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CASE REPORT

INTERMEDIATE

CLINICAL CASE

Recurrent Cardiac Sarcoma Resection With Modified Partial Autotransplantation

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ABSTRACT

Primary cardiac sarcomas are very aggressive, being a challenge to cardio-oncology specialty. Surgical planning and innovative techniques have enhanced the possibility of resection. We have described a case of recurrent primary left atrium angiosarcoma, successfully resected with a modified partial autotransplantation technique, planned using a 3-dimensional model created from computed tomography. (**Level of Difficulty: Intermediate.**) (J Am Coll Cardiol Case Rep 2021;3:1694–1699) © 2021 The Authors. Published by Elsevier on behalf of the American College of Cardiology Foundation. This is an open access article under the CC BY-NC-ND license (http://creativecommons.org/licenses/by-nc-nd/4.0/).

HISTORY OF PRESENTATION

A previously healthy 32-year-old man had a history of sudden-onset atypical exertional chest pain, with no associated symptoms. Pain was usually relieved with nonsteroidal anti-inflammatory drugs. There was no dyspnea, loss of appetite, weight loss, or fever. After approximately a month with recurrent episodes of chest pain that occurred weekly, the patient sought medical attention. On physical examination, the patient was in good health status and hemodynamically stable, breathing comfortably with no rales.

LEARNING OBJECTIVES

- To revise a case of primary, recurrent cardiac sarcoma through diagnostic imaging.
- To understand the surgical planning using CT with 3-dimensional printing.
- To learn the technique of modified partial autotransplantation versus complete autotransplantation in the treatment of primary cardiac sarcomas.

PAST MEDICAL HISTORY

The patient had a hospital admission elsewhere 7 months earlier, due to an episode of syncope. At that time, he was diagnosed by 2-dimensional echocardiography (2D TTE) with a large intra left atrial mobile mass, which was presumably interpreted as being a cardiac myxoma, particularly because of embolization, and for being the most likely primary cardiac tumor at this location. He was submitted to a minimally invasive thoracoscopic resection under cardiopulmonary bypass (CPB). However, the surgeon who performed this operation described that the tumor was larger than expected with invasion to the right and left superior pulmonary veins, and complete resection was not possible. Pathology revealed a

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high-grade pleomorphic primary cardiac sarcoma. Neoadjuvant chemotherapy with epirubicin and ifosfamide was initiated with the intention of tumor downstaging. On the other hand, there was tumor progression despite cytotoxic therapy.

DIFFERENTIAL DIAGNOSIS

In the presence of a malignant cardiac tumor, the differential diagnosis between primary or secondary cardiac tumors is imperative. A complete workup excluded the differential diagnosis of a metastatic tumor.

INVESTIGATIONS

The 2D TTE revealed the presence of an intracardiac mass adherent to the left atrium roof, divided into 2 components: a fixed component measuring 29 mm, and another mobile component measuring 41 mm that prolapsed into the left ventricle through the mitral valve during diastole.

Cardiac magnetic resonance imaging (CMRI) (**Figure 1**) showed an infiltrative mass in the superior, posterior, and lateral walls of the left atrium, corresponding to approximately 40% of the left atrial cavity. The mass measured 48 mm × 26 mm in diameter, being very close to the cardiac fibrous body. Computed tomography (CT) revealed that there was invasion to the noncoronary aortic annulus and the anterior mitral annulus. The mass also invaded circumferentially the left superior pulmonary vein, and it was very close to the orifice of the right superior pulmonary vein. There was invasion of the epicardial fat of the superior left atrial wall, extending close to the pulmonary trunk posteriorly. This particular case was turned down by the original surgeon who operated this patient, and by 3 other surgeons who judged the case to be inoperable.

Based on findings of the CT, a 3-dimensional reconstruction (**Figure 2**) was performed using a commercial workstation (Intellspace Portal, Philips), and the model was exported in STL format. Different 3-dimensional cardiac models (**Figure 3**) were printed using an ABS filament printer (ForceONE, 3dLAB). Those provided our team with a thorough assessment of tumor size, location, and relation to adjacent structures. The models guided us in deciding to operate, to obtain patient consent, and to determine prognosis. The advanced imaging suggested that there was invasion of the fibrous trigone and aortic annulus, which would create a low likelihood of complete resection with safe margins. This issue was discussed by the cardio-oncology team. We considered that cytoreduction was justified in this scenario because there were no other alternatives, particularly with the potential mitral valve obstruction or tumor embolization and sudden death. The patient was young with no comorbidities and no metastasis.

