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Case Report

Adrenocortical Carcinoma with Extension into the Inferior Vena Cava: Case Report

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Abstract

Adrenocortical carcinoma (ACC) is a rare abdominal malignancy. A 68 years old female suffered of lower limbs edema, in the investigation she was diagnosed with a mass in the abdomen, with infiltration of the Inferior Vena Cava IVC, which was later found to be ACC. The mass was large, with more than 15 centimeters at longest dimension. A successful resection of the tumor and (IVC) tumor emboli was performed.

Keywords: *Adrenocortical Carcinoma; Vena Cava Inferior Thrombectomy; Syria*

Introduction

Adrenocortical carcinoma (ACC) is a rare case with an annual incidence of 0.7 - 2 cases per year and a worldwide prevalence of 4 - 12 cases per million/year. ACC patients have a poor prognosis with a 5-year overall survival (OS) below 30% in most series [1].

Most ACCs cause extended abdominal mass, hypersecretion of steroids including glucocorticoids and androgens, and 15% of the ACC cases are discovered accidentally. Most cases of ACC accurate at women at the first decade of life with a second peak between (40 - 50) years of age [2].

ACC is found to be associated with inherited disorders accompanied with gene mutations: Li Fraumeni syndrome (LFS), Multiple endocrine neoplasia type 1 (MEN1), Beckwith-Wiedemann syndrome (BWS), Familial adenomatous polyposis (FAP), Neurofibromatosis type 1 (NF1) [1].

Complete surgical resection is the known curative therapy [3].

Case Presentation

A sixty eight-years-old woman presented to the hospital with lower limbs edemas. She was in good statues with no other complaints. Edema evolved in the course of one month.

Patient was, other than described, healthy with no comorbidities, or any remarkable medical history.

Abdominal Ultrasound (US) was performed in the course of edema studying and it revealed a large mass on the right adrenal gland pushing the diaphragm on the right side.

An Magnetic Resonance Imaging (MRI) showed a solid mass with clear borders measures (94 x 95 x 110) millimeters (mm) located above the right kidney with central necrosis it pushes the right kidney downwards, the lower aspect of the mass is adjacent to the right kidney artery and vein, the above and lateral aspects of the mass are pushing on the liver and the medial aspect is partially surrounding the (IVC) with an emboli, that measures 6 mm inside the lumen (Figure 1).

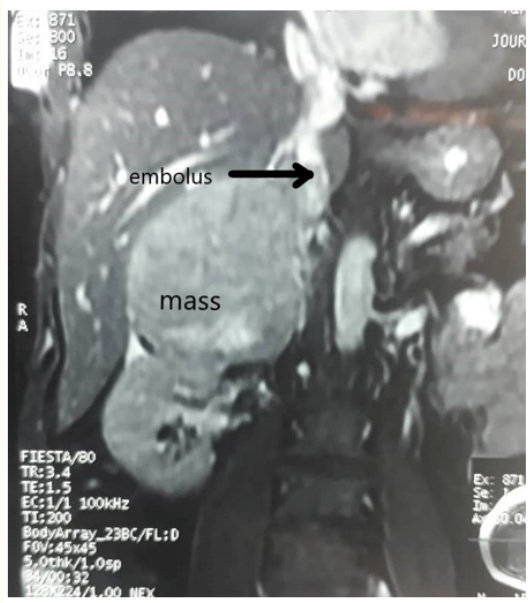

Figure 1: *MRI shows mass and IVC embolus.*

Serum laboratory analysis came in normal ranges, so did the urine analysis.

The surgery took almost 160 minutes, not only the mass was excluded from the surroundings without harming any of the organs associated to it; we were also able to extract the IVC emboli. The patient was stable through. Two units of blood were transfused during (Figure 2-4).

The patient then was released to intensive care unit, got well and then discharged from the hospital 6 days later in a good stable state.

Following the surgery, a Doppler ultrasonography was performed to the lower limbs and it appeared normal. As the reason for the edema was the mass compressing the main veins and the tumor embolus in the IVC.

Post-operative Pathology confirmed the diagnosis as it revealed a mass of 14 centimeters, that weighs 516 Grams. It appeared to be ACC grade III with lymph angioinvasion, lymph nodes were free of disease, and a tumor embolus of the IVC. KI 67 scored 3 - 5% (low proliferative index) which is a good prognostic factor (Figure 5 and 6).

