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CASE REPORT

A Rare Case of Catecholamine-Secreting Adrenal Myelolipoma

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Abstract

Adrenal myelolipoma (AML) is a rare, benign, asymptomatic, nonfunctioning tumor of the adrenal cortex detected incidentally. AML can be accompanied by several other endocrine disorders simultaneously. Here, we report a case of a 36-year-old female with primary hypothyroidism and metabolic syndrome accompanied by severe hypertension and pheochromocytoma. However, the histopathological examination of the excised adrenal gland confirmed myelolipoma. Following surgery, her plasma nor-metanephrine levels decreased to normal values and the patient became normotensive, which suggested that the mass was functioning.

Key words: adrenal myelolipoma, hypertension, normetanephrine

INTRODUCTION

Adrenal myelolipoma (AML) is a rare, benign, asymptomatic, non-functional, incidentally detected tumor of the adrenal cortex, composed of mature adipocytes and hematopoietic tissue. It was first described by Gierke in 1905 and the term 'myelolipoma' was given by Oberling in 1929.[1] It is sporadic with an overall incidence previously reported to be 0.08%–0.4% on autopsy, but is recently reported to have an increased incidence of 10%–15% due to the widespread use of imaging.[2] Men and women are equally affected with its occurrence being more common between the fifth and seventh decades of life.[2] These lesions are usually unilateral and incidentally detected.[2] A certain number of bilateral tumors have been described in the literature. Myelolipomas are often smaller than 4 cm in diameter but can reach wider sizes.[3] Those measuring more than 10 cm in diameter are termed giant adrenal myelolipomas. The largest adrenal myelolipoma reported in the literature weighed 6 kg and measured 31 cm × 24.5 cm × 11.5 cm.[4,5] Surgical intervention is reserved for patients who attain a substantial size, considering the risk of rupture and retroperitoneal hemorrhage. These tumors can rarely be functional, leading to endocrinopathies.[6,7] Primary hypothyroidism is a relatively common condition in the Indian population, with the prevalence being 11%, compared with only 2%-4.6% in the Western population.[8] As such, there is no common pathogenesis between adrenal myelolipoma and thyroid dysfunction. Hence, the co-occurrence of hypothyroidism and the adrenal mass in our case is likely coincidental. However, certain autoimmune conditions can impact both the thyroid gland and adrenal cortex resulting in combined hormone deficiencies as part of autoimmune polyglandular syndrome type 1 (APS-1).

CASE

A 36-year-old Indian female, known case of primary hypothyroidism for the last 11 years, on levothyroxine (100 mcg daily), with anti-TPO levels suggestive of Hashimoto's thyroiditis (496 IU/ml; N.V. <10 IU/ml), presented with complaints of oligomenorrhea, hirsutism, and weight gain over the last 2-3 years without any cushingoid features. Initial weight before hypothyroidism was 65 kg and her current weight is 80 kg with body mass index is 29.4 kg/m^2. The patient was suspected to have polycystic ovarian syndrome, with ovarian ultrasound and fasting insulin levels consistent with PCOS. She satisfied the criteria for metabolic syndrome according to the NCEP ATP-III criteria (Table 1). She was found to have very high blood pressure readings (200/110 mm Hg) on multiple hospital visits with home BP monitoring values also suggestive of severe hypertension. Retrospectively, she also reported a history of intermittent palpitations, headaches, and sweating for the last 3-4 months. Based on her clinical history, she was evaluated for secondary causes of hypertension. There was no history of adrenal gland disease in any of the patient's family members.

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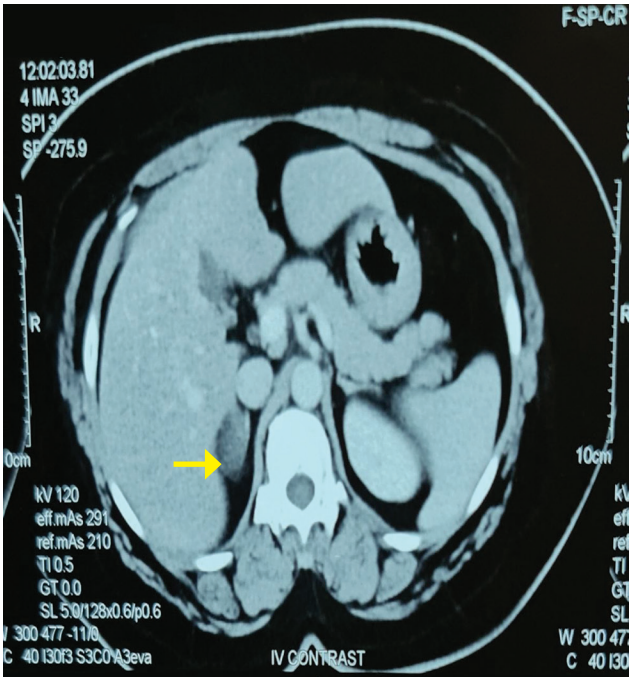

