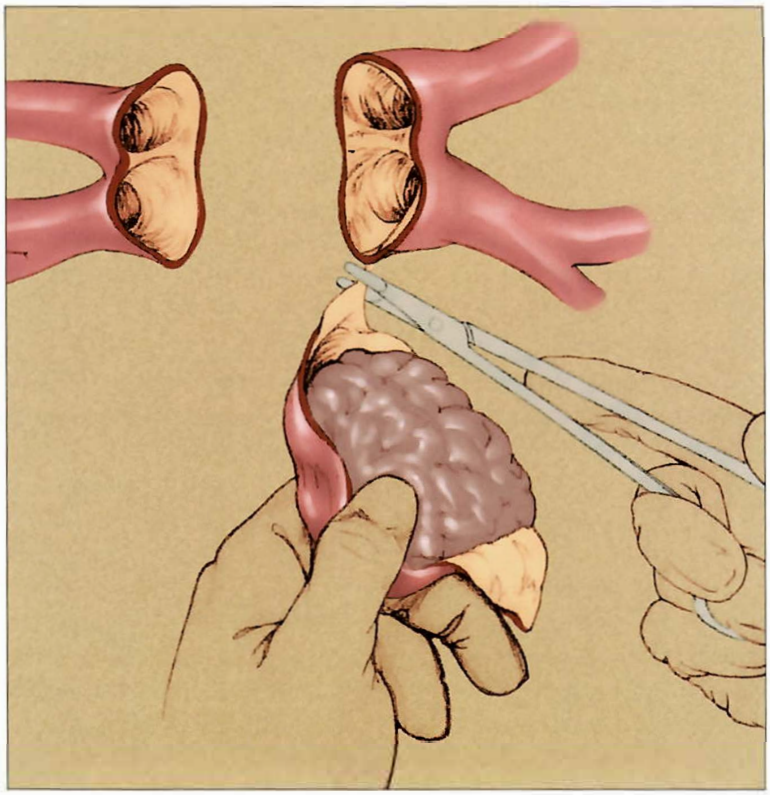

Figure 2. Complete exision of tumor mass

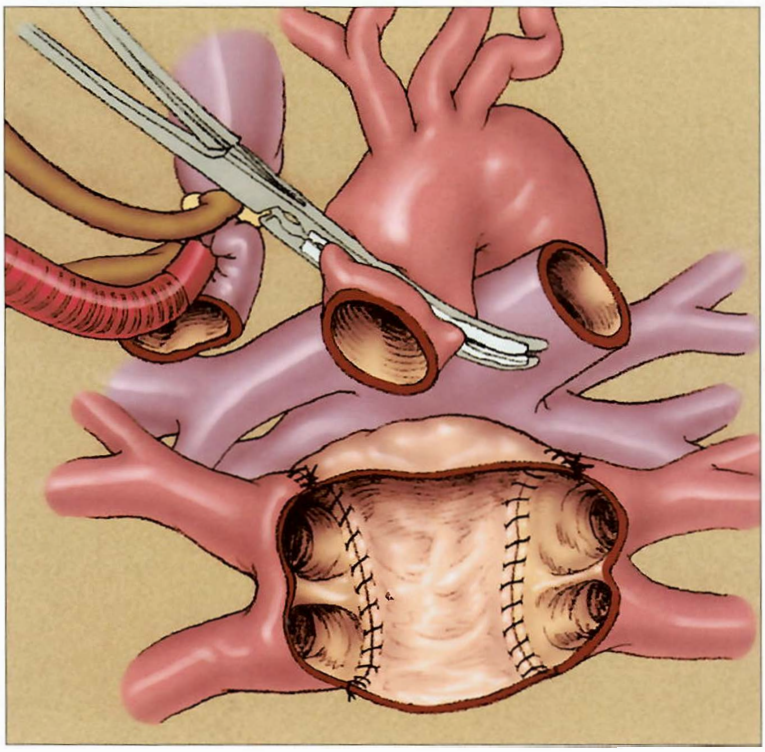

Figure 3. Reconstruction of the left atrium with bovine pericardium

at that time revealed a large 4.5 x 3.5 cm mass extending from rhe right superior pulmonary vein and involving the previously unresected left aerial roof (Figure 5). Evaluation revealed no evidence of metastatic disease, and the patient was referred for resection.

