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Case report

Adrenal Myelolipoma Associated with Focal Segmental Glomerulosclerosis: A Case Report

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Abstract

Adrenal myelolipoma is a rare and usually slow-growing benign tumour. It is composed of hematopoietic and fatty tissue in different proportions, and it is frequently an asymptomatic finding during radiologic tests. We present the case of an unusual association, nephrotic syndrome secondary to focal segmental glomerulosclerosis with concomitant adrenal myelolipoma.

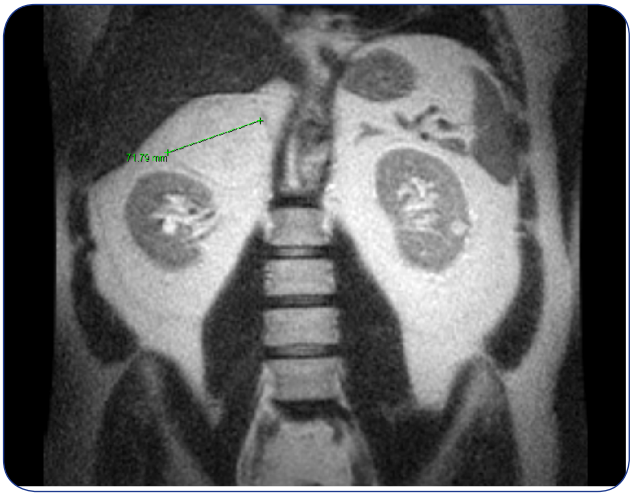
Keywords: Adrenal myelolipoma; Nephrotic syndrome; Focal segmental glomerulosclerosis

Case Report

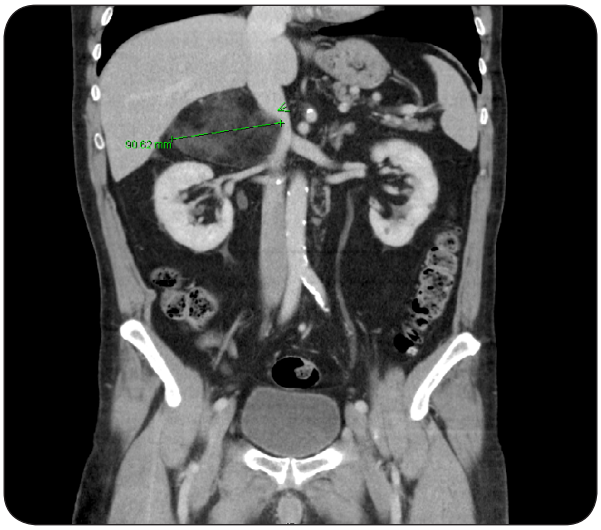
A 66-year-old male who was referred to our Department of Urology from the Nephrology Clinic for evaluation of an incidentally discovered right adrenal mass.

The patient presented the following medical history: non-insulin dependent diabetes mellitus, obesity grade II, Dyslipidemia and ischemic heart attack revascularized by triple aortocoronary bypass. His usual home treatment included Adiro, Atorvastatin, Enalapril, Atenolol, Metformin and Torasemide. Because of suboptimal control of blood pressure, the patient was referred to a nephrologist, who detected proteinuria (1.5-3 g/24h) and intermittent microhaematuria.

As part of his study, magnetic resonance imaging (MRI) revealed a right adrenal myelolipoma measuring 7 cm diameter, so that a functional study of the adrenal mass was performed at such time with negative results.



At that time it was decided an expectant attitude and periodic monitoring by Magnetic Resonance Imaging or Computed Tomography (CT). One year later and still the patient asymptomatic, CT revealed a lesion growth, reaching diameters of 9 x 8 cm and causing mass effect and compression of the inferior vena cava, so the patient was referred to us.



After preoperative evaluation which did not contraindicate surgery, laparoscopic right adrenalectomy and renal biopsy were performed. Postoperative recovery was favourable, being discharged on the third postoperative day.

