

# Giant Bilateral Adrenal Myelolipoma: Case Report

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## Abstract

Adrenal myelolipomas are nonfunctional tumors that are usually asymptomatic; however, they have been known to coexist with other endocrine disorders, such as Cushing's syndrome, congenital adrenal hyperplasia (CAH), Conn's syndrome, and pheochromocytoma. We report a case of a 49-year-old man with hypertension and diabetes mellitus who complained of chronic abdominal pain, vomiting, and early satiety. Preoperative contrast-enhanced computerized tomography (CECT) was performed, and adrenal myelolipoma was considered, lab investigations revealed a nonfunctional tumor. CECT also revealed bilateral renal cortical cyst, right renal calculi, and hepatic cyst. A left open cortical sparing adrenalectomy was performed, pathological examination confirmed the diagnosis, and a radiological surveillance was planned for the right tumor. Four years following this, the patient came back with a similar presentation. Right adrenalectomy was performed after preoperative workup, and subsequently steroid replacement therapy was initiated. We suggest adequate follow-up of a patient presenting with adrenal myelolipoma and to explore the possibility of establishing a syndromic diagnosis such as autosomal dominant polycystic kidney disease (ADPKD).

**Keywords:** ADPKD, adrenal myelolipoma, adrenalectomy, bilateral adrenal tumor, hepatic cyst, hypertension, renal calculi, renal cyst, Von Hippel–Lindau syndrome

## INTRODUCTION

Adrenal myelolipoma is an uncommon entity, which has a relatively increasing incidence due to the widespread use of imaging modalities such as computed tomography (CT) and magnetic resonance imaging (MRI) scans. Despite its benign biology, when presented as a giant tumor, the surgeon can face diagnostic dilemmas. We describe a case of a bilateral giant adrenal myelolipoma, a unique case compared to those reported in the literature courtesy to the associated features, and discuss issues concerning the diagnosis and management.<sup>[1-3]</sup>

## CASE REPORT

A 49-year-old man, hypertensive with type 2 diabetes, came with a complaint of diffuse insidious, sporadic, abdominal pain associated with recurrent vomiting and early satiety for 3 months, with inconclusive family history. General examination was unremarkable. On abdominal examination, there were palpable bilateral retroperitoneal masses. Contrast-enhanced computerized tomography (CECT) abdomen [Figure 1] showed retroperitoneal heterogeneously enhancing lesion with hemorrhagic foci in the suprarenal

region with a predominance of fatty tissue with -26 to 17 HU density, measuring 30 × 18 × 13 cm in the left suprarenal region and 8 × 5 × 6.7 cm in the right suprarenal region. The differential diagnoses considered were adrenal myelolipoma, exophytic angiomyolipoma, and retroperitoneal sarcoma.

Other findings were bilateral renal cortical cysts (Bosniak type 3) and hepatic cyst in the IVa Couinaud segment. There were no abnormalities in laboratory tests, as well as in the hormonal investigations (24 h urinary metanephrines and creatinine to exclude pheochromocytoma, 1 mg overnight dexamethasone test to exclude Cushing's syndrome, and plasma aldosterone renin ratio to exclude Conn's Syndrome as advised by the endocrinologist.) for adrenal masses (nonfunctional tumor). In view of the large size of the tumor and the compressive symptoms, the patient was advised for left

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**Figure 1:** Left sided lesion at first presentation

adrenalectomy. After informed consent, the patient was taken up for left cortical sparing adrenalectomy under general anesthesia. Postoperative period was uneventful. Histopathology report of the adrenalectomy specimen showed myelolipoma, and the patient was kept under regular follow-up. The patient was asked to follow up with annual CT imaging. He was advised of the possible need for right adrenalectomy in case of persistent symptoms, new onset of symptoms, or significant change in imaging or laboratory investigations. Three years later the patient presented with right-sided loin pain for 15 days for which a CT scan of abdomen was performed, which showed bilateral nephrolithiasis with no increase in the size of the right adrenal tumor. The patient underwent right double J (DJ) stenting under spinal anesthesia on 25/12/19. Six months later, the patient complained of dull aching pain in the right loin, for a period of 1 month. In view of the similar presentation and laboratory investigations like the left-sided lesion, he was advised right adrenalectomy after necessary clearances were obtained from the relevant departments.

Preoperative urology consultation was taken for intraoperative assistance for right renal calculi. After informed consent, the patient was taken up for right adrenalectomy with right nephrolithotomy with adhesiolysis under general anesthesia on 20/6/2020. Postoperatively, the patient was shifted to the Intensive Treatment Unit. Steroid replacement was started as advised by the endocrinologist. The patient improved symptomatically. Histopathology report of the resected tumor showed myelolipoma.

The steroid dose was reduced and on postoperative day 9, the patient was switched to oral steroid replacement. The patient was discharged on hydrocortisone and fludrocortisone supplement and was asked to follow up regularly with the endocrinology department to consider tapering the steroid dose.

## DISCUSSION

This case has an unusual presentation as it is not just an isolated bilateral adrenal myelolipoma, but associated

with other conditions such as bilateral renal cysts, with right-sided renal calculi, hepatic cysts, and that the patient is hypertensive on medication, though the hypertension having been diagnosed much before the tumor was diagnosed. The patient's tumor fell under the category of nonfunctional adrenal tumor as was proved by the laboratory investigations. This patient's clinical signs, symptoms, CT findings, and associated features warranted concern for a syndromic diagnosis such as a possible association with ADPKD or as a coexistent pathology, which would explain the condition. Keeping that in mind, a PubMed search with the words "adrenal gland," "ADPKD," "hepatic cysts," and "renal cysts" was carried out. Gejyo *et al.* described a case of a 42-year-old woman with autosomal dominant polycystic kidney disease complicated by adrenal malfunction (primary aldosteronism).<sup>[4]</sup> Bilateral myelolipoma mostly was associated with congenital adrenal hyperplasia (CAH) or Cushing's disease,<sup>[5,6]</sup> whose pathogenesis could be explained by chronic *adrenocorticotrophic hormone* (ACTH) stimulation.<sup>[7]</sup> Non-functional, bilateral tumors such as this one, with the associated conditions, to best of our knowledge has not been described up until now.

## CONCLUSION

The adrenal myelolipoma is a rare pathological entity, its bilateral nature may be associated with other conditions such as Cushing's syndrome or CAH. In this case, a large bilateral adrenal myelolipoma is described, which had other associated features with a possibility of association with ADPKD. Although the myelolipoma being large in size, it is not something to be worried of except for the rare complication of rupture, and it is rather slow in its disease progression. Surgical management was advised as the condition was causing mass effect and compressing the surrounding structures. However, even with the contemporary imaging modalities, precise diagnosis may be difficult, a CT scan is very helpful as a primary imaging modality. We advocate regular monitoring of the tumor size and symptomatology before going ahead with the surgical resection of the tumor. We also recommend to look for associated features in case a diagnosis of an adrenal myelolipoma is made and to elicit a family history to increase the chances of identifying its possible coexistence with other syndromes. In the setting of the surgical treatment, it is important to preserve in some way the hormonal function by cortical sparing adrenalectomy if possible.

## Declaration of patient consent

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

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### Conflicts of interest

There are no conflicts of interest.

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