case presented, mitral regurgitation was tolerated well until the sixth decade. To our knowledge, this is the first reported case in the literature. Posterior wall motion, a long, mobile anterior leaflet, and annular geometry were able to compensate somewhat for this abnormality. We consider that decompensation occurred when the anterior leaflet degenerated and annular dilatation occurred after left ventricular and atrial enlargement. Ring annuloplasty is an alternative surgical procedure used for similar disorders [3]. This, and mitral valve repair, are particularly preferred in newborns and children to prevent the disorder from compromising children’s development and growth. However, in our case, the patient was a 62-year-old with a missing posterior leaflet and degenerated anterior mitral leaflet. Therefore, we preferred to use mitral valve replacement, which was suitable for this patient.

References

Large Cardiac Tumor Managed With Resection and Two Ventricular Assist Devices
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Symptomatic cardiac tumors can lead to a rapid clinical deterioration and death. Prompt surgical resection is ideal in this situation as it is the only proven treatment to date. We report the radical resection of a large malignant cardiac tumor that obstructed the right ventricular outflow tract. Extensive resection precluded reconstruction and limited the ability to implant a total artificial heart; thus, 2 paracorporeal devices were implanted instead. (Ann Thorac Surg 2014;97:321–4) © 2014 by The Society of Thoracic Surgeons

Soft tissue sarcomas are the most common malignant neoplasm of the heart, pericardium, and great vessels [1]. In some instances, they can obstruct the intracardiac outflow tract, which can lead to sudden circulatory collapse [2, 3]. Because roles for neoadjuvant therapy are not well defined, complete surgical resection of these tumors remains the mainstay treatment for palliation and improvement in clinical outcomes. In this report, we describe our first surgical experience using 2 paracorporeal ventricular assist devices (PVADs) after total cardiectomy in a patient with metastatic cardiac sarcoma.

A 28-year-old male patient with a history of right thigh mesenchymal chondrosarcoma was admitted with a chief complaint of increasing shortness of breath at rest. Echocardiogram showed a dilated right ventricle with a large obstructing mass and a posterior pericardial effusion. Ejection fraction was estimated to be 0.40. Cardiac magnetic resonance imaging clearly demonstrated a severely enlarged heart and an obstructed right ventricle outflow tract (RVOT) (Fig 1). All evidence indicated that the massive right-sided obstruction was the primary cause of the acute biventricular failure and that the remaining metastatic disease was under adequate control. The case was discussed at our multidisciplinary tumor board conference. The consensus was that resection of the tumor causing outflow obstruction was the top priority in this high-risk setting and that long-term survival was feasible, including eventual consideration for cardiac transplantation. Emergency approval from our Institutional Review Board and Ethics Committee was granted, and the patient underwent total heart replacement 2 days later.

Total cardiectomy was planned with concomitant biventricular reconstruction and placement of two Thoratec PVADs (Pleasanton, CA). The Syncardia Total Artificial Heart (TAH; Syncardia, Tucson, AZ) device could not be considered because native atrial tissue and ventricular cuffs would be resected to ensure a complete resection. Intraoperatively, we found a severely enlarged heart and near total replacement of the right ventricle by the tumor (Figs 2A, 2B, 2C). Upon opening the right atrium, we noted tumor extension into the vena cava. A separate large mass (5 × 8 cm) originating from the interventricular septum was the main cause of obstruction at the right ventricular outflow tract (Fig 2). The heart was excised, including the right atrium. Each atrium was reconstructed with a 32-mm diameter Hemashield platinum graft (MAQUET, Inc, Wayne, NJ) (Figs 3A, 3B). To prevent collapse, slight tension was placed on the graft and tissue glue was applied until stiffness was achieved. Also, a vacuum suction was employed at a negative pressure of 10 mm Hg on the right VAD and no suction on the left VAD. The outflow grafts were sewn to the remnants of the pulmonary artery and aorta (Fig 3C). With the pumps set to a fixed rate of 70, flows were generally kept between 3.5 to 4.5 l min⁻¹. The left side VAD flows were adjusted to maintain slightly higher flows (approximately 0.3 l min⁻¹) than the right side VAD. To manage filling...
pressures, a central venous line was placed with the goal of maintaining the central venous pressure between 10 and 15 mm Hg.

