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Case Report

Surgery Section

A Rare Presentaion of Adrenocortical Carcinoma

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ABSTRACT

Adrenocortical Carcinoma (ACC) is a rare tumour, with an annual incidence of approximately one per million. Almost all cases occur in patients aged 40 to 50 years, but there is a minor peak in occurrence among children younger than 5 years. It demonstrates no significant gender predilection. At the time of presentation, ACC tend to be very large and have usually spread beyond the confines of the adrenal gland. The presentation of this tumour may vary, either it can present as virilisation or Cushing's syndrome or both. The authors present their experience of a rare presentation of an asymptomatic ACC with a large size of tumour mass with no metastasis with just complain of burning micturition in a 55-year-old male patient which was difficult to diagnose clinically. The patient is doing well after surgical management.

Keywords: Computed tomography, Cushing's syndrome, Radical nephrectomy, Serum cortisol

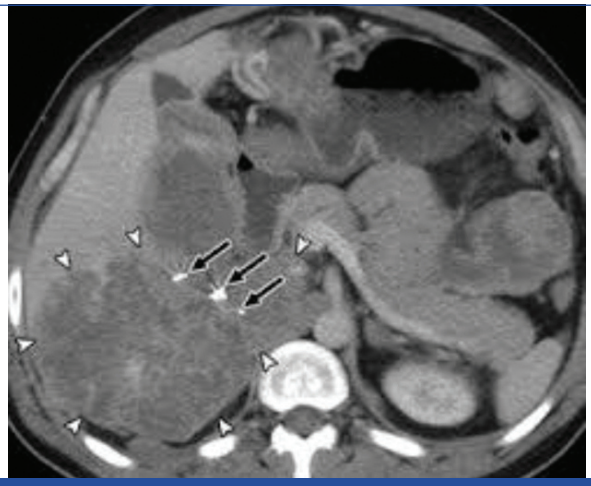
CASE REPORT

A 55-year-old male patient had complains of burning micturition with acute retention of urine since 15 days. The patient had no history of fever, pain or lump in abdomen, haematuria or polyuria or nocturia, loss of appetite or loss of weight, breathlessness or jaundice. Patient had neither cushingoid features nor any co-morbid conditions (hypertension, diabetes, tuberculosis or bronchial asthma). The patient also had no history of any previous medical or surgical interventions.

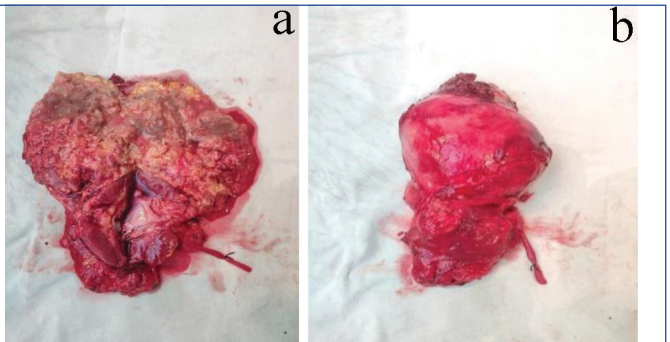
On clinical examination, patient was moderately built with no pallor, icterus, cyanosis, clubbing or lymphadenopathy. Abdomen was soft, non-tender, bowel sounds present in all four quadrants with no distension, guarding or rigidity. Patient underwent Ultrasound Sonography test (USG) of abdomen and pelvis which was suggestive of a large lesion in the right adrenal gland with heterogenous enhancement. Differentials considered were; metastasis or ACC. Fat between the lesion and surrounding structure was well-maintained. There was a calculus at right vesico-ureteric junction with resultant proximal mild hydroureter and hydronephrosis with non-obstructive calculus in lower calyx of the right kidney.

Patient then underwent Contrast-Enhanced Computed Tomography (CECT) abdomen and pelvis which revealed a right sided Vesicoureteric Junction (VUJ) calculus of 5×5 mm in size and an astonishing finding of a large solid cystic lesion of size 12×9.5×10 cm in the right adrenal gland with heterogeneous enhancement [Table/Fig-1]. It showed mild vascularity on doppler and compressed kidney downward and also inferior surface of liver parenchyma which was suspicious of adrenocortical tumour. Fat plane between the lesion and the surrounding structures were well-maintained. Blood Investigations reports were as follows: Serum Cortisol (8 AM) 17.2 µg/dL; Serum Cortisol 6.17 µg/dL; Serum Dehydroepiandrosterone Sulfate (DHEAS) 7200 µg/mL; Urine Metanephrines 220 µg/24 hours. Hence, the patient was provisionally diagnosed with ACC with subclinical Cushing's syndrome. Based on the CT finding, a decision of exploratory laparotomy was taken. Intraoperatively, a large mass was found on upper surface of right kidney arising from adrenal gland with infiltration to inferior surface of the liver and upper pole of kidney [Table/Fig-2].

Histopathological examination of the adrenal tumour showed a well-vascularised tumour. There were large, polygonal tumour cells, with basophilic, eosinophilic, or vacuolated cytoplasm, focally with intracytoplasmic hyaline globules [Table/Fig-3]. Hence, Right Adrenalectomy with Right Nephrectomy and wedge resection of liver was done. The postoperative period of the patient was



[Table/Fig-1]: CT scan image of Adrenocortical carcinoma of the right adrenal gland (marked with arrow).

