MRI with a Pheochromocytoma

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Objectives

- Discuss the origin of pheochromocytomas
- Discuss the receptors primarily activated with catecholamine release from pheochromocytomas
- Discuss the effect of long-standing hypertension on the heart
- Discuss the medications that can aid in reduction of hypertension associated with pheochromocytomas both preoperatively and intraoperatively

Introduction

A pheochromocytoma is a catecholamine secreting tumor that is derived from chromaffin cells of the adrenal medulla. The excess release of Epinephrine and Norepinephrine typically presents as hypertension, tachycardia, headaches and sweating. Symptoms can range from severe to asymptomatic. When left untreated, pheochromocytomas can lead to a muscular reconstruction of the heart, termed cardiomyopathy. To safely take care of patients with pheochromocytomas, recognition and proper preoperative and intraoperative medications can be necessary.

Preanesthetic Evaluation

A 4.7 kg, 4-month-old male presented to the emergency department 17 days prior to the MRI for an aspiration event prompting intubation and assessment of cardiac function. An echocardiogram was performed and showed an ejection fraction of 36% with elevated BNP and troponin.

Another echocardiogram was performed 7 days later and showed left ventricular hypertrophy with ejection fraction remaining at 31% with blood pressure 130/90 mmHg. Patient was started on Milrinone at 0.7 mcg/kg/min and Nicardipine at 2 mcg/kg/min at this time. After controlling the hypertension, another echo was performed on the day of the MRI, showing an ejection fraction of 69% with a blood pressure 103/51 mmHg. The Milrinone was planned to be weaned off the day before the MRI. The patient was noted to have elevated renin levels, metanephrine levels were elevated 4-5 times the upper limit of normal, with concurrent elevated normetanephrine. Nephrology was consulted for suspected pheochromocytoma, though nephrologist stated, “there is not a good established range for an infant this young”.

The patient was put on the schedule for an urgent MRI of both the abdomen and brain, in hopes of confirming suspected pheochromocytoma and evaluating new onset seizures.

The patient’s Captopril was held the morning of MRI, but the Isradipine was taken. The patient presented to the MRI holding bay with Nicardipine at 3.5 mcg/kg/min. A subclavian central venous catheter was available to use. Patient was able to hold his saturations in the mid 90’s on room air.

Anesthetic Plan

The anesthesia staff planned for a slow mask induction to induce general anesthesia. The airway would be secured with an LMA size 1. Additional access to be obtained included a peripheral IV and arterial line.

With heart failure, the goal was to maintain systemic vascular resistance and ensure adequate preload to ensure end organ perfusion. However, with the pheochromocytoma, the goal was to reduce blood pressure so as not to add to the hypertensive state produced by the excess catecholamines. These two goals were compromised with the end goal of remaining normotensive throughout induction and the procedure.

Anesthesia staff had Epinephrine, Phentolamine, and other rescue drugs ready. An ETT and blade were also prepared with anticipation of possible intubation necessary.

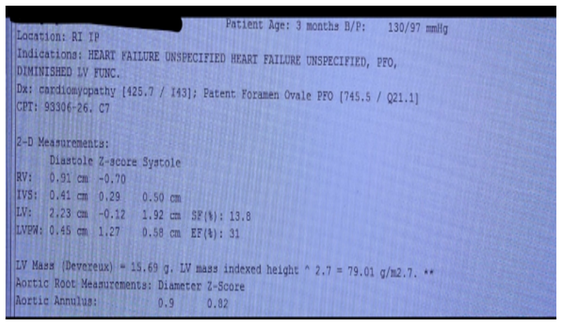

Image 1. Echocardiogram results before attempting to control the hypertension.

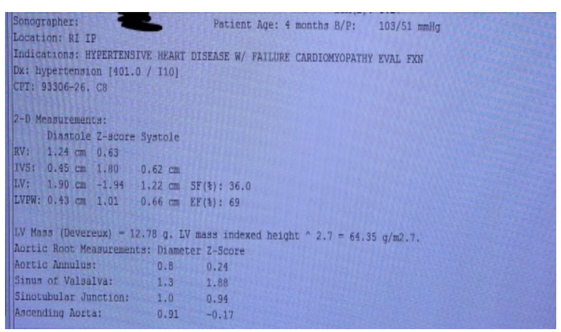

Image 2. Echocardiogram results after obtaining control of the hypertension.

