ejpmr, 2021,8(11), 420-422

SJIF Impact Factor 6.222

EUROPEAN JOURNAL OF PHARMACEUTICAL AND MEDICAL RESEARCH

www.ejpmr.com

Case Report

ISSN 2394-3211

EJPMR

ADRENOCORTICAL CARCINOMA- A RARE CASE REPORT

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Article Received on 27/08/2021 **Article Revised on 17/09/2021** **Article Accepted on 07/10/2021**

ABSTRACT

Adrenocortical carcinoma (ACC) is an uncommon and rare malignancy which arises from adrenal cortical cells. Half of the adrenocortical tumours are functional being associated with cushings syndrome or virilisation and rest of them present as a mass in abdomen or discovered incidentally during unrelated imaging procedures. We report a 46 year old female with complaints of heaviness and pain in abdomen. The patient had history of paroxysmal hypertension. The laboratory data showed no abnormality except for marginally raised Vannilyl mandelic acid levels. Ultrasonography Abdomen showed a solid echogenic mass measuring 8 x 5 x 5 cm in the right hypochondriac region in the upper pole in right kidney. The patient underwent nephrectomy with removal of adrenal mass and specimen sent for Histopathological examination. The diagnosis of adrenocortical carcinoma was made by morphology and was confirmed by Immunohistochemistry using specific markers.

KEYWORDS: Adrenocortical carcinoma, paroxysmal hypertension, Cushing`s syndrome, Mass in abdomen.

INTRODUCTION

Adrenocortical carcinoma(ACC) is a rare neoplasm, which arises from adrenal cortical cells and its incidence is about 0.5 to 2 cases per million population.[1] Bimodal distribution is noted with peaks in first and fifth decade with predilection for females, female to male ratio is2.5:1.[1] The patient presents with signs and symptoms related to hormone excess or non specific symptoms related to tumour growth and some are discovered incidentally. The 5- year survival rate varies from 16 to 85% of patients, however recurrence after complete resection is common and occurs in 23% to 85% of patients.[2] Death occurs in first two years of diagnosis.

CASE REPORT

A 46 years Female came with complaints of abdominal pain and fullness since 4 to 5 months to surgery OPD. Patient had a history of paroxysmal hypertension. On physical examination there were no features of virilisation or cushing`s syndrome. Abdominal examination showed a palpable mass in right lumbar region. Ultrasonography(USG) of abdomen was done which showed a solid echogenic mass measuring 8 x 5 x 5 cm in the right retroperitoneal region in the upper pole of right kidney, suggestive of malignancy. The value of vanillyl mandelic acid (VMA) in urine in 24 hrs sample was 9.1mg/24hrs (Reference range= Less than 8.0) and VMA to creatinine ratio was 10.53mg/gm creatinine (Reference range= Less than 8.0).The values were only marginally raised. The patient underwent nephrectomy with concurrent adrenalectomy. The specimen was sent to histopathology department.

GROSSLY nephrectomy specimen with adrenal mass was received. Kidney was unremarkable and adrenal mass measured 7.5 x 5x 4.4 cm. The tumour was partially capsulated with areas of capsular invasion, multinodular, with large areas of haemorrhage & necrosis.(Fig 1) Histologically- (Fig 2)The tumor was showing neoplastic cells arranged in nodules , cords and trabeculae. The individual cells showed mild to moderate pleomorphism, prominent basophilic nucleioli (Fuhrman grade III),bizzare nuclei, clear cells less than 25%,necrosis and mitoses. Mitosis were more than five per ten high power field. Tiny cystic ares, areas of necrosis, haemorrhage,capsular breech also seen (Modified weiss score-5) Immunohistochemistry(IHC) - IHC was done with markers (Fig 3) Synaptophysin, chromogranin, calretinin, S100 and Ki 67.

Tumor cells showed positivity with Calretinin (negative in pheochromocytoma) and negative for chromogranin A and S100 (both positive in pheochromocytoma), Ki67 showed more than 20% proliferaion index.

Above findings proved the morphological diagnosis of Adrenocortical carcinoma.

