ORIGINAL RESEARCH PAPER

Oncology/Radiotherapy

RECURRENT ADRENOCORTICAL CARCINOMA – A CASE REPORT

KEY WORDS: Adrenocortical Carcinoma, Adrenocortical Tumour.

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ABSTRACT

Incidence of Adrenocortical Carcinoma approximately 1 to 2 (1, 2) case/ million Population per year. It represents only 0.02% of all Neoplasm. It has two peak of incidence 0-5 years and 40 to 50 years of age. (3) It predominantly occurs in Females with Male: Female ratio of (1:1.5-2.5). (4,5) Some studies suggested proliferative effects of oestrogen hormone on malignant adrenal cells (3) The National Institute of Health office of Rare Disease, research defines rare disease as having prevalence of less than 2,00,000 (two lakh) patients in the United States of America (8). A 37-year-old female presented with pain in abdomen for the last 3 months, she had undergone surgery in 2019, Histopathology report came as adrenocortical tumor. Biopsy was done which had shown malignant neoplasm, Immunohistochemistry report has confirmed as Adrenocortical tumour. Melan-A diffuse moderate strong positive in tumour cells. Adrenocortical carcinoma is found to be more common in the Children (11) Similar to our case report Else and colleagues also reported unilateral involvement with the normal contra lateral adrenal gland and female preponderance. The adjuvant therapies are advised to decrease the chances of recurrence. For unresectable or metastatic disease treatment is advisable with palliative purpose.

BACKGROUND INFORMATION

Incidence of Adrenocortical Carcinoma approximately 1 to 2 (1, 2) case/ million Population per year. It represents only 0.02% of all Neoplasm. It has two peak of incidence 0-5 years and 40 to 50 years of age. (3) It predominantly occurs in Females with Male: Female ratio of (1:1.5-2.5). (4,5) Some studies suggested proliferative effects of estrogen hormone on malignant adrenal cells (3) 80% of adrenocortical carcinoma were found hormone secreting (6). Poor prognosis associated with non-secreting adeno corticoid tumor because of delayed due to indigenous character (7).

The National Institute of Health office of Rare Disease, research defines rare disease as having prevalence of less than 2,00,000 (two lakh) patients in the United States of America.(8) The surveillance epidemiology and end result database (SEER) reported an estimation of incidence is 0.72 per million cases per year which results 0.2% of all cancer related deaths in United States of America(2) Complete surgical removal of the tumor can provide the best chance of cure beside high probability of recurrence (9) Hereby we present a case of Advanced and Recurrent case of adrenocortical carcinoma.

Case Report

A 37-year-old female presented with pain in abdomen for the last 3 months, she had undergone surgery in 2019, Histopathology report came as adrenocortical tumor. Post operatively she was on follow-up till 2020, Due to COVID-19 pandemic she lost to follow up. Now she is presented at our hospital with pain and heaviness in abdomen for the last 3 months CECT Abdomen report shown.

Triphasic CT Scan of whole abdomen

The CT imaging features reveal 11.3cm (CC) × 8.7cm (AP) s 8.4cm (TR) sized well defined heterogeneously enhancing retro peritoneal mass lesion seen in the right lumbar region as described above.

Histopathological evaluation is recommended to differentiate recurrence of post right nephrectomy neoplasm from other differentials.

Ultrasound Abdomen And Pelvic

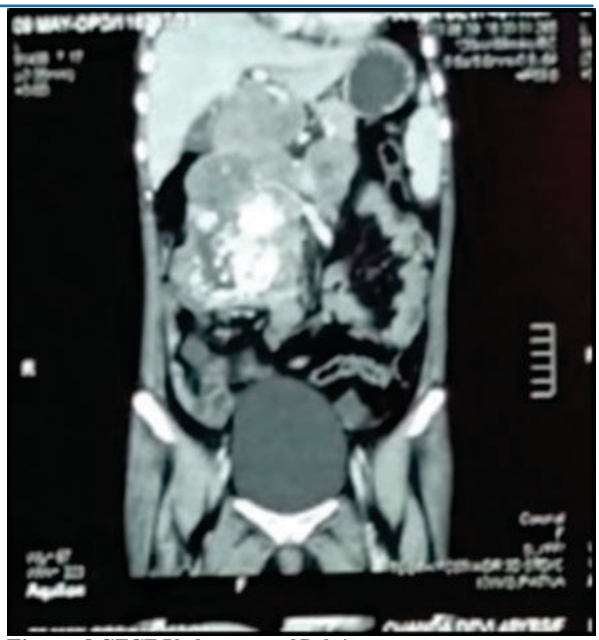

Figure.1 CECT Abdomen and Pelvis

H/O of surgery for abdominal tumor. No details were available at the time of scanning.