Pathology showed:

- Focal segmental glomerulosclerosis

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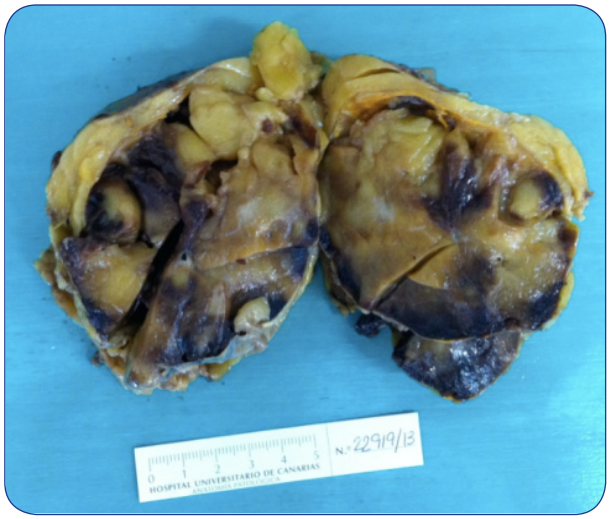
- Adrenal myelolipoma

Macroscopic Description

Adrenalectomy piece measuring 10 x 8.5 x 8.5 cm and weighing 366 grams. The cut is identified as a tumour, which displaces the glandular parenchyma to the periphery and is approximately 8 x 8 x 6.5 cm (T) When dissected this lesion has predominantly brown-yellow sectional planes of greasy interspersed with hematologic - coagulated areas and soft consistency, reaching 0.1 cm from the surface. (TS). No limphadenopathies.

Microscopic Description

Tumour lesion consists of mature adipose tissue and bone marrow hematopoietic elements. It is made of unilocular cells with optically empty lipid vacuoles which reject the periphery nuclei without atypia or mitosis; between these foci an adipocyte hematopoietic cellularity is discovered, including megakaryocytes and eritrocytic extravasation areas. Adrenal gland is compressed by this tumour, without significant histologic features.



The patient came to control visit one month after surgery. His blood and urine analysis showed stable glomerular filtration rate, proteinuria improvement (1 g/24h) and a better control in blood pressure (130-140/60-70 mmHg) Due to this good response to surgery, the patient was not considered candidate to immunosuppressive therapy. He should continue his regular treatment and be re-evaluated in three months.

Discussion

The adrenal myelolipoma is a rare, benign and slow growing tumour. Its finding in autopsy series ranges from 0.08-0.2 % [1].

First described by Gierke in 1905, was Oberlin who coined the term to describe these lesions. Histologically, myelolipoma is a tumour composed of mature adipose and hematopoietic tissue in different proportions, often with representation from all three hematopoietic series (red cells, white cells and megakaryocytes) [2].

Aetiology or risk factors are unknown, being the most accepted theory proposed by Meaglia and Schmidt in 1992, with a phenomenon of metaplasia of adrenocortical cells in response to stimuli such as infection or necrosis resulting from chronic diseases such as hypertension, arteriosclerosis or diabetes [3, 4, 5].

Experiments by Selye and Stone in rats, published in 1950, showed transforming reticular layer cells of the adrenal cortex in mature hematopoietic tissue and fat tissue, in response to a sustained stimulation with adenohypophysis extract injections and testosterone [6].

Other authors have described a chromosomal translocation (3; 21) (q25, p11) associated with adrenal myelolipoma, indicating that it could be a neoplasm arising from hematopoietic tissue [7]. The adrenal myelolipoma usually appears at middle age, between the ages of 40 and 70, with predilection for males (ratio male / female 1.75 / 1) [8].

