Anesthetic Management of a Child with Malignant Hypertension Secondary to a Renal Paraganglioma and Concomitant Renal Artery Stenosis

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Objectives:
1. Identify the differential for hypertension in a child.
2. Select the appropriate workup for a child with malignant hypertension.
4. Provide goals of therapy for intraoperative management of a child with a catecholamine-secreting paraganglioma.
5. Appreciate the postoperative considerations in the management of catecholamine-secreting paraganglioma.

Case History:
Your patient is a 7-year-old female who initially presented to an outside hospital with headaches, dizziness, and vomiting. Her blood pressure on admission was 170/122, and a head CT revealed hypertensive encephalopathy. Blood chemistry revealed an elevated serum creatinine of 1.1 mg/dL. Urinalysis was significant for proteinuria. A renal ultrasound scan revealed a 1.8 x 2.5 cm mass in the right renal hilum. Doppler studies were significant for right renal artery stenosis. The left renal artery was patent.

The abdominal MRI demonstrated a solid mass with attenuation similar to liver tissue in the right renal hilum with visible narrowing of the adjacent right renal artery. The mass showed slight homogeneous enhancement and was given a tentative diagnosis of renal cell carcinoma.

Initial lab values revealed an increase in renin, normal aldosterone, normal epinephrine, and a moderate elevation in norepinephrine.

Questions:
What is the significance of these lab values? What is the differential diagnosis for hypertension in a child? Do you recommend any further workup for her hypertension? What are your recommendations for a drug regimen for antihypertensive therapy?

Case History (continued):
Your patient’s 24-hour urine catecholamine levels are checked and the results show: Epinephrine and dopamine levels are normal. Norepinephrine and metanephrine levels are modestly elevated.
Further workup involved a whole body nuclear medicine scan with MIBG (iodine-131 metaiodobenzylguanidine). This was negative for any evidence of an adrenergic tumor.

Questions:
What is the significance of these findings? Does this alter your choice of drug therapy for hypertension?

Case History and Physical Examination (continued):
Your patient is scheduled for an exploratory laparotomy, tumor excision, and possible right renal artery repair. She was initially placed on phenoxybenzamine, which caused symptomatic hypotension. She was switched to doxazosin and eventually placed on atenolol. Review of systems other than previously mentioned in the case history is negative. She has no known drug allergies. Vital signs are as follows: HR 73, BP 142/97, T 36.6, SPO2 99%, Wt 28KG. Physical exam, including airway exam, is unremarkable other than a soft systolic murmur. Pre-operative lab values are as follows: Na 137, K 3.6, Cr 1.1, BUN 15, Hgb 13.8, Hct 41.3, Plt 369,000.

Questions:
Do you recommend any further preoperative testing? If so, what studies would you order? Should we involve any other health care providers in her perioperative management? If so, whom should we involve? Given that the surgeon is planning on performing a laparotomy, would your plan have changed if he/she had planned on performing a laparoscopic approach instead? Please comment on any preoperative recommendations to the family for the morning of surgery. What risks and possible complications should be disclosed to the family? How would you present these complications to them?

Case progression:
You see your patient on the morning of surgery. She is upset and crying, and her mother states that she is deathly afraid of “getting a shot.”

Questions:
Will you pre-medicate this child? Will you require a pre-operative peripheral intravenous line? Will she undergo a mask or intravenous induction? Will you place an arterial line before or after induction? Will you place a central venous line? How will you maintain anesthesia? What will you use for analgesia? Do you recommend an epidural for postoperative pain control? If so, how do you plan on placing the catheter? Will you run an epidural infusion during the case as a method of balanced anesthesia? What are your anesthetic hemodynamic goals for this patient? What pharmacologic agents would you use for blood pressure control and/or for blood pressure support? Are you concerned about the adequacy of alpha and beta blockade, and if so, how will you determine it?
**Intraoperative Care:**
The patient had an uneventful induction of anesthesia and intubation. An epidural catheter was placed for postoperative pain control, and test dose was negative for intravascular placement. Surgical excision was made by a right subcostal approach. Examination during surgery showed the tumor had no involvement with neighboring organs or with the renal vein. During dissection of the tumor, the patient’s blood pressure increases to 210/121. The heart rate remains unchanged at 80 beats per minute.

**Questions:**
Why do you think the heart rate remained unchanged? What is your differential diagnosis for this episode of hypertension? What is your recommended treatment?

Your intervention corrects the blood pressure, and the surgeon continues with the dissection. There is another blood pressure surge along with ST segment elevation.

**Questions:**
How do you respond? What is your recommended treatment? What is the cause of the ST segment changes? The surgeon is asking whether you want to call for help. Do you require a cardiology consultation?

Your intervention brings the blood pressure under control and the EKG normalizes. The surgery continues. Soon after resection of the tumor, the patient’s blood pressure drops to 56/34.

**Questions:**
What is your differential for the sudden hypotension? What treatment intervention do you employ? What vasopressor(s) would you use to bring up the blood pressure? Will she require continuous vasoactive support (i.e. an infusion)?

Your patient’s blood pressure recovers. Following tumor removal, the surgeon determines that revascularization or correction of the renal artery stenosis would carry a significant chance of failure and resultant recurrent hypertension. After extensive discussion and consultation with the parents, the decision was made to remove the right kidney. Total blood loss was approximately 150 mL. The total operative time was 4.5 hours. Pathological review of the tumor confirmed the diagnosis of paraganglioma.

Final ABG is 7.33/35/215/20, BE: -6.

**Questions:**
What is your plan for postoperative management of this patient? Will you extubate at the end of the case? Where should this patient go after the surgery - ICU, step-down unit, monitored bed? If you placed an epidural catheter, what will you give for pain management? Is the patient still at risk for hypertension or hypotension in the post-operative period?
Postoperative Care:
Your patient was successfully extubated at the end of the surgery and admitted directly to the ICU. Pain control was achieved with an epidural infusion of low concentration local anesthetic and opiate. She required low-dose continuous vasoactive support in the early postoperative period. In the afternoon of postoperative day #0 (approximately 4 hours after arrival to the ICU), a respiratory code blue is called, and the patient is found to be obtunded and significantly hypoglycemic. 1gm/kg of dextrose 50% is given with rapid reversal of her mental status. After this episode, her blood sugar remained stable. She was transferred to the ward in the evening of postoperative day #2.

Questions:
What is the differential diagnosis of her hypoglycemia? How is this related to her paraganglioma, if at all? Are any further labs or studies needed?

She had an otherwise uneventful hospital course and was discharged home.

References: