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Case Report

Incidental Diagnosis of An Adrenal Myelolipoma

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Abstract

Introduction: *Adrenal myelolipoma is a rare benign tumor that develops of mature adipose and hematopoietic tissue in adreanl gland.*

Case report: *A 59-year-old male patient, referred for a right adrenal incidentaloma discovered during the workup for bilateral chronic kidney stones. After a negative secretory workup, the non-functional caracater of the mass was retained. The patient was operated on with a histological criterion of myelolipoma.*

Discussion and conclusion: *The prognosis for these tumors is good, due to its local scalability despite its challenging clinical and radiological diagnosis.*

Keywords: *Adrenal myelolipoma, benign, surgery, prognosis.*

INTRODUCTION:

Adrenal myelolipoma is a rare benign tumor that arises from the adrenal gland and containing a mixture of mature adipose and hematopoietic tissue (1). Usually small (<4 cm) and asymptomatic, adrenal myelolipoma can reach large size exceeding 10 cm named as such giant myelolipoma which is an indication for surgery (1). The largest myelolipoma reported in the literature reachted 31 cm and weighted 6 kg (2).

We present a case of an adrenal incidentaloma whose exploration revealed a myelolipoma, to illustrate clinical, radiological and therapeutic features.

CASE REPORT:

A 59-year-old male patient, diabetic on insulin and hypertensive on angiotensin converting enzyme inhibitor, follow-up for hyperuricemia on allopurinol, complicated by uretic bilateral renal calculi, having benefited from a percutaneous nephrolithotomy with right double J probe mounting, with family history of renal calculi and breast and liver neoplasia.

A Uroscan performed as part of the diagnostic work-up of his lithiasis pathology revealed a right adrenal incidentaloma at the expense of the external arm, roughly rounded and well-limited with a thin wall and triple component predominantly fatty, with a small tissue component and central organoid calcifications, measuring 7*5.3 cm. It contacts the liver anteriorly and the right diaphragmatic pillar posteriorly, without infiltrating them (Figure 1).

On admission, the patient reported intermittent right low back pain. He was stable, blood pressure (BP) at 120/70 mmHg, Heart Rate (HR) at 82 bpm, Respiratory Rate (RR) at 18 cpm, in moderate obesity with BMI at 34,5 kg/m2, an acanthosis nigricans with abdominal obesity but without purple stretch marks. The abdomen was soft without sensibility or palpable mass.

Biology: Blood count: White blood cells at 7640/mm3, normocytic normochromic anemia with 12 g/dl of hemoglobin, natremia at 137 mmol/l, kalaemia at 4.4 mmol/l, creatinine clearance at 21 ml/mn/1.73, corrected calcemia at 88 mg/l. **Hormonal secretion tests:** measuring 24-hour urinary methoxylates derivatives (UMD) and urinary free cortisol (UFC), both of which returned normal. Minute braking test with dexamethasone 1 mg was negatif at 2,8 ug/dl.

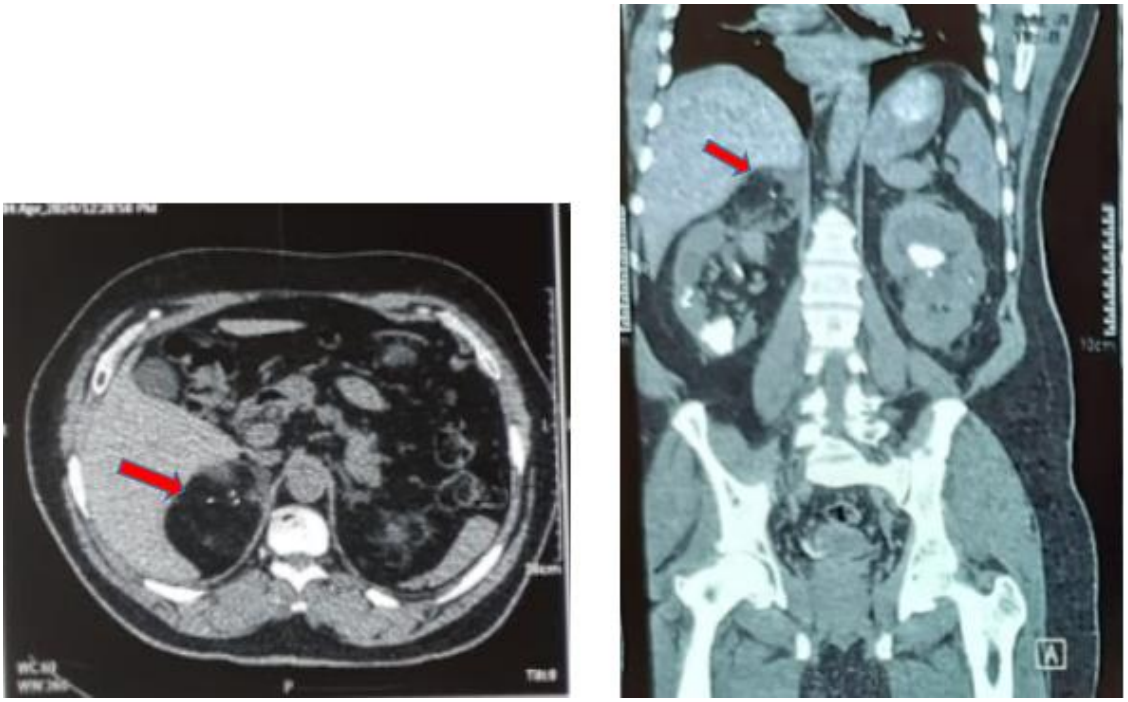

Figure 1: Uroscan: Right adrenal mass (Red arrow).

