

Suprarenal Mass with Hypertension: A Diagnostic Dilemma

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Suprarenal mass with hypertension points towards endocrine etiology like pheochromocytoma, primary aldosteronism and Cushing's disease. Here we report a 50 year old female who presented with adrenal mass and hypertension. On radiology and histopathology, she was reported as having benign adrenal myelolipoma. Her pre-operative and post-operative vanillylmandelic acid in 24 hrs urine were within normal limits. Rarely adrenal myelolipoma can be associated with catecholamine secretion. This case is reported here to highlight the importance of good clinical workup when faced with the diagnostic dilemma of a functional adrenal myelolipoma or pheochromocytoma as a differential to suprarenal mass associated with hypertension along with brief review of few rare case reports.

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Introduction

Adrenal myelolipoma are benign tumor of the adrenal cortex with fat and myeloid components. They were first described by Gierke in 1905.¹ They are usually unilateral and as most cases are asymptomatic and non-functional, are found incidentally on imaging or autopsy. Symptoms are generally related to their increasing size and pressure effects on adjacent tissue or organs which can present as pain abdomen or as a palpable mass in the flank.

Case Report

We received specimen labeled as right adrenal mass. The patient was a 50 year old female with history of giddiness for last one year and complaints of right abdominal pain for the past two months along with vomiting. She was on treatment for hypertension for the last 2 months, and her blood pressure at admission was 150/90 mm of Hg.

Her routine blood investigations showed mild microcytic hypochromic anemia with normal white cell counts and platelet count. Urine examination showed no proteinuria or glucosuria. Her thyroid hormone profile showed elevated thyroid stimulating hormone (TSH) with slightly raised total triiodothyronine (TT3) and

normal total thyroxine (TT4). Her serum anti-TPO was 159 AU/ml with >20 AU/ml considered as positive/raised levels (Table 1).

On CECT abdomen, a 39x39x52 mm size well-defined soft tissue lesion with predominantly fat attenuation was seen in right adrenal gland with peripheral irregular soft tissue component and no obvious calcification or increased vascularity, consistent with benign myelolipoma.

In view of hypertension with right adrenal mass, a provisional diagnosis of pheochromocytoma as a possibility was suspected. On subsequent workup, her 24-hour urine vanillylmandelic acid (VMA) by high-performance liquid chromatography was 5.66 mg (reference range is 1.60 to 7.30 mg in 24 hours urine). Her 2D echocardiography showed left ventricular ejection fraction of 60% and was reported as a normal echo doppler study at rest (Table 1).

Thus, she was successfully managed laparoscopically with excision of the right suprarenal mass, leaving the rest of the right suprarenal gland behind. In histopathology, we received multiple soft to firm yellowish tissue pieces, aggregate measuring 5 x 4 x 1.5 cm (Fig 1). Sections submitted showed a residual compressed part of the adrenal cortex under the adrenal gland capsule with

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Table 1: Results of investigations (in chronological order)

S.No	Name of test	Date of test	Result	Reference range
1	Hemoglobin	15-11-2021	10.6	12–15 (gm/dl)
2	Total white cell count	15-11-2021	8.70	4–10 (thousand/ml)
3	Platelet count	15-11-2021	285	150–410 (thousand/ml)
4	CECT abdomen	16-11-2021	Benign myelolipoma- right adrenal	-
5	Echo doppler	24-11-2021	Normal at rest	-
6	Thyroid profile	30-11-2021	TSH: 8.26 TT3: 2.16 TT4: 10.6	0.35–5.5 (μIU/ml) 0.8–1.9 (ng/ml) 4.5–12.6 (μg/dl)
7	Anti- TPO	11-12-2021	159.0	< 20 (AU/ml)
8	Thyroid profile	28-12-2021	TSH: 4.85 FT3: 3.28 FT4: 0.878	0.35–5.5 (μIU/ml) 2.77–5.27 (pg/ml) 4.5–12.6 (ng/dl)
9	24 hr urine VMA	04-01-2022	5.66	1.60–7.30 (mg/24 hrs)
10	Post op 24 hr urine VMA	22-02-2022	1.71	1.60–7.30 (mg/24 hrs)

prominent zona fasciculata. Rest of the sections showed mature adipocytes with islands of hematopoietic cells showing trilineage differentiation. Numerous megakaryocytes were seen (Fig 2, 3). No portion of the adrenal medulla was seen. There were no features of hyperplasia or pheochromocytoma. Histopathology diagnosis of right adrenal myelolipoma was confirmed on the basis of these findings.

