

Resection of a synovial cell sarcoma by cardiac autotransplantation: A case report



Danielle M. Mullis, BS,^a Yuanjia Zhu, MD, MS,^{a,b} Brandon A. Guenthart, MD,^a Spencer A. Bonham, BA,^a Winston L. Trope, BE,^a Gerald J. Berry, MD,^c Y. Joseph Woo, MD,^{a,b} and John W. MacArthur, MD,^a Stanford, Calif

From the Departments of ^aCardiothoracic Surgery, ^bBioengineering, and ^cPathology, Stanford University, Stanford, Calif.

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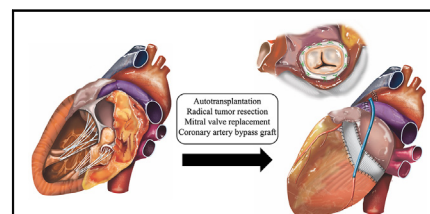
Address for reprints: John W. MacArthur, MD, Department of Cardiothoracic Surgery, Falk Cardiovascular Research Center, Stanford University School of Medicine, 300 Pasteur Dr, Stanford, CA 94305 (E-mail: jwm2108@stanford.edu).

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Patient with a large cardiac synovial sarcoma underwent a cardiac autotransplantation.

CENTRAL MESSAGE

Autotransplantation can be used as a technique to allow for safe tumor resection when a cardiac tumor involves both the left atrium and left ventricle.

▶ Video clip is available online.

The patient provided written informed consent for publication of this report. A previously healthy 25-year-old male patient presenting with sharp pleuritic chest pain was found to have an intracardiac mass. A transthoracic echocardiogram (TTE) revealed a 3- × 2-cm echolucent structure within the posterolateral myocardium at the level of the mitral annulus and a 1.1- × 1.1-cm rounded echo density adherent to the atrial surface of the posterior mitral valve (MV). Cardiac magnetic resonance imaging (MRI) showed a 4- × 5-cm ovoid mass in the left atrium (LA), extending into the left ventricle (LV) and into the pericardial space. Computed tomography angiography with intravenous contrast was also performed (Figure 1, A). A positron emission tomography scan showed a fluorodeoxyglucose-avid hypermetabolic cardiac mass, subcarinal nodes, and lung nodes.

The patient underwent a transbronchial biopsy showing synovial sarcoma in the LV (Figure 2, A), but no evidence of metastatic disease, with negative level 4R and level 7 lymph nodes. A fluorescence in situ hybridization study showed SS18 gene rearrangement, which is indicative of synovial sarcoma (Figure 2, B). The patient's case was reviewed at a tumor board by a multidisciplinary team including a sarcoma

oncologist. Their recommendation was upfront surgery followed by adjuvant chemotherapy. The cardiac surgery team determined that an autotransplantation would likely be the best approach given the extent of the tumor and the patient's age.

The patient was taken to the operating room and underwent a median sternotomy and pericardiotomy (Video 1). A saphenous vein was harvested due to the proximity of the tumor to the left circumflex coronary artery. The distal ascending aorta and superior vena cava were directly cannulated, and an inferior vena cava (IVC) cannula was placed via the right femoral vein. Cardiopulmonary bypass was initiated, and the LA vent was placed. After aortic cross-clamp and cardiac arrest via antegrade cardioplegia, the heart was resected by dividing the superior vena cava, right atrium/IVC junction, aorta, and pulmonary artery. When the LA was divided, enough atrial tissue was retained between the left side of the pulmonary vein and the tumor to allow for anastomosis during reimplantation.