Figure 1. The axial image of computed tomography scan showing a right-sided adrenal mass with fat attenuation value 1 *(yellow arrow)*.

Table 1. NCEP ATP III Criteria for Metabolic Syndrome

Features	Patient's value
Waist Circumference (cm)	96
Fasting Triglyceride Level (mg/dl)	200
Blood Pressure (mm Hg)	200/110
HDL Cholesterol (mg/dl)	40
Fasting Blood Sugar (mg/dl)	95

The patient's serum cortisol after 1 mg overnight dexamethasone suppression test (ONDST) was suppressible (1.0 mcg/dl), and plasma normetanephrine was very high at 836 pg/ml (N.V. <196 pg/ml) with normal metanephrine levels by liquid chromatography with tandem mass spectrometry method (samples were obtained with protocol). The patient was not on any medication or diet which might have raised catecholamine levels. Her potassium level was normal (3.8 meq/L) with normal plasma aldosterone level and normal plasma renin activity (PRA). Her testosterone and dehydroepiandrosterone sulfate (DHEAS) were within normal limits. Contrast-enhanced computed tomography (CECT) of the abdomen showed a right-sided well-defined ovoid hypodense lesion with enhancing soft tissue components arising from the right adrenal gland measuring 3.1 x 1.8 x 1.3 cm with few coarse calcifications and fat attenuation value of 1 with normal left adrenal gland (Figure 1). Therefore, a diagnosis of pheochromocytoma was considered and the patient was shifted to prazosin (alpha-blocker). After achieving adequate alpha blockade, metoprolol (beta blocker) was added for controlling tachycardia.

The patient underwent right adrenalectomy via laparoscopic transperitoneal approach. The descending colon was mobilised, and the left renal vein was identified. The adrenal vein was clipped and divided. The mass was separated from the upper pole of the kidney and its lateral attachments using blunt and sharp dissection. There were no significant blood pressure fluctuations intra-operatively.

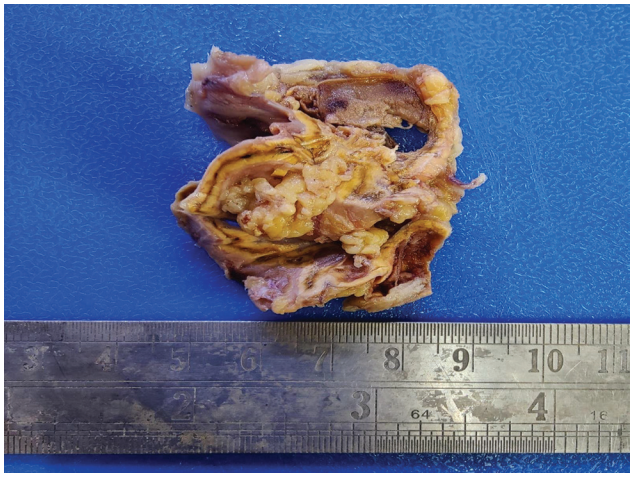

Figure 2. Gross specimen. Compressed adrenal at the periphery with fatty tissue in the center with specks of dark brown area.

