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## Clinical presentation and surgical resolution of atypical non-adenomas adrenal tumors

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**Abstract**

Adrenal tumors are very common in medical practice, including a wide spectrum of etiologies. Although most of them are adenomas, infrequent lesions can become a diagnostic and therapeutic challenge for clinicians. Here we present two atypical cases of non-adenomas adrenal tumors with specific imaging, biochemical profile, and surgical resolution.

The first case is a 54-year-old woman with a history of hypertension and type 2 diabetes, with an incidentally discovered 7 cm adrenal mass on CT, compatible with a myelolipoma. Biochemical work-up was remarkable for hypersecretion of both cortisol and aldosterone. She underwent laparoscopic adrenalectomy. During follow-up, biochemical cure of hormonal hypersecretion was confirmed.

The second case is a 44-year-old woman in whom a 7 cm left non-adenoma adrenal mass was diagnosed incidentally by magnetic resonance. Complete hormonal screening was normal. Laparoscopic adrenalectomy was performed. Biopsy informed a solid neoplasm separated from normal adrenal tissue, with immunohistochemistry compatible with a Schwannoma.

These case vignettes describe atypical adrenal cases that should be considered in the differential diagnosis of non-adenomas adrenal tumors. They represent uncommon presentations and illustrate the importance of an appropriate multidisciplinary evaluation of adrenal tumors, in order to improve the surgical and clinical management of these patients.

**Keywords:** adrenal incidentaloma; adrenal tumor; adrenal myelolipoma; adrenal schwannoma.

**Case report**

Adrenal tumors are very common in medical practice since abdominal imaging is more commonly performed [1]. Most of these tumors are apparently “non-functioning” adenomas, but less common lesions can be found which can generate difficulties among healthcare professionals [2]. Hence, clinicians need to be aware of these infrequent entities to determine an accurate diagnosis, hormonal profiling, and proper management.

**Case I**

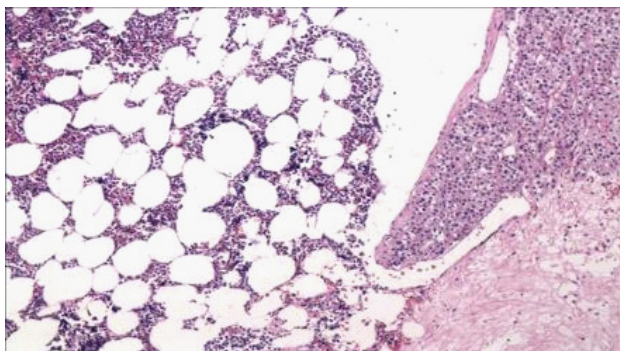
A 54-year-old woman with a history of arterial hypertension and Type 2 Diabetes Mellitus (T2DM). Abdominal ultrasound showed a right adrenal mass with solid and cystic components. A Computed Tomography (CT) was performed (Figure 1) showing a 7.3 X 5.5 cm very low-density lesion consistent with adrenal myelolipoma. Pre-operative blood tests were requested and impressively displayed mild hypersecretion of both cortisol and aldosterone (Table 1). Laparoscopic right adrenalectomy was performed.

Twenty-four hours after surgery, blood tests revealed hypocortisolism, which was successfully supplemented, confirming mild Autonomous Cortisol Secretion (ACS). The patient did not require antihypertensive therapy after surgery and biochemical cure with normalization of renin values was documented.



**Figure 1:** CT of adrenal myelolipoma.

Biopsy (Figure 2) showed an ovoid formation of 7.5 X 6.5 X 3.3 cm, compatible with adrenal co-secreting myelolipoma (mild ACS and mild primary aldosteronism).



**Figure 2:** Biopsy of Adrenal Myelolipoma. Adrenal tumor zone; neoplasm composed of discohesive cells interspersed with mature adipose tissue is recognized on the left of the image. On the right, separated by a thin fibrous septum, preserved adrenal cortical tissue with transition towards hyaline areas of the tumor (hematoxylin and eosin, x250).

**Case II**

A 44-year-old woman with a four-month history of low back pain, without any other symptoms nor any findings in physical examination. Spinal Magnetic Resonance Imaging (MRI) showed an adrenal mass and an abdominal CT scan confirmed the presence of a homogeneous, non-adenomatous, juxta-adrenal 7 cm mass (Figures 3 and 4). No hypersecretion was observed in the screening tests (Table 1).

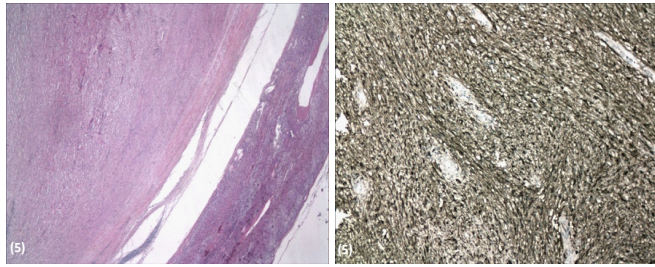
**Table 1:** CT scan showing epidural hematoma with associated diffuse subarachnoid hemorrhage.