Once the heart was placed on ice, the LA mass adjacent to the posterior mitral annulus was identified. The LA was transected towards the anterior and posterior commissures of the MV and its chordal attachments. The posterior MV and its chordal attachments were excised. After close examination, the mass inside the LV was meticulously dissected away from healthy-appearing tissue. The resection spanned

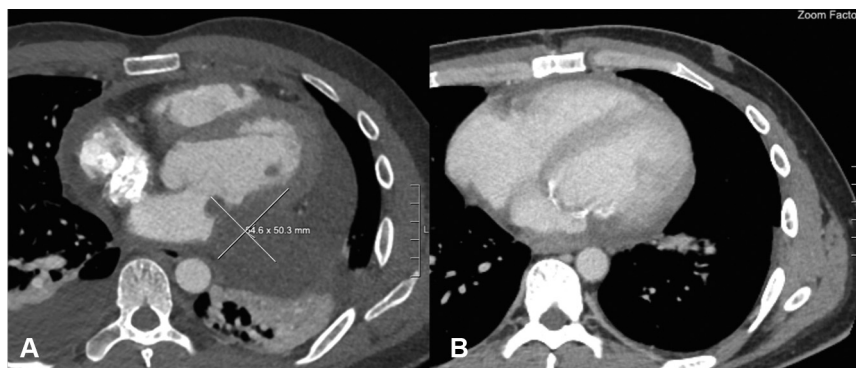


FIGURE 1. CT of the chest with intravenous contrast. A, CT angiography with intravenous contrast showing the 5.5- × 5.0-cm mass before surgery. B, CT of the chest with intravenous contrast taken after surgery. CT, Computed tomography.

from the anterior commissure of the MV, under the mitral annulus of the atrioventricular groove, to the posterior commissure of the MV. The specimen was sent to pathology.

The left circumflex artery was ligated proximally near the left atrial appendage. A 1-cm piece of felt was placed along the resection line inside the LV to reconstruct the mitral annulus and the neoatrioventricular groove. Interrupted 2-0 sutures were placed through the felt, through the endocardial LV, out of the epicardial LV, and through the Dacron sheet. The Dacron sheet was used to reconstruct the posterior LV, LA, and atrioventricular groove (Figure E1). The anterior leaflet was removed from the annulus and positioned onto the neoposterior annulus. Then, pledgeted polyester sutures were placed around the annulus and placed through the sewing ring of the 33-mm Epic MV ring (St Jude Medical, Inc). The valve was lowered onto the annulus and the sutures were secured. The Dacron sheet was sutured with running 4-0 polypropylene suture. An end-to-side distal anastomosis was performed between the first obtuse marginal artery and an endoscopically harvested saphenous vein graft. The sutures were tied, and the heart was placed back into the chest.

The left atrial anastomosis was completed with a running 3-0 polypropylene suture, using the Dacron sheet as the posterior LA. The IVC was anastomosed, followed by the pulmonary artery and aorta, while also deairing the heart. The crossclamp was removed, and the heart was reperfused. The heart string device was inserted into the ascending aorta and the saphenous vein graft was anastomosed proximally in an end-to-side fashion. There was no intraoperative bleeding from the atrial or ventricular suture lines. The heart was optimized, and the patient was carefully weaned from cardiopulmonary bypass. The patient was transferred to the intensive care unit in sinus rhythm.

The total cardiopulmonary bypass time was 193 minutes, and the total crossclamp time was 141 minutes. The cardiac mass biopsy report showed that the tumor was a grade

310 × 6.5 × 4.5-cm monophasic synovial sarcoma weighing 119.1 g (Figure 2, C). The neoplasm was microscopically present at the resection margins. One week after the procedure, TTE revealed no mitral regurgitation with an estimated left ventricular ejection fraction of 58%. The patient was discharged 8 days after his surgery. One month later, TTE demonstrated no mitral regurgitation, normal-sized ventricles, and a left ventricular ejection fraction of 58%. Given the positive margins (Figure 2, D), 5 cycles of adjuvant chemotherapy (doxorubicin and ifosfamide) were initiated at 1-month follow-up. As of 8 months after the operation, the patient was doing well. He was physically active and walking 2 to 3 miles each day.

