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Case Report

Pheochromocytoma: A Rare Culprit of Uncontrolled Hypertension

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Abstract

Pheochromocytoma is a rare cause of hypertension that is curable. We present a case report of a 24-year-old female who presented with severely elevated blood pressure and palpitations. Investigations revealed a left adrenal mass that was successfully excised. A high index of suspicion is required to make a diagnosis of pheochromocytoma in young patients with severely elevated blood pressure.

Keywords: *Pheochromocytoma; Paroxysmal Hypertension; Metanephrines*

Background

A case report of a patient with a rare clinical condition: A left neuroendocrine tumor of adrenal origin with typical presentation of hypertension, palpitations, severe headaches, and sweating resulting from hormone excess. Pheochromocytomas are rare tumors, mostly benign and are catecholamine-producing tumors of the chromaffin of the adrenal medulla or of a paraganglion origin [1]. These tumors are estimated to occur in 2 - 8 out of 1 million people per year [2]. Patients with the disease may develop potentially devastating effects on multiple body systems (e.g. cardiovascular, cerebrovascular), and can lead to death if untreated [3]. The main aim of this case report is to have a high index of suspicion for pheochromocytomas.

Case Report

24-year-old female diagnosed with hypertension three months prior to her presentation. Primary complaint of episodic headaches mostly at the occiput and forehead, worse at night, associated with intermittent dizziness, palpitations and sweating. She was on amlodipine 10 mg, no history of alcohol or tobacco use and no familial history of hypertension.

Her physical examination was unremarkable apart from her blood pressure being elevated at 184/118 mmHg. The haemogram, renal function tests, glucose, and thyroid profile were normal. Endocrine tests revealed normal levels of aldosterone, renin and serum cortisol.

The plasma metanephrines was normal, however, the plasma normetanephrine levels were markedly elevated. The 24-hour urine metanephrine levels were also significantly raised along with an increased Normetanephrine: Creatinine ratio.

A bilateral renal Doppler ultrasound scan showed patent renal arteries. However, a left perinephric mass with pressure effect on the left renal artery resistive index of 0.18 to 0.82.

Helical axial CT scans of the abdomen and pelvis with oral and IV contrast revealed a large lesion with cystic areas in the left adrenal gland. The lesion had no calcifications or hemorrhage, displaced but did not invade the pancreatic tail, left kidney and the surrounding vessels. The left renal artery and left renal vein were well opacified with no filling defects suggestive of thrombus formation.

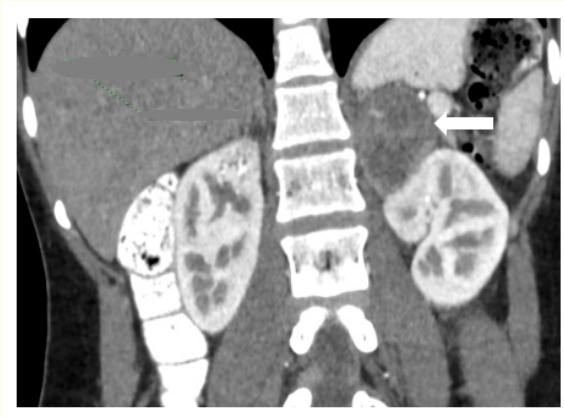

Figure 1: *Computed tomography scan of left adrenal cystic mass.*

Blood pressure was controlled with perindopril 5 mg, amlodipine 5 mg and indapamide 1.5 mg. Phenoxybenzamine 10 mg twice daily was administered to optimize the patient for a complete left adrenalectomy which was successfully performed a month later. She was nursed at the High Dependency Unit where she was given vasopressor support for a day before being weaned off and was later transferred to the surgical ward where she was later discharged in a stable condition.

The histology results of the excised left adrenal and perinephric mass showed: a dark biopsy aggregating to 5 cm x 2 cm x 2 cm on gross examination. The cut surface showed hemorrhage and yellow coloration in some areas. Microscopy sections showed a highly cellular tumor with round to oval cells arranged in nests exhibiting mild to moderate pleomorphism. No mitosis or necrosis was noted.

Discussion

Pheochromocytomas are catecholamine producing neuroendocrine tumors that can be adrenal or extra-adrenal in origin [4]. High circulating levels of catecholamines can lead to severe hypertension and can have devastating effects on multiple body systems (e.g. cardiovascular, cerebrovascular) [3]. They are often referred to as one of the 'great mimics' in medicine [5]. It is usually fatal if unrecognized. Manifestations can mimic many diseases and cause erroneous diagnoses.

In Africa, few case reviews have been reported. Availability of biochemical assays for catecholamines and their metabolites has improved our ability to establish or exclude the diagnosis of pheochromocytoma in a suspected patient. The tumor can occur at any age with equal gender distribution [4].

Pheochromocytoma can be associated with certain genetic syndromes such as multiple endocrine neoplasia type 2 (MEN 2), neurofibromatosis (NF) and von Hippel-Lindau (VHL) syndrome [4]. Approximately 15% are malignant, 18% extra-adrenal, and 20% familial [6]. Typical clinical manifestations are sustained or paroxysmal hypertension, severe headaches, palpitations and sweating resulting from hormone excess.

Plasma or urinary metanephrines are approximately 98% sensitive for detecting pheochromocytomas. They can be localized by MRI and CT; 131I- and 123I-metaiodobenzylguanidine (MIBG) are highly specific and 81% to 90% sensitive, respectively [6]. Diagnosis depends on clinical suspicion, demonstration of high levels of free catecholamines in the plasma or urine, or high localization of the tumor by appropriate imaging techniques that include CT scanning, MR imaging, and 131I-MIBG scintigraphy [7]. Because approximately one out of four pheochromocytomas turn out to be hereditary entities, screening for genetic alterations is important [1].

Surgical extirpation is the treatment of choice unless the risk of operation is overwhelming or distant metastasis has already occurred [7]. Laparoscopic and adrenal sparing surgical intervention following preoperative α-blockade is the treatment of choice and usually curative. In malignant pheochromocytomas, radiotherapy and chemotherapy are palliative treatment options [1].

Conclusion

The paroxysmal release of catecholamines leads to a potentially serious and lethal state which warrants a high index of suspicion for Pheochromocytoma and prompt diagnosis of the disease. A multidisciplinary approach in the process of management is crucial in ensuring appropriate hemodynamic control in the preoperative optimization of patients with the disease. Although surgical extirpation represents the main modality of ultimate cure, pharmacological perioperative management remains the main stay of successful outcome.

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