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A hormone secreting adrenal myolipoma in an asymptomatic woman

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ABSTRACT

Adrenal myolipoma is a benign adrenal tumor which contains macroscopic amount of adipose tissue and usually hormonally inactive. However, functional adrenal myolipoma has also been reported in the literature.

In this article, we present an interesting case of hormone secreting adrenal myelolipoma in an asymptomatic pregnant woman.

1. Introduction

Adrenal myelolipoma (AML) is an uncommon benign neoplasm composed of mature adipose and myeloid cells which is usually asymptomatic and incidentally found on imaging by detecting macroscopic fat component in an adrenal mass. They account for 6–16 % of incidentalomas. However, they may produce symptoms due to the mass effect on adjacent structures, intratumoral hemorrhage or rupture. Endocrine dysfunction is rarely encountered in patients with AML and there are some reports of overproduction of adrenal hormones as glucocorticoids, aldosterone, androgens and catecholamines which first were misdiagnosed as secreting adrenal adenoma or pheochromocytoma.[1]

Here, we present a case of young woman referred for an incidentally found adrenal mass with abnormal high level of urine catecholamines who underwent surgery with impression of pheochromocytoma despite imaging findings suggestive for AML. The histopathology confirmed the diagnosis of AML.

2. Case presentation

A 30 year-old pregnant woman was referred to our clinic because of an adrenal mass incidentally found in her routine sonographic examination. The patient declared no related symptoms as abdominal discomfort, hypertension or hormonal disturbance. In clinical examination, blood pressure was normal and no abnormal clinical finding besides pregnancy related symptoms was observed. Hormonal assay was done and high level of catecholamines was detected that confirmed with repeated testing (normal level urine 24 h metanephrine = 33.7 and high level urine 24 h normetanephrine = 23,337). The patient was admitted with impression of adrenal pheochromocytoma. In the contrast-enhanced abdominal MRI, a large fat containing mass with mild heterogenous enhancement was reported at right adrenal gland measuring 170*120 mm suggestive for adrenal myolipoma (Fig. 1). During her administration, the patient was normotensive. The underwent abdominal surgery and the mass was resected. Following surgery, urine catecholamines returned to near normal level. (Urine 24 h normetanephrine = 610). Pathology confirmed the diagnosis of adrenal myolipoma (Figs. 2 and 3).

3. Discussion

Myelolipoma is a benign-bahaviouring neoplasm arising from adrenal cortex and was first described in 1905 by Gierke EGierke E(2). Adrenal myelolipoma (AML) is a slow growing tumors consists of adipose tissue and hematopoietic cells. The presumed pathophysiology attributed to the role of fetal adrenal glands in extramedullary hematopoiesis and stimulation of metaplasia in reticuloendothelial cell in adrenal cortex secondary to stress factors. Some studies emphasize the role of ACTH in developing AML. However, other studies have not found any association between the level of ACTH and increased risk of AML and this issue is still under debate.[2,3]

AML is most commonly asymptomatic and discovered incidentally in adults with a mean age of 55–65 years, equivocally in male and female. It accounts for the second most common adrenal incidentaloma with a prevalence of about 6–16 % of incidentalomas and 3·3–3·6 % of all adrenal tumors.[3] However, despite perpetually benign behavior, these neoplasms may present with endocriologic and non-endocrinologic

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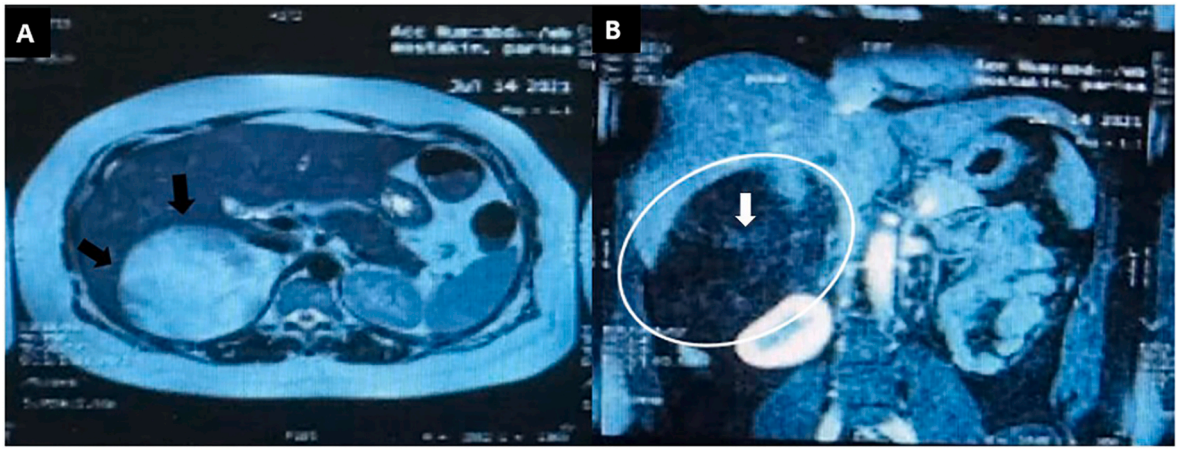

