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CASE REPORT

Adreno Cortical Carcinoma – A Rare Entity

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ABSTRACT

Adrenocortical carcinoma is a rare neoplasm of 1-2 per 1 million people. We report a case of 34yrs male patient presented with swelling and occasional pain in the upper abdomen since 2 months without any functioning tumor symptoms.The clinical, biochemical,histological features along with differential diagnosis are discussed below.This case is presented because of its rarity and also highlighting the importance of differentiating it as functional and nonfunctional tumor and preparing a case for laparoscopic and open surgery.

KEY WORDS : Adrenal, VanillylMandelic acid,Adrenalectomy

Introduction

Adrenocortical carcinoma[1,2] is a rare neoplasm with incidence of 2 per 1 million people, bimodal age distribution in children below 10 yrs and in fourth and fifth decade of age. Approximately 60% of tumors are functional that secretes hormones like cortisol [30%], androgens[20%], estrogens[10%], aldosterone[2%] and mostly it presents with clinical features like Cushing's syndrome. Virilization is more frequently seen in children and has better prognosis after complete resection than in adults.

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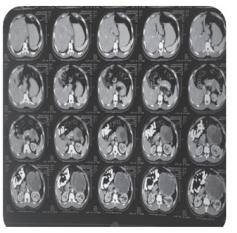
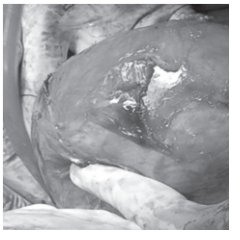
Case Report

A 34yr male presented[3, 4]with a complaints of swelling and occasional pain in the upper abdomen since 2 months.On examination,an oval shaped swelling of appro15x10cm involving left hypochondrium and epigastric region with well-defined margins, firm consistency and side to side mobility is seen. Clinically differential diagnosis of Retroperitoneal sarcoma or retroperitoneal cyst or pseudocyst of pancreas or left adrenal mass was made. CECT ABDOMEN shows necrotic mass lesion arising anterior to left kidney,contiguous with anterior limb of left adrenal gland pushing pancreas and descending colon. Bone scan shows no bony metastatic deposits. 24 hr urine for Vanillyl Mandelic acid was normal. Provisional diagnosis [5] of LEFT ADRENAL MALIGNANCY was made and preceded with open adrenalectomy. Histopathology report shows it as ADRENO CORTICAL CARCINOMA.

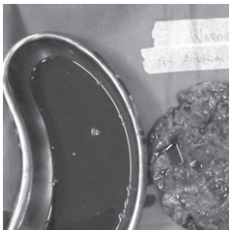
Ct Scan Abdomen



Interoperative Picture



Cut Section Of The Specimen



Discussion

ACC is a rare neoplasm of incidence 2 per 1 milion population with a slight female predilection with bimodal age distribution common in children, 4th and 5th decade of life. ACC occurs with increased frequency in children[6] with Beckwith-wideman syndrome and Li Fraumani syndrome.

60% of ACC are functional tumors often present with clinically pure endocrine symptoms like cushings syndrome and sometimes may be mixed with virlization. Virlization in children indicates malignancy in 70% of cases. In this case patient presented as a non functional tumor with vague symptoms.

The size of the adrenal mass in CT SCAN & MRI is the single most important criteria in diagnosing malignancy. Tumor of >6cm in size is malignancy in almost 92% of cases.

Biochemical tests like serum electrolytes, urinary caetocholamines, 24 hr urine for cortisol, overnight 1 mg dexamethasone suppression test are diagnostic tests to confirm a functional and nonfunctional tumor and also for medical management, post-operative status. Histopathologically Weiss criteria introduced to differentiate benign and malignant tumors.

For staging of the tumor we followed TNM staging:

Stage 1:
Tumor<5cm .with no spread to surrounding tissues or lymphnodes without distant metastasis.

Stage 2:
Tumor>5cm with other stage1 characteristics.

Stage 3:
Tumor invading to nearby tissue and or spread nearby lymphnodes

Stage 4:
Distant metastasis.

Medical management is by Adrenostatic drugs like ketoconazole, aminoglutathimide, metyrapone and Mitotane+Ectoposide+Doxy rubicin[7] as chemotheraphy.

Adrenalectomy either by open or laparoscopic [8]is the treatment of choice for adrenal tumours depending on the size. Highest rate of local recurrences [9]is seen in 60-80% of cases. If tumor of >6cm and local infiltration is seen better to do open adrenalectomy than laproscopic.

Radiotheraphy is for local recurrence and in bone mets.5 yr survival rate was very poor of <5% in stage 4 disease.

Conclusion

This case is presented because of the occurrence of 15 * 10 cms size stage IV nonfunctioning adrenal mass in a 34 year old male is a rarity.

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