Most cases are unilateral, with a slight predilection for the right adrenal gland. In a recent contribution of 6 cases and review of the literature, Sz- Wen found a total of 131 cases reported to date (2012), with 12% of cases including another contralateral adrenal tumour. These tumours are primarily found in the adrenal gland (85 %), although myelolipomas have also been described in the presacral region (50 % of extra-adrenal myelolipomas) , retroperitoneal , gastric , liver , spleen and lymph nodes [9].

Frequently, adrenal myelolipoma is asymptomatic and it is found as an incidental finding in abdominal imaging studies (ECO, CT, MRI) requested for another reason. In other cases, especially when they exceed 6-7 cm, can produce symptoms such as abdominal pain, haematuria, renovascular hypertension and even spontaneous retroperitoneal hemorrhage (Wunderlich Sd.), especially those bigger than 7 cm [10].

The association of adrenal myelolipoma with hypertension and obesity is common, although it is unknown whether there might be a pathogenic mechanism that relates it. Obesity could also be the link between adrenal myelolipoma and focal segmental glomerulosclerosis, as Alexopoulos et al. suggested it in 2003 [11]. To explain hypertension in these patients, some authors postulate a mechanism of renovascular hypertension by renal compression exerted by the tumour, while others see it as an incidental finding in obese and elderly. Its association with various endocrine diseases such as Cushing Sd., adrenal insufficiency or Conn Sd., has also been described, which has led some authors to include this tumour in an atypical variant of multiple endocrine neoplasia [12].

By definition, these tumours are considered to be non-secreting, since they are formed by fatty tissue and mature hematopoietic tissue that is not in contact with the sinusoids. However, there is a case described in the literature in which the tumour was associated with hypersecretion of catecholamines. After an open surgery, catecholamines in the patient's blood normalized without histologically features of the origin of catecholamines

hypersecretion with the study of the piece [13].

In the diagnosis, a study of adrenal secretion must be done in all patients, as well as a determination of catecholamines and metanephrines in blood and 24 hours-urine, basal cortisol suppression after 1 mg of dexamethasone, and kaliemia. As imaging test, Computed Tomography (CT) is considered to be the best choice for adrenal myelolipoma. Usually there is no problem for diagnosis using this test, since it is the only tumour of the adrenal gland with well defined encapsulation, containing a significant amount of adipose tissue, easily recognizable in CT because of its low attenuation coefficient (≤20 Hounsfield Units) [14].

MRI with fat suppression can also be used for a proper classification and delineation, as it is the best test to demonstrate the presence of adipose tissue. If despite imaging tests doubts persist, it should be indicated in a CT-guided needle aspiration, which is rarely performed unless a differential diagnosis with liposarcoma or metastasis is proposed. The key issue is previously to rule out the presence of a pheochromocytoma, as the puncture could trigger a severe hypertensive crisis.

Differential diagnosis of myelolipoma includes tumours with fat component (angiomyolipoma, lipoma and liposarcoma), adrenal carcinoma, pheochromocytoma and adrenal metastases. Usually, CT or MRI, based on the fat content of the mass together with its heterogeneity and the presence of regular and well-defined edges [15], easily diagnoses adrenal localization myelolipomas.

However, to diagnose retroperitoneal myelolipomas is much more difficult, given its similarity to teratomas and retroperitoneal liposarcomas. In these cases, obtaining tissue for histological examination is essential for surgical planning, because while myelolipomas and teratomas are usually well-defined tumours without invasion of neighbouring organs, retroperitoneal liposarcoma is a tumour that can be very aggressive (especially its pleomorphic variant) and it usually requires more extensive resections that in many cases should include adjacent organs (bowel, kidney...).

Myelolipoma treatment should be individualized, based on the characteristics of the tumour and the patient. Most authors advocate regularly perform imaging tests to asymptomatic myelolipomas under 6 cm [16, 17].

Surgical resection is recommended in those bigger than 6 cm, symptomatic or when malignancy is suspected. Laparoscopic approach in experienced centers should be the norm, as was in our case, being technically feasible and providing very favourable results from the oncological and functional standpoint [18].

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