Fig. 1. In axial T2 weighted image(A) there is a large well-defined heterogenous hyperintense mass in right suprarenal region (black arrows). In coronal post-contrast fat sat image(B) the mass show dominant signal loss (circle) due to fat suppression with mild heterogenous enhancement (white arrow).

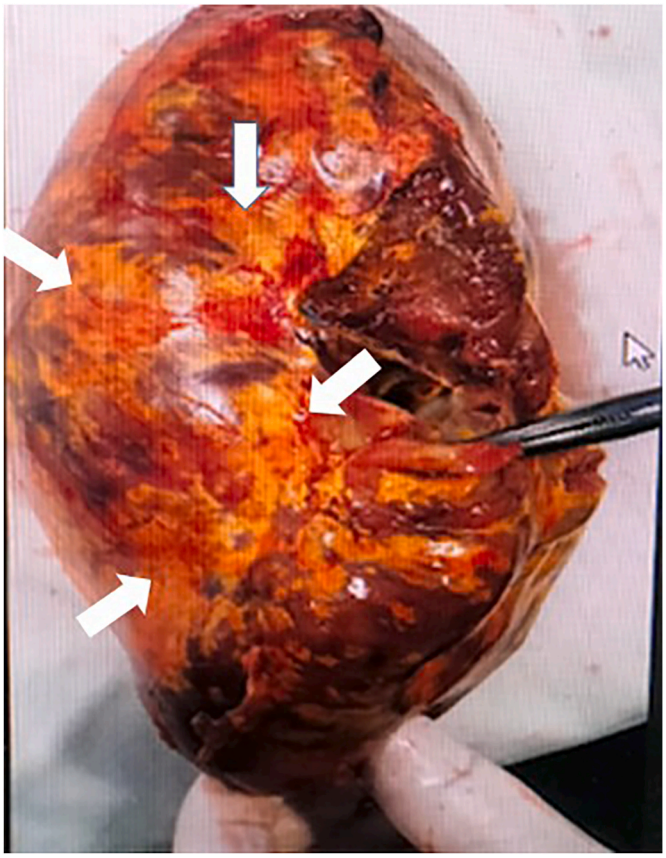

Fig. 2. Gross evaluation of the resected mass demonstrates intralesional yellow macroscopic fat (white arrows). (For interpretation of the references to colour in this figure legend, the reader is referred to the Web version of this article.)

symptoms. AML diagnosis is made on cross-sectional CT or MR imaging by detecting variable amount of macroscopic fat component in a well-defined adrenal mass. Macroscopic fat shows very low (−30, −100) Hounsfield units in CT and hyper-intensity in T1 and T2 weighted MR images which is suppressed in fat-sat image.[2,4]

Large AMLs, especially when giant (called when exceeding 10cm) can cause abdominal discomfort and flank pain due to mass effect. Rarely, intratumoral hemorrhage or tumoral rupture can also occur in large AMLs.

