Urology Case Reports 35 (2021) 101541

Contents lists available at ScienceDirect

Urology Case Reports

journal homepage: http://www.elsevier.com/locate/eucr

Functional medicine

Geant adrenal myelolipoma: A case report with literature review

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ARTICLE INFO

Keywords:
Retroperitoneal mass
Myelolipoma. adrenal gland. incidental

ABSTRACT

Adrenal myelolipoma is a rare, benign, non-functioning tumor, composed of mature adipose tissue and hematopoietic cells. We present the case of a 26-year-old woman who presented with abdominal distention and right back pain radiating to the right hypochondrium. Computed tomography of the abdomen revealed a large retroperitoneal mass, which was suspected to be a retroperitoneal liposarcoma. All hormonal studies related to adrenal gland were within normal limits. Open surgery resection was performed. Histopathology showed an adrenal myelolipoma with 39 × 21.5 × 8.5 cm and weighting 4930 g h. The patient evolved with an uneventful postoperative period.

Introduction

Adrenal myelolipoma is a rare, benign, non-functioning tumor of the adrenal gland. It was first described by Gierke in 1905 and named by oberlinng in 1929.[1]

This tumor is often asymptomatic, discovered incidentally and falls within the scope of adrenal incidentalomas. Histologically, it is characterized by the presence of mature adipocytes and normal hematopoietic tissue.[2]

Radiologically, adrenal myelolipomas may mimic other fat containing retroperitoneal lesions, like liposarcoma, teratoma, and extramedullary hematopoiesis.[3]

The therapeutic attitude depends on the size of the tumor, whether it is symptomatic or not and its possible complications. We report the case of a symptomatic large adrenal myelolipoma discovered in a 26-year-old woman who required surgical treatment and we discuss the diagnostic and therapeutic aspects.

Case presentation

A 26-year-old female patient presented with pain in the right upper abdomen for last one month. The pain was continuous, dull aching, gradual in onset, not associated with fever. There was no history of nausea, vomiting, fever, jaundice, hematemesis, or melena. On physical examination, a vague lump was evident in the right lumbar region, hard in consistency, immobile with a smooth surface. The abdominal CT scan revealed a large well-defined retroperitoneal tumor, which compressed and displaced the right kidney downward, the mass was heterogeneous with areas of soft-tissue attenuation (Fig. 1). The approximate dimensions of the lesion were 35 cm × 20 cm. Routine and the 24 hours urinary excretion of cortisol and VMA level were normal.

Based on the results of these imaging studies and the enormous size of the tumor, we made a preoperative diagnosis of a liposarcoma originating in the retroperitoneal space. The patient underwent surgical exploration through a midline incision. A large retroperitoneal mass was found to have a large well-encapsulated, did not invade contiguous structures and was completely resected. The renal vasculature was not compromised. The specimen size was 39 × 21.5 × 8.5 cm (Fig. 2) and weighed 4930 g. The patient has evolved satisfactorily. The histopathological diagnosis was an adrenal myelolipoma (Fig. 3).

Discussion

Adrenal myelolipoma was described for the first time in 1905 by Gierke, and owes its name to Oberling. This neoplasm is composed of variable proportions of adipose tissue and bone marrow represented by all three lineages of hematopoietic elements [1,2]. Clinically it can be found associated with Cushing's syndrome, hyperaldosteronism, or congenital adrenal hyperplasia.[4]

The incidence of these tumors ranges from 0.08 to 0.4%.[3] They are

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https://doi.org/10.1016/j.eucr.2020.101541
Received 24 November 2020; Received in revised form 13 December 2020; Accepted 15 December 2020
Available online 18 December 2020

usually discovered incidentally in autopsies, surgeries or in imaging studies (ultrasound or CT) performed for other reasons.[4,5]

They occur frequently between the fifth and seventh decade of life, without a predominance of sex,[5] mostly unilateral and rarely exceeds 4 cm. The right adrenal gland is more commonly involved than the left. However, very large and bilateral tumors occur in about 10% of cases.[3]

Spontaneous retroperitoneal hemorrhage is the most significant complication, especially with large myelolipomas. It can manifest with local pain in the back, epigastrium, or flanks, accompanied by nausea, vomiting, hypotension, and anemia. The most important differential diagnosis for myelolipomas is retroperitoneal (perirenal) well-differentiated liposarcoma.[3,5]

The Computed tomography is the most sensitive test because it can show an encapsulated adrenal mass with tissue density equivalent to fat. However, myelolipoma only shows this relatively characteristic aspect on CT if there is a large area of adipose tissue or if the fat cells and myeloid elements are distributed heterogeneously. The CT differential diagnosis is difficult when the mass is composed of myeloid tissue with low fat content. Distinguishing between myelolipoma and liposarcoma is impossible radiologically. To confirm the diagnosis of a myelolipoma, Some authors recommend CT-guided fine-needle aspiration biopsy.[3–5]

Treatment is generally defined by the size and symptoms of the tumor. For those greater than 7–10 cm, surgery is recommended due to its high risk of spontaneous rupture retroperitoneal hemorrhage. If there are symptoms (regardless of size), or there is no clear diagnosis, it must be surgically extracted, or at least a biopsy must be obtained to rule out malignancy, previously ruling out functionality, as with any other adrenal tumor.[3,4]

