Challenges in Pheochromocytoma Diagnosis: A Case Study with Unusual MRI Findings and Mild Symptoms.

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INTRODUCTION

Pheochromocytomas are catecholamine-secreting tumors originating from the chromaffin cells of the adrenal medulla.

The diagnosis of pheochromocytoma is often prompted by the classic triad of symptoms: episodic headache, sweating, and tachycardia.

Additionally, patients may have sustained or paroxysmal hypertension.

Although sustained or paroxysmal hypertension is the most common sign of pheochromocytoma, a noteworthy minority (around 5-15%) of patients present with normal blood pressure.

While some radiographic features are commonly seen in pheochromocytomas, imaging lacks diagnostic specificity.

Biochemical evaluation is required to make the diagnosis.

We present a case of a pheochromocytoma presenting with mild symptoms and unusual features on MRI.

CASE PRESENTATION

A 73-year-old African American female with a history of hypertension and atrial fibrillation presented to urgent care with a few weeks' history of daily headaches and generalized abdominal discomfort.

She was advised to continue treating symptoms.

Two weeks later, upon follow-up with her primary care physician, her headaches and abdominal pain had improved but were still occurring every few days.

An abdominal CT scan revealed a retroperitoneal mass, and further investigation with MRI identified a 5.2 x 2.9 cm mass compressing the posterior wall of the inferior vena cava and approximating the right adrenal gland.

She was referred to Surgical Oncology for consideration of biopsy versus resection of the mass; however, biochemical testing demonstrated elevated plasma normetanephrine and metanephrine and she was thus referred to Endocrinology for further evaluation.

Updated imaging was performed to ascertain more detailed information about the radiographic features of the mass.

Contrasted MRI demonstrated slight enlargement of the mass over the preceding 6 months, now measuring 5.7 x 3.1 cm.

Notably, the mass was not found to be hyperintense on T2-weighted images.

Contrasted CT demonstrated a heterogeneous mass of intermediate density (pre-contrast 32 Hounsfield units) arising from right adrenal gland. Absolute contrast washout was 66%.

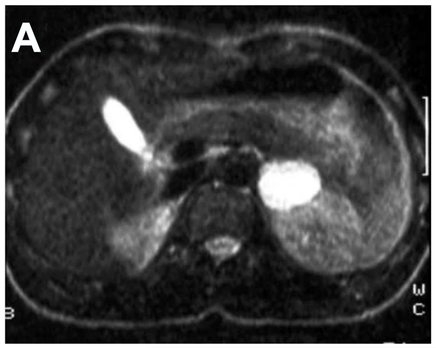
Further biochemical workup revealed elevated levels of epinephrine, norepinephrine, normetanephrine and metanephrine.

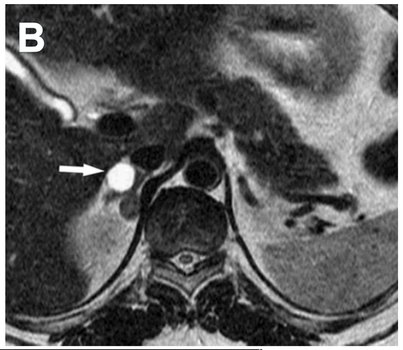
Levels of plasma dopamine, plasma aldosterone, plasma renin activity, random serum cortisol, and ACTH were all normal, and her 8am serum cortisol was appropriately suppressed (1.1 ug/dL) after an overnight 1 mg dexamethasone suppression test.

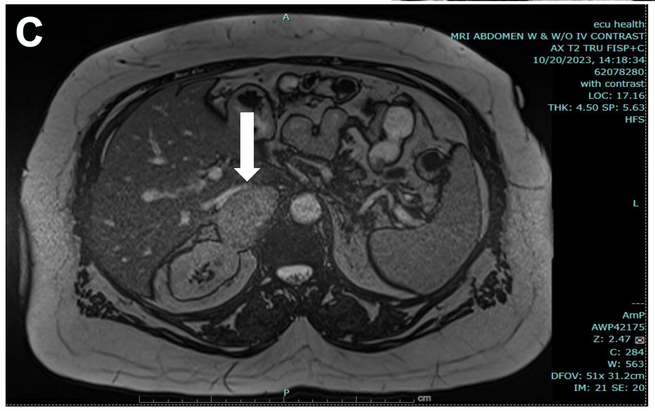
After pre-treatment with alpha blockade, she underwent successful laparoscopic right adrenalectomy, with surgical pathology confirming a 6.0 cm pheochromocytoma.

