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Adrenal Myelolipoma Associated with Contralateral Secretory Adenoma: A Case Report

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Abstract **Case Report**

Adrenal myelolipomas are benign lipomatous tumors containing myeloid cell elements, most of which present as adrenal incidentalomas and account for 6% of all adrenal masses. Adrenal myelolipomas are usually unilateral in size, most often discovered in midlife, and affect both sexes almost equally. On imaging, adrenal myelolipomas have pathognomonic imaging features consistent with the presence of gross fat. Large adrenal myelolipomas may cause symptoms of mass effect and may occasionally be complicated by hemorrhage. If there is a concomitant adenoma or adrenal hyperplasia, excess adrenal hormones may be detected in patients with adrenal myelolipoma. We report the case of an adrenal myelolipoma associated with a contralateral secretory adenoma, responsible for Cushing syndrome in the patient.

Keywords: adrenal myelolipoma, Cushing syndrome, CT, adrenal hyperplasia.

INTRODUCTION

Adrenal myelolipoma is a benign, non-secreting tumor; most often discovered incidentally, it is the second most frequent incidentaloma after adrenal adenoma [1, 2].

It is a pathology that concerns subjects between the fifth and sixth decades. Its pathophysiology remains poorly understood, involving theories of cellular metaplasia under the influence of stimuli, notably hormonal [2].

Associations with endocrine pathologies are also reported, notably congenital adrenal hyperplasia [3].

The management is done on a case-by-case basis and consists of therapeutic abstention with simple radiological monitoring or elective surgical procedure [1, 2].

However, following a tumor rupture with intra-lesional or retroperitoneal hemorrhage, which is certainly a rare complication of adrenal myelolipoma, is nonetheless formidable, requiring rapid management to avoid hemorrhagic shock. Trans arterial embolization then seems to be of great interest [4].

We report the case of an adrenal myelolipoma associated with a contralateral secretory adenoma, responsible for Cushing syndrome in the patient.

CASE REPORT

This is a 60-year-old woman, without any particular pathological history, who presented to the emergency room for intermittent left flank and hypochondrium pain evolving for a few months with an acute recrudescence of her pain since 3 days.

On clinical examination the patient was conscious with a Glasgow score of 15/15, her blood pressure was elevated to 160/10 mm Hg, normocardial, eupneic, and her conjunctiva was normal colored.

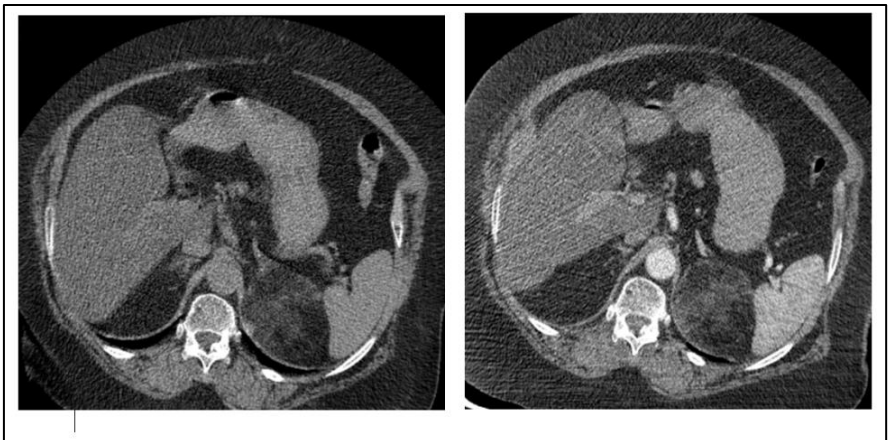
Facial and torso obesity was noted.

The biological workup showed hyperglycemia at 2.5 g/l.

An abdominopelvic CT scan was performed, showing a well-limited, rounded formation on the left adrenal gland, with regular contours, fatty density, and dense areas, enhancing after injection of contrast, measuring 7.7x6.7 cm, compatible with an adrenal myelolipoma.

A nodular lesion, well limited, with regular contours, dense, homogeneous, with an estimated spontaneous density of 14.5 HU with a relative washout measured at 50%, was associated with the diagnosis of adrenal adenoma.

The patient's workup was completed by hormonal measurements: cortisol, aldosteronemia, and renin levels returned in favor of hypercortisolemia, attesting to the secreting nature of the right adrenal adenoma.



Axial section CT images before and after contrast injection shows a rounded left adrenal lesion, well limited with regular contours, hypodense, the seat of large areas of fat density associated with dense areas enhancing after contrast injection, compatible with a left adrenal myelolipoma.

The patient was referred for a right adrenalectomy with simple surveillance of her left adrenal myelolipoma.

