Primary malignant cardiac tumors are quite rare. The incidence is only 0.02%. These cancers have a poor prognosis because of early metastases and a high recurrence rate. Rhabdomyosarcomas are the second most commonly occurring malignant primary cardiac tumors. Surgical resection is the treatment of choice for primary cardiac sarcomas. Tumor size and location greatly influence the anesthetic and hemodynamic management strategies during surgical resection. The case of a 38-year-old woman with a recurring primary rhabdomyosarcoma is discussed.

Keywords: Malignant, primary cardiac tumor, rhabdomyosarcoma.
nal jugular vein using sterile technique and ultrasound guidance to allow for rapid administration of crystalloids, colloids, or blood products as needed. Anesthesia was maintained with 1% isoflurane, fentanyl boluses as needed (for a case total of 1.5 mg), and administration of rocuronium for muscle relaxation with a target train-of-four of 1 of 4. Cefuroxime, 1.5 mg, was administered with an IV piggyback infusion within 1 hour before incision and was redosed 4 hours into the case.

A preoperative computed tomography (CT) scan determined there were no anatomic cardiac structures that were close to the sternum. A sternotomy was performed using an oscillating saw. In preparation for aortic cannulation, the patient was placed in a reverse Trendelenburg position to achieve a mean arterial pressure of 60 mm Hg and thus prevent dissection of the aorta. The aorta was cannulated 3 minutes after central IV administration of 30,000 U of heparin (400 U/kg). Individual venous cannulas were placed in the superior vena cava and the inferior vena cava rather than a 2-stage right atrial cannula to prevent inadvertently dislodging any tumor from that chamber. A 1% concentration of isoflurane was administered during cardiopulmonary bypass (CPB) by a certified perfusionist. Cardiac arrest was achieved 1 minute after application of the aortic cross-clamp and administration of del Nido cardioplegia solution. Del Nido cardioplegia has 2 unique properties in which it differs from standard Buckberg cardioplegia; it lacks a glucose component and is administered as a single dose. It is dosed as 20 mL/kg, with a maximum volume of 1,000 mL for patients weighing greater than 50 kg.

The right atrium was opened, and a mass was visualized coming through the tricuspid valve, while the base of the mass was in the right ventricle. The anterior aspect of the right ventricle was then opened, and the endocardium at the base of the ventricular septum appeared white (Figure 2). All grossly visible tumor was resected, and biopsy specimens were obtained from the pedunculated right ventricular mass, papillary muscle, endomyocardium, and septum. Frozen-section pathology specimens determined that the excised mass was a high-grade sarcoma with a negative margin in the muscle but a positive margin at the endocardium. All pathology specimens except the tricuspid valve biopsy showed recurrent, poorly differentiated sarcoma with rhabdomyoblastic differentiation. The right ventricle was closed using a bovine pericardium patch because of the size of the cardiac chamber incision. The total time for CPB was 1 hour 18 minutes; aortic cross-clamp time was 44 minutes, and separation from CPB was uneventful. Heparin administration was reversed with 250 mg of protamine, with a final activated clotting time of 112 seconds (baseline, 137 seconds).

A postoperative echocardiogram revealed that the right ventricular mass was eliminated, with mild right ventricular dysfunction and 3+ to 4+ tricuspid regurgitation (Figure 3). The tricuspid valve was not repaired or replaced because the surgical team suggested that this would resolve itself over time with the tumor removed. The patient’s chest was closed with sternum wires, with...
no major change in hemodynamics. The patient was transported to the cardiovascular intensive care unit while hemodynamically stable, intubated, and sedated with a propofol infusion. Perioperatively 1,200 mL of crystalloid was administered, with a total blood loss of 500 mL and urine output of 950 mL.

The patient’s immediate postoperative recovery and 30-day recovery remained uneventful. One month postoperatively, the patient was started on a chemotherapeutic regimen of etoposide, mesna, and ifosfamide. After initiation of chemotherapy, the patient experienced upper respiratory tract infections, urinary tract infections, profound anemia (hemoglobin, 6.7 g/dL), and febrile leukopenia (white blood cells, 20.7 k/µL). The most recent mammography and chest CT results have not revealed any metastasis. However, the most current cardiac MRI has shown the return of a right atrial thrombus and soft-tissue mass, which reappeared less than 45 days after the surgical resection and continues to grow with each surveillance imaging.

