VOLUME-8, ISSUE-11, NOVEMBER-2019 • PRINT ISSN No. 2277 - 8160 • DOI : 10.36106/gjra

Original Research Paper

General Surgery

A RARE CASE OF ADRENOCORTICAL CARCINOMA: A CASE REPORT

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ABSTRACT Adrenocortical carcinoma (ACC) is a rare, heterogeneous malignancy with a poor prognosis. ACCs may be hormone functioning and non-functioning , sign and symptoms develop depending upon the excessive hormone secreted. Surgical resection is the preferred treatment option . Presenting a 60 year old male patient with complaints of abdominal pain , weight loss , hypertension and hyperglycemia diagnosed as malignancy in adrenal gland in CECT abdomen , operated for right open adrenalectomy. Histopathological examination suggestive of : adrenocortical carcinoma

KEYWORDS : Adrenocortical Carcinoma , Malignancy , Surgical Intervention.

INTRODUCTION

Adrenocortical carcinoma is a rare disease (incidence 0.72 person per one million) in which malignant cells form in the outer layer of the adrenal gland (adrenal cortex).A tumor may be functioning (makes more hormones than normal) or may be non-functioning (does not make more hormones than normal). Most ACC are functioning and cause signs and symptoms according to the increase hormonal levels. Early diagnosis and removal of the tumor has better survival rate.

CASE REPORT

A 60 year old male patient was admitted with chief complain of abdominal pain and sudden weight loss in last one month and newly detected diabetes mellitus (RBS : 375) and hypertension (164/92).

INVESTIGATIONS

Routine workup revealed an unremarkable hemogram and serological investigations were within normal limits
HbA1C: 16.1
24 hrs urinary VMA levels: 1.6 mg/24 hrs (2.0-13.2)
S.Aldosterone level: <0.97 ng/dl (1.76-23.2)
S.cortisol: 17.93 (2.68-10.5)
Free metanephrine level: 1.20 pg/ml (0-65)

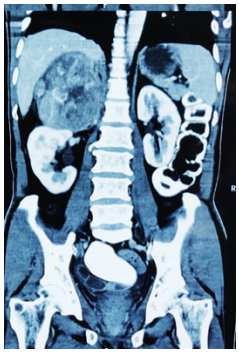
USG ABDOMEN :

Right kidney : 90 x 50 mm.
No evidence of hydronephrosis or calculus
Approximately 10x8x8 cm sized nixed echogenic lesion with internal vascularity in noted in right suprarenal region, superiorly it abuts segment VI of right lobe of liver with loss of fat plane , Anteriomedially the lesion abuts the infrahepatic IVC with preserved fat plane.
No evidence of hydronephrosis or calculus
Impression : Malignant mass arising from right adrenal gland.

TRIPLE PHASE CECT ABDOMEN AND PELVIS :

Large well defined heterogenous density lesion within right suprarenal region measuring 87 x 96 mm in axial plane with craniocaudal extension of 95mm. Right adrenal gland is not visualized separately. Lesion shows few internal hyper dense hemorrhagic areas. On contrast administration, lesion shows mild heterogenous enhancement with internal traversing pathological vessels, predominantly supplied by inferior adrenal artery. Lesion shows early washout with internal hypodense non enhancing necrotic area.

Superiorly : lesion abuts right lobe of liver and right hemi diaphragm
Anteriorly : abuts 1st and 2nd part of duodenum
Inferiorly : abuts cortex of upper pole of right kidney causing its caudal displacement. Lesion abuts hepatic flexure of colon
Anteromedially , abuts intra and infrahepatic inferior vena cava.

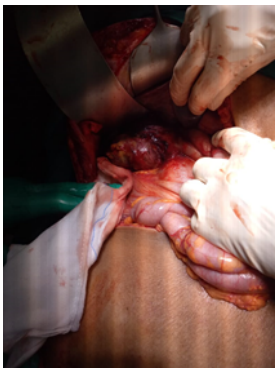


MANAGEMENT

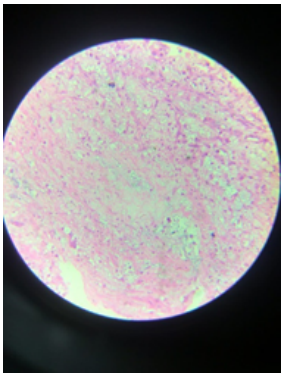
Patient was planned for exploratory laparotomy and right adrenalectomy.

After opening the peritoneal cavity , diffuse right adrenal mass of approx. 12x10 cm of size , irregular in shape abutting liver , IVC and duodenum. Mass was necrotic with friable consistency.
Patient was discharged on POD : 7 . Post op was uneventful

and patient was normotensive and euglycemic on discharge



HISTOPATHOLOGICAL EXAMINATION:
Adrenal cortical carcinoma of the right adrenal gland . T3NxMx



DISCUSSION
Adrenal cortex has 3 parts: Zona glomerulosa which secretes Aldosterone (Mineralocorticoids), Zona fasciculata which secretes Cortisol, Corticosterone and Cortisone (Glucoc orticoids), Zona reticularis which secretes Dehydroe piandrosterone (Androgen) . ACC mainly affects adults, children may be affected too. The median age at diagnosis is 46 years.

3 important prognostic factors:
- Completeness of resection
- Stage of disease
- Pathological grade

Most common sites of metastasis are the lung , liver , peritoneum and less commonly bones and major veins . Palliation of metastatic functioning tumors may be achieved by resection of both the primary tumor and metastatic lesions. Unresectable or widely disseminated tumors may be palliated by adrenolytic therapy with mitotane antihormonal drugs , systemic chemotherapy and/or radiation therapy . 5 year survival with stage 4 tumor is usually less than 20%.

Radical open surgical excision is the treatment of choice for patients with localized malignancies and remains the only method by which long term survival is approximately 38-46%
Various differential diagnosis of ACC includes adrenal adenoma, pheochromocytoma, neuroblastoma , ganglio neuroma , stromal neoplasm like lipoma or myelolipoma which can be differentiated on the basis of biochemical radiological and HPE investigations.

CONCLUSION
ACC is a very rare tumor of adrenal cortex and should be suspected in individuals with adrenal mass and newly detected hypertension, hyperglycemia and various other features due to increase hormonal levels. CECT abdomen pelvis revealing central areas of necrosis and hemorrhage with relative contrast retention and calcification points more towards ACC. Surgical resection remains the mainstay of treatment. Chemotherapy and radiotherapy to bone metastasis with surgical removal of localized functioning metastases may be attempted in Stage IV tumor but 5 year survival is less than 20 %. Although chemotherapy (mitotane) has shown disease free progression , but no overall survival have been demonstrated so far for Stage I and II disease.

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