DISCUSSION

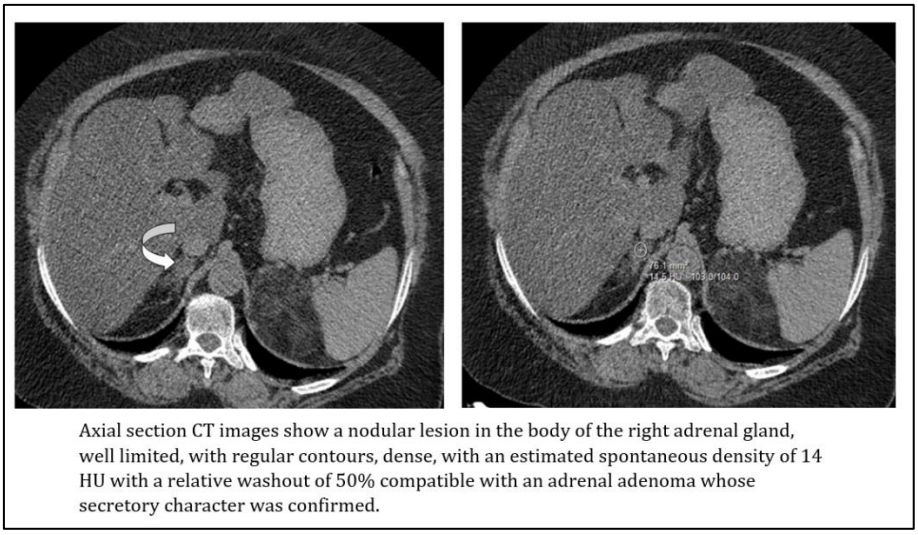
Adrenal myelolipoma is a benign, dual contingent tumor composed of adipose tissue and mature hematopoietic cells [1, 2].

These two contingents are present in variable proportions, hence the variability of the appearance of this tumor macroscopically and on imaging. Nevertheless, in its typical form myelolipoma has a predominant proportion of adipose tissue, which allows the diagnosis to be made radiologically [1].

This tumor is the second most frequent adrenal incidentaloma after adrenal adenoma and constitutes 2-4% of all adrenal tumors [4].

The average age of discovery of these tumors is 51 years with no predominance among one of the genders [5].

Often unilateral, with a predominance of the right side at 59.2%. Bilateral forms have been reported in 12% of cases, which should be investigated for an associated underlying pathology, in particular congenital adrenal hyperplasia [5].



Axial section CT images show a nodular lesion in the body of the right adrenal gland, well limited, with regular contours, dense, with an estimated spontaneous density of 14 HU with a relative washout of 50% compatible with an adrenal adenoma whose secretory character was confirmed.

The pathophysiology of this tumor is not yet clearly understood. Several hypotheses have been put forward.

The metaplasia of the reticuloendothelial cells is caused by various stimuli such as infection, necrosis, trauma, or oxidative stress [1, 2].

Exposure to high ACTH levels has also been implicated given the association of often bilateral and giant myelolipomas with congenital adrenal hyperplasia, mainly in its non-classical form given the delay in diagnosis in the face of frustrating clinical pictures that do not appear in the neonatal period but at an older age [3].

This has been supported by the development of extramedullary adrenal hematopoietic tissue in laboratory rats from which the pituitary gland has been removed and hormonal extracts from the anterior pituitary gland have been administered. On the other hand, the absence of overexpression of ACTH receptors on the surface of some giant myelolipomas challenges this theory [2].

Although myelolipoma is a non-secreting tumor, associations with secreting tumors, particularly adrenal adenomatous tissue, within the same entity responsible for Cushing's syndrome or Conn's syndrome have been reported [2].

Nevertheless, given the rarity of these associations, systematic hormonal assays are not indicated except for clinical signs [2].

We have not found any case report of concomitant myelolipoma and contralateral functional adrenal adenoma in the same patient.

Anatomically, and macroscopically, a myelolipoma presents as an oval or elliptical mass, more or less voluminous, ranging from a few millimeters to 43 cm with an average of 10.2 cm. It presents to describe a smooth, regular surface with a capsule or pseudo capsule [2].

A giant myelolipoma is defined by one of these diameters greater than 10 cm [2].

On cutting, yellowish portions can be distinguished, reflecting the adipose contingent of the tumor, and reddish-brown portions related to the hematopoietic contingent of the tumor [2].

Microscopically, we find adipose tissue as well as mature hemopoietic tissue similar to that of the bone marrow with the three cell lineages. Megakaryocytes, erythrocytes, and myeloid cells are found [2].

Immunohistochemical analysis is not routinely performed for the diagnosis of myelolipoma but may be used in cases of diagnostic doubt, especially in the case of well-differentiated retroperitoneal liposarcoma [1].