Discussion

- **Tumor Prevalence and Pathophysiology.** Primary cardiac tumors occur rarely in adults, with a reported incidence rate in the general population of 0.02%.\(^5\)\(^,\)\(^6\) Seventy-five percent of primary cardiac tumors are benign, and the remainder of the tumors are malignant.\(^5\)\(^,\)\(^6\) Malignant primary cardiac neoplasms are uncommon because most tumors of the heart are metastatic in origin.\(^1\)\(^,\)\(^3\) Sarcomas, including angiosarcoma, rhabdomyosarcoma, and AIDS-related sarcoma, are the most frequently occurring primary malignant tumors of the heart, with angiosarcoma being the most prevalent in adults.\(^1\)\(^,\)\(^3\)\(^,\)\(^5\)

Rhabdomyosarcomas are the second most prevalent primary cardiac malignant tumors.\(^1\)\(^,\)\(^2\)\(^,\)\(^4\)\(^,\)\(^6\)\(^,\)\(^7\) Because rhabdomyosarcomas are rare, epidemiologic data, literature on its presentation, medical management, and anesthetic management for surgical resection is limited.\(^3\)\(^,\)\(^7\) Seventy-five percent of rhabdomyosarcomas occur in infants less than 1 year of age, with equal frequency in boys and girls.\(^1\)\(^,\)\(^2\)\(^,\)\(^4\)\(^,\)\(^6\)\(^,\)\(^7\) In adults, rhabdomyosarcomas occur most often in individuals between 30 and 50 years of age.\(^5\)

Although malignant primary cardiac tumors are mainly found on the right side of the heart, rhabdomyosarcomas may originate in any chamber.\(^1\)\(^,\)\(^2\)\(^,\)\(^5\)\(^,\)\(^6\) They are aggressive, invasive tumors, with the high probability of infiltrating into the cardiac valve leaflets with extension into the pleura, pericardium, or mediastinum.\(^2\)\(^,\)\(^5\)\(^,\)\(^6\) Primary characteristics of rhabdomyosarcomas are that they (1) arise from striated muscle, (2) grow rapidly, (3) may be large and bulky, and (4) can fill a cardiac chamber (Figure 4).\(^3\)\(^,\)\(^7\) Macroscopically the tumor may appear as soft, gelatinous nodes with central necrosis.\(^5\)\(^,\)\(^7\)

- **Patient Presentation and Diagnosis.** Malignant cardiac tumors are often difficult to diagnose because of their rarity and variable, nonspecific clinical presentation.\(^3\)\(^,\)\(^5\)\(^,\)\(^7\) Heart failure, dyspnea, and hemorrhagic

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Figure 3. Postoperative Transesophageal Image, Four-Chamber View
Right ventricular mass is eliminated, and there is mild right ventricular function and 3+ to 4+ tricuspid regurgitation.

Figure 4. Gross Photograph of Rhabdomyosarcoma Showing Grayish-White, Poorly Circumscribed Tumor With Areas of Hemorrhage
(Source: Kumar et al.\(^8\) Reprinted with permission from Elsevier.)
Abbreviations: PA, pulmonary artery; TEE, transesophageal echocardiography.

**Major intraoperative concern and anesthetic considerations**

- Hemodynamic collapse, severe hypotension and arrhythmias caused by a large tumor in right atrium, restricting venous return. **(Anesthetic considerations: Careful positioning of patient on operating room table, Preparation for major volume resuscitation, central intravenous access, large-bore peripheral intravenous access, Vasoactive medication prepared for immediate use, Surgeon present for induction of anesthesia, Cardiopulmonary bypass machine primed and ready for immediate use)**

- Worsening of myocardial depression, further decrease in coronary perfusion pressure, systemic vascular resistance, and stroke volume. **(Anesthetic considerations: Minimal use of preoperative sedative medication, Slow and smooth titration of induction medication)**

- Tumor located in the atria or atrioventricular valves, obstruction of intracardiac blood flow. **(Anesthetic considerations: Judicious use of inotropic agents; do not want to enhance contractility to a point at which it worsens passage of intracardiac blood flow)**

- Hepatic cells are often a metastatic target. **(Anesthetic considerations: Preoperative assessment for hepatic impairment; adjust medications as needed)**

- Tumor embolization. **(Anesthetic considerations: Consider using TEE for PA catheter placement, Consider placing PA catheter at femoral site)**

