(International Print/Online Journal)

SJIF IMPACT FACTOR: 5.565
PUBMED-National Library of Medicine ID-101739732

ISSN (Print): 2209-2870
ISSN (Online): 2209-2862

International Journal of Medical Science and Current Research (IJMSCR)
Available online at: www.ijmscr.com
Volume 6, Issue 4 , Page No: 152-55
July-August 2023

IJMSCR

Adrenocortical Carcinoma Masquerading As Pheochromocytoma: A Case Report

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Type of Publication: Case Report
Conflicts of Interest: Nil

Abstract

Adrenocortical carcinoma (ACC) is a rare and aggressive disease with an incidence of approximately one case per million.The diagnosis of ACC is often challenging and is based on careful investigations of clinical, biological and imaging features before surgery and pathological examination after tumor removal. We report a case of 56 year old female with adrenal cortical carcinoma which was diagnosed as pheochromocytoma clinicoradiologically.

Keywords: Adrenalectomy , Adrenal tumor, modified weiss criteria immunohistochemistry

Introduction

Adrenal cortical carcinoma (ACC) is a rare cancer and is the most common primary carcinoma of adrenal gland[1].The reported annual incidence is 1 case per million in united states according to a recent study done in united states[2].The previous reported annual incidence was 0.5 -2 cases per million with female preponderance[3].The diagnosis of ACC is often challenging and is based on careful investigations of clinical, biological and imaging features before surgery and pathological examination after tumor removal[4].

Case Report

A 56 year female presented with abdpmonal pain since 2 months along with headache and palpitations . CT scan abdomen showed a left suprarenal mass. Serum creatinine, 24 hour urine vanillylmandelic acid and metanephrine levels were raised.

A clinical diagnosis of pheochromocytoma was made and patient underwent left adrenalectomy which was sent for histopathological examination. We received an adrenal mass measuring 9.5 x 7.5 x 5.5 cm. Outer surface was encapsulated and cut section showed a solid grey brown mass with areas of necrosis and hemorrhage (Figure 1). On microscopic examination , tumor cells arranged in sheets, cords, trabeculae and focal alveolar pattern showing pleomorphism, high N/C ratio, coarse granular chromatin, intranuclear pseudoinclusion and moderate to abundant eosinophil granular cytoplasm separated by fibrovascular septa (Figure 2A). Mitotic activity (6/50 hpf) was increased. Capsular , sinusoidal and lymphovascular invasion (Figure 2B) was present along with areas of necrosis.< 25 % clear cells and multinucleated cells seen (Modified Weiss score of 6). IHC revealed positive Melan A, calretinin (Figure 3), inhibin, vimentin, synaptophysin and negative chromogranin, and NSE. A final diagnosis of adrenocortical carcinoma of low grade was established.

FIGURE 1: Gross image showing encapsulated tumor with solid grey brown appearance

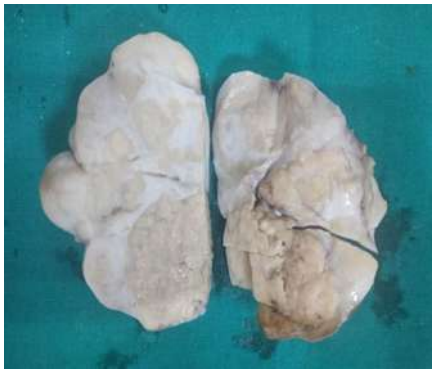


FIGURE 2: Photomicrograph showing sheets and trabeculae of tumor cells showing pleomorphism, high N/C ratio, coarse granular chromatin and moderate to abundant eosinophil granular cytoplasm separated by fibrovascular septa with sinusoidal invasion in 2B