DISCUSSION

Synovial sarcoma is an extremely rare soft tissue tumor more commonly found in the lower limb.¹ Cardiac synovial sarcoma typically manifests in the pericardium and right atrium, which means there is little precedence for how to treat left-sided cardiac synovial sarcomas.¹ Furthermore, a tumor that expands from the LA into the LV poses an extreme challenge to surgeons due to poor exposure in the chest.²

Autotransplantation is a technique that allows for enough visibility to remove complicated and large left heart tumors.³ However, it is still an extremely rare procedure that requires tremendous surgical expertise and a medical center equipped to handle complex cases. The gold standard to treat cardiac synovial sarcoma is a complete surgical resection.⁴ In our patient's case, the attempt was to achieve a curative R0 resection. During the procedure, a significant portion of the LV was excised, and surgeons felt they would be unable to resect any further LV muscle without drastically reducing cardiac function. Margins were not sent during the operation to minimize ischemic time to the myocardium. Ramlawi and colleagues³ reported similar 2-year survivals for patients with both positive and negative surgical margins after autotransplantation to treat cardiac

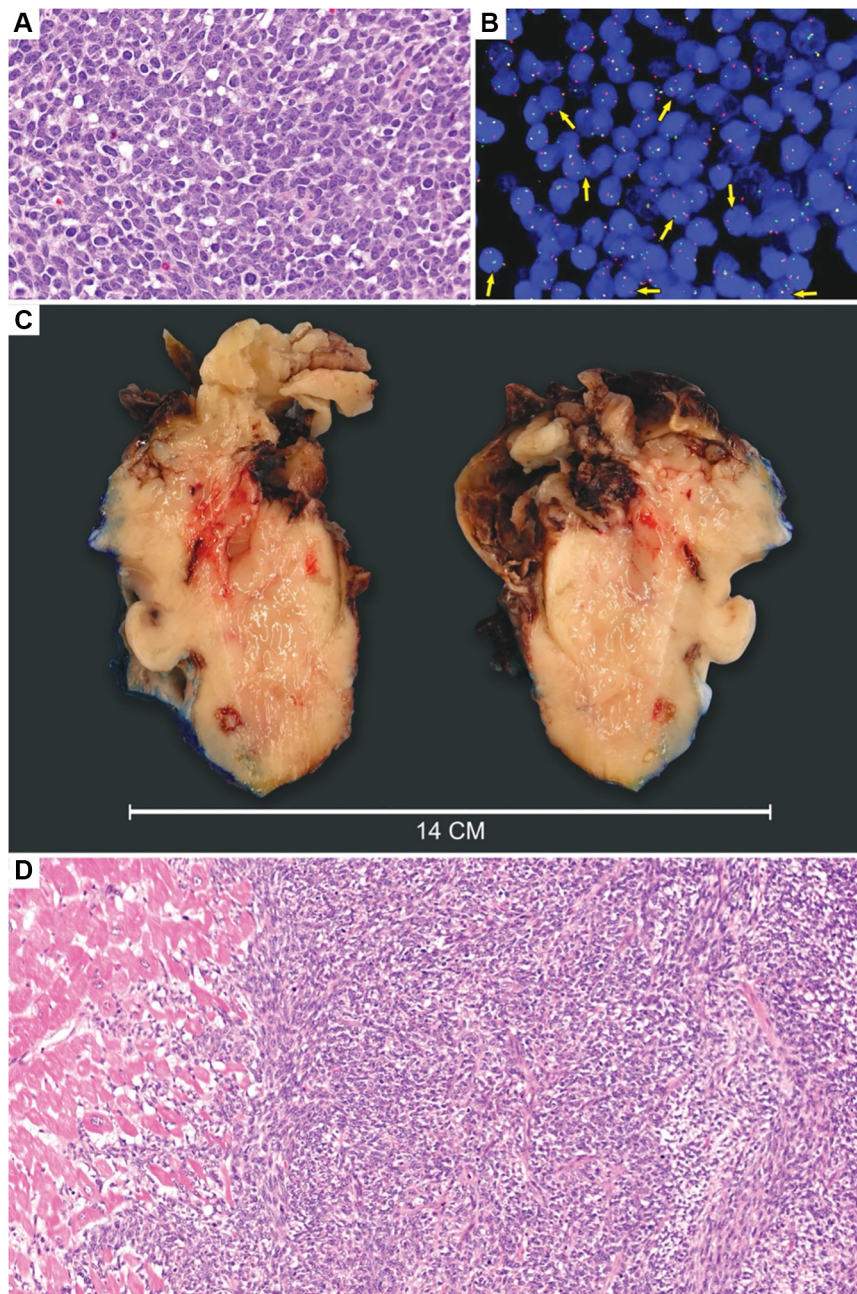
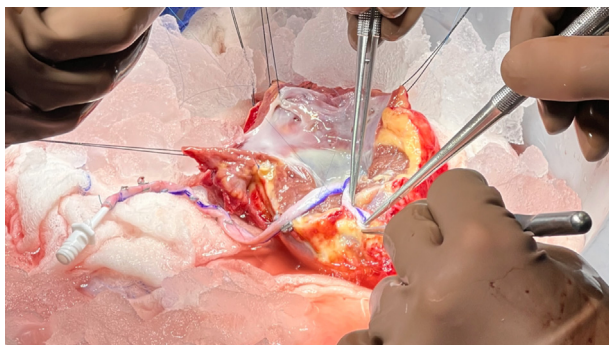