Discussion

ACC is a rare retroperitoneal tumor of the, At the time of presentation, ACCs are generally large tumors, measuring on average 10 to 13 cm. Patients with ACC usually present with signs and complaints associated with hyper hormonal secretion and some are found accidentally, our patient suffered from symptoms caused by the involvement of the mass to the IVC [7].

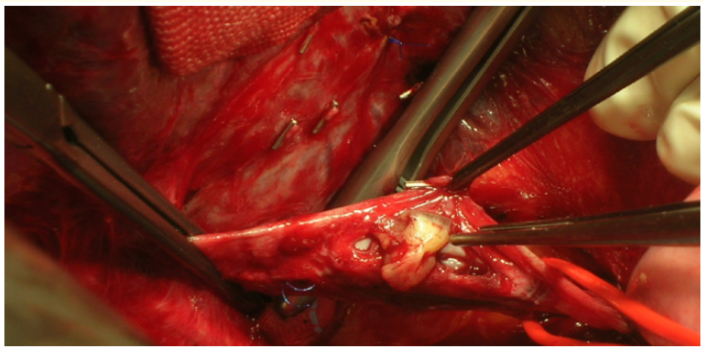

Figure 2: *Shows IVC embolus.*

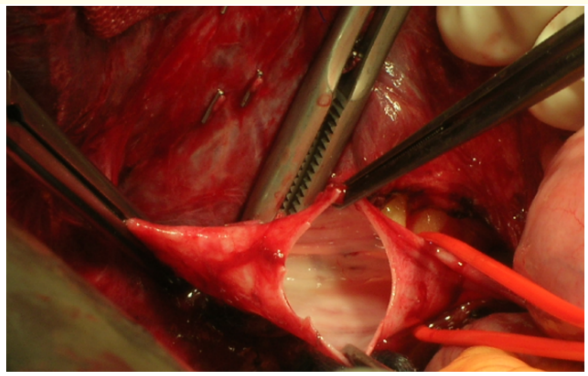

Figure 3: *Shows IVC embolus extraction.*

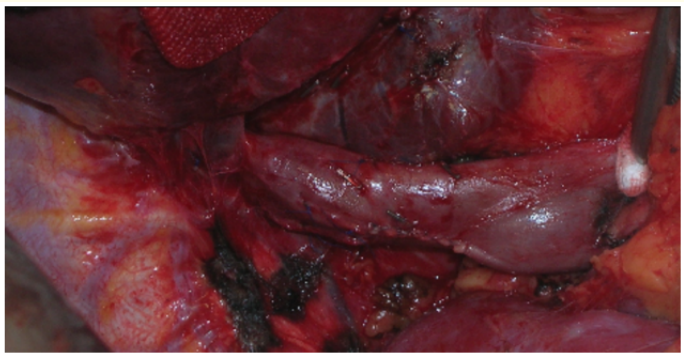

Figure 4: *Shows IVC post embolus extraction.*

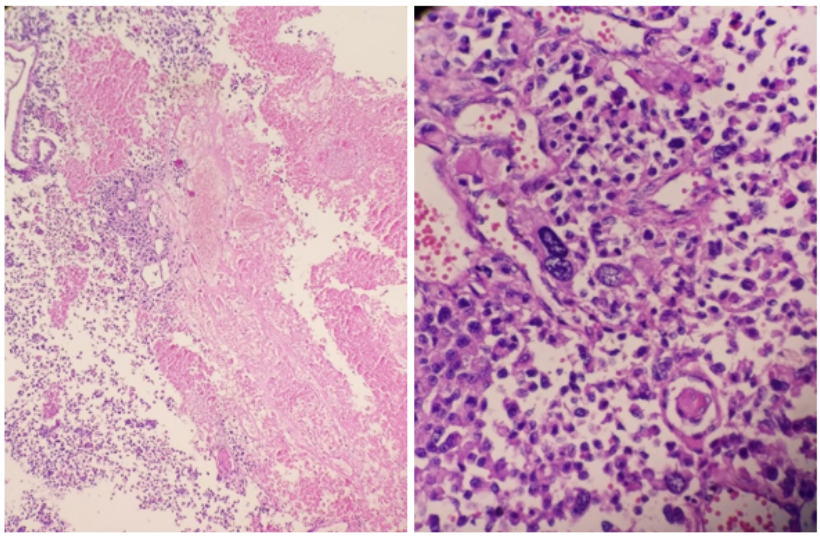

Figure 5 and 6: *Showing the pathological findings.*

ACC has some characteristics in imaging signs, it is frequently > 6 cm, heterogeneous and irregularly shaped. On CT: density > 10 HU without contrast; less than 40 to 50% washout or density > 35 HU 10 - 15 minutes after contrast injection. And on MRI: hyperintense on T2-weighted sequences; < 30% loss of signal intensity on out-of-phase images [1].