RESULTS

Test	Patient Result	Reference Range
Normetanephrine	47 nmol/L	(<0.90 nmol/L)
Metanephrine	1.7 nmol/L	(<0.50 nmol/L)
Epinephrine	107 pg/mL	(<95 pg/mL)
Norepinephrine	3939 pg/mL	(217 – 1109 pg/mL)
Dopamine	< 40 pg/mL	(< 40 pg/mL)
Renin	< 0.6 ng/mL/h	(2.9 – 10.8 ng/mL/h)
Aldosterone	6.5 ng/dL	(<21 ng/dL)
ACTH	31 pg/mL	(7.2 – 6.3 pg/mL)







A-B: MRI T2 Pheochromocytoma.

C: Patient MRI T2 Pheochromocytoma.

DISCUSSION

The presented case highlights the critical role of biochemical investigations in the evaluation of adrenal masses.

While imaging modalities such as CT and MRI provide valuable anatomical information, they often lack specificity in distinguishing functional from non-functional lesions.

In our patient, imaging did reveal some non-reassuring features including pre-contrast density of > 10 Hounsfield units on CT scan but did not demonstrate the cardinal feature of hyperintensity on T2-weighted MRI.

In fact, only one-third to two-thirds of cases exhibit this characteristic of hyperintensity on T2-weighted MRI.

It is crucial to avoid biopsy of an adrenal mass that has not been evaluated for functionality, as inadvertent biopsy of a pheochromocytoma can lead to serious complications from a catecholamine surge, including acute hypertension and hypertensive crisis.

Additionally, it is crucial to remember the increased risk of catecholamine surge during surgery. Therefore, pre-treatment with alpha blockade and volume expansion is always necessary before surgery.

KEY POINTS

Paroxysmal hyperadrenergic spells (headache, diaphoresis, palpitation, pallor, and anxiety) in a hypertensive patient suggest pheochromocytoma.

The best screening test is either fractionated urinary or plasma-free metanephrines.

Tumor localization is performed by CT or MRI.

Surgical resection is the definitive treatment of pheochromocytoma.

Exercise caution with pheochromocytomas. Biopsies pose a risk of triggering catecholamine release or crisis.

REFERENCES

McDermott, S., McCarthy, C. J., & Blake, M. A. (2015). Images of pheochromocytoma in adrenal glands. Gland Surgery, 4(4), 350–358. https://doi.org/10.3978/j.issn.2227-684X.2014.11.06

Radulović, M. K., & Strojan Fležar, M. (2022). Pheochromocytoma presenting as a pancreatic mass: Avoiding a dangerous pitfall in endoscopic ultrasound-guided fine needle aspiration biopsy using GATA3 immunostain. *Cytopathology*, 33(1), 127-131. **https://doi.org/10.1111/cyt.13060**

Casola, G., Nicolet, V., vanSonnenberg, E., Withers, C., Bretagnolle, M., Saba, R. M., & Bret, P. M. (1986). Unsuspected pheochromocytoma: risk of blood-pressure alterations during percutaneous adrenal biopsy. *Radiology*, 159(3). Retrieved from https://doi.org/10.1148/radiology.159.3.3517958

Zhang, L., Åkerström, T., Mollazadegan, K., Beuschlein, F., Pacak, K., Skogseid, B., & Crona, J. (2023). Risk of complications after core needle biopsy in pheochromocytoma/paraganglioma. Endocrine-Related Cancer, 30(7), e220354. https://doi.org/10.1530/ERC-22-0354