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Case Study

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AN INTERESTING CASE OF SECONDARYHYPERTENSION DIAGNOSED AS PHEOCHROMOCYTOMA

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ABSTRACT

Pheochromocytomas are rare tumors arising from chromaffin cells of the adrenal medulla. The clinical features result from excessive secretion of catecholamines. These tumors can be benign or malignant and are frequently associated with familial syndromes like neurofibromatosis type 1, multiple endocrine neoplasia type II, and Von-Hippel Lindau disease. In addition, sporadic pheochromocytomas are among the most frequently overlooked causes of secondary hypertension. Here, we delineate the presentation and imaging observations of a 45-year-old female patient where we diagnosed herwith pheochromocytoma while evaluating for secondary hypertension.

KEYWORDS: Pheochromocytoma, Chromaffin cells, Adrenal tumor, ZELLBALLEN appearance.

INTRODUCTION

Pheochromocytomas are an uncommon tumor of the adrenal gland, with characteristic clinical, and to a lesser degree, imaging features. The tumors are said to follow a 10% rule:

- ~10% are extra-adrenal
- ~10% are bilateral
- ~10% are malignant
- ~10% are found in children
- ~10% are not associated with hypertension
- ~10% contain calcification

The estimated prevalence of pheochromocytomas in hypertensive adults is thought to range from 0.1-0.6%. The incidence in the general population is believed to be around 0.05% based on autopsy series

The majority of cases are sporadic. In 25% of cases, a pheochromocytoma is a manifestation of an underlying condition, often familial, including:

- Multiple Endocrine Neoplasia Type Ii (Men2): both MEN IIa and MEN IIb
- Von Hippel-Lindau Disease
- Neurofibromatosis Type 1
- Sturge-Weber syndrome
- Carney triad: for extra-adrenal pheochromocytoma
- Tuberous Sclerosis
- Familial Pheochromocytoma

CASE REPORT

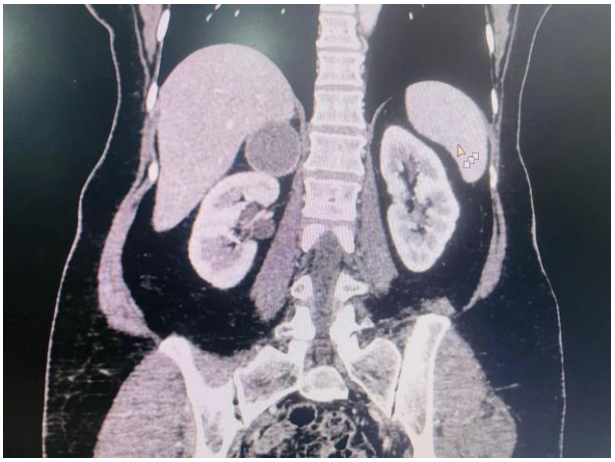
A 45 yr old female Came to ED with SOB with Accelerated HTN (BP of 180/120 mmhg) and Acute LVF presentation requiring Non-Invasive Ventilation. She wasrecently diagnosed as HTN (6 months back) and on irregular treatment. No Family h/o HTN, She is married & having 2 children (No h/o PIH). She has a H/O palpitations, headache on & off chest discomfort.

After stabilization with NTG, Diuretics, NIV. She was worked up for secondary HTN.
Hb was 12.2g/dL, CUE - no proteinuria & Creat was 1.4.

On USG Abd Both kidneys normal in size, Grade - I RPCs are present On further evaluation, Renal artery doppler was found to be negative2D ECHO revealed mild concentric LVH with Good LV Function.

CT Aortogram was normal. 24hr urinary free Cortisol, overnight dexamethasone suppression test was normal.

Thyroid profile, S.calcium, PTH levels were within normal limits Further, the patient underwent an abdominal CT scan which revealed 44 x 43 mm well defined homogeneous right suprarenal mass.