MANAGEMENT

The patient was submitted to reoperative cardiac surgery (**Figure 4,** Videos 1 and 2) under general anesthesia and median sternotomy. CPB was instituted through aortic and bicaval cannulation.

ABBREVIATIONS AND ACRONYMS

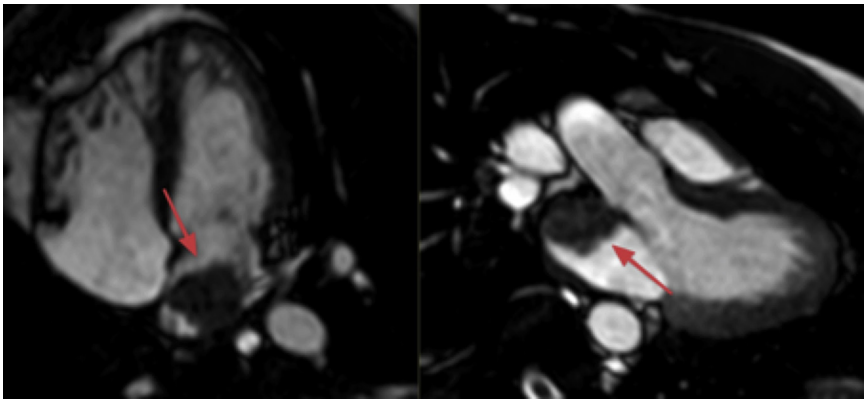
CMRI = cardiac magnetic resonance imaging

CPB = cardiopulmonary bypass

CT = computed tomography

2D TTE = 2-dimensional echocardiography

FIGURE 1 Magnetic Resonance



Magnetic resonance 4-chamber view and the left ventricular outflow tract showing lesion in the left atrial posterior wall **(arrows)**, in close relation to the fibrous trigone.

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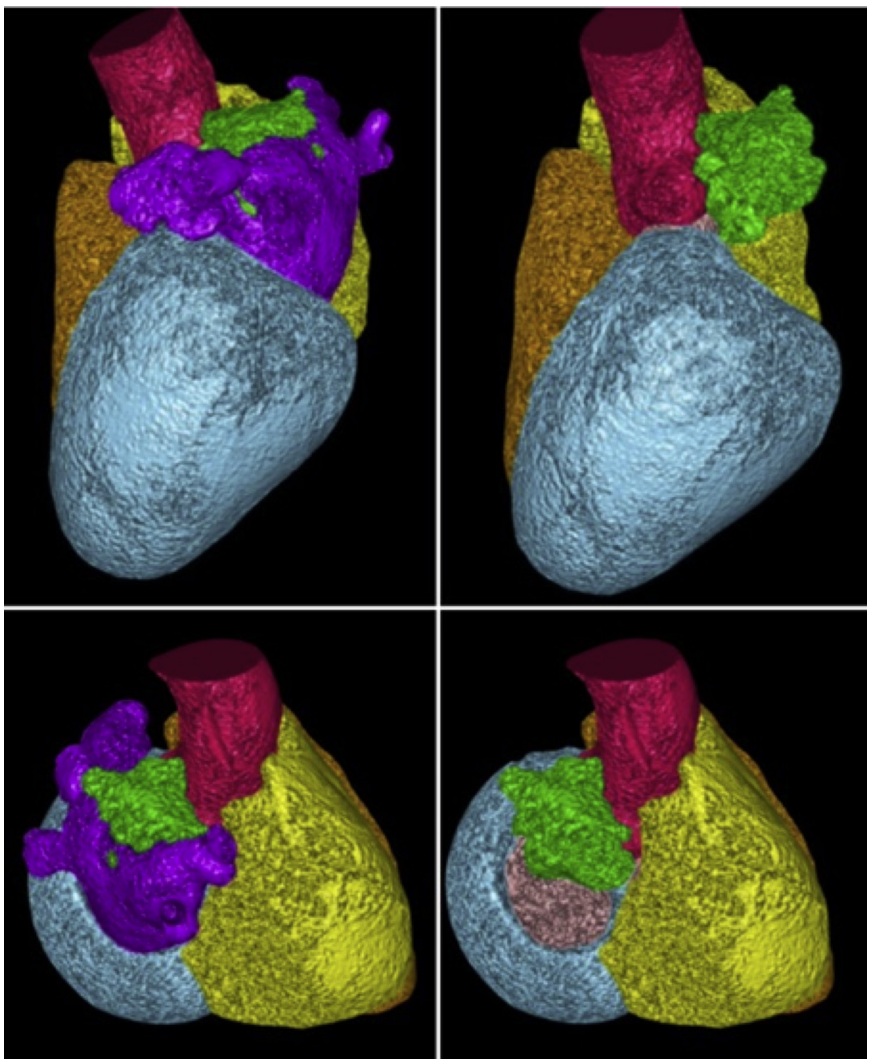
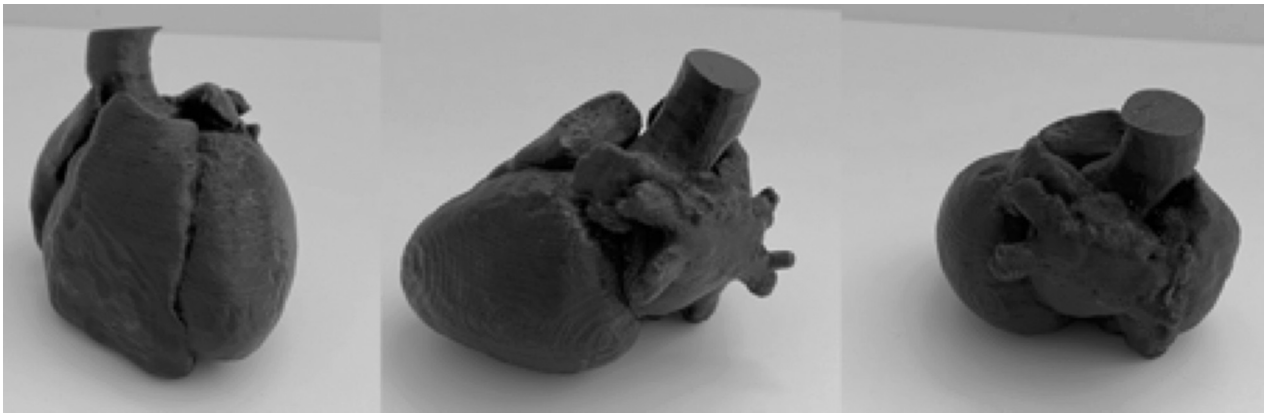
FIGURE 2 Computed Tomography