Myelolipoma is the second most common adrenal incidentaloma after adrenal adenoma, which is most often discovered incidentally during a CT scan for another reason. Nevertheless, patients may report compressive signs in the context of large tumors, which will be expressed by pain in the hypochondrium and or right flank, respiratory discomfort, or simple discomfort [5].

More rarely, the patient may report nausea or vomiting, indicating a possible tumor rupture, which may be spontaneous or secondary to even a minimal trauma [6-9].

The physical examination is most often normal, sometimes abdominal curvature may be noticeable.

In rarer cases (4.5%) complicated by tumor rupture leading to hemorrhage, most often intra-lesional or more rarely retroperitoneal, the patient presents with a hemorrhagic shock state with mucocutaneous pallor, hypotension, and tachycardia [10].

Complications related to myelolipoma occur when it is large (> 6 cm) due to its compressive nature, but also due to the risk of tumor rupture and therefore of hemorrhage. Hemorrhagic risk has been reported with myelolipomas of size ≥7cm [10].

The diagnosis of myelolipoma on imaging relies primarily on CT given the typical majority extracellular fat component characteristic of myelolipoma. In the adrenal cavity, a more or less large, rounded or oval, well-limited formation with regular contours, hypodense with negative densities between -20 and -120 HU, testifying to the fatty nature of the lesion, is found, with dense areas enhancing after contrast about the areas of hematopoietic tissue [4, 5, 10, 11].

In less typical cases, rare fatty islands can be identified. The diagnosis is then difficult to evoke radiologically and histopathological evidence is required. A fine needle biopsy with ultrasound or CT scan may be useful [11].

In the case of intra-lesional hemorrhagic changes, spontaneously dense or even hyperdense areas are found, with densities ranging from 50 to 90 HU, which is not enhanced after contrast [10].

Retroperitoneal hemorrhage will be seen as dense effusion shooting through the retroperitoneal

compartments, especially in the perirenal area, with possible hematoma formation. Extravasation of contrast is evidence of active bleeding.

Ultrasound is a first-line examination for flank or right hypochondrium pain and is not specific. A mass is found at the upper pole of the kidney with a heterogeneous echo structure, hyperechoic, with hypoechoic areas, with blurred contours, embedded in the surrounding retroperitoneal fat [2].

MRI shows a retroperitoneal mass, in contact with the upper pole of the kidney, well limited, with regular contours, T1 and T2 hyper signal, T1 hypo signal, and T2 intermediate signal, enhancing after injection of contrast in relation with the hematopoietic contingent of the tumor[5]. In case of hemorrhagic changes, the signal will vary, and there are generally areas of T1 hyper signal, and T2 hypo signal, not enhanced after contrast, which indicates the subacute nature of the bleeding [10].

The management of myelolipoma will depend on the complications or the risk of complications caused by the volume of the tumor and must be adapted on a case-by-case basis.

In the case of an asymptomatic tumor or a tumor smaller than 4 cm, therapeutic abstention is required [5].

Annual or biannual radiological follow-up over a period of 5 years is indicated by the American Association of endocrine surgery to evaluate the increase in tumor size [4].

If the tumor is symptomatic (signs of compression) or asymptomatic but greater than ≥ 7 cm, surgery is indicated due to the risk of rupture and hemorrhage [5, 10].

Currently, laparoscopy is indicated except in cases of giant myelolipoma greater than 10 cm where an open technique by retroperitoneal approach preferably given the lesser postoperative complications remains justified [5, 10].

Nevertheless, cases of giant myelolipomas successfully operated by laparoscopic approach have been reported [5].

In case of retroperitoneal hemorrhage secondary to tumor rupture with possible hemodynamic instability, endovascular treatment with transartériel catheterization for selective arterial embolization is required to stop the bleeding, stabilize the patient, and allow an elective surgical procedure to avoid the complications of emergency surgery [10].

Cases of successful embolization have been reported [4].

CONCLUSION

Adrenal myelolipoma is a benign tumor, most often discovered by chance or following compressive signs. A choice is made on a case-by-case basis, between therapeutic abstention and surgery.

More rarely, it may be complicated by intra-tumoral or retroperitoneal hemorrhage, usually secondary to spontaneous tumor rupture or, more unusually, post-traumatic hemorrhage, requiring rapid management, which ideally would involve embolization by selective endovascular arterial catheterization to overcome the emergency.

Myelolipoma is defined as a non-functioning tumor, although there are reports of hormonal disorders in myelolipomas associated with secretory adrenal adenomatous tissue coexisting within the same tumor. In this article, we report the case of a woman with an adrenal myelolipoma and a contralateral adrenal adenoma with clinical Cushing's syndrome.

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