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Oncology

A rare case of virilizing adult ectopic adrenal tumor

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ABSTRACT

Ectopic adrenal gland is an atypical medical condition with a prevalence of less than 1%. It occurs due to an abnormal gland development during embryonic life and can be found in different parts of the body. Rarely, these ectopic glands can be hormonally active and present as bulky masses. Herein we report a case of a patient who underwent laparoscopic surgery for a large myelolipoma associated with an androgen producing adenoma in an ectopic adrenal gland.

Introduction

The adrenal glands are retroperitoneal endocrine organs usually located over the kidneys and responsible for production of several substances including mineralocorticoids, glucocorticoids, androgenic steroids, and catecholamines. The gland develops between the third and fourth week of embryonic life and is composed by two types of tissues, the cortex and the medulla.[1]

Ectopic adrenal tissues or ectopic adrenal glands are rare, with prevalence around 1% and occurs due to an abnormal gland development.[2] Ectopic tissue is usually found near the topic adrenal glands while ectopic gland occurs along the path of embryonic migration in the urogenital tract.[2] Eventually, these ectopic adrenals may present as bulky masses that secret hormones leading to a florid clinical presentation or sometimes as invasive neoplasia due to malignant transformation.[2]

Adrenal myelolipoma is a rare benign tumor that occurs at the adrenal cortex.[2] It is composed of hematopoietic and adipose tissue and is usually a non hormone-secreting tumor. Herein we report a rare case of a bulky androgen-producing ectopic adrenal myelolipoma treated laparoscopically.

Case Report

We report a case of a 28 years old, female patient with a history of amenorrhea for the last 3 years. During a clinical visit it was noticed signs of virilization. Blood tests and imaging were ordered. After that the patient was referred to our clinic for evaluation. On physical exam, the patient presented with android muscle growth and android body fat distribution (Fig. 1). A masculinized voice was also noticed, as well as abundant hair. On genital exam clitoromegaly was present. The rest of physical exam was unremarkable and vital signs were normal. Blood tests results demonstrated increased serum levels of testosterone (3.5 ng/ml, reference 0.1–10 ng/ml) and DHEA-sulphate (3698 μg/dl, reference 10–160 μg/dl). The abdominal computed tomography (CT) scan identified a retroperitoneal mass, measuring 7.0 × 6.0 × 8.4cm, with regular contours, mixed density of soft tissues, fat and calcification, with a heterogeneous contrast enhancement. (Fig. 2). Angiotomography showed that the mass was in intimate contact with left renal hilum. Lymph node enlargement was not detected and chest tomography was normal.

All treatment options, its risks and benefits, were discussed and it was opted for a transperitoneal laparoscopic surgical approach. The patient was placed in the right lateral decubitus position at 30° and all pressure points were padded. The first 10 mm trocar was placed 2 cm superior the umbilicus to the left of midline using Hasson technique and a 30-degree scope was inserted. After completion of the pneumoperitoneum another two trocars were inserted at left midclavicular line. One 10 mm caudal trocar was placed 5 cm superior the umbilicus and a 5 mm trocar was inserted 5cm bellow the costal margin. The line of Toldt was identified and left colon was carefully reflected medially. A bulky mass was visualized in intimate contact with the anterior face of the lower pole of the left kidney and left renal vein. Left ureter and left gonadal vein were individualized. Several anomalous vessels coming from renal and left gonadal veins towards the tumor were identified. The lesion was carefully dissected preserving its capsule and the