In June of 2004, the patient underwent redo cardiac excision, ex vivo resection of the left atrial tumor, amputation of the right superior pulmonary vein tumor and oversewing of the vein, reconstruction of the left atrium with bovine pericardium, and redo cardiac autotransplantation. Because of the patient's renal failure and the complexity of the case, it was elected to delay right lung resection. Although he initially did well, he developed evidence of superior vena caval syndrome. Two days postoperatively he was returned to the operating room, where a 20 mm Dacron interposition graft was inserted at the site of the previous superior vena caval anastomosis after venacavagram revealed a 50% stenosis and pressure transduction showed a 12 mm mean gradient across the superior vena cava.

The patient underwent another day of dialysis and returned to surgery the following day to undergo a righc pneumonecromy. His post-operative course was eventful due to development of deep venous thrombosis, for which he was placed uneventfully on anticoagulation. Doing well, che patient was discharged home 21 days post-operatively. The cardiac echo prior to discharge showed ejection fraction of 60% with no evidence of residual tumor. Pathology revealed high-grade sarcoma of approximately 7 cm in size in the lung in addition to the 4.5 cm tumor removed from the atrium. All lymph nodes were negative.

DISCUSSION

Primary cardiac sarcomas are rare entities with an autopsy incidence of 0.001% to 0.00%.1 Of these tumors, about 75% are benign and 25% are malignant; of the primary cardiac neoplasm that are malignant, about 75% are sarcomas.2 While chemotherapy and radiotherapy have been used, neither of these relieves symptoms as well as surgical resection, and neither shows a significant increase in survival when used alone.3,4,5,6 Left-sided sarcomas represent a special problem because of the posterior location of the commonly seen left atrial site and difficult approach to intracavitary left ventricular tumors. Surgical approaches include a standard left atrial approach through the interatrial groove, orthotopic cardiac transplantation or cardiac explamation, tumor removal and cardiac autotransplantation. Standard left atrial approaches usually work for benign disease but rarely give adequate exposure for complete radical resection of left atrial or left ventricular malignant tumors or very large benign tumors. Orthotopic cardiac transplantation is hindered by the lack of donor organs, the standard morbidity and mortality associated with orthotopic transplantation and immunosuppression, and the unknown effect of immunosuppression on possible residual malignant disease. Cardiac autotransplantation has been used successfully for benign tumors,7 mitral valve surgery8 and atrial fibrillation,9 and in our opinion, it offers the best approach to complex tumors, as well.

In 1985, Cooley and Reardon attempted to use this procedure for primary malignant cardiac tumor, but that patient did not survive.10 At the Methodist DeBakey Heart Center, the first