Three-dimensional model created from computed tomography acquisition, delineating relations between the tumor and the cardiac structures. Left atrium in **purple** and the tumor in **green**.

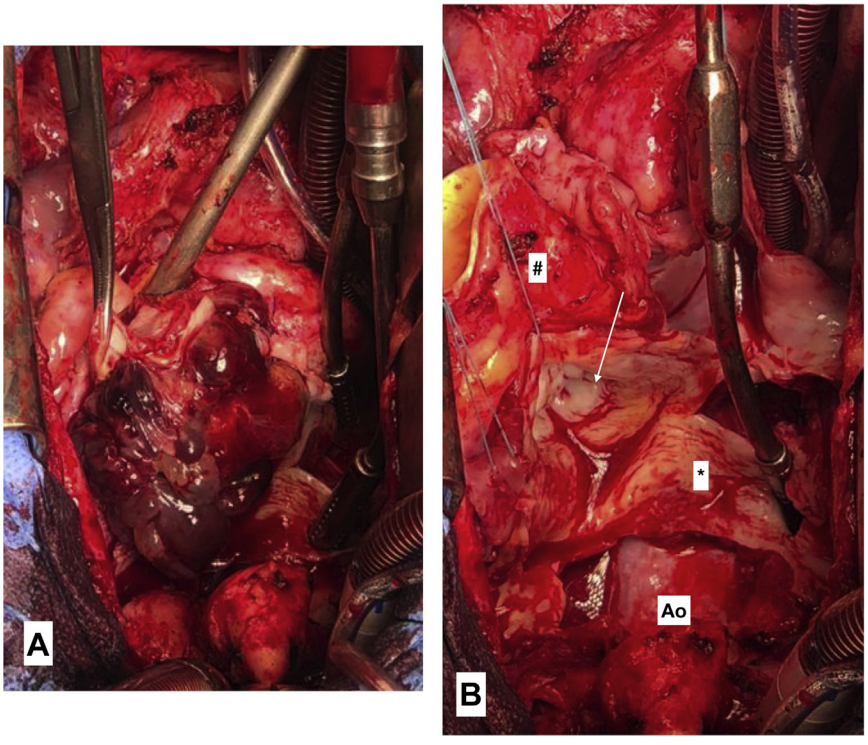
Myocardial protection was through intermittent cold blood cardioplegia delivered every 20 minutes in the coronary ostia. The intracardiac tumor (Figure 5) was disclosed easily after left atriotomy through the right superior pulmonary vein. To adequately expose the left atrium, we performed complete transection of the superior vena cava, ascending aorta, and pulmonary trunk. We then identified that the tumor was adherent to the superior wall of the left atrium invading the lumen of the right and left superior pulmonary veins and to the interatrial septum, with a slight free margin in the anterior mitral valve annulus. The tumor did not invade the heart skeleton, or the aortic root, which allowed us to completely resect it en bloc with free margins. Reconstruction of the left atrium was performed with 2 pieces of bovine pericardium: one quadrangular-shaped in the superior wall up to both superior pulmonary veins and a second one diamond-shaped in the superior interatrial septum up to the anterior mitral valve annulus. Afterward, there was primary closure of the right atrium, pulmonary artery, aorta, and superior vena cava. Weaning from CPB occurred without difficulty. Intraoperative transesophageal