1. Right kidney not well visualized. There is large heterogeneous echotexture mass with coarse calcification seen in the right renal-suprarenal region measuring approximately 9.6x6.6cm suggestive of tumor recurrence. Suggest CECT evaluation for assessment.
2. No focal lesion identifiable in liver.
3. Mildly bulky uterus. No adnexal mass or cyst seen.

Impression:

Poorly differentiated tumour; favour adrenal origin.

ADVISE:

Clinico-radiological correlation & IHC for definite categorization.

CT Scan OfWhole Abdomen:

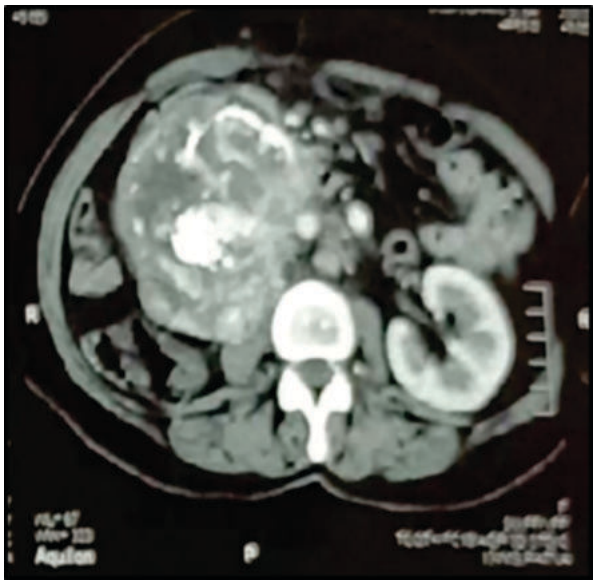

Figure.2 CECT Image

IHC final diagnosis

Adrenal cortex tumour

Histopathology

Microscopy:

Section shows tumor composed of cords and insulae of polygonal cells closely surrounded by network of thin-walled blood vessels. Tumor cells show mild to moderate nuclear atypia, round to oval nuclei, coarsely clumped chromatin, prominent nucleoli and large amount of granular eosinophilic cytoplasm. Few cells show eosinophilic whorled intracytoplasmic inclusions. Foci of necrosis are seen. Mitoses are occasional.

Status post right nephrectomy with malignant retroperitoneal mass in the right lumbar region on chemotherapy. Follow up scan. Comparison done with the previous CT scan dated 07/09/2020.

There is lobulated outline soft tissue attenuation retroperitoneal mass showing significant amorphous calcification seen in the right lumbar region with epicenter in the anatomical suprarenal region and it measures ~ 8.3 x 7.8 x 9.7cm (AP × TR x SI) in the present seen.

The mass shows heterogeneous contrast enhancement in the post contrast study.

There is severe compression of IVC noted. Loss of fat planes appreciated with the adjacent aortic segment. Loss of fat planes also appreciated with the right diaphragmatic crura. The duodenum is draped over the mass, however the fat planes with the duodenum are preserved. In comparison to previous CT study dated 07/09/2020, The mass shows mild size reduction.

Immunohistochemistry:

IHC Markers	Result
PAX-8	Negative
Inhibin	Negative
Ki-67	1-2 %
Chromogranin	Negative
Melan A	Negative
CK	Negative
Synaptophysin	Negative
S 100	Negative
Calretinin	Positive
BerEP4	Negative
WT-1	Negative
D2-40	Negative