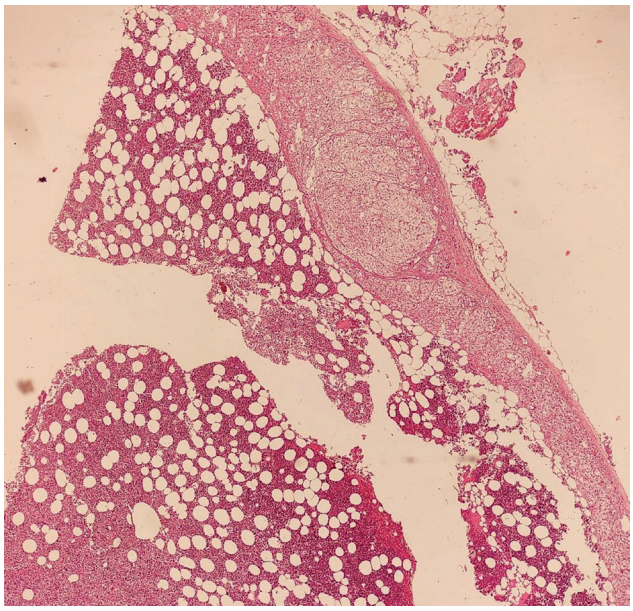

Figure 3. Compressed adrenal parenchyma at periphery, the center of the lesion showing cellular lesion with entrapped adipocyte [Hematoxylin & Eosin (H & E), 4X].

The patient did not require antihypertensive therapy after surgery, and plasma normetanephrine returned to normal (156 pg/ml) after 10 days of surgery. Gross and histopathological examination revealed myelolipoma (Figures 2–4) with no evidence of pheochromocytoma or medullary hyperplasia with negative chromogranin A staining. These features suggested that the mass was functioning and secreting catecholamine. She remains normotensive without treatment in the follow-up of three months after surgery.

Table 2. Comparison Between Index Case and other Case Reports

Features	Tamidari H et al.	Udupa S et al.	Jakka N et al.	Jindal T et al.	Adapa S et al.	Index Case
Age (Years)	48	58	40	55	40	37
Gender	Female	Male	Male	Female	Male	Female
Side of Tumor	Right	Left	Right	Left	Right	Right
Chief Complaints	Right upper quadrant discomfort, Anxiety attacks with sweating x 45 days	Left Hypochondrium and Left Lumbar pain, BP - 170/110	Hypertension, Adrenal mass	Heaviness in Left Flank	Testicular and Bilateral pedal edema, BP -234/119	Oligomenorrhoea, Hirsutism, Weight gain for 5 years, Episodic paroxysm x 4 months
Past History	Hypertension x 12 years, Controlled on Beta blocker	Diabetes Mellitus, Sustained Hypertension on treatment	Hypertension x 3 years (on treatment)	Hypertension x 2 years controlled on treatment	Hypertension, Chronic Kidney Disease, Morbid obesity, Adrenal mass	Primary hypothyroidism x 8 years
Imaging Findings	11 x 10.5 x 7 cm, well encapsulated, non homogenous, low density (-100 to -200 HFU) suggestive of (s/o) Right Adrenal Mass	18.4 x 10.1 x 9 cm, intermixed area of fat and mildly enhancing soft tissue component s/o left adrenal myelolipoma	9.8 x 8.5 cm, well defined, heterogenous, low density (-80 to -100 HFU) s/o Myelolipoma	10 x 8 x 6 cm, supra renal hypoattenuating mass with macroscopic fat s/o Adrenal Myelolipoma	9.7 x 7.7 x 6.1 cm, hypoattenuating right adrenal mass with attenuation value is 0	3.1 x 1.8 x 1.3 cm, well defined hypodense lesion with enhancing soft tissue component with areas of fat attenuation, Average attenuation value is 1
Investigations	24 hour urine metanephrine increased	24 hour urinary vanillylmandelic acid increased	24 hour urine metanephrine increased	24 hour urinary metanephrine increased	norepinephrine and dopamine increased	Plasma free Normetanephrine increased
Post op value	Normal	Normal	Normal	Normal	Not mentioned	Normal
Post op BP	Normal	Normal	Normal	Normal	Not mentioned	Normal
Biopsy	AML	AML	AML, Chromogranin A +ve	AML	AML	AML, Chromogranin A -ve

