Incidentally Discovered Nonfunctioning Adrenal Cortical Carcinoma: A Case Report And Review Of Literature

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Abstract

Adrenocortical carcinomas are extremely, rare, heterogeneous malignancies that arise from adrenal cortical cells and carries poor prognosis. We are presenting a case of highly progressive, metastatic adrenocortical carcinoma diagnosed incidentally during routine investigation procedure as a mass in right suprarenal gland .After thorough clinical, biochemical and radiological examination patient diagnosed as a case of Nonfunctioning Adrenal Cortical carcinoma of right adrenal gland with extension to renal parenchyma and peritoneal dissemination. Patient underwent surgical excision of mass, histopathology of the mass confirmed adrenocortical carcinoma. Patient was planned for combination chemotherapy with carboplatin, etoposide and adriamycin three weekly up to six courses.

INTRODUCTION

Adrenal cortical carcinomas (AC) are rare malignant tumor with an annual incidence of about 0.6-1.67 cases per million persons per year. [1] Overall, these tumors accounts for only 0.2% of the causes of deaths from cancer and due to this extremely low incidence rate, it is considered rare. [2] Clinically silent or biochemically and clinically asymptomatic adrenal masses also known as incidentaloma are incidentally discovered lesions, when noninvasive imaging methods such as USG, CT, MRI scan are performed for reasons other than known or suspected adrenal disease. [3] The detect ability of adrenal incidentaloma estimated in whole population at 0.1%; 0.41% in non – endocrine patients and at 4.3% in oncologically diagnosed ones. [4] The prevalence of tumors with a diameter > 1.5 cm is 1.8 % and with diameter > 6 cm is 0.025 %. [5]

CASE REPORT

A 39 year old male patient underwent ultrasound examination of abdomen for routine investigation procedure while complaining of weakness and loss of weight, diagnosed having a mass in right adrenal gland. Ultrasound (USG) examination showed a heterogeneous mass of size 4.1x 3.1 cm in relation to upper pole of right kidney posterior to inferior vena cava, suggestive of adrenal mass. Detailed history of the patient was taken and found there was no significant history of pain abdomen, diarrhea, vomiting, urinary incontinence, frequency, fever. There was no hypertension, diabetes mellitus, asthma and tuberculosis. No family history of malignancies detected. General and physical examination of the patient was normal. On local examination there was no mass detected in lumber region except tenderness. Blood investigations, liver function and renal function tests were within normal limits. Urine complete and microscopic examination was negative for albumin and sugar. Urinary vanillyl mandelic acid was 2.20 mg%. Serum aldosterone was 23.4 units. The results of pheochromocytoma work up were negative. On CECT scan a small enhancing (2x2.2 cm) lesion seen at the right suprarenal gland. A 3x4 cm enhancing mass lesion was seen to the midpole of the right kidney extending into the renal parenchyma and pushing vessels at the renal hilum, enhancing lesion with surrounding fluid 2.2x2.3 cm seen intraperitoneally into right iliac fossa. (Fig 1) The patient underwent exploratory lapratomy with excision of the mass.

Perioperative findings was 3 cm size adrenal tumor. Histopathological examination revealed a tumor measuring 3 cm weighing 200gm, adrenocortical carcinoma, revealing extensive areas of necrosis, high mitotic rate, and capsule invasion. (Fig 2) Peritoneal seedling also revealed infiltration by tumor. Diagnosis was made a tumor of adrenal cortex with metastases.

Figure 1

Figure 1: CECT scan showing an enhancing lesion at right suprarenal gland.

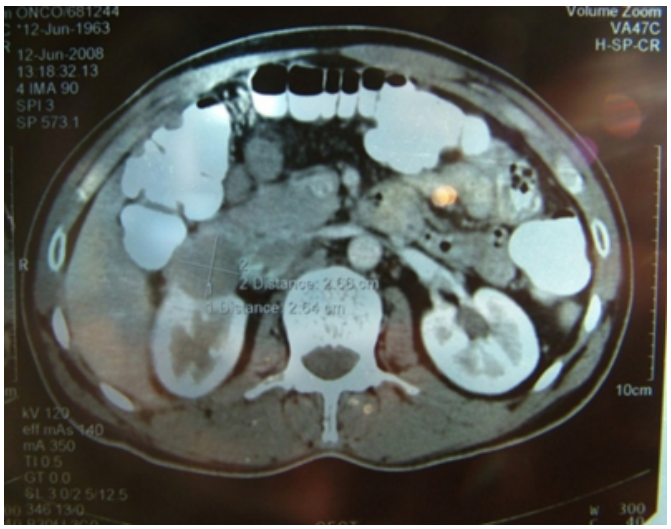


Figure 2

Figure 2: Photomicrograph showing atypical mitotic figures (400x H&E)