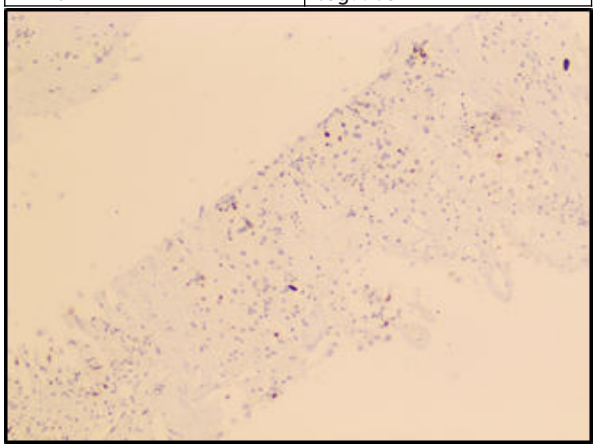

Figure.3 Ki 67 – 1%-2%

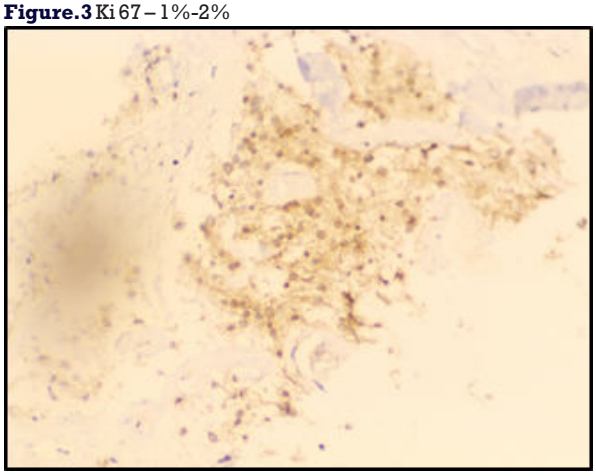

Figure.4 calretinin ihc 40x- Positive

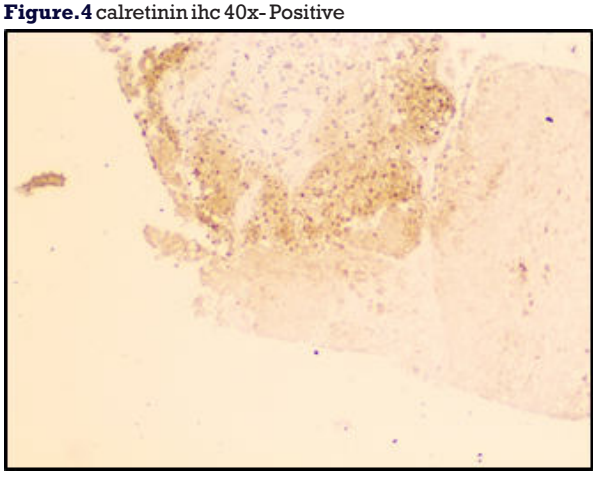

Figure.5 calretinin- Positive

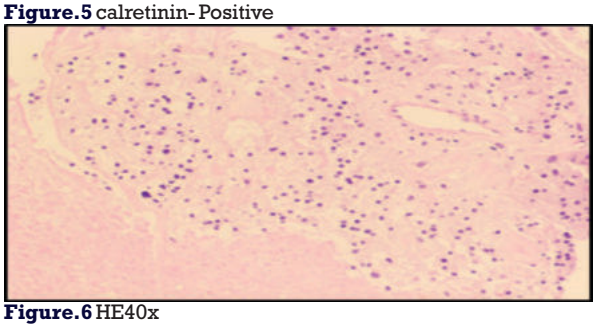

Figure.6 HE40x

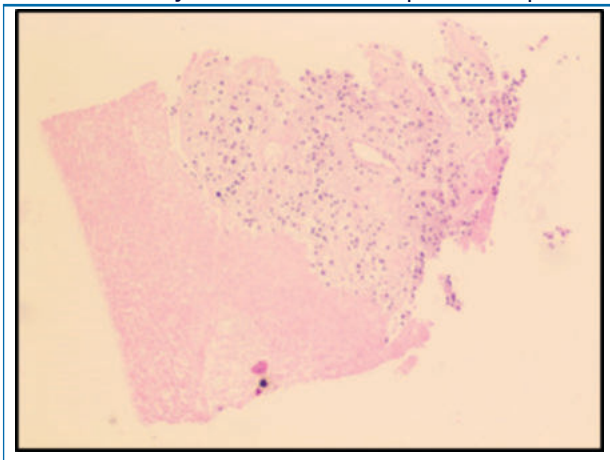

Figure.7 HE

Biopsy was done which had shown malignant neoplasm, Immunohistochemistry report has confirmed as Adrenocortical tumour. Melan-A diffuse moderate strong positive in tumour cells. After 3 years she has presented with recurrence, Biopsy has shown poorly differentiated tumour from Adrenal origin. Immunohistochemistry report shown Adrenocortical tumour Ki 67-1-2 calretinin positive.

DISCUSSION

Adrenocortical carcinoma is found to be more common in the Children (11) Similar to our case report Else and colleagues also reported unilateral involvement with the normal contra lateral adrenal gland and female preponderance (10) It is usually presented as large tumour more than 6cm in diameter (11) with internal haemorrhage, necrosis, and calcification, they have presented as bilateral disease in 2%-10% of the cases (12) The early detection of recurrence by clinical presentation on Imagining techniques is the Impact factor for prognosis and survival (13) Detection of early recurrence at the early stage may allow a treatment with the curative intent. The adjuvant therapies are advised to decrease the chances of recurrence. For unresectable or metastatic disease treatment is advisable with palliative purpose. The outcome after surgical resection has remained sub-optimal Studies suggested that the patients of adrenocorticoid tumour have high risk for tumour recurrence despite after complete surgical resection. Cytotoxic chemotherapy has a rate for advanced and metastatic adrenocortical carcinoma.

Initial studies in 1970 and 1980's shown less effective, Response rate with single regiment chemotherapy have shown 10% to 20% (10)

Recent phase III trial compared Doxorubicin, Cisplatin, mitotane versus streptozocin.

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