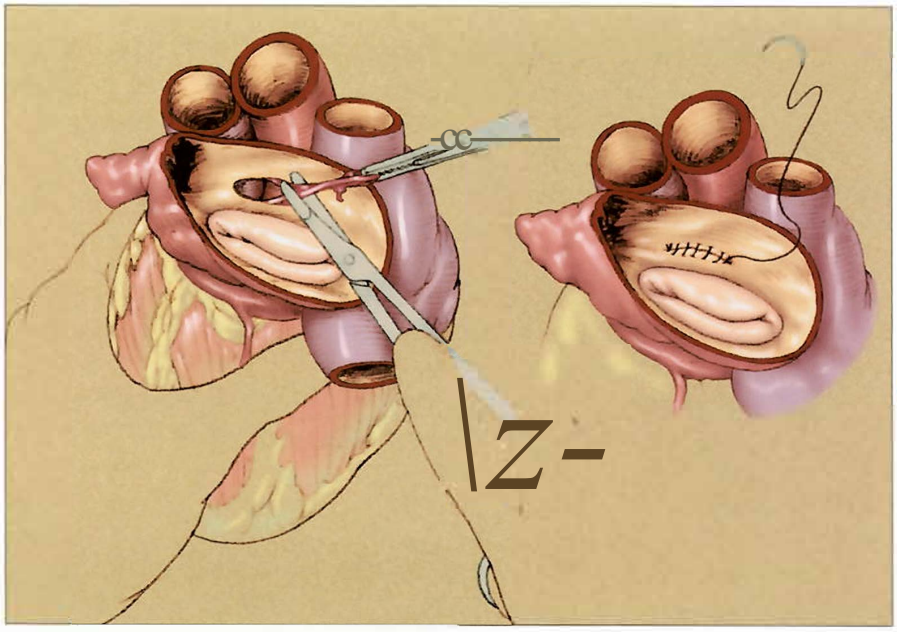

Figure 4. Additional anterior left atrial wall implant resected

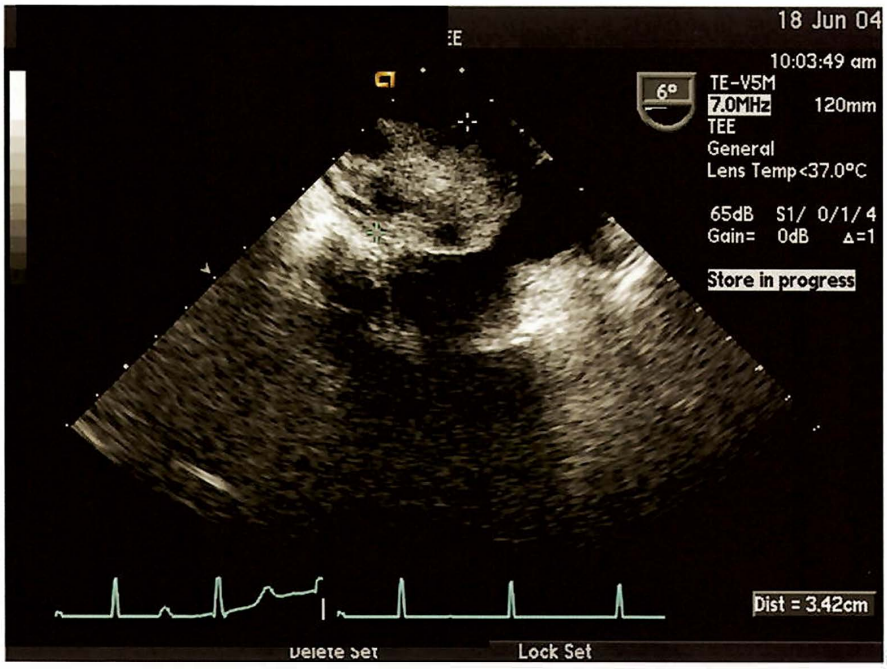

Figure 5.
4.5 cm x 3.5 cm immoblile mass adherent to the interatrial septum

successful cardiac autorransplantation for primary malignant cardiac rumor occurring in the left atrium was done in 1999,2 and the first aucocransplant for intracavitary left ventricular sarcoma was done in 2003 with the patient doing well and currently free of tumor. We successfully have performed 12 consecutive cardiac aucouansplantations for left atrial and left ventricular tumors with no hospital mortality and with nine current survivors. The current patient represents a five-year survival from his initial aurotransplantation for sarcoma, the first successful redo cardiac autotransplantation, and the first cardiac autocransplantarion combined with pneumonectomy.

In conclusion, we believe that primary cardiac sarcoma should be resected to relieve symptoms, control local tumor growth, and enhance the potential for long-term survival when combined with appropriate adjunctive chemotherapy. Complete radical excision is greatly aided by the technique of cardiac autotransplancation.

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