On post-operative follow-up, her blood pressure on medication was under control (130/80 mm of Hg) and her 24-hour urine VMA was 1.71 (within normal reference range).

Discussion

Myelolipoma of the adrenal gland is a rare benign, usually unilateral tumor. It is generally seen in the 5th to 7th decade of life and shows no gender predilection.² They are composed of mature adipocytes (fat) and hematopoietic precursors cells (megakaryocytes, myeloid



Fig 1: Photograph of gross specimen of laparoscopically removed right adrenal mass.

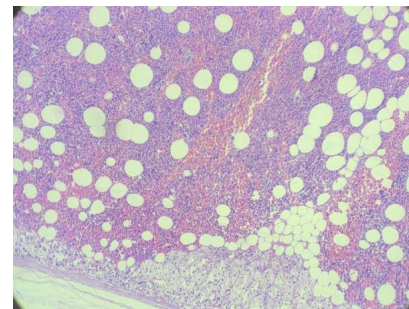


Fig 2: Photomicrograph showing adrenal cortical capsule with zona fasciculata, mature fat and normal haematopoietic elements in section from adrenal mass (H and E stain, x100)

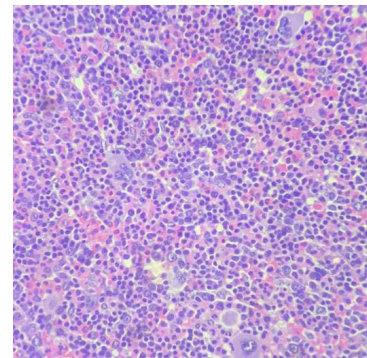


Fig 3: Photomicrograph showing normal haematopoietic elements in the section from adrenal mass (H and E stain, x 400)

and erythroid cells) and are thought to arise as a result of metaplastic change in reticuloendothelial cells in response to stress like infection or necrosis or endocrine stimulus.³

It is usually an incidental finding on ultrasonography or computed tomography (CT) of abdomen or on autopsy as most of them are non functional and cause no hematological disorder. Their size is generally less than 5 cm in greatest dimension and scan is most sensitive in

diagnosing them.⁴ It shows a well-encapsulated adrenal mass of low density with negative attenuation values with or without calcification and hemorrhage.^{5,6} In our study, on CT, the diagnosis was adrenal myelolipoma which was further affirmed by normal 24-hour urine VMA test. Histopathology showed no microscopic features of pheochromocytoma (Zellballen pattern) or adrenal hyperplasia. In cases with diagnostic challenges, immunohistochemistry (IHC) for chromogranin would help in confirming a catecholamine-secreting adrenal myelolipoma. Other differential diagnoses for adrenal mass can be angiomyolipoma and lipoma or liposarcoma because of its location and fat content. In angiomyolipoma, which is again a benign tumor, there is the characteristic microscopic finding of spindle cells, thick-walled hyalinized blood vessels with fat and maybe an epithelioid component (in epithelioid variant). Our case had very low vascularity on imaging, which did not favor angiomyolipoma. Only mature fat or lipoblasts with atypia are seen in lipoma or liposarcoma. On follow-up, the patient's blood pressure was under control on medications and her post-operative 24 hrs urine VMA was also within normal reference range.

The literature review showed four reported cases of functional adrenal myelolipoma. Tamidari et al and Jakka et al have separately reported case of right adrenal catecholamine-secreting myelolipoma with increased 24-hour urinary metanephrines.^{7,1} Udupa *et al.* have reported a large myelolipoma with increased 24-hour VMA levels.⁸ Brogna *et al.* reported a giant cortisol-secreting adrenal myelolipoma.³ In all these patients, blood pressure and biochemical abnormalities normalized on follow-up after surgery. Jakka et al performed IHC on their case which was positive for chromogranin A.

This case emphasizes the diagnostic dilemma faced when there is a suprarenal mass with hypertension and confirms the diagnostic utility of preoperative urine

VMA. Hypertension in our patient could have been an associated disease not related to the suprarenal mass.

Learning points

Myelolipomas are rare and usually non-functional but rare case reports of functional myelolipomas have been seen. Good evaluation and follow-up, sometimes with the help of immunohistochemistry, might help in giving appropriate adequate treatment to these patients.

Conflict of interest

None

Source of Funding

None

Ethical Approval

All procedures were in accordance with the ethical standards at the authors' institution and with the 1964 Helsinki declaration and its later amendments or comparable ethical standards.

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