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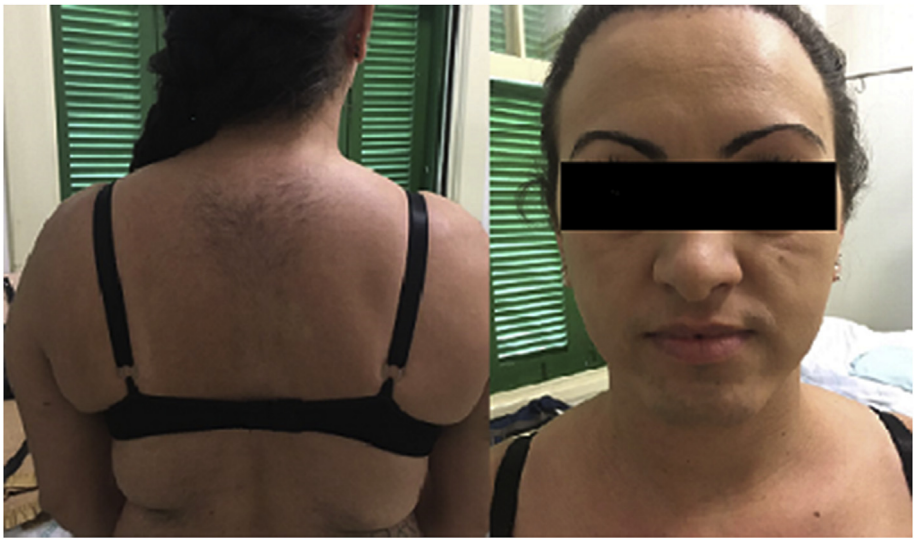

Fig. 1. Android hair and body fat distribution.

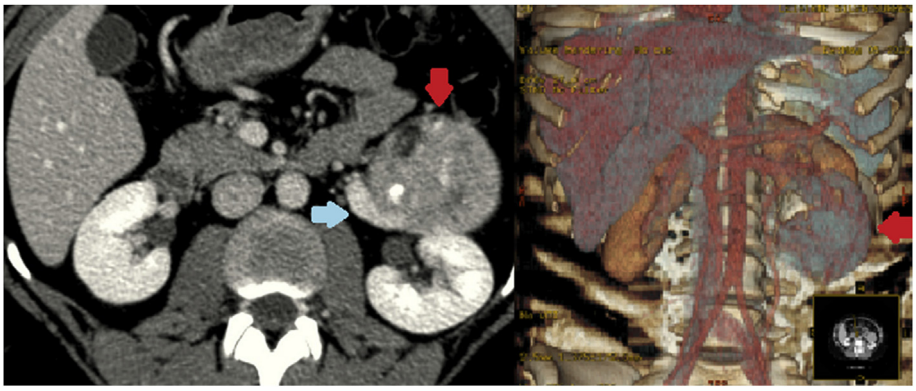

Fig. 2. Angiotomography and CT scan.

anomalous vessels were cauterized with ultrasonic scalpel. An arterial branch from aorta, was visualized, dissected and controlled under metal clips, as well as large venous vessels coming from the gonadal and renal left veins. The specimen was removed trough a 6cm low Pfannenstiel incision.

The surgery duration was 270 minutes with an estimated blood loss of 25ml. The tumor was resected successfully without capsular damage (Fig. 3). Patient was prescribed with intravenous non-opioid analgesics for the first 24 h and after that with oral analgesics. She was discharged 48 hours after the procedure. No minor or major complications were presented. Final pathology was categorized as an andrenocortical neoplasia with oncocytic cells associated with myelolipoma, of probable benign behavior by the modified Weiss criteria (Fig. 4). Immunohistochemical staining confirmed the diagnosis. Forty-five days after the surgery, the patient came to the clinic presenting with weight loss, decreased hirsutism, and had already started with her menstrual cycle. Serum hormonal levels were normal.

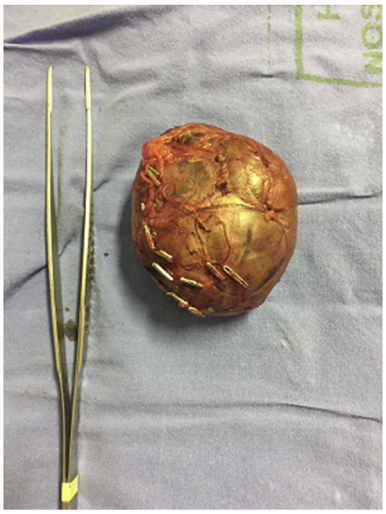

Fig. 3. Ectopic adrenal mass.