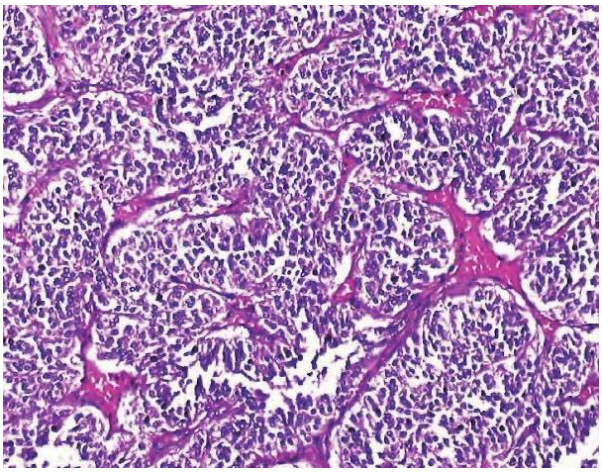


We sent samples for PRA, Aldosterone, ACTH levels which turned out to be negative.

On further evaluation 24hr urinary metanephrines were sent and levels are found to be 428 micrograms/24hrs (high). Finally a diagnosis of Pheochromocytoma was made.

Two weeks later after being prepared according to the guidelines for pheochromocytoma resection. She underwent endoscopic retroperitoneal adrenalectomy.

Histopathologic examination of the adrenal showed that it was a typical pheochromocytoma, with large polygonal cells arranged in a thick nest & separated by rich capillary network (ZELLBALLEN appearance).



Post surgery there was again turbulence in BP management for which Betablockers, Nitroprusside was given and managed conservatively.

On discharge patient was kept on Bisoprolol 5mg and she was on follow upsince 2 months and doing well.

DISCUSSION

Pheochromocytoma is a rare but classical cause of uncontrolled secondary hypertension, with a minority having superimposed paroxysmal hypertensive crises. A broad spectrum of potential presenting symptoms includes the classic triad of headaches, palpitations, and profuse sweating.

The presence of hypertension resistant to multiple antihypertensive medications therapies should raise suspicion for pheochromocytoma.

- Hypertension occurs in about 70% to 90% of patients
- Sustained hypertension-50% because of persistent vasoconstriction d/tnorepinephrine
- Paroxysmal hypertension -45% d/t hyperadrenergic spells of epinephrine
- Normotensive 13% d/t small size tumours or predominantly dopaminesecreting tumors

Pheochromocytoma should be suspected in patients who have one or more ofthe following:

1. Hyperadrenergic spells (e.g., self-limited episodes of nonexertional forceful palpitations, diaphoresis, headache, tremor, or pallor)
2. Resistant hypertension
3. A familial syndrome that predisposes to catecholamine-secreting tumors (e.g., MEN2, NF1, VHL.)
4. A family history of pheochromocytoma
5. An incidentally discovered adrenal mass with imaging characteristics consistent with pheochromocytoma
6. Pressor response during anesthesia, surgery, or angiography
7. Onset of hypertension at a young age (<20 years)
8. Idiopathic dilated cardiomyopathy

CONCLUSION

Clinical suspicion is almost always present before imaging: secondary hypertension and positive urinary catecholamines, usually large and heterogeneous adrenal masses, with cystic and necrotic components.

Patient education is critical to the management of pheochromocytomas. It is important for them to understand the episodic nature of symptoms prevalent in the condition. They should be informed that episodes of severe hypertension may be precipitated by medications like dopamine receptor antagonists, beta-blockers (typically non-selective), tricyclic antidepressants, corticosteroids, sympathomimetics, and neuromuscular agents, or may be noted in the setting of surgery or induction of anesthesia. Several blood, urine, and imaging tests are available to assist medical professionals in diagnosing pheochromocytoma. Options for treatment include medications to control blood pressure and other symptoms. Options for surgical removal of the tumor are also available depending on several factors.

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