FIGURE 3 3-Dimensional Model



Three-dimensional printing showing the tumor in relation to the cardiac structures.

FIGURE 4 Intraoperative Photographs



Intraoperative photographs showing the excised primary cardiac sarcoma **(A)**. **(B)** Surgical aspect of the heart after tumor removal, showing the mitral valve **(arrow)**, posterior left atrium **(*)**, orifice of the right pulmonary veins (metallic sucker), posterior wall of the transected ascending aorta **(#)**, and the distal portion of the ascending aorta (Ao).

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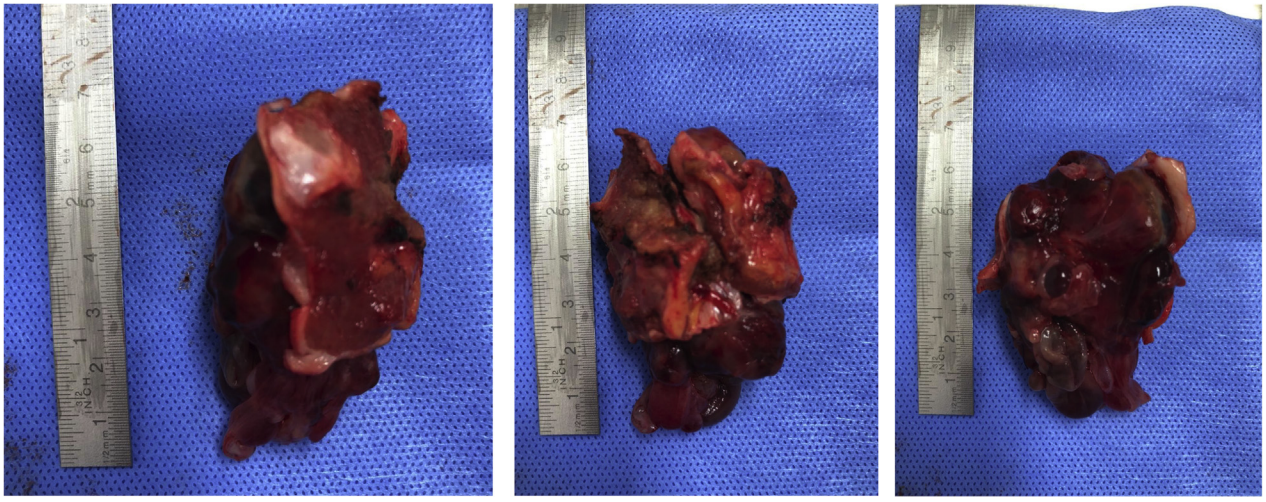
FIGURE 5 Excised Cardiac Sarcoma



Different views of the excised cardiac sarcoma with the attached atrial wall (whitish tissue).

echocardiography confirmed the good surgical result with complete tumor resection with no significant residual problems.

Pathology confirmed the diagnosis of high-grade pleomorphic sarcoma, exhibiting osteoid formation and extensive necrosis.