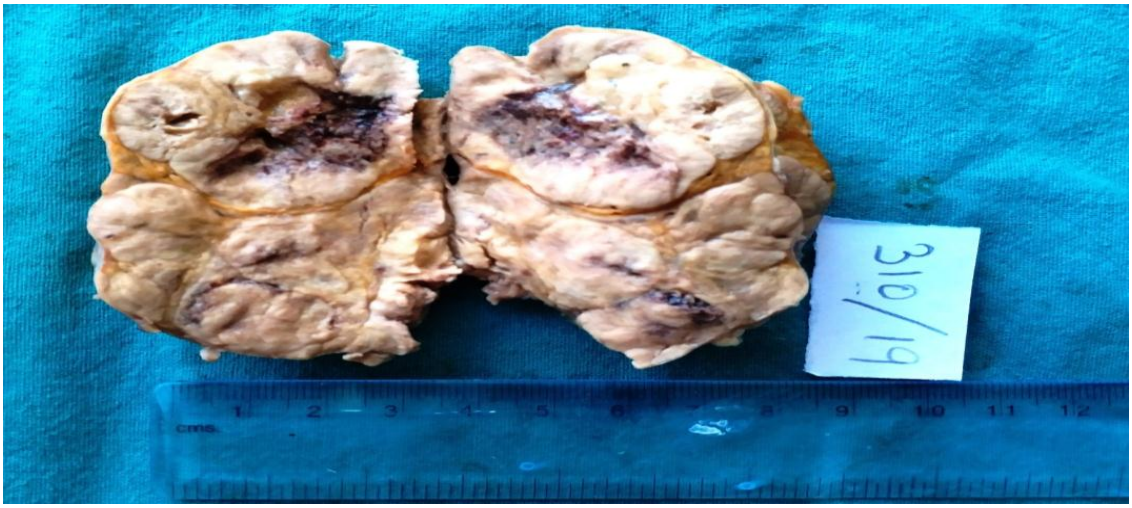

Figure 1: Gross.

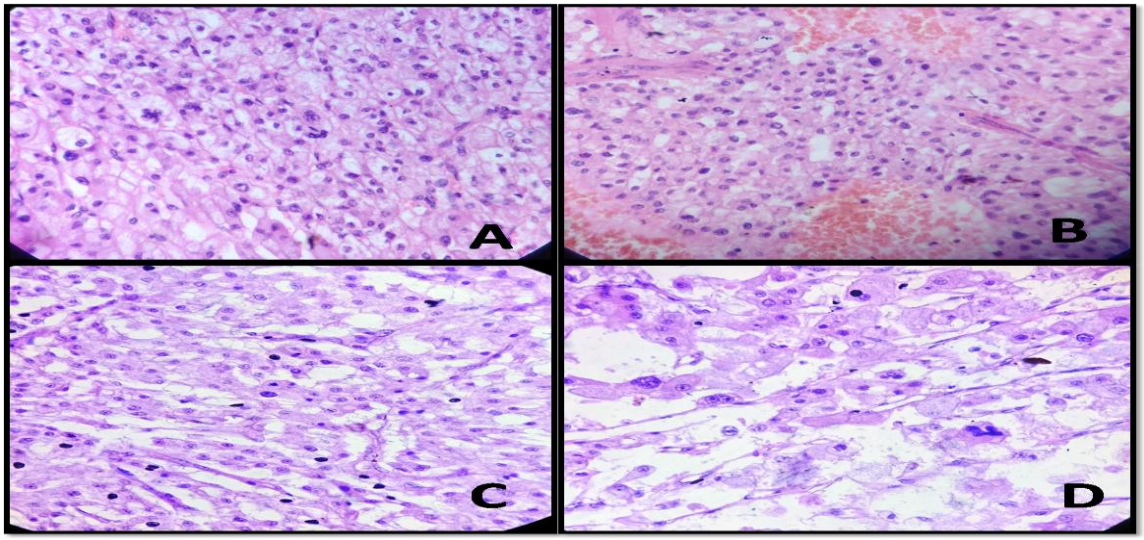

Figure 2: Adrenocortical carcinoma. A) Tumor cells showing irregular and pleomorphic nuclei B)Tumour showing haemorrhage C) Tumour cells arranghed in trabeculae and sheets D)-Mitotic figure(40X).