Given this clinical, biological and radiological finding, the patient underwent right adrenalectomy. Macroscopic study revealed a block weighing 74,5 g, measuring 7,2*5*3 cm. On section, a well-limited, encapsulated neoplasm measuring 5*4.5*4 cm, buff-yellow in color, firm in consistency, with hemorrhagic changes.

Histological examination revealed a benign tumor proliferation, consisting of lobules of regular adipocytes associated with a hematopoietic tissue of granulocytes, erythroblasts and megakaryocytes, with fibrous interstitial tissue, vascular congestion, polymorphous inflammatory infiltrate and some calcifications. The diagnosis of Adrenal Myelolipoma was retained.

DISCUSSION:

Adrenal myelolipoma is a rare tumor with an incidence ranging from 2 to 4%. The first case of adrenal myelolipoma was reported in the literature by Gierke in 1905. This tumor owes its name of Myelolipoma to the researcher Oberling, who described a proliferation associating a mixture of adipose and hematopoietic tissues (3). Adrenal myelolipoma usually occurs between the ages of 50 and 70 without a predominance in either sex (4).

Several theories have been proposed to explain the etiopathogenesis of this tumor; the most widely credited of which is the existence of factors such as chronic stress, infection, necrosis or prolonged exposure to ACTH (5,6). The main theory explaining its tumorigenesis: the fatty part derives from mesenchymal stem cells of the stromal fat of the adrenal cortex under certain stimuli. Once the adipocytes have matured, they accumulate and become inflammatory, stimulating the surrounding adrenal cortical tissue. At the same time, the hematopoietic cells in the central part of the fatty tissue dedifferentiate and divide, giving this type of tumor its fatty and hematopoietic nature (7).

Other myelolipoma's locations have been reported in the literature, including splenic, pulmonary, hepatic and mediastinal (8).

Several studies have reported benign adrenal tumors as factors favoring type 2 diabetes and hypertension. At the same time, screening for a background of mild autonomous cortisol secretion (MACS), which represents an additional cardiovascular risk factor is needed (9).

Our patient had 59 years old at the time of diagnosis of this benign tumor, diabetic and hypertensive with MACS retained for minute braking at 2.8 ug/dl. These data are consistent with those in the literature.
Adrenal myelolipomas are usually non-secreting and pauci-sympotomatic. In most cases, the main symptom is a moderate abdominal pain and discomfort, which was the case for our patient. The majority of adreanl myelolipomas

reported in the literature were diagnosed in the setting of adrenal incidentalomas with imaging techniques performed on non-adrenal motifs.

Morphological diagnosis is based mainly on enhanced CT scan, ultrasound and, to a lesser extent MRI (10).

The maximum size of these tumors reported in the literature varies between 10 and 30 cm (11). These considerable sizes can be explained by the often-non-functional nature of these tumors and the absence of any specific symptomatology. In our patient the size was 7,2*5*3 cm with nonspecific symptom and with an incidental detection.

In general, when it comes to adrenal tumours, CT and MRI are very useful for discussing differential diagnoses, but it is the histological study that provides etiological confirmation. In our case, at the clinico-biological stage, we didn't have enough evidence to suggest an etiology, which is why surgery was planned for both therapeutic and diagnostic purposes.

For Adrenal Myelolipoma the main treatment is surgery, especially when the tumour exceeds 7 cm. Others indications are presence of symptoms of a secreting tumour and/or a mass larger than 6 cm, and in the case of a mass less than 4 cm and/or non-functional requiring post-operative follow-up (12).

The surgical approach can be either laparoscopic or laparotomic, depending mainly on the size of the tumour and its anatomical relationship, especially with the large vessels (inferior vena cava, aorta...). In our case, surgery was performed laparoscopically given the size of the tumour, the absence of infiltration of neighbouring organs or large vessels.

The follow-up schedule for adrenal myelolipomas has not been codified; in general, authors report that abdominal ultrasound or CT scans are performed at 12 and 24 months, but there is no consistency (13).

CONCLUSION:

Adrenal myelolipoma is a rare benign tumour usually discovered as an incidentaloma. Despite its benign nature and characteristic fatty and hematopoietic composition, it poses many diagnostic difficulties, requiring discussion of other differential diagnoses of retroperitoneal masses, mainly pheochromocytoma, adrenocortical carcinoma and liposarcoma.

Declaration of interest:

The authors declare that they have no direct or indirect interest (financial or in kind) in any private, industrial or commercial organization related to the presented subject.

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