Postoperatively, acute renal failure was the only major organ dysfunction; liver and pulmonary functions were maintained with normal function. The patient was heparinized postoperatively with the goal of keeping partial thromboplastin time twice normal. The patient required postoperative hemodialysis therapy and was successfully discharged from the intensive care unit for further long-term, in-hospital care. The patient was transferred to the intermediate care unit that regularly cares for both single and biventricular supported patients. Physical rehabilitation was underway and he gradually improved his oral intake. However, on postoperative day 33, the patient's biventricular assist device (bVAD) flow suddenly decreased and circulatory shock occurred. A change in flow signal on the right VAD was noted. He had inadequate filling of the PVADs due to compression and collapse of the newly constructed atria. The patient expired despite all attempts to establish hemodynamic stability. Autopsy revealed large blood clots in the mediastinum and pericardial cavity compressing the atrial graft reconstructions, which resulted in cessation of pump flow.

Comment

Our case illustrates the concept of radical resection and total cardiac replacement with biVAD implantation, which we felt was the best surgical option for our patient. Resection in our case included significant portions of affected left and right atria, both ventricles, and septum. Because the Syncardia TAH requires the
presence of both native atria to achieve optimal implantation, we were left with the following limited biventricular support options: (1) to place biVAD support with 2 PVADs; or (2) employ biVAD support with either 2 HeartMate II VADs (Thoratec Corp, Pleasanton, CA) or HeartWare HVADs (Framingham, MA). We elected for the former option for various reasons. The Thoratec PVAD is Food and Drug Administration approved for biventricular support, has multiple literature reports in this setting, and is ideal for short to midterm support. For example, Silverman and colleagues demonstrated a similar approach in a 14-year-old girl, which resulted in a successful bridge to transplant after 118 days with biVAD support [4]. Our group had previously demonstrated the successful placement of 2 Thoratec PVADs implanted as cardiac replacement therapy in a patient with massive thrombus formation in 4 cardiac chambers [5]. Thus, we were relatively experienced with this device when employed in an emergency setting. For example, Silverman and colleagues demonstrated a similar approach in a 14-year-old girl, which resulted in a successful bridge to transplant after 118 days with biVAD support [4]. Our group had previously demonstrated the successful placement of 2 Thoratec PVADs implanted as cardiac replacement therapy in a patient with massive thrombus formation in 4 cardiac chambers [5]. Thus, we were relatively experienced with this device when employed in an emergency setting. The latter options had limited reports as biventricular support and were not deemed optimal because of the extensive resection required and lack of our institutional experience with these devices as an emergency biVAD support system [6, 7].

The rapid onset of the patient’s complication limited our ability to carry out any time consuming diagnostic evaluations of the pump malfunction. Prior to the system collapse, the patient had been ambulating and self-feeding, and appeared to be recovering well. However, the patient may have benefited from serial imaging studies such as non-contrast computed tomography or transthoracic echo, which may have helped identify mediastinal cloting or graft compression.

The use of this radical technique as a destination therapy, bridge to candidacy, or bridge to transplantation in patients with metastatic disease has never been established by any evidence-based literature report. However, solid organ transplantation may be feasible for well-localized primary malignant tumors, assuming a low chance of recurrence and that life expectancy can be significantly extended. In our case, if the patient had demonstrated at least a 2-year tumor-free period, he could have been considered a candidate for transplantation. Also, as the mechanical devices are “immune” to the chemotherapy-related cardiac toxicity, treatment can be tailored irrespective of this effect with a goal of optimizing complete systemic remission. Future improvements in the clinical setting will require additional in-depth discussions and review of similar cases.

The onset of acute renal failure requiring dialysis was an indicator of poor outcome in our patient. The need for dialysis alters the patient’s volume status and can lead to inadequate filling of the atrial grafts and pump chambers. These volume shifts, especially hypovolemia, can result in “underfilling” and more susceptibility to compression from mediastinal hematoma. Our experience supports the notion that dialysis is a risk factor or indicator for poor outcome in assist-device patients.

Surgical resection of cardiac tumors can be curative, or palliative, especially if symptomatic at presentation.
Primary Leiomyma: A Rare Space Occupying Lesion in the Right Ventricle

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A case of primary cardiac leiomyoma arising from the infundibulum of the right ventricle of a 24-year-old woman is presented. The mass protruded into the cavity of the right ventricle and caused severe right ventricular outflow tract obstruction. We resected the tumor under cardiopulmonary bypass successfully, and confirmed it histologically to be a benign leiomyoma. To the authors’ knowledge, this is the first case report of a primary cardiac leiomyoma in an adult woman.

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