Pediatric Anesthetic Management of the Surgical Resection of Recurrent Pheochromocytoma in a Patient with Resolution of Severe Dilated Cardiomyopathy

Jenna Helmer Sobey, MD Than Thuan Nguyen, MD Humphrey Lam, MD Thomas Austin, MD

Introduction

- Pheochromocytoma is a rare tumor in the pediatric population
- Secretes a combination of norepinephrine/epinephrine, only norepinephrine, or only epinephrine
- In 10% of patients with pheochromocytoma, the tumor is part of a familial disorder such as MEN syndrome, von Rechlinghausen, or von Hippel-Lindau syndrome
- Patients may present with episodic headaches, sweating, and palpitations
- Prolong exposure of catecholamines may result in a dilated cardiomyopathy
- Recovery of heart function has been reported in many adult cases but there are much fewer cases reported in children

Case History

- 11 year old boy with history of Von Hippel Lindau syndrome and bilateral pheochromocytoma s/p right sided adrenalectomy and partial left adrenalectomy at age 9 presented with recurrent bilateral pheochromocytomas
- Significant dilated cardiomyopathy with an LVF of 15% before initial surgery with very little improvement from a cardiac standpoint several months following initial resection
- Two years post surgery, patient again found to have elevated VMA and normetanephrine at outside facility
- VMA 24 HR urine: 8.8 mg/24 hr (Normal < 3.4)
- Normetanephrine 3755 mcg/24 hr (Normal 67-503)
- Referred to Vanderbilt Medical Center for further workup
- Investigation at our institution:
  - Echo: RV wall motion normal, LVEF mildly depressed at 46%, much improved function compared to older echo
  - Nuc. Med Scan: Solitary left adrenal MIBG-avid mass, consistent with recurrent residual pheochromocytoma
  - MRI abdomen: 3.4 x 2.3 x 5.6 cm (AP, transverse, CC) T2 hyperintense mass superior and anterior to the left kidney
  - Started on phenoxybenzamine, a long acting alpha receptor antagonist, 15 days prior to surgery
  - After adequate alpha blockade, the patient was boarded for resection
  - Induction was performed with high dose fentanyl and vecuronium
  - Blood pressure was 100-120/80-90 pre induction and post induction
  - 2 large bore PIVs, radial arterial line and a triple lumen CVL were placed
  - A thoracic epidural was placed for postoperative analgesia
  - Anesthetic maintenance was achieved with sevoflurane, dexmedetomidine infusion and epidural infusion
  - Hemodynamics were stable except for BP spikes with tumor manipulation
  - Sodium nitroprusside and esmolol were utilized for hypertension and tachycardia
  - After tumor removal, the patient required norepinephrine and vasopressin for hypotension
  - Patient was extubated in the operating room and transported to PACU.
  - In PACU, patient required a 5% dextrose infusion to correct hypoglycemia

<table>
<thead>
<tr>
<th>Condition</th>
<th>What is it?</th>
<th>Anesthetic Considerations</th>
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</thead>
<tbody>
<tr>
<td>Cystic Fibrosis</td>
<td>-mutation on the long arm of chromosome &amp; malfunction of the CFTR chloride channel on exocrine glands -affects airway, pancreas, hepatobiliary system, gastrointestinal tract, bones, and the genitourinary tract</td>
<td>Airway/Pulmonary: - airway reactivity - loss of airway cartilaginous support - chronic hypoxia -mucous plugging -pulmonary hypertension</td>
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<tr>
<td>Factor V Leiden</td>
<td>-most common cause of thrombophilia -mutation that confers factor V resistance to protein C degradation -increased risk for deep vein thrombosis, preeclampsia, placental abruption, and abortions</td>
<td>Hepatobiliary: -abnormal liver function -cirrhosis -portal hypertension -altered metabolism of drugs</td>
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<td>Factor XI deficiency</td>
<td>-rare -prolonged activated partial thromboplastin time -severity of bleed does not correlate well with factor levels -levels &lt;15% may require fresh frozen plasma for surgical procedures -levels &gt;50% are less likely to cause significant bleeding -can develop Factor XI inhibitor</td>
<td>Hypercoagulable state - antiocoagulated</td>
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</tbody>
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Discussion

- Clinical presentation
  - Hypertension, headache, palpitations, diaphoresis, pallor, orthostatic hypotension, syncope, tremor, anxiety, blurred vision, abdominal pain, diarrhea, and vomiting
- Complications from pheochromocytoma
  - Hypertensive crisis, pancreatitis, stroke, seizures, cardiomyopathy
- Medical management
  - Initiate alpha blockade prior to surgery
  - Calcium channel blockers- nifedipine
  - Alpha blockers- prazosin, doxazosin, phenoxybenzamine
  - Beta blockers- propranolol, atenolol, metoprolol
  - Signs of adequate alpha blockade
  - Arterial blood pressure not > 165/90 mmHg 36hrs before surgery
  - Orthostatic hypotension <15%
  - ECG free from ST segment and T wave abnormalities for 2 wks
  - No more than 1 PVC every 5 min
- Anesthetic Considerations
  - Volume status
  - Drugs to avoid: droperidol, morphine, atracurium, pancuronium, ketamine, ephedrine, halothane, cocaine, metoclopramide
  - Invasive Monitoring- Arterial BP, CVP, Foley catheter
  - Consider PAC and/or TEE if myocardial dysfunction is present
- Pheochromocytoma Associated Cardiomyopathy
  - Results from excessive alpha receptor stimulation by norepinephrine
  - Reduction in coronary blood flow and increased afterload may play a role
  - Severity of injury appears to be proportional to catecholamine surge
  - Alpha blockade is the mainstay of treatment but the most dramatic restoration of cardiac function seen after removal of tumor
  - Complete restoration of cardiac function usually takes 3.5-18 months
  - Our patient’s function did not return to normal until 2 years after initial presentation

References

- Eisenhofer, Graeme et al. Pheochromocytomas in von Hippel-Lindau Syndrome and Multiple Endocrine Neoplasia Type 2 Display Distinct Biochemical and Clinical Phenotypes