- Atrial fibrillation or flutter occurs in 25% of patients, and ventricular tachycardia is not uncommon. **(Anesthetic considerations: Placement of defibrillator pads before induction of anesthesia for immediate cardioversion/defibrillation)**

- Acute rise in right atrial pressure that could induce an atrial arrhythmia. **(Anesthetic considerations: Adequate depth of anesthesia during laryngoscopy, incision, and sternotomy)**

**Table. Major Intraoperative Concerns Associated With Surgical Resection of Cardiac Tumors and Anesthetic Considerations**

<table>
<thead>
<tr>
<th>Consideration</th>
<th>Anesthetic Considerations</th>
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</thead>
<tbody>
<tr>
<td>Pericardial effusion is some of the more common presenting symptoms with malignant primary cardiac tumors.6,9</td>
<td>Adequate depth of anesthesia during laryngoscopy, incision, and sternotomy</td>
</tr>
<tr>
<td>Depending on the tumor size and the location of infiltration, other nonspecific symptoms such as weight loss, fatigue, muscle pain, coughing, and leukocytosis may occur.1,5,6,9</td>
<td>Adequate depth of anesthesia during laryngoscopy, incision, and sternotomy</td>
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<tr>
<td>A cardioembolic stroke can result from a detached tumor that migrates to the peripheral vasculature system or pulmonary artery.2-4,6</td>
<td>Adequate depth of anesthesia during laryngoscopy, incision, and sternotomy</td>
</tr>
<tr>
<td>Cardiac arrhythmias, such as atrioventricular block, atrial fibrillation, and atrial flutter may occur from tumor infiltration of the neural electrical pathways.2-3,6</td>
<td>Adequate depth of anesthesia during laryngoscopy, incision, and sternotomy</td>
</tr>
<tr>
<td>The most common cause of acute symptoms is usually from obstruction of blood flow to the heart.2,4,6</td>
<td>Adequate depth of anesthesia during laryngoscopy, incision, and sternotomy</td>
</tr>
<tr>
<td>Tumors located in the right atrium may cause the patient to exhibit signs and symptoms of right heart failure, including ascites, peripheral edema, hepatomegaly, and venous hypertension.2,3</td>
<td>Adequate depth of anesthesia during laryngoscopy, incision, and sternotomy</td>
</tr>
<tr>
<td>Ventricular outflow obstruction or subvalvular stenosis may occur from large tumors invading the ventricles.2</td>
<td>Adequate depth of anesthesia during laryngoscopy, incision, and sternotomy</td>
</tr>
<tr>
<td>Echocardiography is usually the first diagnostic procedure performed once assessment of clinical presentation has occurred.4,6</td>
<td>Adequate depth of anesthesia during laryngoscopy, incision, and sternotomy</td>
</tr>
<tr>
<td>Anatomic information about the tumor’s size, location, and extent of cardiac infiltration is obtained from a cardiac CT scan and a cardiac MRI.3-5</td>
<td>Adequate depth of anesthesia during laryngoscopy, incision, and sternotomy</td>
</tr>
<tr>
<td>Surgical resection is imperative, and it is the primary mode of treatment for primary cardiac tumors.</td>
<td>Adequate depth of anesthesia during laryngoscopy, incision, and sternotomy</td>
</tr>
</tbody>
</table>

**Treatment.** Surgical resection helps to relieve symptoms and avoid hemodynamic and embolic sequelae.4,6 Unfortunately, recurrence rates remain high and the prognosis remains poor after complete resection of a cardiac sarcoma.3 The median survival rate with a complete resection is 27 months, compared with only 10 months with incomplete resection.3 Orthotopic heart transplantation has been considered for hearts with isolated unresectable tumors; however, because of the high potential of early systemic metastasis, the survival is still only 1 to 2 years.4,5 Heart transplant may also ensue an exacerbation of undetected micrometastases because of the required immunosuppression.3,4,6 Auto heart transplantation is usually not an option and is controversial because of the high potential for early metastases even with low-grade sarcomas.4,5 Auto transplantation is when the heart is explanted for ex vivo surgical resection of the tumor, then implanted back into the patient. Chemotherapy and radiation are considered ineffective, and these treatment modalities do not demonstrate significant beneficial effect on survival for patients diagnosed with cardiac sarcomas.4,6 Most patients often die of distant metastases.5,6

