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Cardiac Autotransplantation for Removal of Left Atrial Hemangioma and a Review of the Literature

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ABSTRACT

We describe the management of a patient who presented with symptoms of severe congestive heart failure. A 48-year-old man was initially seen in the emergency room, admitted to the hospital, and worked up with a transthoracic echocardiogram, a transesophageal echocardiogram, and a computer tomography scan of the chest. All cardiac valves were normal, as was the left ventricular ejection fraction. A mobile left atrial tumor measuring 6 × 4 × 5 cm was found attached to the left atrial dome, left atrial cuff, and left pulmonary veins. With each systolic atrial contraction, the mass prolapsed into the left ventricle across the mitral valve annulus, inducing a gradient of 19 mm Hg. The workup of the patient was negative for malignancy. The only feasible therapy for this patient was to excise the mass on cardiopulmonary bypass and cardioplegic arrest. At the time of surgery, the findings confirmed that the mass was attached broadly to the left atrial dome wall–epicardium, and the attachments were similar to those of the transesophageal echocardiographic findings. Atrial attachments extended from the base of the heart, along the atrioventricular groove, the left dome of the left atrium, the left atrial cuff, and the anterior aspect of both left pulmonary veins. The tumor could not be adequately excised, and reconstruction of the defect was not feasible with the heart in situ. We therefore decided to explant the heart and excise the tumor with a 0.5-cm margin of healthy tissue. The broad left atrial defect was reconstructed with bovine pericardium. The reconstruction encompassed the dome of the left atrium, the left atrial cuff, and the pulmonary veins. The heart was reimplanted back into the pericardial cavity. The superior vena cava with the retained sinus node was also anastomosed. The pathology diagnosis was a benign cavernous hemangioma. The sinus rhythm recovered following removal of the aortic cross-clamp and reperfusion of the heart. The patient had a rapid recovery and was discharged home on the 12th postoperative day. Placement of a pacemaker was not required because the patient retained the sinus rhythm. A review of the literature on cardiac autotransplantation revealed that this type of surgery has been performed frequently in centers that have a cardiac transplantation program or a surgeon who has cardiac transplantation experience. To our knowledge, this report is the first of cardiac autotransplantation for benign hemangioma.

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INTRODUCTION

Primary cardiac tumors are rare clinical entities. Autopsy series reveal that primary tumors of the heart occur with an incidence of 0.0017% to 0.28% [Lam 1993; Reynen 1996]. Surgical resection is a standard treatment for patients who have obstructive symptoms and in whom complete resection can be achieved [Reardon 2006a]. In the largest Mayo Clinic series, 323 of 329 patients with primary cardiac tumors were surgically treated, with only 6 (2%) being treated conservatively [Elbardissi 2008]; however, some left-sided atrial tumors with a complex anatomy present a challenge to the surgeon [Reardon 2006a], who has to resect the tumor with a margin of healthy myocardium while preserving overall cardiac function [Okita 1994; Colucci 2001].

Cardiac autotransplantation (cardiac explantation, ex vivo tumor resection, reconstruction, and reimplantation) is a possible solution for selected cases. It was introduced for complex cardiac tumors of the left heart by Cooley in 1985. We present a case of cardiac autotransplantation for excision of an anatomically complex benign hemangioma of the left atrium.

CASE PRESENTATION

A 48-year-old man presenting to the emergency room with severe shortness of breath and orthopnea was admitted to the hospital. The patient's medical history was consistent with human immunodeficiency virus serology, hepatitis C, and diabetes. A physical examination confirmed the signs and symptoms of congestive heart failure. Auscultation of the heart revealed a loud pulmonic P2 and a soft first sound at the apex of the heart. Heart failure did not respond to the standard therapy with diuretics and angiotensin-converting enzyme inhibitors.

Transthoracic and transesophageal echocardiograms clearly identified the presence of a mobile cardiac tumor