Our patient had an MRI that complied with the above mentioned. We preferred the MRI over the CT as it gives us a better idea of the vessels as we had already suspected of the IVC involvement and it is crucial to perform at least one of them and not only the ultrasound [6].

Fine needle biopsy of a suspected ACC is almost never justified because of anticipated tumor spill [4] and for that and the definitive necessity for the surgery we proceeded to the tumor complete excision right away.

Most of the adrenal tumors are hormone secreting tumors, Hypercortisolism is the most common presentation of patients presenting with hormone excess (50% - 80% of hormone-secreting ACCs), causing classic symptoms including plethora, diabetes mellitus, muscle weakness/atrophy and osteoporosis. Frequently, very high cortisol levels in ACC saturate the renal HSD11B2 system, resulting in glucocorticoid-mediated mineralocorticoid receptor activation. Therefore, hypokalemia and hypertension are commonly observed in ACC patients with hypercortisolism. Together with pronounced muscle weakness, these symptoms of rapidly progressive Cushing's syndrome are generally indicative of a malignant adrenal tumor. Whilst, adrenal androgens are the second most common in secreting ACCs, causing symptoms like rapid onset male pattern baldness, hirsutism, virilization, and menstrual irregularities in women. Concurrent androgen and cortisol production is evident in roughly half of all ACC patients with hormone excess [7].

Our patient was other than the edema completely healthy, we completed a full serum and urine analysis on our patient and it all came in normal ranges.

The staging system proposed by European Network for the Study of Adrenal Tumors (ENSAT) is the best available system, as this system clearly identifies patient prognosis [4].

Stage I	Tumor restricted to the gland and < 5 cm in size
Stage II	Tumor restricted to the gland and > 5 cm in size
Stage III	Adjacent tissue invasion and/or neoplastic thrombus to IVC
Stage IV	Presence of distant metastasis

Table 1: *ENSAT ACC staging system [4].*

Our patient accordingly, was stage three due to the IVC involvement.

Most ACC patients have respectable disease at presentation; however, more than half of the patients who have undergone complete removal of the tumor are destined to have a relapse, often with metastases [5].

Patient underwent surgery, which was successful as we were able to ablate the mass completely from the surroundings without harming any of the organs or the main vessels, and completely resect the IVC emboli.

Pathology revealed clear borders in a mass of ACC, and ki67 score was (3-5%) which is a good prognostic factor.

Following surgery comes the role of adjuvant therapy, usually Mitotane is advised as it has the best outcome in increasing the disease-free period, because of its toxicity it should be considered mainly in patients with high risk of reoccurrence (stage III disease or tumors > 8 - 10 cm in size, those with high mitotic rates or a proliferative index Ki67 > 10% and those with microscopic evidence of vascular or capsular invasion) [1].

Patients with ACC who had complete surgical resection should be monitored and revaluated every three months, that should include physical examination, hormonal evaluation, and CT of the abdomen and thorax, after 2-3 years period disease free, evaluation can be distended to twice a year [4].

Our patient is still healthy since 7 months, she recovered from the edema, had normal CT. and we will go on reevaluating her every three months.

Conclusion

Although ACC tumors are rare, and unlikely to be found accidentally, it is also possible to be found in relatively healthy looking individual and demonstrate itself in unusual characteristics. Surgery remains the corner stone in the treatment of the ACC tumors.

Follow up therapy with mitotane, should be considered in high risk patients. As it elongates the disease free period.

Patient Consent

Consent to publish the case report was not obtained because this report does not contain any personal information that could lead to the identification of the patient.

Authorship

All authors attest that they meet the current ICMJE criteria for Authorship.

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Conflicts of Interest

The authors declare that there are no conflicts of interest regarding the publication of this paper.

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