Wasn't MEN2B: A Case of Pheochromocytoma

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Abstract

Introduction

Pheochromocytoma is a rare neuroendocrine tumor that originates from the chromaffin cells of the adrenal medulla and is characterized by intermittent surges of catecholamine secretion. The majority of pheochromocytomas occur sporadically, however, they can be associated with genetic disorders (1). The clinical presentation of pheochromocytoma is broad and can range from asymptomatic to sudden cardiac arrest, therefore, it is important for clinicians to know when to suspect and how to manage pheochromocytoma.

Case Presentation:

A 24 y.o. Caucasian man with a medical history significant for childhood Henoch Schönlein Purpura was referred to the emergency department from optometry due to findings of hypertensive retinopathy after evaluation for blurry vision. His blood pressure was elevated to 240/180mmHg. He reported recurrent palpitations associated with facial pallor, headaches, and diaphoresis. These episodes increased in frequency and severity were associated with weight loss. His family history was negative for hormonal dysfunction or malignancies.

He was admitted to the ICU for treatment of malignant hypertension. 24-hour urine normetanephrines returned elevated at 5715 ug/24 hr. CT abdomen and pelvis with contrast demonstrated a right adrenal mass measuring 5.1 x 3.8 cm with 28 Hounsfield units consistent with a pheochromocytoma. He was started on alpha blockade and underwent right adrenalectomy once full alpha blockade was achieved. Histology revealed a neuroendocrine neoplasm consistent with pheochromocytoma. Biochemical testing completed 2 weeks later revealed urinary normetanephrine levels remained elevated post-operatively as did the patient's blood pressure. A meta-iodobenzylguanidine(MIBG) scan is planned to evaluate for possible extra-adrenal pheochromocytoma or residual tissue.

Discussion:

Pheochromocytomas release intermittent surges of catecholamines which can present with the classic triad of episodic headache, tachycardia, and diaphoresis although most patients do not present with the triad (2). Evaluation begins with measurements of plasma metanephrines and 24 hour urine metanephrines. Abdominal imaging with CT is warranted if metanephrine values are greater than two times the upper limit of normal. Treatment of pheochromocytoma begins with alpha blockade for at least seven days, followed by beta blockade, prior to adrenalectomy. Adequate alpha blockade must be achieved to prevent unopposed alpha vasoconstriction leading to hypertensive crisis or pulmonary edema. Although adrenalectomy is usually curative, to ensure the resection is complete, biochemical testing is completed 2-6 weeks after surgery. Extra-adrenal tumors predominantly secrete norepinephrine causing sustained hypertension, as was seen in this case, while adrenal pheochromocytomas predominately secrete epinephrine and are associated with paroxysms of hypertension (4).

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