In case of bilateral myelolipoma, a staged tumor removal is preferable, removing the larger one and continuing to observe the contralateral myelolipoma as long as possible in an effort to avoid adrenal insufficiency and a lifetime of steroid replacement.[4]

Larger tumors require greater exposure, and it may be necessary to make large incisions such as Chevron (bilateral subcostal) or thoracoabdominal incisions as our case. Good exposure is crucial to avoid damaging the vena cava on the right or the aorta on the left, among other structures. The fact that myelolipomas are encapsulated allows them to be easily separated from the surrounding structures.[4,5]

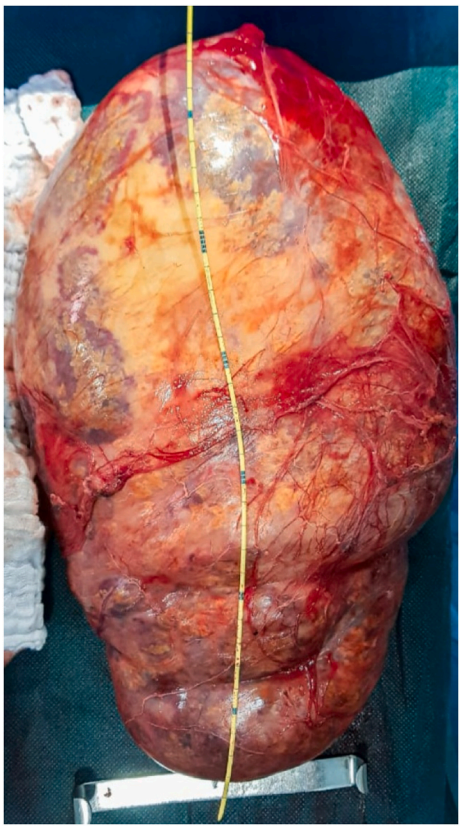

Fig. 2. Postoperative photo demonstrating the surgical specimen.

Conclusion

Adrenal myelolipomas are generally asymptomatic benign tumors whose size can vary from a few millimeters to tens of centimeters; they can compress neighboring organs, becoming symptomatic. In small tumors, medical and imaging control is recommended, ideally with CT to objectify stability or changes in size. If they are large, surgical removal is necessary to demonstrate benignity, prevent spontaneous hemorrhage,

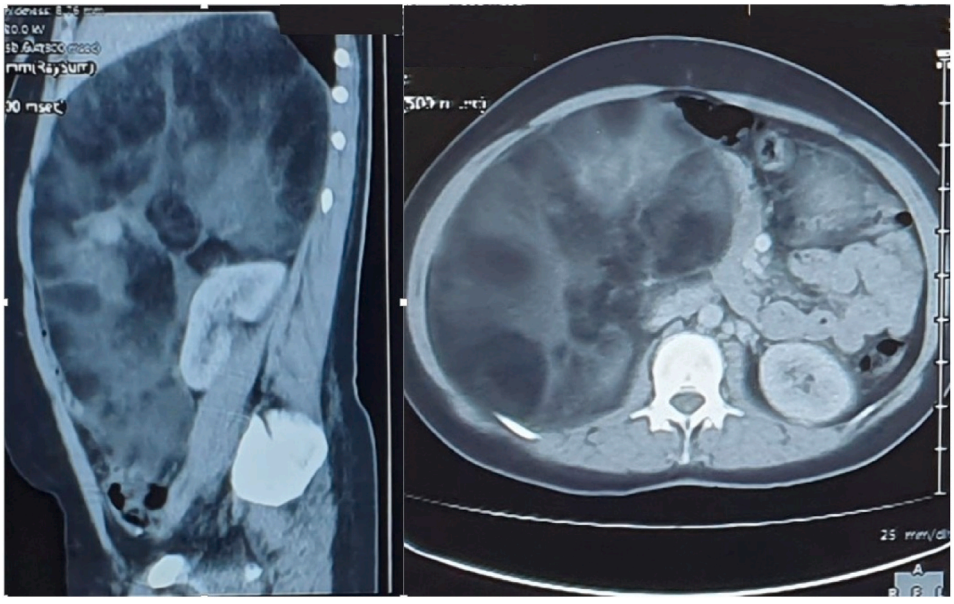

Fig. 1. Preoperative abdominal CT scan demonstrating an encapsulated, hypodense, right retroperitoneal mass, containing fat, interspersed soft tissue attenuation components and few septa.

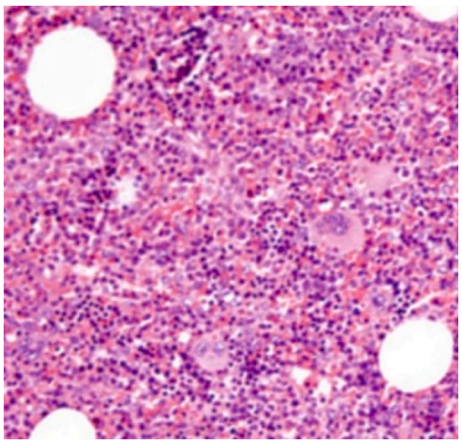

Fig. 3. Histopathologic picture of myelolipoma composed of mature fat cells mixed with hematopoietic elements.

and treat compression of neighboring structures.

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