Case Report

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A case report on adrenocortical carcinoma

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ABSTRACT

Adrenocortical carcinoma is a rare entity and usually diagnosed at later stages which poses a treatment dilemma and usually results in a bad prognosis. It is the second most aggressive endocrine malignancy after anaplastic thyroid carcinoma. Here we report a case in a young male diagnosed incidentally on an ultrasound scan done for abdominal pain. On examination his abdomen was soft with no obvious mass palpable. We are reporting a brief summary of the diagnosis, work up and management of this rare entity in the hope that it may shed some more light on the appropriate management of this elusive condition.

Keywords: Adrenocortical carcinoma, Adrenal malignancy, Abdominal pain

INTRODUCTION

Adrenocortical carcinomas are extremely rare with an incidence of around 1-2 per million population. It is the second most aggressive endocrine malignancy following anaplastic carcinoma of the thyroid.² A female predilection has been observed in the pattern of cases, with a bimodal age distribution. 60% tumours are functioning with Cushing's syndrome being the most common manifestation Followed by cushing's and virilisation syndrome.^{3,4} It is the non-functioning tumours that pose a diagnostic dilemma and treatment difficulty as they are late in the course of the disease. Here we present one such case report of a non-functioning adrenocortical carcinoma diagnosed incidentally on radiographic screening. The diagnostic and pre-treatment work-up and management is being summarized here.

CASE REPORT

A 27-year-old male patient presented to the surgery OPD with complaints of abdominal pain of one month duration. It was an intermittent stabbing type of mild ache, over the left lumbar region. There were no exacerbating or relieving factors. No diurnal or postural variation. He also

complained of occasional low backache. There were no associated symptoms such as headache, palpitations, diaphoresis, anxiety, tremulousness, fatiguability, giddiness, dyspnea, vomiting, alteration of bowel habits, blood in stools, haematuria, mucus diarrhea, loss of weight or appetite, lower limb weakness or numbness.

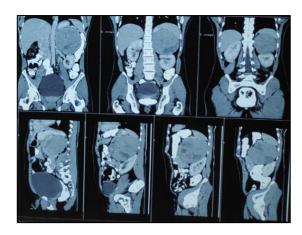


Figure 1: CT film showing a heterogeneously enhancing mass lesion in the left adrenal gland.

He did not have any significant past medical history or family history of any malignancies.



Figure 2: Intraoperative image showing a large adrenal tumour measuring 15 cm.



Figure 3: Gross cut section of the tumour with multiple areas of internal haemorrhage and necrosis.

General examination findings: patient was moderately built and nourished with no pallor, icterus, cyanosis, clubbing, generalised lymphadenopathy, or dependant edema. Pulse rate 84 per minute, BP: 130/90mmHg all 4 limbs, respiratory rate: 16 per minute and afebrile

His abdominal examination was unremarkable. No masses could be palpated per abdomen or on digital rectal exam. Routine blood investigations were within normal limits.

Biochemical tests revealed the following: 24-hour catecholamine and metanephrine levels were normal in the serum and urine. Serum cortisol and overnight dexamethasone suppression tests were normal. Serum aldosterone and electrolytes were within the normal range. Thyroid function tests were also normal.

On abdominal imaging, ultrasound revealed a large well-defined mass lesion measuring 12.7×9.8×7.4 cm. It was a heterogeneously hypoechoic mass lesion within the left adrenal mass, inferior to spleen, indenting on left kidney with internal cystic areas and calcified foci possibly: non-functioning phaeochromocytoma or adrenocortical

carcinoma. Contrast enhanced abdominal CT revealed a large heterogenous lesion measuring 11.6×11.4×9.7cm in the left suprarenal region with calcification and hypodense foci, heterogenous post contrast enhancement. The lesion was displacing the left kidney inferiorly, posteriorly abutting abdominal wall, medially the aorta, anteriorly abutting the pancreas, stomach, descending colon.

Intraoperatively, a left lateral curvilinear incision was placed to gain access to the tumour approaching it intraperitoneally, and left adrenalectomy was performed. A highly vascular left adrenal tumour weighing 635g was extracted. It was abutting but not infiltrating the surrounding structures and compressing the left kidney. Open surgery was performed as the tumour size was more than 6 cm in size which would make a laparoscopic approach more difficult. The postoperative period was uneventful and the patient was discharged on the 5th postoperative day, at which point he could take a normal oral diet, mobilise comfortably and his pain could be controlled with oral analgesics.

