The clinical conundrum of a catecholamine secreting giant adrenal myelolipoma

Tarun Jindal¹, Satyadip Mukherjee¹, Rajan Koju¹, Sandip Giri²

¹Department of Uro-Oncology, Tata Medical Center, Kolkata, West Bengal, India, ²Department of Radiology, Tata Medical Center, Kolkata, West Bengal, India

Abstract

Adrenal myelolipomas are uncommon tumours of unknown aetiology. They arise from the adrenal cortex and comprise lipomatous and myeloid elements. They are considered to be functionally inert, and metabolic evaluation is not mandatory for them. Adrenal myelolipomas can rarely be functionally active, and patients may present with hypertension, electrolyte imbalance or features of Cushing’s syndrome. The association of these tumours with catecholamine secretion is exceptionally rare. We describe a case of a functional adrenal myelolipoma associated with catecholamine secretion in a 55-year-old female patient with a history of hypertension. The surgical excision of the mass resulted in normalisation of the urinary catecholamine levels and resolution of the hypertension.

Keywords: Adrenal, catecholamine, myelolipoma

INTRODUCTION

Adrenal myelolipomas are tumours of unknown aetiology. These tumours do not cause any specific symptoms and are often diagnosed incidentally.[1,2] They are considered to be benign and functionally inert; hence, surgical intervention is reserved for patients in whom they attain a substantial size considering the risk of rupture and retroperitoneal haemorrhage. These tumours can rarely be functional, leading to endocrinopathies.[1,2] We describe an extremely unusual case of a large adrenal myelolipoma associated with catecholamine secretion.

CASE REPORT

A 55-year-old female presented to us with a history of heaviness in the left flank. She was a known hypertensive, well controlled for the last 2 years on a combination of telmisartan and hydrochlorothiazide. There was no history of palpitations, headache or excessive sweating. The past, personal and family history were non-contributory. The routine blood investigations including haemogram, serum electrolytes and renal function tests were within normal limits. The electrocardiogram showed a left-axis deviation. Ultrasound examination revealed a large left suprarenal mass. Contrast-enhanced computed tomography (CT) scan was performed which revealed a 10 cm × 8 cm × 6 cm supra-renal hypodense mass with macroscopic fat, suggestive of adrenal myelolipoma [Figure 1a and b]. The contralateral adrenal was normal, and there were no other lesions demonstrable on the CT scan. Endocrine consultation was obtained as a part of the institutional protocol, and functional assessment was performed. The
24-h urinary metanephrines were elevated to 989 µg/day (normal range 74–297 µg/day), while the cortisol levels were normal. Ga-68 DOTATATE positron emission tomography/CT scan did not show any abnormal uptake. A clinical diagnosis of functional, catecholamine-secreting, adrenal mass was considered. Her antihypertensive medications were changed to prazosin (alpha-blocker) followed by metoprolol (beta-blocker). A laparoscopic transperitoneal left adrenalectomy was performed. The descending colon was mobilised, and the left renal vein was identified. The adrenal vein was clipped and divided [Figure 1c]. The mass was separated from the upper pole of the kidney and its lateral attachments using blunt and sharp dissection. The mass was bagged and delivered through a muscle cutting incision in the left iliac fossa. There were no significant blood pressure fluctuations intra-operatively. On gross examination, the mass was well encapsulated and measured 10.5 cm × 8 cm × 6 cm [Figure 1d]. The cut surface was yellowish in colour with areas of haemorrhage. The microscopic examination revealed mature adipose tissue with haematopoietic tissue, suggestive of myelolipoma. A small rim of normal adrenal tissue was also seen. There was no histological evidence of pheochromocytoma or adrenal hyperplasia. The patient made an uneventful recovery and could be discharged on the 3rd post-operative day. The blood pressure normalised and antihypertensive medications were stopped after surgery. The patient has completed 18 months of follow-up and is doing well. The 24-h urinary metanephrines, repeated 1 month after surgery, were normal.

**DISCUSSION**

Adrenal myelolipomas are uncommon, often detected incidentally. They originate from the adrenal cortex and comprise lipomatous and haematopoietic elements. The exact aetiology is unknown, but it has been postulated that the lipomatous elements originate from the fat containing mesenchymal stromal cells of the adrenal cortex while the haematopoietic elements are derived from the reticuloendothelial cells of the blood capillaries. They are more common on the right side, and the patients tend to be in their fifth decade at the time of diagnosis.\(^1-3^\) Cross-sectional imaging by a CT or magnetic resonance imaging can confirm the diagnosis due to the presence of macroscopic and microscopic fat. These tumours are

![Figure 1](image-url)
considered to be non-secretory, and functional evaluation was not considered to be mandatory during their work-up.\textsuperscript{[1-3]} This recommendation has however been questioned. In a review of the literature, it was observed that nearly 7% of the adrenal myelolipomas may be functionally active.\textsuperscript{[2]} The majority of these ‘functional’ myelolipomas were associated with hypercortisolism followed by hyperaldosteronism. To the best of our knowledge, only six cases of adrenal myelolipoma have been reported which were associated with catecholamine secretion.\textsuperscript{[2-6]} It is interesting to note that all of these patients had giant adrenal masses (≥10 cm), similar to the present case. Surgical excision resulted in normalisation of blood pressure and urinary metanephrines in all these cases.\textsuperscript{[2,4-5]} It is also noteworthy that ours is the first case of a catecholamine-secreting giant myelolipoma which could be laparoscopically excised with excellent outcome.

**Declaration of patient consent**

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given her consent for her images and other clinical information to be reported in the journal. The patient understand that name and initials will not be published and due efforts will be made to conceal identity, but anonymity cannot be guaranteed.

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**Conflicts of interest**

There are no conflicts of interest.

**REFERENCES**

1. Decmann A, Perge P, Tóth M, Igaz P. Adrenal myelolipoma: A comprehensive review. Endocrine 2018;59:7-15.
2. Shenoy VG, Thota A, Shankar R, Desai MG. Adrenal myelolipoma: Controversies in its management. Indian J Urol 2015;31:94-101.
3. Vaidya A, Hamrahian A, Bancos I, Fleseriu M, Ghayee HK. The evaluation of incidentally discovered adrenal masses. Endocr Pract 2019;25:178-92.
4. Udupa S, Usha M, Visweswara RN, Desai MG. Left-sided giant adrenal myelolipoma secreting catecholamine. Indian J Pathol Microbiol 2012;55:389-91.
5. Jakka N, Venkateshwarlu J, Saryavani N, Neelaveni K, Ramesh J. Functioning adrenal myelolipoma: A rare cause of hypertension. Indian J Endocrinol Metab 2013;17:S249-51.
6. Adapa S, Naramala S, Gayam V, Gavini F, Dhingra H, Hazard FK, et al. Adrenal incidentaloma: Challenges in diagnosing adrenal myelolipoma. J Investig Med High Impact Case Rep 2019;7:2324709619870311.