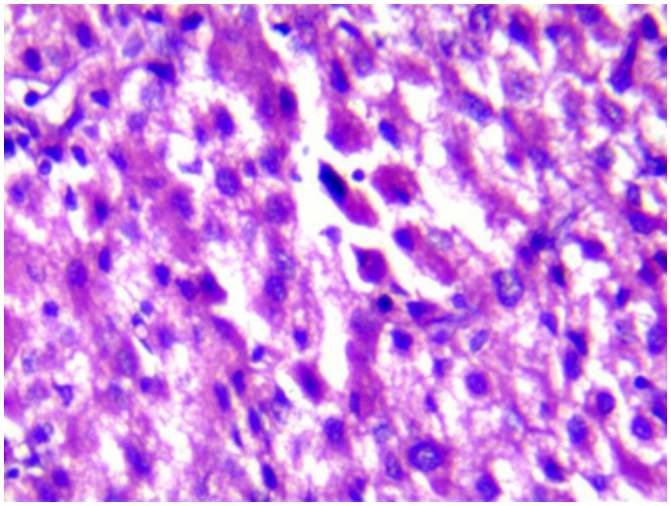


Figure 3

Figure 3: Photomicrograph showing large areas of necrosis (100x H&E)

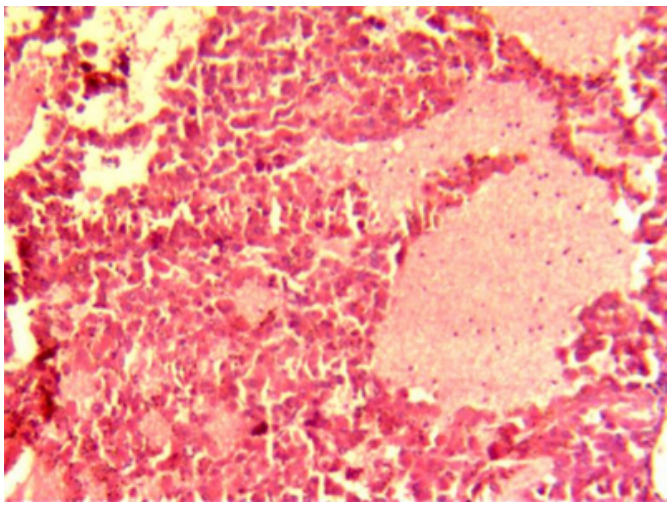
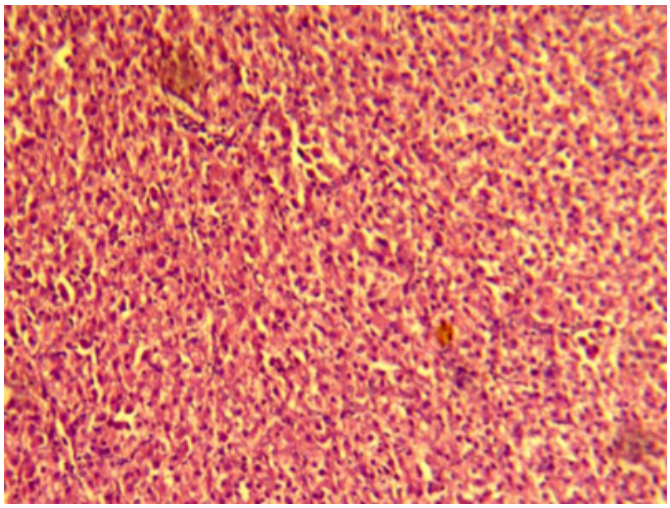


Figure 4

Figure 4: PhotomicrographShowinganaplasticcells(100xH&E)



DISCUSSION

Adrenocortical carcinoma is a rare, aggressive and heterogeneous tumor with poor prognosis. [6] There is a bimodal age incidence of these tumors, with the disease peaking in the first and in fourth to fifth decade of life. [7] ACCs may be classified in to functional and nonfunctional. Non functional ACCs are less common only 10% of masses exhibit evidence of hormone over activity. ACCs are slightly more common in females than males. Non functional carcinomas occur in an older age population (>30 years) and are more common in men (3:2 male to female ratio), while functional tumors are more common in women (7:3 female

to male ratio). [8] The present case is 39 year old male with nonfunctional ACC, correlates with data given in literature.