The histopathology report came as Adrenocortical carcinoma. Section showed adrenal tissue with an encapsulated neoplasm composed of cells arranged in solid diffusely large nests and in alveolar pattern. individual cells were, round/oval cells with abundant eosinophilic cytoplasm and large vesicular nuclei, some showing prominent nucleoli, 4 mitosis/hpf. There was necrosis, capsular and vascular invasion. Lymphoid aggregate was observed surrounding the tumour.

DISCUSSION

Adrenocortical carcinoma is a rare entity (incidence of 2 per 1 million).² It can occur at any age But bimodal distribution is more commonly observed in the first and fourth decades. Approximately 60% are functional with hormone overproduction.⁵ Cushing's syndrome is the most common manifestation among functioning varieties.⁵ It is mostly sporadic. This condition may be associated with Li Fraumeni syndrome, MEN 2 syndrome, Carney complex, Beckwith Weideman syndrome, to name a few.⁶ The pathogenesis remains elusive, with possible molecular mechanisms including Tumour protein TP53 inactivating mutations, IGF-2 overexpression, ZNRF 3 gene mutation, and Beta catenin activating mutations.

These tumours may manifest themselves with abdominal pain, Cushing syndrome, virilization, feminisation, and Hypertension: due to excessive circulating mineralocorticoids or glucocorticoids. Non-functioning tumours (50%) may be asymptomatic and go undetected until late stages. Even functioning tumours may be asymptomatic. According to WHO classification ACC variants include Oncocytic ACC, Myxoid ACC and ACC with sarcomatous areas.

When ACC is suspected, the diagnostic blood investigations to be performed are 24 hours urine samples

for catecholamines and their metabolites, clonidine suppression test, plasma metanephrine, low dose (1mg) dexamethasone suppression test, Plasma renin and aldosterone. These tests will help establish the functional status of the tumour. Further characterization should be done by performing imaging. CECT Abdomen has 85 to 95% sensitivity and 70-100% specificity. Benign tumours appear well encapsulated, with smooth and regular margins.

Malignant tumours are hyperattenuating, inhomogeneous, with irregular borders, evidence of local invasion and lymphadenopathy. MRI Abdomen is 95% sensitive, and 100% specific. Adenomas appear as low signal intensity lesions, while carcinoma has moderate intensity on MRI. A tumour of size more than 5 cm on imaging is usually malignant. Other imaging modalities include radionuclide scanning with NP-59 and FDG-PET scan.

Indications for surgery include tumour size more than 4 cm, functioning status and imaging features such as more than 20 HU, delayed contrast washout, areas of haemorrhage, necrosis, and peripheral enhancement. Radical surgery is the treatment of choice in tumours which haven't metastasized as it gives a potential for cure. Adrenalectomy, resection of surrounding lymph nodes with en bloc resection of the surrounding involved structures is the surgical procedure that must be followed.

PRA (posterior retroperitoneoscopic adrenalectomy) –is regarded as the standard approach currently.

Newer modalities of treatment include Mitotane which is a steroid synthesis inhibitor and may be given following resection as an adjuvant therapy, to reduce recurrence rates, or as a primary modality in metastatic cases. Suramin which is a Growth factor inhibitor is another newer treatment modality. Chemotherapy may be instituted in cases which have metastasis on presentation. Commonly used chemotherapeutic agents are Etoposide, Doxorubicin and cisplatin combination or streptozocin. Chemotherapy can be administered as adjuvant treatment also.

Radiotherapy may be given for palliation in bone metastasis or in unresectable local recurrence cases. Other than its usage in these conditions, RT doesn't confer any additional survival benefit.

CONCLUSION

ACC is a rare entity, associated with a poor prognosis. The median survival rates depending on the stage are as follows, with stage I-II:159 months (95% C.I), Stage III: 26 months (95% C.I), and Stage IV: 5 months (95% C.I). Here we have discussed an extremely rare case of Adrenocortical carcinoma. The patient in question has a

relatively better prognosis as the patient is a young male with no distant metastasis and having had an R0 resection. But still ACC is considered to have overall low progression-free survival rates, hence needs constant vigilance and close follow up post surgery.

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