Discussion

Ectopic adrenal tumors are rare conditions that routinely are detected in the gland migration path during fetal life. Although more frequently found in the abdomen, ectopic adrenal lesions can be found in different body locations.[1,2] Moreover, sometimes these lesions can present as malignant tumors or even large hormonally active masses.[2]

Myelolipoma is a rare and usually asymptomatic benign tumor.[2] Autopsy studies found that these lesions have an incidence lower than 0,1% and are routinely located at topic glands.[2] Bishop et al. postulated that this tumor can emerges from a clonal proliferation of stem cells, however its origin is not completely understood.[3] Nonetheless, extra-adrenal myelolipomas may be found in different body locations, for example near the liver, lymph nodes and spleen.[2]

Myelolipomas are lesions composed by a variable proportion of

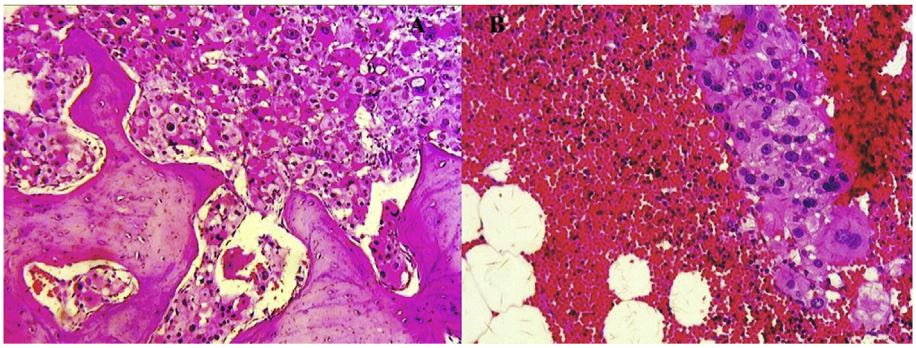

Fig. 4. 4A-adrenal tissue with oncocitic areas and osseous metaplasia; 4B- adrenal tissue with hematopoietic tissue and mature adipocits (HE, 20 X).

mature adipose tissue and active hematopoietic elements wrapped in a pseudocapsule.[3] Traditionally, the CT scan demonstrates a heterogeneous well-circumscribed fat containing-lesion and MRI shows a T1 hiperintense sign and a variable T2 signs depending on the hematopoietic component.[4]

Few cases of hormonally active ectopic adrenal tumor have been described. For instance, Mazza et al. found an ectopic aldosteronoma that was located in the retrocava location that were successfully treated by a conventional open approach[5]

Although the laparoscopic technique is the first choice for adrenal surgery, to the best of our knowledge this is the first case of virilizing ectopic adrenal myelolipoma treated laparoscopically. The benefits of a minimally invasive approach are multiple. Less pain and bleeding and lower length of hospitalization stay are all well-established improvements. In our case there was a minimal bleeding and patient was discharged in the second postoperative day.

Benign ectopic adrenal tumors are rare. In this case the tumor was located over the left renal hilum. As in topic adrenal tumor, the minimally invasive approach is safe and feasible for ectopic adrenal lesions.

Consent section

Written informed consent was obtained from the patient for publication of this Case Report and any accompanying images. A copy of the written consent is available for review by the Editors-in-Chief of this journal.

Author's contribution

1. Concept and design: Ricardo Segabinazzi Dotto, Daniel Melecchi de Oliveira Freitas
2. Provision of study materials or patients: Guilherme Marx, Juliana Lohman Machado.
3. Collection and assembly of data: Guilherme Marx, Mateus Bastos
4. Manuscript writing: all authors
5. Final approval of manuscript: all authorsAcknowledgements Tatiana Witt Nunes

Conflicts of interest

The authors have no conflicts of interest to declare.

Appendix A. Supplementary data

Supplementary data to this article can be found online at https://doi.org/10.1016/j.eucr.2019.100907.

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