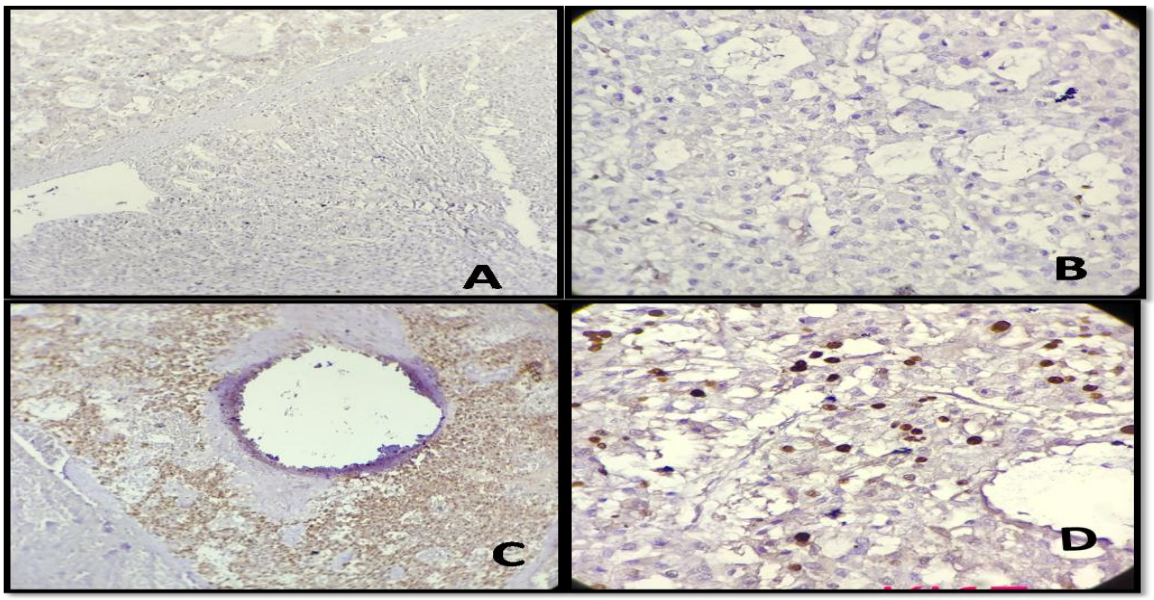

Fig. 3: Immunhohistochemistry A) Chromogranin A negative B) S100 negative C) Calretinin positive D)Ki 67 Index> 20%.

DISCUSSION

ACC is a rare neoplasm with grave prognosis and with an incidence of 0.5 to 2 cases per million population. Bimodal distribution is noted with peaks in first and fifth decade with predilection for females, female to male ratio is2.5:1.[1] The patient presents with signs and symptoms related to hormone excess or abdominal pain. 30% cases may also have palpable abdominal mass.[3] The differential diagnosis of adrenocortical carcinoma is Adrenocortical adenoma (ACA), renal cell carcinoma (RCC) and adrenomedullary tumours.

Pathologically malignant adrenal tumours differ from benign lesions by their gross characteristics of tumour weight, haemorrhages, capsular involvement and diagnostic score of Weiss based on microscopic parameters. The nine Weiss histologic criteria of malignancy to be assessed are nuclear grade, mitotic rate, atypical mitosis,<25% clear cells,>1/3 diffuse pattern, necrosis, venous invasion,sinusoidal invasion, capsular invasion.[4] Each Weiss criterion is scored 0 when absent and 1when present. Thus total Weis score lies between 0 to 9. A score of three or less is classified as ACA and those with four or more as ACC. In our case, we noted that the Weiss score was five-(nuclear grade Fuhrman grade III, Mitosis>5/10hpf, necrosis, Haemorrhage, capsular invasion) hence confirming as ACC.

The second differential diagnosis is pheochromocytoma. Features as zellballen pattern on histology, elevated VMA,IHC markers positive for chromogranin favour the diagnosis of pheochromocytoma.Our case showed Chromogranin A was negative, Calretinin was positive, S100was negative and Ki 67 Index >20%.These IHC findings confirmed the diagnosis of ACC.

Third differential diagnosis is renal cell carcinoma(RCC),which can directly infiltrate the adrenal gland or metastasize in it.Features suggestive of RCC are presence of glands,neoplastic cells positive for cytoplasmic glycogen with hameorrhage, IHC markers positive for Cytokeratin and EMA.

The primary treatment modality adrenocortical neoplasms is surgical excision.[5]

There is high rate of recurrence postoperatively. The patients usually die within two years of onset of symptoms.[6] There exists a correlation between microscopic degree of differentiation in carcinoma and survival.[7] Proliferative activity in form of mitoses or other means seem to be pathologic feature of greatest prognostic significance.[8,9,10]

CONCLUSION

ACC is an extremely rare tumour and it is essential to differentiate it from adrenocortical adenoma and pheochromocytoma by correlating with clinical,biochemical,imaging and histological features as they differ prognostically.

ACKNOWLDEGEMENT

None

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