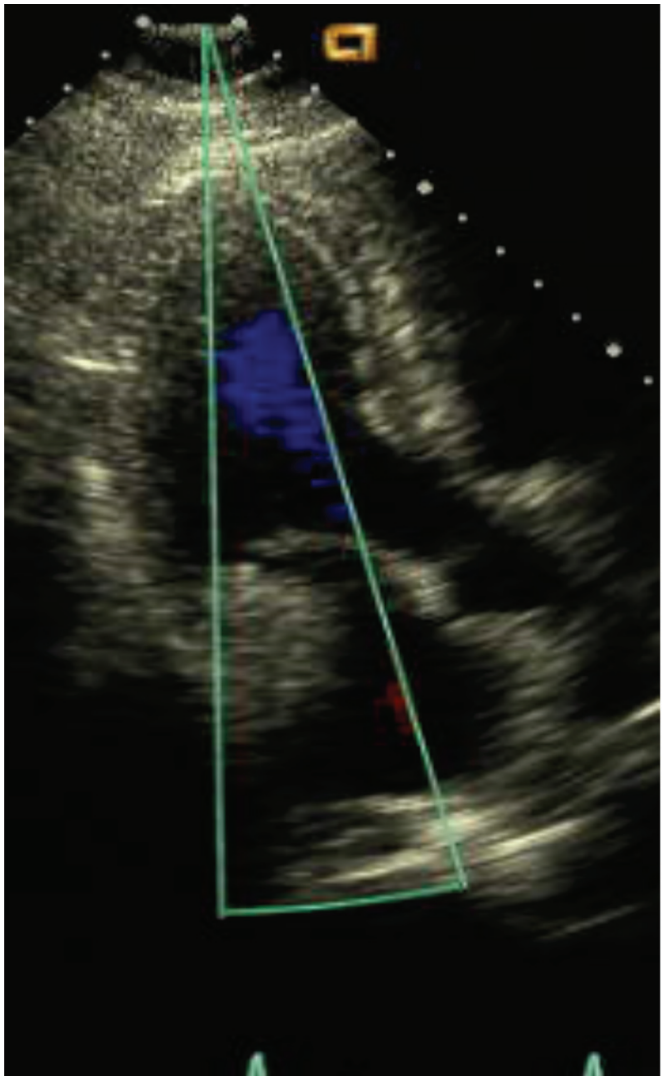

Figure 1. Apical long axis view demonstrating the left atrial tumor above the mitral valve.

measuring 6 × 4 × 5 cm (Figures 1 and 2). The tumor prolapsed into the left ventricle through the mitral valve with each atrial systole, creating a mitral stenosis physiology and inducing a mitral valve gradient of 19 mm Hg. The mass was located in the left atrium, with the attachment of the base extending from the dome of the left atrium into the left atrial cuff and anterior to the left pulmonary veins. The left ventricular ejection fraction was normal.

Cardiac catheterization revealed the patient to have normal coronary arteries, an elevated pulmonary pressure, an elevated right ventricular systolic pressure, and mild elevation of the right atrial pressure. He was referred for excision of the cardiac tumor.

The main concern was the extent of the tumor implantation, which transesophageal echocardiography revealed to extend from the posterior aorta along the dome of the left atrium into the lateral aspect of the annulus of the mitral

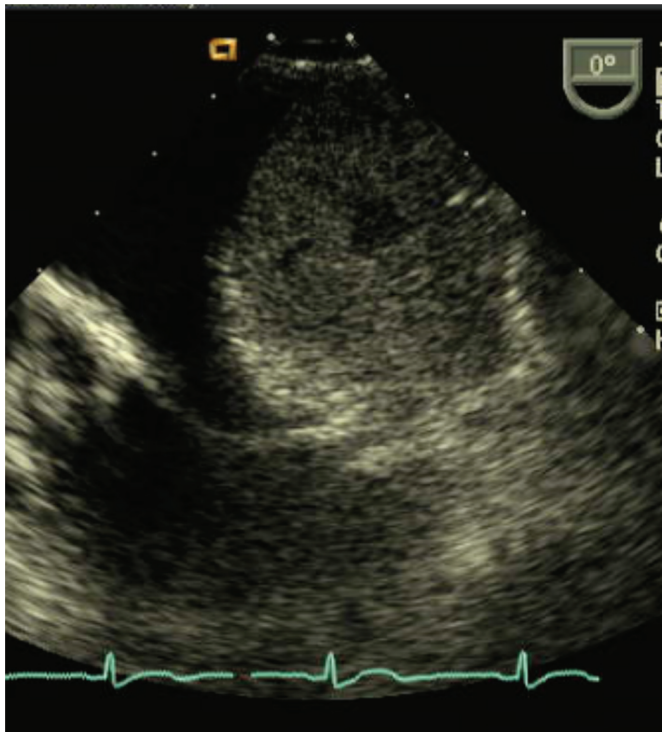

Figure 2. Transesophageal echocardiographic view of left atrial mass seen obstructing the mitral valve.

valve. The extent of the tumor would appear to preclude excision and reconstruction with the heart in situ.