Patients with AML may present with endocrinologic manifestations especially hypertension and cushing disease symptoms. Endocrine dysfunction in patients with AML are encountered in different settings. Association of congenital adrenal hyperplasia (CAH) and AML has been described in different studies. The proposed ethiopathology for this association is the constant elevation of ACTH level in CAH patients which stimulate tumor growth. The incidence of CAH in patients with AML was estimated about 10 % in a literature review established by Decmann Á(1, 4). In addition, development of bilateral AML in patients with CAH can also emphasize the ACTH role in evolution of AMLs. Coincidence of other functioning adrenal tumors with AML has been reported as the other cause of endocrine disturbances in these patients. Secreting adrenal myelolipoma have also been diagnosed in 7 % of cases presenting with overproduction of adrenal hormones including aldosterone, glucocorticoids and rarely sex hormones and catecholamines.[5]

The pathophysiology of hormonal excess in patients with AML is not fully understood. However, some studies suggest irritation of normal adrenal parenchyma by the mass can be responsible for hormonal disturbance. There are just a few reports of catecholamine secreting AML presented with refractory hypertension and high level of 24h-urine VMA and the diagnosis were confirmed with imaging and histopathologic findings.[2,5] Regarding to the established guidelines, there is no need for metabolic evaluation and hormonal work up before surgery in the case of typical AML diagnosed with imaging modalities.[5]

4. Conclusion

In our case, the patient was asymptomatic despite striking high level of urine catecholamines which dropped to normal level after surgical resection suggestive for secreting AML. This study suggests that routine hormonal assay may be warranted in patients diagnosed with AML even asymptomatic.

Conflicts of interest/Competing interests

The authors declare no conflicts of interest.

Consent to participate & consent for publication

The patient in this case report signed informed consent. The context of the consent forms included that the patient's images and clinical information would be reported in a journal without mentioning his/her name.

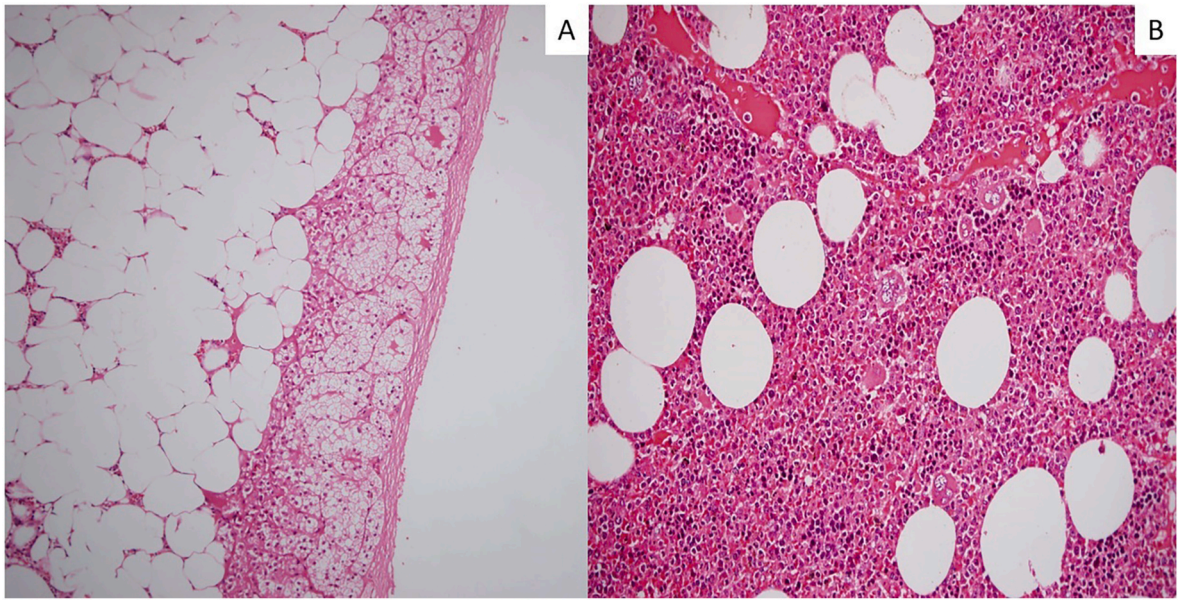

Fig. 3. Histopathologic examination revealed mature adipose tissue adjacent to adrenal cortex(A) hematopoietic cells and mature fat are demonstrated in (B).

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Declarations of interest

The authors declare that they have no conflict of interest.

CRediT authorship contribution statement

Abbas Basiri: Writing – original draft, Supervision, Conceptualization. **Ala Torabi:** Writing – original draft, Conceptualization. **Seyedhossein Rabani:** Writing – original draft, Data curation, Conceptualization.

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