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Case Report

Giant non-functioning adrenocortical carcinoma: an unusual tumour presentation

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ABSTRACT

Adrenocortical carcinoma is a rare tumour with incidence of 1 per million all over the world. Age distribution is bimodal with peaks occurring at 5-20 years and 40-50 years. Tumours greater than 9 cm commonly involve inferior vena cava (IVC) or right side of heart. 80 percent of the carcinomas are functional. We describe a case report of a rare tumour of a non-functioning adrenocortical carcinoma in a 40 year old female having incidence of 0.5/million world over with no extra-adrenal spread at presentation without IVC or right heart involvement in spite of having a large size of 12 cm. Patient underwent successful surgical extirpation with adjuvant radiotherapy with regular follow ups and disease free survival since the surgery. It is imperative to determine hormone levels in symptomatic and asymptomatic patients with adrenal masses. The possibility of adrenocortical carcinoma should not be ruled out in a functioning/non-functioning tumour. All solid incidentalomas on computed tomography (CT)/magnetic resonance imaging (MRI) greater than 5 cm in size should be removed surgically with adjuvant therapy consideration after histopathological reporting.

Keywords: Adrenocortical carcinoma, Non-functioning, Inferior vena cava

INTRODUCTION

Adrenocortical carcinoma (ACC) is a rare, aggressive malignancy with incidence of 0.7 to 2 cases per million annually. ACC presents across the age spectrum with peaks in children less than 5 years and in adults in their fourth and fifth decade of life. The prognosis is poor, as the majority of patients present with regional and distant metastasis at the time of diagnosis. Long-term survivors, however, are occasionally reported.[1] It is slightly more common in women than men. Given the advanced stage at diagnosis, the overall survival remains poor between 20 to 45%.[2] Adrenal masses can be broadly classified into hyperfunctioning (hormone secreting) and non-functioning (non-hormone secreting). Functioning ACC presents earlier with hormonal manifestations such as virilisation, feminisation or Cushing's syndrome. However, non-functioning tumours pose a diagnostic challenge because they are diagnosed incidentally due to a mass effect or metastatic disease. Moreover, successful radical management is seldom achievable because diagnoses are made when the tumour has either invaded the local structures or metastasised, explaining the poor prognosis associated with these masses.[3] We herein present a case of giant non-functioning adrenocortical carcinoma without any pressure effects which is a rare entity.

CASE REPORT

A 40 year old Indian woman presented with right flank pain since a year. The pain was intermittent, sharp, non-radiating and of moderate severity. The patient was vitally stable and non-hypertensive on presentation. On physical examination, minimal tenderness over right flank and a palpable right upper quadrant mass was present. Rest of

clinical examination revealed normal findings with no evidence of cushingoid features, hypertension or sex hormone excess seen.

An ultrasonography of abdomen and pelvis revealed normal size and cortical echogenicity of both kidneys with evidence of well-defined heterogeneous lesion seen superior to right kidney abutting right lobe of liver and showing minimal internal vascularity within it, measuring 12×10 cm with possibility of neoplastic aetiology of right adrenal gland.

A contrast enhanced computed tomography (CECT) of chest, abdomen and pelvis revealed a heterogeneously enhancing lesion with non-enhancing necrotic areas within and surrounding fat stranding in right adrenal gland with indentation of segment VI and VII of liver with maintenance of fat planes measuring 12×10×8 cm in size. There was mild right perinephric fat stranding. No other abnormality was noted in chest or abdomen .The above described lesion was most likely suggestive of adrenocortical carcinoma.

A functional adrenal work up was performed which included serum aldosterone, potassium, renin, metanephrine and adrenocorticotrophic hormone levels; a low dose dexamethasone suppression test, 24 hour urinary metanephrine, urinary potassium and vanillyl mandillic acid levels. All results were within reference ranges.

A metastatic work up included computed tomography of head chest and bone scan, all of which revealed no significant abnormality. En bloc excision of the mass was thus carried out.

Operative findings

A thoracoabdominal incision was used. After initial general inspection of the abdominal cavity, hepatic flexure of colon was mobilised by dividing the lateral part of the gastrocolic ligament and the peritoneal reflection over the ascending colon and duodenum was kocherised.

The liver was mobilised in order to facilitate dissection of tumour by dividing the falciform ligament and lateral triangular ligament. The tumour was visualised to be of size 12×10×8 cm with extension up to posterior aspect of liver and abutting of inferior vena cava (Figure 1). However, Gerota's fascia was intact and no liver and renal metastases were noted. No enlarged lymph nodes could be appreciated. Thus, right adrenalectomy was accomplished with paracaval and perihilar node clearance on right side. Appropriate closure with insertion of right abdominal drain and intercostal drainage tube on right side was done. Patient tolerated the procedure well and was shifted to intensive care unit.

In post-operative period, patient was given injectable antibiotic cover for two days followed by oral antibiotics for seven days. Immediate mobilisation and resumption of feeds was started on day 2. Abdominal drain was removed on post-operative day 3 and intercostal drain tube and urine catheter on day 4. Patient had minimal post-operative pain which was treated with non-opioid analgesics with monitoring of renal function. Patient was shifted to general ward on day 4 and discharged on day 6. Patient was followed up after 4 weeks and showed no postoperative complications. Patient was referred to radiotherapy department for further management.

Figure 1: Intra operative image showing adrenal mass.

Figure 2: Post mass excision image of adrenal fossa.