Intraoperative Course

Sevoflurane was turned to 3% for induction, but breath holding still occurred with a jaw thrust. When the Sevoflurane was turned up above 3%, the child would go apneic. Even when apneic the patient would squirm with attempted arterial line placement. To ensure the patient was deep enough for placement of the LMA and arterial line, 8 mg of Propofol and 5 mcg of Fentanyl were given.

Upon transfer from the MRI induction room to the scanner the patient began to cough and buck causing desaturation. The anesthetic was deepened, but the LMA was still not seating appropriately. The decision was made to switch the airway to an ETT by giving 8 mg of Propofol and 3.5 mg of Rocuronium. The airway was secured with a grade 2a view.

The MRI was uneventful and the patient was reversed with Sugammadex before being transported to the PACU. The patient remained normotensive throughout.

Discussion

Pheochromocytomas hypersecrete Epinephrine and Norepinephrine, which in turn have several downstream cardiac effects. Norepinephrine preferentially binds to alpha 1 and alpha 2 receptors, while Epinephrine prefers to bind to Beta 1 and Beta 2 receptors. Activation of alpha 1 receptors causes smooth muscle constriction, preferentially constricting the arteries as compared to veins. Therefore, the excess Norepinephrine secreted by the pheochromocytoma can cause an increase in afterload that the heart must pump against.

When cardiac muscle is pumping against increased pressure, it begins to increase the strength and size of cardiac muscle, termed hypertrophy. Because of the pressure overload, the heart experiences increased systolic wall stress causing the heart to add muscle in parallel to increase the thickness and strength of the ventricle, termed concentric hypertrophy. Concentric hypertrophy is the hearts way of compensating to be able to overpower the added pressure it must now pump against. Concentric hypertrophy can lead to diastolic heart failure due to a loss of compliance of the ventricles. In diastolic heart failure, the heart struggles to relax to allow adequate filling of the ventricles due to the increased muscle mass, thus decreasing cardiac output with preserved ejection fraction.

Once a patient is known to have a pheochromocytoma, premedication is commonly started to control hypertensive episodes intraoperatively. It can also be used in hopes of preventing heart failure by reducing the afterload the heart is pumping against. This is accomplished by starting an alpha receptor antagonist, such as Phenoxybenzamine or Phentolamine.

Discussion Continued

Phenoxybenzamine is irreversible, noncompetitive, and nonselective at the alpha receptors, and most commonly started between 7-14 days before surgery. Phentolamine is another nonselective alpha receptor agonist, but it is competitive and therefore can be used intraoperatively to treat the hypertension. Both of these drugs are commonly given with a beta blocker to avoid reflex tachycardia through the baroreceptor reflex. However, it is important to make sure that alpha blockade is always initiated first, as unopposed alpha stimulation could cause a severe hypertensive crisis in a patient with a pheochromocytoma.

		Upper limit for normotensive individuals		Diagnostic cut-off values for pheochromocytomas
Metanephrine (μg/24 h)	Females	180		400
	Males	261		
Normetanephrine (μg/24 h)	Third decade	390		900
	Eight decade	560		

Source: Mayo Medical Labs and Quest Diagnostic/Nichols Institute.
[a]In the normotensive population and in pheochromocytoma, separated by a grey zone that could include medullary hyperplasia.

Table 1. Metanephrine and Normetanephrine levels for indication of pheochromocytoma

Summary

When a pheochromocytoma is not properly diagnosed, the excess Epinephrine and Norepinephrine can lead to a chronically hypertensive state. When the heart has to beat against an elevated pressure for an extended period of time, the heart begins to remodel leading to hypertrophy and eventually heart failure. However, with proper recognition and management, the receptors that catecholamines work on can be blocked to allow adequate time for the removal of the pheochromocytoma.

References

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