FIGURE 2. Histologic findings of the resected cardiac tumor. A, Photomicrograph from the endobronchial ultrasound-guided needle biopsy performed 1 week before surgery. B, Fluorescence in situ hybridization study with arrows pointing to SS18 gene rearrangement. C, Photo of the resected specimen. D, Photomicrograph of the tumor infiltrating the myocardium.

sarcoma. Although our patient had positive surgical margins, he has responded very well to chemotherapy, and as of 8 months after the operation, has not required radiation.

Echocardiography and cardiac MRI enable physicians to roughly estimate the size of the tumor,⁵ but it is challenging to fully understand the extent of the tumor until the heart is explanted. In our case, there was some discrepancy between the size interpreted from imaging and the size of the

resected mass, which could be due to the difficulty to truly appreciate both the extent of the tumor and the necessary margins for resection with 2-dimensional MRI. When dealing with LA and LV tumors, the team should plan for a possible radical reconstruction, which may include MV replacement and possible coronary artery bypass graft. In conclusion, autotransplantation can allow for safe tumor resection for large, left-sided tumors.



VIDEO 1. Presentation of the case and surgical techniques. This video shows the preoperative imaging and surgical techniques used to resect the large synovial cell sarcoma and reconstruct the heart. Video available at: [https://www.jtcvs.org/article/S2666-2507\(22\)00476-X/fulltext](https://www.jtcvs.org/article/S2666-2507(22)00476-X/fulltext).

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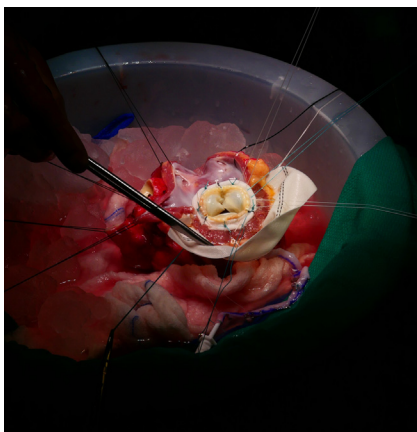


FIGURE E1. An intraoperative photograph of the *left* atrium, which was reconstructed with a Dacron sheet, and the biological mitral valve, which was successfully implanted onto the mitral annulus.