Perioperative Management of a Pediatric Patient with Catecholamine-Induced Cardiomyopathy Undergoing Laparoscopic Paraganglioma Excision Requiring Biventricular Assist Device Support

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PHEOCHROMOCYTOMAS ARE catecholamine-secreting tumors that arise from chromaffin cells in the adrenal medulla. Likewise, paragangliomas are extra-adrenal tumors of the sympathetic and parasympathetic paraganglia, which may be located anywhere from the base of the skull to the pelvis. Sympathetic paragangliomas may secrete vasoactive catecholamines while parasympathetic paragangliomas typically are non-functional. Both pheochromocytomas and functioning paragangliomas are rare neuroendocrine tumors, distinguished only by location. Classic presenting symptoms include headaches, palpitations, and sweating. Cardiac sequelae may include hypertension, tachycardia, and arrhythmia, with the possibility of progression to catecholamine-induced cardiomyopathy. The authors present the perioperative course of an adolescent who developed severe cardiomyopathy secondary to a catecholamine-secreting paraganglioma, requiring biventricular assist devices, who was treated successfully with multidisciplinary care including laparoscopic tumor resection.

CASE REPORT

The patient was a 12-year-old previously healthy male who originally presented with 4 weeks of rash, low-grade fever, cough, and worsening lower extremity edema. A comprehensive diagnostic workup was initiated, including a transthoracic echocardiogram significant for an ejection fraction of 20% to 23% and markedly depressed left ventricular wall motion. He was diagnosed with congestive heart failure and initially managed with inotropic support including dopamine and epinephrine. However, his condition rapidly deteriorated, with subsequent respiratory failure and bradycardic arrest requiring cardiopulmonary resuscitation. He was transferred to the authors' tertiary care facility, and venoarterial extracorporeal membrane oxygenation (ECMO) support was initiated. Further workup, including cardiac biopsy, was negative for an infectious etiology of the patient's cardiomyopathy. After 3 days of ECMO, he was transitioned to a Thoratec® (Thoratec Corporation, Pleasanton, CA) paracorporeal left ventricular assist device (LVAD) support. While initial echocardiograms showed normal right ventricular wall motion, after LVAD placement, the right ventricular function declined, ultimately requiring inhaled nitric oxide and milrinone. Seven days following LVAD placement, he developed a large pericardial hematoma with significant tamponade effect. Despite surgical evacuation of this hematoma, his right ventricular function remained poor, so a Thoratec® right ventricular assist device (RVAD) also was placed. His intensive care unit course was marked by persistent hypertension requiring multiple vasoactive drugs including nicardipine, amlodipine, hydralazine, milrinone, esmolol, and labetalol. Systolic blood pressures persisted in the 120s to 150s, with diastolic blood pressures in the 60s and 70s. Further review of his medical history with family members revealed that the child had occasional episodes of flushing and palpitations. An abdominal computerized tomography (CT) scan was performed, revealing a 4.6-by-5 centimeter juxtarenal mass adjacent to the left adrenal gland, which was concerning for pheochromocytoma or paraganglioma (Figs 1 and 2). Plasma and urine catecholamine testing confirmed the diagnosis, with urine catecholamines of 465 micrograms/24 hours (normal < 116 micrograms/24 hours), plasma metanephrines 0.71 nmol/L (normal 0-0.49), and plasma normetanephrine 22.3 nmol/L (normal 0-0.89).

After several days of alpha-adrenergic-receptor blockade, with a phentolamine infusion and phenoxybenzamine, subsequent beta-blockade was achieved with esmolol. A laparoscopic adrenalectomy was planned with ongoing biventricular assist device (BiVAD) support. A laparoscopic-assisted partial colectomy also was planned during this procedure because of a site of mild lower gastrointestinal bleeding identified at the splenic flexure. Given the multiple cannulae in his upper abdomen, port placement for this laparoscopic surgery required careful planning (Fig 3). The decision was made to include the VADs in the sterile field, with port placement arranged around the VADs and tubing. Cardiopulmonary bypass and a cardiac surgery team remained on standby if needed. Invasive arterial blood pressure monitoring and central access were available for the procedure. Transesophageal echocardiography was avoided due to actively bleeding peptic ulcers in addition to the aforementioned colonic bleeding. The patient was intubated the day prior to the procedure, so airway manipulation was not necessary. He was kept in the supine position with a wedge used to elevate his left side. A cardiac surgeon was present during this positioning to ensure the ventricular assist devices remained in proper position without undue traction on the cannulae. After sterile preparation, abdominal insufflation was initiated carefully to ensure continued adequate preload. Pneumoperitoneum was achieved at 15 mmHg pressure without significant hemodynamic change. Maintenance of anesthesia was achieved with isoflurane and fentanyl, with vecuronium for neuromuscular blockade. His phentolamine infusion was continued intraoperatively for blood pressure control, and nitroglycerin boluses were available for acute increases in blood pressure not attributed to pain. He required 1 unit of packed red blood cells at the time of dissection of the neoplasm due to surgical bleeding and moderate hypotension (80/40), with prompt resolution after transfusion. The patient otherwise

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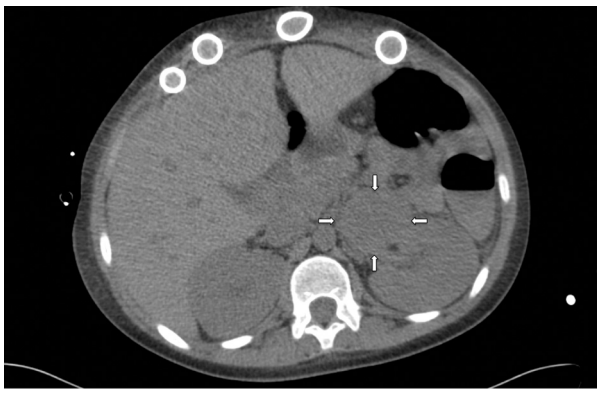

Fig 1. Transverse abdominal non-contrast CT image of juxtarenal mass. Arrows indicate borders of the mass.

remained hemodynamically stable throughout the procedure, and his phentolamine infusion was stopped after tumor removal.