**Anesthetic Management.** There is limited literature outlining the anesthetic management of cardiac surgery for resection of primary cardiac tumors. The anesthetic management strategies for surgical resection of cardiac tumors that are described in the existing literature are management strategies that do not deviate significantly from anesthetic management strategies of other open-cardiac chambers. However, there are specific intraoperative concerns pertaining to surgical resection of cardiac tumors for which the anesthesiology provider should be prepared (Table). Anesthetic management for surgical resection of cardiac tumors begins with a comprehensive review of the preoperative cardiac imaging by the anesthesiology provider with the cardiologist and surgeon. This information will enable the anesthesia provider to anticipate the probable and possible intraoperative physiologic
implications of the tumor, based on size and location. For example, if a large tumor is present in the right atrium, restriction of venous return could potentially occur and cause severe hypotension and arrhythmias. The anesthetic considerations in preparation for such an event would include judicious positioning of the patient on the operating table to minimize the risk of impaired venous return. The anesthesia provider would also need to be prepared for volume resuscitation and immediate administration of vasoactive medication in case the patient became hemodynamically unstable because of decreased venous return or increased valvular outflow obstruction due to a large tumor in the right atrium restricting flow through the tricuspid valve. Because of the possibility of hemodynamic collapse in such an instance, there should be a surgeon present and readiness for CPB at all times, including during induction of anesthesia.

The anesthesia provider should minimize the use of sedative medications outside the operating room, and induction medications should be titrated cautiously in this patient population. The goal is to avoid further myocardial depression and to maintain systemic vascular resistance and coronary perfusion pressure. Preservation of right ventricular filling pressures, stroke volume, and normal sinus rhythm minimizes hemodynamic compromise during induction. Although the use of inotropes may be warranted to improve myocardial contractility, it is important to recognize that enhanced contractility from inotropic support could increase outflow tract obstruction and decrease passage of intracardiac blood flow, especially if the tumor is located in the region of the atria or the atrioventricular valves. Large atrial tumors can mimic valvular stenosis by obstructing atrioventricular flow. Because hepatic cells often are a metastatic target, the anesthesia provider should assess the patient for hepatic impairment preoperatively in patients with a malignant tumor so that medication doses can be adjusted if indicated.

Small cardiac tumors increase the risk of embolization during tumor resection. Tumor embolization can lead to acute pulmonary venous hypertension with or without pulmonary edema. The risk of placing a pulmonary artery catheter also must be weighed against the risk of causing possible tumor embolization. Because most malignant tumors are located on the right side of the heart, use of TEE guidance for placement of a pulmonary artery catheter may be warranted. Additionally, the anesthesia provider should consider placing a central venous catheter at the femoral site instead of the internal jugular or subclavian vein, as a safer alternative.

Because atrial fibrillation or flutter arises in 25% of patients with cardiac tumors, defibrillation pads should be placed before induction so that the patient can be immediately cardioverted if needed. Reports of atrioventricular block and ventricular tachycardia are not uncommon during the periorioperative period, especially in patients with intramyocardial or intracavity tumors that infiltrate or cause inflammation of conduction tissue. The anesthesia provider must also ensure that an adequate depth of anesthesia is maintained, especially during stimulating events in the surgical procedure, including laryngoscopy, incision, and sternotomy, to prevent an acute rise in right atrial pressure that may induce an atrial dysrythmia. Use of TEE enables the anesthesia provider to (1) detect tumor dislodgement and/or fragmentation, (2) postresection examination, and (3) patient volume status, especially if the central venous pressure is unreliable because of the presence of a tumor in the right atrium that causes severe tricuspid regurgitation.

Conclusion

The existing literature regarding the anesthetic management for surgical cardiac tumor resection is very limited because of the rare occurrence of primary cardiac tumors. Surgical resection is the treatment of choice for cardiac tumors. This case presentation and discussion aim to identify a gap in knowledge and provide an outline of major intraoperative concerns and anesthetic considerations associated with surgical resection of cardiac tumors for the anesthesia provider. It is imperative for the anesthesia provider to have a clear understanding of the tumor’s pathology, size, and location in order to anticipate any major untoward intraoperative concerns that may arise during surgical resection. A comprehensive review of the preoperative cardiac imaging and coordination of the intraoperative approach with the surgical team and cardiologist will also facilitate appropriate anesthetic management. However, despite a successful surgical resection, chemotherapy, and radiotherapy, the prognosis usually remains poor, and the tumor’s local involvement and stage of presentation determines the ultimate outcome.

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