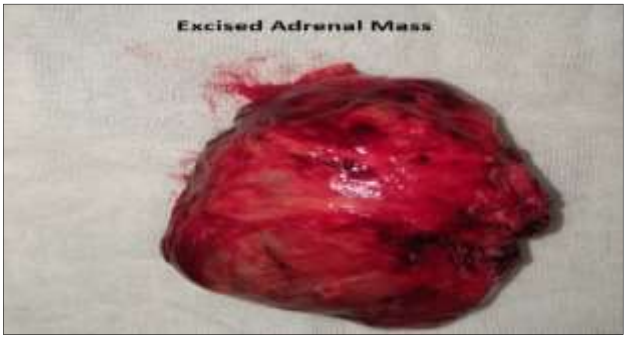

Figure 3: Excised adrenal mass.

Histological findings

Histopathological evaluation showed an adrenal mass of size 12×9×8 cm with attached fibrofatty tissue of size 6×4

cm. External surface was bosselated and cut surface showed few yellowish white areas on gross examination.

Microscopic examination showed adrenocortical neoplasm of polygonal cells with mild to moderate pleomorphic hyperchromatic nuclei focally prominent nucleoli and granular eosinophilic to clear cytoplasm. Clear cells accounted for <25% of tumour volume (2). Increased number of mitoses 30-38/50 hpf (2) and abnormal mitosis (1) were seen. Large areas of tumour necrosis were seen (1). No definitive evidence of capsular invasion was seen. Paracaval and perihilar nodes were free of tumour.

All these features were suggestive of adrenocortical carcinoma. Aubert et al modification of Weiss system score was 6.[4]

Immunohistochemistry panel

Immunohistochemistry testing showed positive result for vimentin, inhibin, tumor protein p53, neuron specific enolase and synaptophysin (weak) suggestive of adrenocortical carcinoma. However no definite comment could be made upon capsule infiltration.

Adjuvant radiation

Tumour bed radiotherapy 50.4 Gy in 24 fractions was given at 1.8-2 Gy per fraction five times a week for six weeks.

Follow up

Contrast enhanced computed tomography done after six months of surgical extirpation revealed few sub centimetre calcifications in operative bed significance of (s/o) post-operative status and patch of consolidation/round atelectasis with surrounding fibrosis in lateral segment of right lower lobe lung parenchyma s/o infective aetiology. This was managed with oral medications.

DISCUSSION

Non-functioning adrenocortical cancers are rare tumours (incidence 0.2 per million) that generally present late in their course.[2] Hormone secretion is not a distinguishing feature between benign and malignant masses. The silent clinical nature of this tumour results in poorer outcomes. Hence majority of non-functioning adrenocortical carcinomas present with locally advanced or metastatic disease.[1] Hormonal evaluation helps in identifying the functional status, origin of the tumour and helps in better perioperative management of the patient.[1,4] Tumours larger than 8 cm in size are seen to spread beyond confines of adrenal gland; however this was not observed in present case. Line of management of the tumours is determined preoperatively by McFarlane classification (Figure 4). In this case, McFarlane stage II is observed hence regional lymphadenectomy was done. Post-operatively, Aubert modification of Weiss system is used to determine malignant nature (Figure 5). In this case, score of six was seen signifying malignant nature of the tumour.

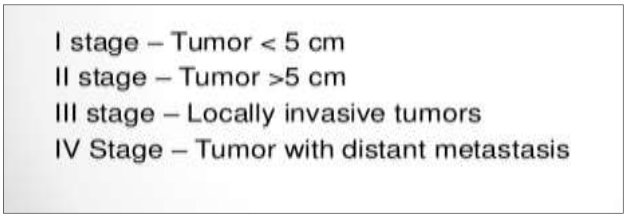

Figure 4: McFarlane classification of adrenal tumours.

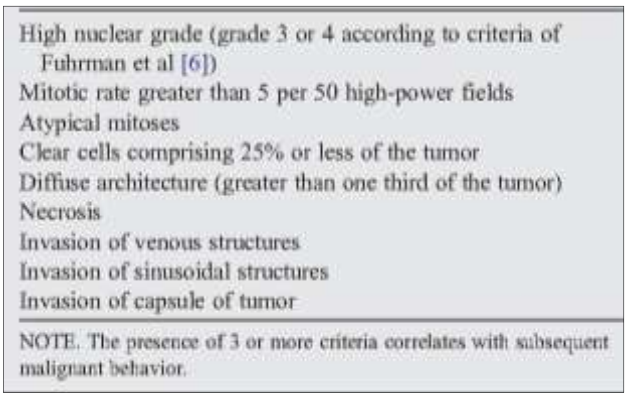

Figure 5: Aubert modification of Weiss classification of adrenal tumours.

CONCLUSION

Although surgical resection offers the only potentially curative therapeutic modality, less than half of these patients present at a stage where resection is possible for cure.[4] Adjuvant radiotherapy may significantly reduce the rate of local recurrence in patients with adrenocortical carcinoma.[5] An aggressive strategy for recurrent and metastatic ACCs is advisable. If the patient presents in an early stage and undergoes surgical excision the survival rate is increased.[6]

In summary, hormonal evaluation is a must in symptomatic and asymptomatic patients alike. The possibility of adrenocortical carcinoma should not be ruled out in functioning and non-functioning tumours. Thus, adrenalectomy for large masses (McFarlane stage II) must be followed with regional lymphadenectomy. After surgical extirpation proper follow up of histopathological reporting should be maintained for chemoradiotherapy consideration. Regular follow up for recurrence should be maintained every 3 months for 2 years followed by every 6 months for 5 years.[1]

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