The patient was taken to the intensive care unit after successful tumor excision, intubated, and mechanically ventilated. Histologic examination of the adrenal mass indicated the specimen was consistent with a paraganglioma not involving the adrenal gland. The patient's hemodynamics improved, and urine and plasma metanephrine concentrations returned to normal values after the tumor resection, but his cardiac function continued to require biventricular support. Twelve days after paraganglioma removal, he was relisted for transplant. Approximately 1 month after resection, the patient underwent an uneventful orthotopic heart transplant. He was discharged from the hospital approximately 80 days after initial admission. The patient continues to do well at the time of this writing.

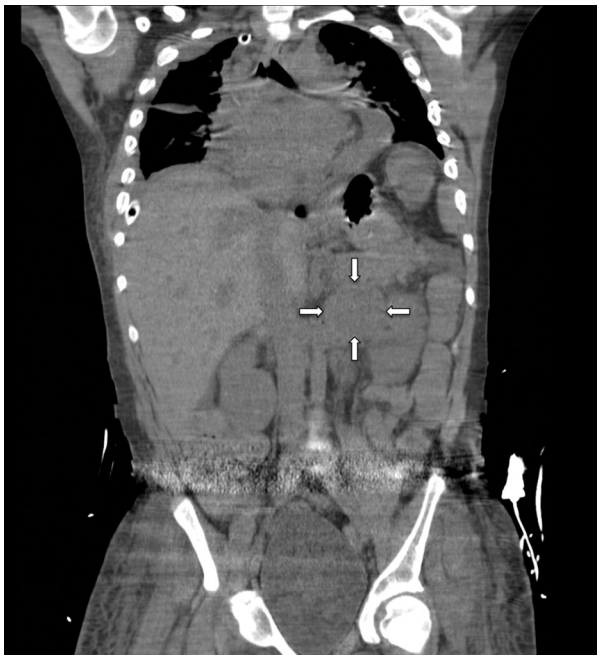

Fig 2. Coronal abdominal non-contrast CT image of juxtarenal mass. Arrows indicate borders of the mass.

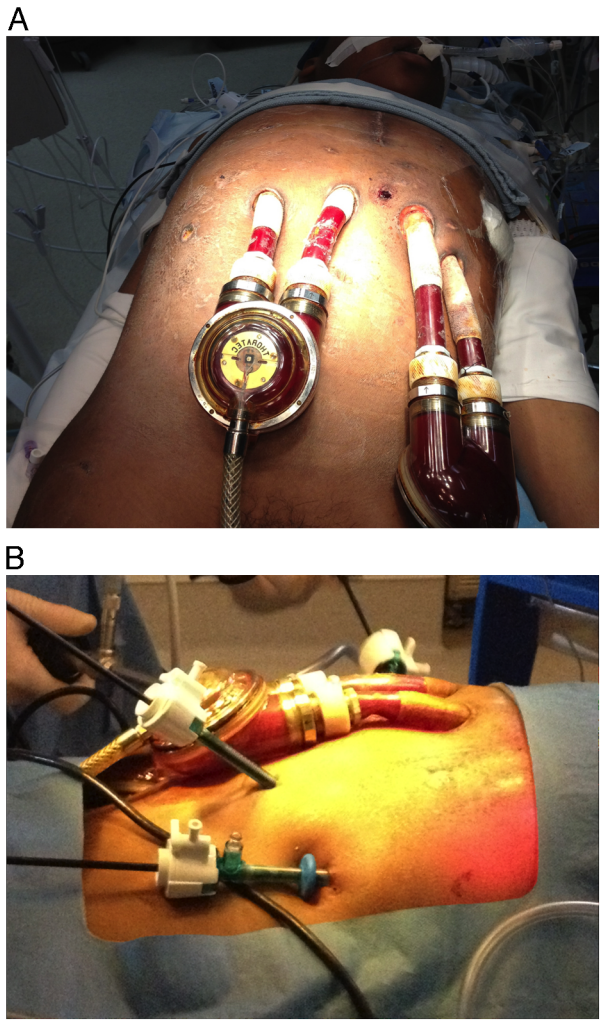

Fig 3. (A) Abdomen with paracorporeal biventricular assist devices in place. (B) Port placement for laparoscopic adrenalectomy shown from patient's left side. The biventricular assist devices are included in the sterile field.

DISCUSSION

Pheochromocytomas and paragangliomas are rare neoplasms arising from neural crest cells called paraganglia and may occur anywhere these cells are found. Their incidence is estimated at 0.3 per million per year, and approximately 10% to 20% of cases are diagnosed during childhood.[1] Approximately 10% to 30% of pheochromocytomas and paragangliomas are familial, most often related to the multiple endocrine neoplasia type 2, neurofibromatosis type 1, Von Hippel Landau, and the familial paraganglioma syndromes. The pheochromocytomas and paragangliomas diagnosed in childhood usually are related to one of the previously mentioned syndromes. There is a slight predominance in males, and the average age of diagnosis is 11 years.[1] The patient in this case was evaluated for these familial syndromes, but genetic screening tests were negative for common mutations associated with these syndromes. Classic symptoms of pheochromocytoma include recurrent headaches, profuse sweating, and palpitations, but they also may cause