SURGICAL MANAGEMENT

A standard midline sternotomy was performed, and the pericardium was incised. Palpation along the left side of the pulmonary artery immediately revealed the presence of a large rubbery mass that compressed the pulmonary artery anteriorly. The heart was displaced anteriorly and rotated toward the right. Lifting of the heart allowed visualization of the entire tumor bulging into the pericardial cavity, and with the retracted sternum, it became evident that the tumor was not intracardiac but rather covered by a smooth epicardial lining. The extent of the base attached to the heart was assessed. Anteriorly, the attachment extended from the left mid dome of the left atrium and posterior to the aorta–pulmonary artery and then extended laterally to the left and in front of the left atrial cuff and the anterior aspect of both left pulmonary veins. The posterior attachment could not be visualized.

The final assessment of this tumor was that it was subepicardial in nature; the edges were well demarcated. Excision of the mass with a 0.5-cm margin would require a wide exposure and reconstruction of the left atrium, for which cardiac explantation was required. Therefore, we decided on (1) cardiac explantation with retention of the sinus node with the superior vena cava, (2) tumor excision, (3) reconstruction of the left atrial defect, and, finally (4) reimplantation of the heart as an autotransplant.

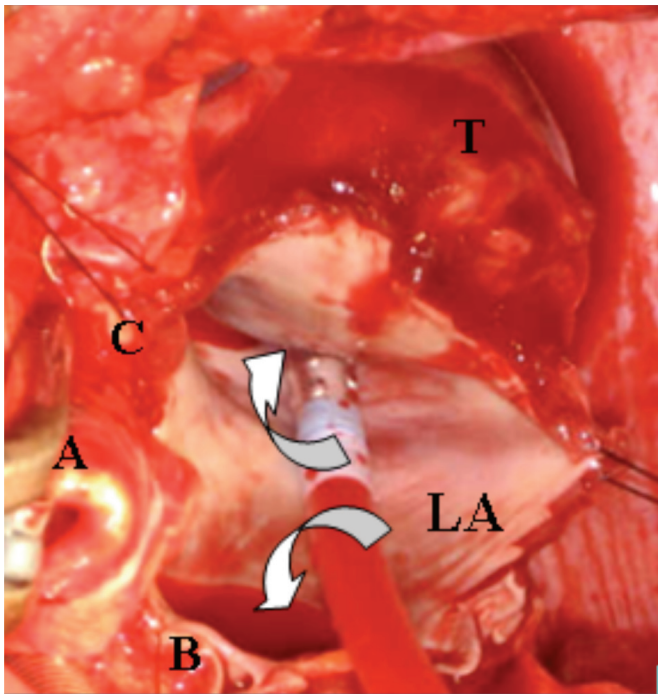

Figure 3. Excised heart removed from the pericardial cavity. The left atrium is widely opened. The tumor (T) is well visualized. The suction device has been placed within the left atrial cuff and the pulmonary vein. The left atrial cuff incision has been completed. Indicated are the aortic clamp and displaced transected aorta (A), interatrial septum (B), retracted left atrial dome (C), and posterior left atrial wall (LA). The arrows indicate openings of the right and left atrial cuffs.

Figure 4. Left atrial view of the completely excised left atrial tumor.

Cannulation of the superior vena cava was distal to the sinus node at the level of the azygos vein. All of the remaining cannulation sutures were placed as for an orthotopic heart transplant. The patient was heparinized, and the distal ascending aorta, distal superior vena cava, and inferior vena cava were cannulated. Cardiopulmonary bypass was initiated, and the core temperature was brought to 28°C. A vent was placed through the right superior pulmonary vein. The aortic cross-clamp was applied, antegrade cardioplegia was administered, and the cardiectomy was performed. The excision encompassed the anterior half of the proximal superior vena cava with retention of the sinus node. Then, the lateral wall of the right atrium was transected, and the mid intra-atrial septum was divided. The mid aorta and mid pulmonary arteries were transected. Anterior displacement of the distal aortopulmonary artery exposed the cardiac tumor. The dome of the left atrium, the pulmonary cuff, and the anterior aspect of the pulmonary veins were transected toward the atrioventricular groove. The heart was removed from the pericardial cavity and placed in a bowl of cold saline.

The second stage of the procedure consisted of excision of the tumor from the left atrium with a 0.5-cm rim of healthy atrium (Figure 3). The tumor was completely removed, with the incision of the left atrium extending along the mid dome of the left atrium posteriorly to the pericardial reflexion. The anterior left atrial cuff and both anterior left pulmonary veins were removed as a single specimen. The frozen section confirmed a benign cavernous hemangioma (Figure 4). The large atrial defect precluded direct cardiac implantation, and an extensive atrial reconstruction was performed.

