Adrenal

ADRENAL CASE REPORTS

Inflammatory Myofibroblastic Adrenal Tumor: An Extreme Rare Cause of Adrenal Incidentaloma

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Introduction: An inflammatory myofibroblastic tumor (IMT) is considered a complex disease with a suggested neoplastic nature. It has a variable histological and clinical presentation. Up to now, the exact etiology is still unknown. Various sites such as the lungs (first site described in 1939), the heart, the gastrointestinal tract, and the genitourinary tract have been reported in the literature. The adrenal glands are considered an extremely rare location with no recurrence reported after surgical management. Up to our knowledge, only 9 cases have been described in the literature. The age of presentation was variable (from newborn to 57 years old). Case Report: A 70 years old female with a past medical history of well-controlled diabetes type 2, and hypertension presented with a progressively increasing chronic right flank pain for the past couple of months, associated with 10 lb weight loss. Workup was done and an incidental right adrenal mass was found on CT scan. The mass was 3.2cm x 3.3 cm partially necrotic with adjacent stranding abutting the right adrenal gland and the liver. The opposite gland was normal and there was no evidence of any other mass lesion in the abdomen. MRI adrenal showed 3.2x2.5x3.6 cm solid heterogeneous mass in the right adrenal gland (Hypointense on T1 and T2 weighted images. She had no sign or symptoms suggestive of Cushing and/or pheochromocytoma. Her hypertension was well controlled on lisinopril 10 mg. Hormonal workup was within normal range except for a mild elevation of DHEAS. Given the size of the mass, the worrisome radiological characteristics, and a family history of pheochromocytoma in her daughter, surgical options were discussed with the patient. Subsequently, the patient underwent the right robotic adrenalectomy. The postoperative course was uneventful. Pathology revealed a 6.5x6x 5 cm IMT of the adrenal. The CT scan done 6 months postoperatively didn't show any recurrence. **Discussion:** and conclusion: Adrenal IMTs are extremely rare but they should be considered in the differential diagnosis of adrenal masses. Follow-up is warranted since its behavior still remains uncertain. More cases are needed to unmask the true biological behavior and pathogenesis of adrenal IMTs.

Adrenal

ADRENAL CASE REPORTS

Interpreting Normetanephrines-the Significance of Clinical Context

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Introduction: Pheochromocytoma ranges in presentation, but the diagnostic hallmarks are increased catecholamine secretion in patients with an adrenal mass. However, interpretation requires consideration of the clinical state.

Case: A 60-year-old man presented with 6 months of episodic flushing, night sweats, fatigue, back pain, and 20-pound weight loss. Vitals were notable for hypotension and mild tachycardia. He was thin with evidence of hyperpigmentation. Contrasted CT showed 15 cm heterogeneous masses of both adrenals and a rib lesion. Plasma normetanephrines were elevated on two separate measures (526 and 1,398 pg/mL, nl <=148). Plasma metanephrines were normal. Urine normetanephrines were increased 4-fold (2,165mcg/g Cr, n 108–524). Labs noted low DHEA-S (2 mcg/dL, n 38–313), high ACTH (204 pg/mL, n 6–50), random cortisol (5.2 mcg/dL, n 3.5–18.3), undetectable aldosterone, and high plasma renin activity (83 ng/mL/h, n 2.9–10.8). Cortisol peaked at 6.7 mcg/dL 1 hour after 250mcg of cosyntropin. MRI revealed over 15cm infiltrating masses arising from adrenals with modest flare on T2 imaging. Dotatate PET/CT showed mild uptake of bilateral adrenals and metastases to liver and bone. He was started on steroid replacement and doxazosin for presumed pheochromocytoma.

He was transferred to our facility where he remained hypotensive despite adequate glucocorticoid and mineralocorticoid replacement. Once stabilized, repeat plasma normetanephrines were insignificant (218 pg/mL, n 0–145) and chromogranin was normal (68 ng/mL, n<93). Biopsy of the rib lesion confirmed diffuse large B-cell lymphoma.

Discussion: Pheochromocytoma classically presents in the outpatient setting with palpitations, diaphoresis, pallor, and paroxysmal hypertension. Rarely, it results in hypertensive emergencies requiring hospitalization. In patients with catecholamine excess and an adrenal mass, pheochromocytoma is usually suspected. 3-fold or more increases of normetanephrines or metanephrines above upper cutoffs are rarely false-positives. However, this assumes clinical stability. Sympathoadrenal activation is a physiologic response to acute illness. Metanephrines in critically ill patients without pheochromocytoma approached those of pheochromocytoma in one study. Levels normalized upon recovery from illness, highlighting the importance of verifying biochemistries once the patient is stable. The degree of elevation in normetanephrines seen in this patient was a physiologic response to adrenal insufficiency and clinical instability. Pathology confirmed lymphoma rather than pheochromocytoma for which he was started on chemotherapy.

Conclusion: Elevated normetanephrines is an appropriate physiological response in the acutely ill. Careful consideration of the clinical picture in conjunction with biochemical data is critical.

Adrenal

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Large Bilateral Adrenal Myelolipomas in the Setting of Congenital Adrenal Hyperplasia

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