Pheochromocytoma: A Rare Etiology of Hypertension in the Pediatric Population

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Supplemental Video

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Introduction:
Pheochromocytoma (PCC) is a neuroendocrine tumor that arises from the adrenal medulla. Most are sporadic but they can also be found in the context of a hereditary syndrome. The symptoms are usually associated with the release of catecholamines such as epinephrine and norepinephrine. Catecholamine release can cause an array of symptoms such as hypertension, tachycardia, headaches, and diaphoresis. Pheochromocytomas are rare tumors in the general population more so in the pediatric population. These tumors are seen in 0.5-2% of pediatric cases.

Case Presentation:
A 13-year-old male presented to the emergency department with hypertension. Finding of malignant hypertension prompted a full work up, of which included evaluation for pheochromocytoma. Due to clinical suspicion, plasma metanephrine levels and a magnetic resonance imaging (MRI) of the abdomen were obtained to evaluate for a possible pheochromocytoma. Results of the abdominal MRI revealed a 2.4cm T2-hyperintense right adrenal mass and laboratory results obtained showed metanephrine, plasma: 86.1 (0-88.0 pg/mL) and normetanephrine, plasma: 4169.1 (0-86.1 pg/mL). The above findings were consistent with and confirmed the diagnosis of pheochromocytoma. The patient was referred to pediatric endocrinology and surgery for a right adrenalectomy. In preparation for this surgery, alpha-blockade was established.

Management/Outcome:
During anesthetic induction, the patient developed a hypertensive emergency, which prompted cancellation of his procedure at that time. The patient was later readmitted for medical optimization of his blood pressure prior to surgery. Once this was properly completed, he successfully underwent laparoscopic right adrenalectomy for his right adrenal pheochromocytoma. Final pathology report showed evidence of a succinate dehydrogenase B (SDHB) mutation, which is associated with a possible malignant potential. In addition, the Ki-67 labeling index, which describes the number of cells dividing, was found to be elevated at 6.4% if >6 is considered intermediate.

Learning Objectives
1. Describe the work up and management of an uncommon finding of pheochromocytoma in a pediatric patient.

2. Discuss the operative management of a pediatric patient after resection of a pheochromocytoma.