Because the excision of the atrial tumor produced a large defect and precluded direct suturing of donor–recipient left atrial cuffs, the reconstruction was performed with 2 bovine pericardial patches and 4-0 polypropylene (Prolene) suture. The first patch was used to reconstruct the anterior aspect of the 2 pulmonary veins and the left atrial cuff. The suture line extended toward the pericardial reflexion. The patch had a B configuration, allowing retention of a wide lumen for both pulmonary veins.

The second pericardial patch was sutured to the edges of the defect left at the dome of the left atrium, which extended from the right side of the atrial incision and along the left atrial pericardial reflexion laterally toward the left. The first and second patches were first trimmed to obtain good apposition, avoiding redundant pericardium, and to provide a circumferential edge to which the excised atrium would be implanted. Both patches were sutured together (Figure 5).

The final result of the repair was a smooth edge that extended from the mid dome of the left atrium toward 1 cm to the caudal aspect of the left inferior pulmonary vein.

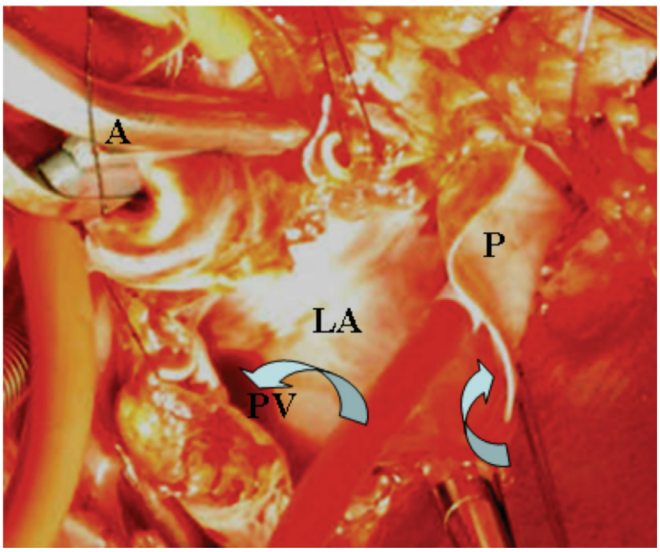

Figure 5. View similar to Figure 3. The tumor has been widely excised, the pericardial patch has been sewn to reconstruct the left pericardial cuff, and part of it has been sutured to the left atrial dome. Indicated are the aortic clamp (A), the posterior left atrial wall (LA), and the pericardial patch (P). The arrows indicate openings of the right and left atrial cuffs. PV indicates pulmonary vein.

Implantation of the heart was done as a standard cardiac autotransplantation. The suturing of the patch to the left atrium was done with extreme care; the first sutures were placed to the preshaped pericardial patch at the level of the atrioventricular groove. The heart was parachuted into the pericardial cavity, continuing along the oblique sinus and the intra-atrial septum, followed by suturing of the remaining portion of the patch to the atrial dome and the cranial aspect of the interatrial septum, completing the left atrial suture. The right atrium anastomosis was initiated at the level of the superior vena cava, tissue bites were small to avoid injury to the conduction tissue, and a second suture was initiated caudally at the interatrial wall and tied in the mid atrium. These pericardial–left atrial suture lines were covered with tissue glue.

The aorta and the pulmonary arteries were sutured in an end-to-end fashion, the heart was deaired, the aortic cross-clamp was removed, and 100 µg of levothyroxine was administered through the heart–lung machine. Atrial and ventricular pacing wires were placed.

Once rewarming was completed, cardiopulmonary bypass was discontinued, and the venous cannulae were removed. The heart regained sinus rhythm, sustaining excellent hemodynamics. The total aortic cross-clamp time was 95 minutes, and the cardiopulmonary bypass time was 128 minutes. The patient was discharged home on the 12th day after surgery.

DISCUSSION

Primary cardiac tumors are uncommon. Approximately 75% to 94% of primary cardiac tumors are benign [Bakaeen 2003; Blackmon 2008; Elbardissi 2008] and include myxoma, papillary fibroelastoma, lipoma, and hemangioma. Six percent to 25% of cardiac tumors, representing mostly sarcomas, are malignant. Hemangiomas are rare. In a Mayo Clinic series, only 7 (2.2%) of 323 tumors were hemangiomas, whereas only 1 of 85 primary cardiac tumors from the Methodist DeBakey Heart Center represented hemangioma [Bakaeen 2003]. Lam et al [1993] reported 2 hemangiomas among 7 primary cardiac tumors. None of 26 patients with benign primary cardiac tumors in the University of California series had hemangioma [Odim 2003].