Case 1- Test (normal value)	Pre-surgery	Post-surgery
Aldosterone (1.8-23.2 ng/dL)	9.35	9.93
Plasma renin activity (1.3-4 ng/ml/hr)	0.26	1.1
Aldo/PRA	35.9	9.02
Dexamethasone Supression Test (<1.8 ug/dL)	2.0	
Nocturnal salivary cortisol I y II (<0.1 ug/dL)	< 0.02	
Plasma cortisol (6.2-19.4 ug/dL)		5.3
ACTH (10-60 pg/mL)	14	72
Urinary metanephrine (52-341 ug/24 hrs)	35	
Urinary normetanephrine (88-444 ug/24 hrs)	144	
<b>Case 2</b>		
Aldosterone (1.8-23.2 ng/dL)	23	9
Plasma renin activity (1.3-4 ng/ml/hr)		6
Aldo/PRA	3	9
Urinary free cortisol (normal < 110 ug/24h)	41	2
DHEAS (< 420 ug/dL)		146
Urinary metanephrine (52-341 ug/24 hrs)	31	9
Urinary normetanephrine (88-444 ug/24 hrs)		242



**Figure 3 & 4:** CT of juxta-adrenal schwannoma.

Left laparoscopic adrenalectomy was performed without incidents. Biopsy (Figures 5 and 6) showed a solid neoplasm with Verocay bodies separated from adrenal tissue by a thin capsule. Immunohistochemistry stained positive for protein S-100 (+) and negative for H-caldesmon, CD34, and DOG-1, compatible with Schwannoma. The adjacent adrenal tissue was normal.



**Figure 5 & 6:** Biopsy of juxta-adrenal Schwannoma.

**Figure 5:** Tumor area (left of the image) that is seen separated by a thin fibrous capsule with respect to the adrenal tissue (on the right). The neoplasm shows solid pattern and multiple small vesels. (hematoxylin and eosin, x20).

**Figure 6:** Tumor zone with immunohistochemical staining against S-100 protein. Intense reactivity is recognized in all the cells that make up the neoplasm, dividing the intercrossed fascicles (x250).

## Discussion

Our case series describes two uncommon adrenal cases of non-adenomas adrenal tumors; a functional co-secreting myelolipoma and a yuxta-adrenal Schwannoma.

Adrenal myelolipomas are predominantly unilateral, formed by hematopoietic and adipose tissue with an incidence of 0.03% [3]. They are rarely functioning and, to our knowledge, co-secretion of aldosterone and cortisol has not been reported in the literature. These tumors are often found incidentally in imaging studies [4] and can co-exist with Cushing syndrome, pheochromocytoma, primary aldosteronism, and congenital adrenal hyperplasia. Bilateral [5] and hormone-secreting masses [6] are scarcely reported. One report of cortisol secreting myelolipoma and another with DHEA-S secretion have been published [6]. Regarding management, surgical removal is recommended in symptomatic adrenal myelolipomas, secreting lesions and in those larger than 6 cm because of hemorrhage risk. Laparoscopy is the preferred surgical approach. Most cases of small myelolipomas should be followed with imaging techniques.

Regarding Schwannomas, these tumors were first described in 1908 by Verocay. Conventional and cellular schwannomas exhibit strong and diffuse immunohistochemical positivity for S-100 protein. They are usually benign, non-secreting and asymptomatic tumors originating from nerve sheaths formed by Schwann cells. They have been scarcely reported as adrenal lesions [7]. Adrenal schwannoma on CT appears as a homogeneous, round, and well-circumscribed lesion, rarely may present with degenerative changes, such as calcification, hemorrhage, or cystic formation [8].

The differential diagnosis must be done with other non-adenomas tumors such as pheochromocytoma, adrenal carcinoma or adrenal sarcoma. Malignant transformation may be found in association with von Recklinghausen disease or neurofibromatosis [9]. CT image typically appears as a homogeneous lesion as our case; however, calcification, hemorrhage, or cystic formation could lead to heterogeneous enhancement [9]. Laparoscopy is the preferred surgical approach.

## Declarations

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## References

1. Barzon L, Sonino N, Fallo F, Palu G, Boscaro M. Prevalence and natural history of adrenal incidentalomas. *European Journal of Endocrinology*. 2003; 149: 273–285.
2. Fassnacht M, Arlt W, Bancos I, Dralle H, Newell, et al. Management of adrenal incidentalomas: European Society of Endocrinology Clinical Practice Guideline in collaboration with the European Network for the Study of Adrenal Tumors. *European Journal of Endocrinology*. 2016; 175: G1–G34.
3. Bishoff JT, Waguespack RL, Lynch SC, May DA, Poremba JA, Hall CR. Bilateral symptomatic adrenal myelolipoma. *J Urol*. 1997; 158: 1517-1518.
4. Hsu SW, Shu K, Lee WC, Cheng YT, Chiang PH. Adrenal myelolipoma: A 10-year single-center experience and literature review. *Kaohsiung J Med Sci*. 2012; 28: 377-382.
5. Zattoni D, Balzarotti R, Rosso R. The management of bilateral myelolipoma: Case report and review of the literature. *Int J Surg Case Rep*. 2015; 12: 31-36.
6. Brogna A, Scalisi G, Ferrara R, Bucceri AM. Giant secreting adrenal myelolipoma in a man: A case report. *J Med Case Rep*. 2011; 15: 298.
7. Limberg J, Stefanova D, Ullmann T, et al. Not all laparoscopic adrenalectomies are equal: Analysis of postoperative outcomes based on tumor functionality. *Surgical Endoscopy*. 2020.
8. Adas M, Ozulker F, Adas G, Koc B, Ozulker T, Sahin IM, et al. A rare adrenal incidentaloma: Adrenal schwannoma. *Case Rep Gastroenterol*. 2013; 7: 420-427.
9. Thomas MB, Stephen J. Adrenal Schwannoma: A rare adrenal incidentaloma. *International Surgery Journal*. 2018; 5.5: 1967-1969.