The risk of developing malignancy in adrenal incidentaloma is about 1/1,000. [9] Differentiating between adrenal adenoma and carcinoma in a small lesion can be challenging, although definitive diagnosis of malignancy in adrenocortical lesions is based on the presence of distant metastasis or local invasion. [10] Depending on these criteria present case diagnosed as in present case local invasion in renal parenchyma as well as disseminated peritoneal metastases were present.

The neoplasm was classified according to histologic criteria of Weiss. [11] Features associated with and increased probability of a malignant clinical behavior included tumor weight (> 400 g), tumor size (> 10.5 cm),vena cava invasion, severe nuclear atypia, >15 mitotic /20 high power fields and the presence of atypical mitotic figures, nuclear grade III or IV, clear cell comprising 25% or less of the tumor, a diffuse architecture, microscopic necrosis and invasion of venous, sinusoidal and capsular structures. Previously, the presence of 4 or more of these histologic findings was defined as indicative of malignancy, later it was modified to 3 or more. [11] In the presented case four criteria were present; high mitotic figures, capsular invasion, necrosis, a diffuse architecture.

Patients with nonfunctioning tumors presents with nonspecific symptoms related to tumor burden, including abdominal fullness, early satiety, pain, weight loss, weakness, fever, or an abdominal mass. [12] In other cases, the mass is found incidentally, during examination or radiologic imaging. As seen in present case.

The differential diagnosis of these adrenal masses include: cortical adenoma, pheochromocytoma, aldosteronomas, metastatic lesions, adrenal cortical carcinomas, and a host of non –neoplastic lesions. [13]

These tumors generally carries a poor prognosis, commonly related to delayed diagnosis, particularly in nonfunctional ACs, as a tendency to grow rapidly and 82% of the patients already had dissemination of the tumor at the time of diagnosis. [14] Common sites of distant metastasis included lung (71%), lymph node (68%), liver (42%), and bone (26%). [12,14]

Prognosis appears more favorable in patients with less than 50 years with localized disease, or non functioning status, complete tumor resection may be associated with improved survival. [15]

Complete surgical excision is the treatment of choice of ACCs . [16] Radical surgical resection of adrenocortical carcinoma offers the best chance for prolonged recurrence free survival. Recurrence of adrenal cortical carcinoma after radical surgery is a common finding although successful reoperations have been reported with encouraging results, most published expressions are anecdotal. [17] Unresectable or widely disseminated tumors may be palliated by antihormonal therapy with mitotane, systemic chemotherapy, or (for localized lesions) radiation therapy. Although the survival for patients with stage IV tumors is usually less than 9 months. [18,19,20] Palliative chemotherapy with cisplatin-based regimens has produced objective responses in approximately 30% of patients treated. [21,22] Decker et al in a study reported that doxorubicin produced objective responses in 3 of 16 patients with poorly differentiated, nonhormone-producing tumors but no responses in 15 patients whose disease did not respond to mitotane. [23] Combination of platinum-based chemotherapy and mitotane achieved a 48.6% objective response and median time-to-progression of 18 months in responders and in 10 of 72 patients subsequent was possible. [24] Depending on these recommendations we gave combination chemotherapy included cisplatin, adriamycin and etoposide three weekly. Patient received six cycle of combination chemotherapy and responded well and there is a marked regression in mass.

ACC is a rare and highly malignant tumor with poor prognosis probably due to presentation with advanced and metastatic disease. Surgical resection remains the principal treatment for stage I to III disease. Adjuvant treatment may improve survival in patients with stage III or IV disease. An aggressive strategy for recurrent and metastatic ACCs is advisable. Although improved survival is seen in patients who present with early stage and have complete primary resection.

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