Most primary cardiac tumors are asymptomatic, although patients may present with thromboembolism, symptoms of congestive heart failure [Rammos 2007], chest discomfort, arrhythmias, conduction disturbances, pericardial effusion, coronary insufficiency, or outflow tract obstruction [Odim 2003]. Sudden cardiac death consequent to conduction disturbances or rupture and tamponade has been reported [Burke 1990]. Our patient's presentation with dyspnea, orthopnea, and signs of congestion was therefore quite typical.

The most useful diagnostic tool is the echocardiogram, which precisely locates the tumor and defines its extent in almost all cases. The echocardiographic appearance may also allow quite accurate prediction of the tumor type and whether it is malignant or benign. Magnetic resonance imaging serves as the next most important test, in which the density of the images may allow identification of tumor cell type [Vander Salm 2000].

Tumor resection is a standard surgical treatment for cardiac tumors, and autotransplantation is rarely used.

Historically, cardiac autotransplantation was initially used for treatment of Prinzmetal angina [Clark 1977; Bertrand 1981]. The idea was to provide a complete denervation of the heart and therefore to eliminate intractable chest pain. This practice was soon discontinued because of high morbidity and mortality, but the technique was used for surgical removal of extensive and posteriorly located cardiac tumors that were difficult for standard surgical resection.

In the experience of the Methodist DeBakey Heart Center, where 85 patients with primary cardiac tumors have been treated, only 5 (5.4%) of 93 operations were autotransplantations, all of which were performed for malignant cardiac tumors, including 3 malignant fibrous histiocytomas, 1 leiomyosarcoma, and 1 undifferentiated sarcoma [Bakaeen 2003]. Another report from the Texas Heart Institute summarized this institution's experience with 21 autotransplantations performed in 20 patients with malignant or complex benign cardiac tumors unresectable by a traditional approach. Of the 20 patients, 17 had malignant lesions, and 3 had benign disease. Two patients had left ventricular lesions, and the rest had left atrial lesions. Histologic analyses revealed 7 malignant fibrous histiocytomas, 5 undifferentiated sarcomas, 3 leiomyosarcomas, 1 malignant osteosarcoma, 1 myxoid sarcoma, 2 paragangliomas, and 1 myxoma [Blackmon 2008]. Other case reports of cardiac autotransplantation have described left atrial sarcoma [Reardon 1999; Iskander 2005; Doty 2006], left ventricular sarcoma [Reardon 2006b], recurrent or large left atrial myxoma [Scheld 1988; Rosetti 2006; Gammie 2007],

and malignant fibrous histiocytoma [Wagner 1999; Wang 2006]. Our report is the first of cardiac autotransplantation performed for benign cavernous hemangioma.

The autotransplantation approach provides wide visualization, which allows complete resection of left atrial tumors [Lam 1993; Reardon 2006a] and achievement of tumor-free margins, as well as good reconstruction of the atrial defect. We therefore performed a complete cardiac explantation, ex vivo tumor resection, reconstruction of the created defect with bovine pericardium, and subsequent cardiac reimplantation.

A transaortic approach has been used for left ventricular tumors [Kawada 1996], but this approach may provide insufficient exposure. Ventriculotomy [Rammos 2007] has been used as in the transmitral valve approach [Kudo 2004; Kurian 2006], with the heart in situ or explanted [Elbardissi 2008], and it has provided excellent exposure for complete resection of intraventricular tumors and reconstruction of the interventricular septum [Troise 2003].

Although orthotopic cardiac transplantation has been performed to treat patients with benign and malignant tumors [Michler 1997], drawbacks include the lack of organ availability and problems related to immunosuppression [Gowdamarajan 2000; Catton 2008]. In an experienced surgical center, cardiac autotransplantation is a viable option for patients who have otherwise nonresectable cardiac tumors, especially those that are posteriorly located.

Cardiac autotransplantation has been used for the management of various conditions, including Prinzmetal angina [Clark 1977; Bertrand 1981], complex congenital cardiac defects [Rosetti 2006], atrial fibrillation with mitral valve surgery [Troise 2003], long QT syndrome [Pfeiffer 1992], mitral valve replacement [Novitzky 2003; Wang 2006], giant left atrium [Livi 1998; Gordeev 2006; Barbukhatti 2009], and atrioventricular disruption after mitral valve replacement [El-Essawi 2007]. Autotransplantation is a useful surgical technique for complete excision of tumors not accessible via standard procedures. This surgery should be performed in heart transplantation centers and/or by surgeons with a cardiac transplantation background.

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