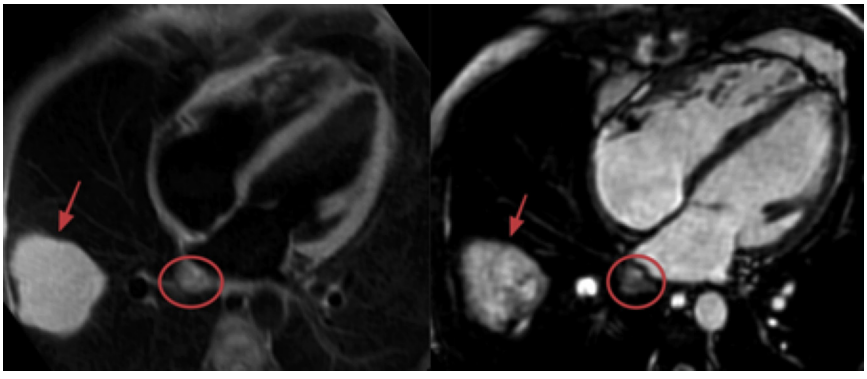
DISCUSSION

Primary cardiac tumors are rare. Autopsy studies (1) show a prevalence of 0.001% to 0.03%, being a quarter malignant. Among malignant tumors, angiosarcoma is the most frequent (1). These tumors are very aggressive, and their prognosis is dismal, particularly by the occurrence of early hematogenic dissemination, local invasion, and resistance to chemotherapy. Surgical resection is the treatment of choice.

Diagnosis of cardiac sarcomas is initially determined by 2D TTE with a sensibility of 75% (2). To provide a more detailed anatomy, particularly if surgical treatment is being considered, CT and CMRI (3) are important to establish tumor characteristics, invasiveness, exact location and tumor relation to adjacent structures, and staging for possible metastases.

There are few reports of the usefulness of 3-dimensional printing after acquisition of processed

FIGURE 6 Recurrent Mass



Magnetic resonance imaging T2 and cine steady-state free precession 4-chamber view demonstrating recurrent mass **(circles)**. **Arrows** represent lung metastasis after the second surgery.

complex radiologic imaging in the surgical planning of cardiac surgery (4). Three-dimensional printing aids in preoperative planning and surgical training, allowing a more focused approach that minimizes CPB times (4). The technology used in this case is considered of low cost. The image was edited in a commercial workstation, highly available in every radiology laboratory. We used a low-cost filament printer, with satisfactory results. On the other hand, discrepancies between imaging and surgical findings occurred in this case. These discrepancies would have been prevented if we had used CMRI with contrast-enhanced steady-state free precession cines in A4C and A2C instead of static images, which would enhance our ability to appreciate possible infiltration into the adjacent structures. Our initial plan was to perform a partial resection because advanced imaging showed tumor invasion of the fibrous skeleton of the heart. In this scenario, partial resection is justified by the impending hemodynamic compromise or tumor embolization in a young patient with no comorbidities and no response to chemotherapy (5). Fortunately, the initial plan was modified during the operation that allowed en bloc resection.

Complete tumor surgical resection remains the treatment of choice in cardiac sarcomas. Autotransplantation, which consists of heart explant, tumor resection with free margins, reconstruction of the left atrium, and heart reimplantation, is a safe and effective technique used by experienced surgeons, with an acceptable hospital mortality (6). In this case, complete heart explant was not necessary. Advanced imaging was crucial for this particular surgical planning. Tumor location free from the posterior left atrium and the inferior pulmonary veins allowed us to proceed with partial autotransplantation, minimizing dissection of the left ventricle and left atrium, and shortening CPB times. Surgical exposure keeping the inferior vena cava and the inferior pulmonary veins intact with retraction of the heart caudally was adequate for a complete excision of the tumor and appropriate reconstruction of the left atrium with bovine pericardium.

FOLLOW-UP

At 6-month follow-up, the patient remains asymptomatic with normal ventricular function and in junctional rhythm, with rare episodes of ectopic atrial rhythm.

Control CMRI revealed no intracardiac mass. There is a recently diagnosed solid nodular mass adherent to the right lung hilum measuring 18 mm × 17 mm that is considered progression of the primary tumor (**Figure 6**).

CONCLUSIONS

Cardiac sarcomas are very aggressive with high local invasion. Surgical treatment is challenging, especially because incomplete resection invariably results in early local recurrence (7).

Complex surgical techniques and innovative diagnostic technology have enhanced the likelihood of tumor resection, with a positive impact on patient survival and better quality of life.

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KEY WORDS cancer, cardiac transplant, computed tomography, 3-dimensional imaging, 3-dimensional printing

APPENDIX For supplemental videos, please see the online version of this paper.