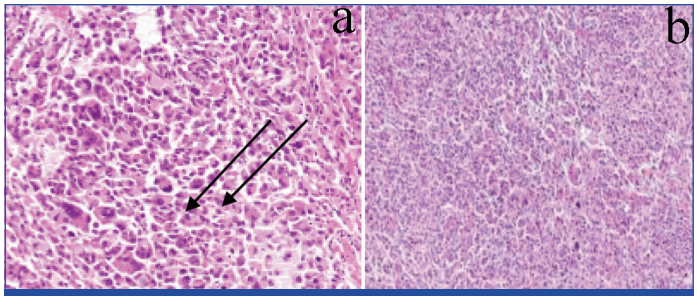


[Table/Fig-2]: Macroscopic aspect- On the cut section, the tumour was nodular, with grayish solid areas, with hemorrhages and necrotic zones, separated by fibrous septa (a and b).

uneventful. Histopathology report revealed oncocytic type of ACC (T.N.M. Staging of pT3, pNx, pMx) with infiltration into the peripheral fat while no infiltration into the kidney, liver, lymph node, blood vessel and ureter. The patient is advised to take tab mitotane 1 gm for five years and follow-up every three months. At present, the patient is asymptomatic and carries out his daily routines.

DISCUSSION

The ACC is a rare endocrine cancer with poor outcome or prognosis. It is usually larger than 4 cm and metastasises to liver, lung or lymph nodes at time of diagnosis. The worldwide incidence



[Table/Fig-3]: a) (40X), b) (10X)- Microscopically, at low power view, the clusters of polygonal-shaped cells seen with eosinophilic cytoplasm (arrows), well-defined nucleoli, and atypical mitoses with haematoxylin and eosin stain.

is 0.5-1 case/million/year. ACC has bimodal age distribution at childhood and the fourth decades of life and incidence of female to male ratio is approximately 1.5:1 [1]. Similarly, in this case, size of the tumour was found to be 12×9.5×10 cm with infiltration into the peripheral fat with no infiltration into the kidney, liver, lymph node, blood vessel and ureter. Mainly, ACC is a non-functioning mass. Patient usually presents with abdominal discomfort or flank pain due to the mass effect. Nevertheless, some of ACC [2] can produce hormones such as corticosteroid, aldosterone [3] or sex steroid [4]. As such, patient with incidental finding of adrenal mass should be evaluated for signs and symptoms of hormone hyper secretion. Patients with functioning ACC may present with clinical symptoms of Cushing syndrome such as round face, easy bruising, truncal obesity, proximal muscle weakness, hyperglycaemias and/or osteoporosis, hyperpigmentation of skin, high blood pressure, headache, depression, decreased libido, decreased fertility, erectile dysfunction and fatigue. Non-functioning ACC patients complain of abdominal discomfort and back pain. The reported patient was comparatively asymptomatic and only had burning micturition.

Diagnosis is done by measurement of serum cortisol, Dehydroepiandrosterone sulphate (DHEAS), catecholamines and dexamethasone suppression test. Imaging studies like ultrasound, CT abdomen pelvis and Magnetic Resonance Imaging (MRI) are useful to evaluate the tumour. ACC frequently presents with metastasis which has to be evaluated by preoperative staging CT scan. CT scan is useful in assessment of central tumour necrosis, calcifications, size, benign adenomas and carcinomas. Typical cases of ACC on CT scan show large unilateral adrenal mass with irregular edges. Macroscopic features of ACC which are suggestive of malignancy include a weight of more than 500 grams, presence of areas of calcification or necrosis and a grossly lobulated appearance. Histologic findings include numerous mitotic scanty cytoplasm. Mainstay of management of ACC is total resection (R0), which is the only potentially curative therapeutic modality [5] for localised ACC. Resection of tumour can ameliorate signs and symptoms of Cushing as well as its complications such as cardiovascular disease and osteoporosis. It has been revealed in several studies that Mitotane can be used as adjuvant therapy in cases of nonoperable ACC or aggressive adrenal gland tumours. It's mechanism of action is unknown but it is believed that Mitotane is cytotoxic to adrenal gland and it selectively damages zona reticularis and zona fasciculate layer of adrenal cortical tissue [6-8].

Thus, medical management is only supportive and is given as an adjuvant therapy in the form of mitotane 1 gm. This drug has been used for more than 50 years, and continues to be the key element of medical treatment of ACC [9]. According to recent data, it probably exerts effects through the down-regulation of the mitochondrial respiratory chain [10]. An author has showed that mitotane treatment allows prolonging patients' survival [11]. In another study, patients in stage II treated with mitotane after surgery had a better prognosis for this stage [12]. In the present case, patient underwent right Adrenalectomy with right Nephrectomy and wedge resection of liver and was advised tab mitotane 1 gm for five years and follow-up every three months. Patient at present is doing absolutely well with his daily routine activities and is asymptomatic.

CONCLUSION(S)

The reported patient did not show any of the clinical symptoms and hence was difficult to diagnose at first. Due to the restricted knowledge reservoir, genuine diagnostic and therapeutic courses are still unclear. Thus, there is a vast portion of medical field to be further explored in order to find an accurate treatment procedure for ACC and a cure is required, so that a patient has minimum chance of secondary tumour infiltration. It would also be good to diagnose asymptomatic cases as early as possible.

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