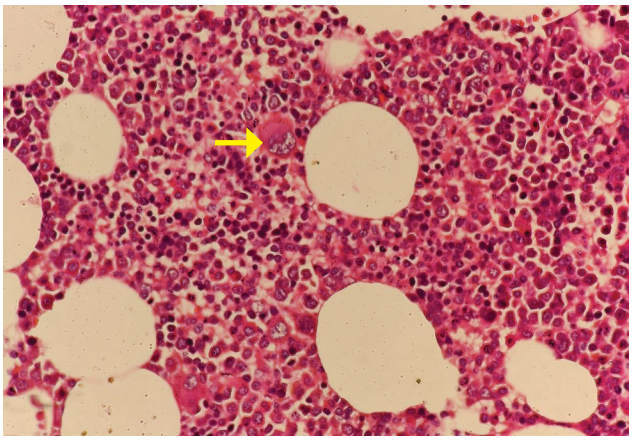

Figure 4. Hematopoietic element comprising megakaryocytes *(yellow arrow)* and erythroid colonies [H & E, 40 X].

DISCUSSION

Adrenal myelolipoma is a nonfunctional benign neoplasm that originates from the adrenal cortex and comprises mature fat and hematopoietic elements. The exact etiology is unknown, but it has been postulated that the lipomatous elements originate from the fat-containing mesenchymal stromal cells of the adrenal cortex, whereas the hematopoietic elements are derived from the reticuloendothelial cells of the blood capillaries. Cross-sectional imaging by computed tomography or magnetic resonance imaging can confirm the diagnosis due to presence of macroscopic and microscopic fat with negative attenuation value with or without calcification. These tumors are considered non secretory, and functional evaluation was not considered mandatory during their work up.[6,7,9] However, this recommendation has been questioned. In a review of the literature, it was observed that nearly 7% of adrenal myelolipomas may be functionally active.[7] The majority of these 'functional myelolipomas' were associated with hypercortisolemia followed by hyperaldosteronism. AML can coexist with other endocrine disorders like Cushing syndrome, congenital adrenal hyperplasia (CAH), primary aldosteronism, and pheochromocytoma. To the best of our knowledge, only seven cases of adrenal myelolipomas have been reported which were associated with catecholamine secretion (Table 2).[7,9-13]

It is interesting to note that all of these patients had giant adrenal masses (>10 cm),[14] but in our case, it is smaller. In our case report, 24-hour urine catecholamine was not measured but we have surrogate evidence that the AML was functional with a >4 times increase in plasma normetanephrine level which was very unlikely to be a false positive result. Also, following surgical excision, the levels decreased to normal. Though the blood pressure normalized after surgery, long-term monitoring by assessing plasma normetanephrine and metanephrine levels annually has been planned. Congenital adrenal hyperplasia has been reported with AML due to chronic adrenocorticotropic hormone (ACTH) stimulation of the adrenal glands.[15] Comorbidities like hypertension, obesity, diabetes mellitus, atherosclerosis, and malignancy have been associated with AML.[16] Few hypotheses have been postulated for the presence of hypertension in a patient with adrenal myelolipoma which include mechanical compression on the renal vessels by the tumor, mechanical irritation of myelolipoma, hemorrhage within the tumor, cortisol or aldosterone hypersecretion

and rarely due to catecholamine hypersecretion, as in the index case. Endocrine workup is beneficial in AML patients with hypertension, in younger patients, in persons with diabetes mellitus or prediabetes, and those with bilateral AML.[7] Thyroid stimulating hormone (TSH) might rise a little in obese patients due to high leptin levels and inflammatory cytokines levels which improve after weight loss.[17] Obesity has been associated with AML but there is no correlation between AML dysfunction and thyroid dysfunction.[16] Genetic studies could not be done due to financial constraints in this index case.

CONCLUSION

Adrenal myelolipoma with hypertension may not be coincidental, it may be due to catecholamine secretion. This case highlights the importance of a detailed workup for adrenal myelolipoma.

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Ethical Consideration
Patient consent forms were obtained before manuscript submission. The potential life-threatening nature of the lesion was explained to the patient and she agreed to surgery.

Statement of Authorship
All authors are certified in fulfillment of ICMJE authorship criteria.

CRediT Author Statement
VJ: Conceptualization; Writing – original draft preparation, Writing – review and editing, Visualization, Project administration; **AA:** Conceptualization, Writing – review and editing, Visualization, Project administration; **BK:** Writing – review and editing; **PS:** Writing – review and editing, Supervision; **HG:** Writing – review and editing, Supervision

Author Disclosure
The authors declared no conflict of interest.

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