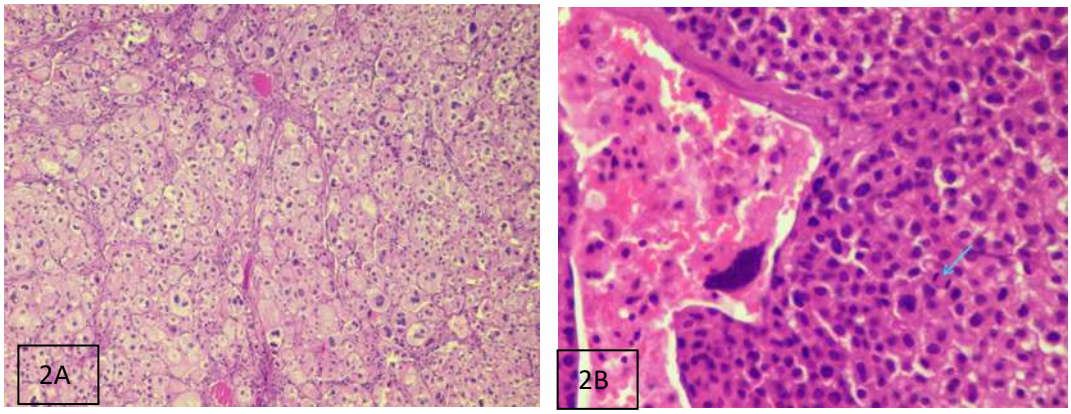
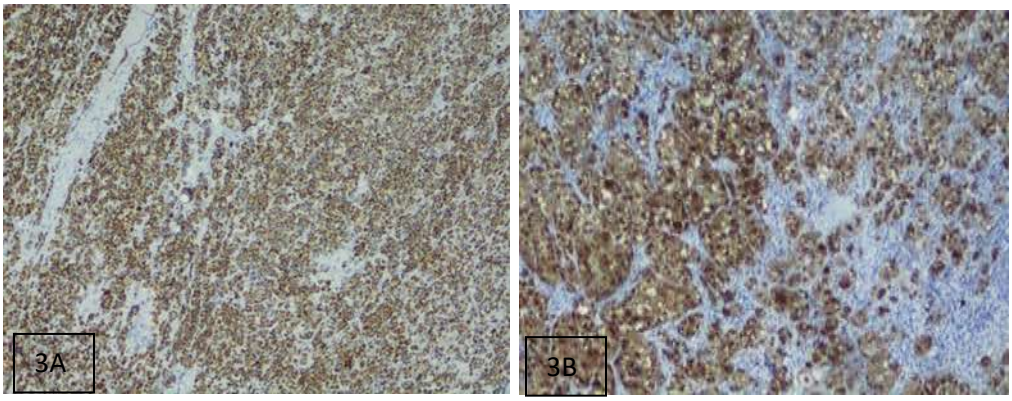


FIGURE 3: Photomicrograph showing cytoplasmic positivity in tumor cells with Melan A (3A) and calretinin (3B)



Discussion

Adrenocortical carcinoma (ACC) is a rare and aggressive disease with an incidence of approximately one case per million[2]. About 60 % are functional tumors secrete hormones and present with clinical features like Cushing's syndrome due to cortisone, virilizing tumor due to androgens, or feminizing tumor due to estrogens[5]. Like all other endocrine tumors, ACC is more common in females with a female to male ratio of approximately 1 to 1.4

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with an average of presentation ranging from 47 to 55 years[2,6].

ACC is often noted on left adrenal (left to right ratio = 1.2 to 1). Bilateral ACC is uncommon and accounted for approximately 1% of the cases[2]. Ectopic ACCs have been reported in the retroperitoneum[7],ovary[8,9] , spinal region[10], liver[11]and abdominall wall[12].

On gross examination, ACC is often large in size with maximum diameter of around 10 to 12 cm. Cut section of the tumor is yellow to tan in color usually. Areas of hemorrhage and necrosis are commonly seen [2].

On microscopic examination, tumor cells of ACC often has eosinophilic cytoplasm. There are thick fibrous bands and capsules with prominent mitotic figures . Necrosis is common in most of the tumors. ACC is subdivided into low grade or high grade depending on mitotic frequency (low grade being ≤20 and high grade >20 mitosis per 50 high power fields/10 mm2) [13]. The most widely accepted system adopted by WHO classification to differentiate ACC from benign cortical adenoma or borderline cortical tumor is Weiss criteria published in 1984 [13]. ACC could be diagnosed on at least three of the 9 histological features—high nuclear grade (Fuhrman III or IV), high mitotic rate (>5 mitoses per 50 high power field, atypical mitotic figures, 25% clear cells, diffuse architecture, tumour confluent necrosis, venous invasion, sinusoidal invasion, and capsular invasion. Modified Weiss has been proposed which is based on 5 of the 9 histological features of Weiss [14]

These systems are helpful in predicting malignant potential of adrenocortical tumours mainly in conventional ACC. Studies have proposed the use of proliferative index (Ki-67 index > 5%) [15] and IGF2 over-expression to confirm the diagnosis of ACC.

In a biopsy specimen, it is important to differentiate ACC from metastatic carcinoma and phaeochromocytoma by clinical history, biochemical studies, radiology, and a panel of immunohistochemical stains [16]. ACC expresses markers specific for steroid producing cells which often include steroidogenic factor-1 (SF-1) and inhibin alpha . The tumour is also positive for markers expressed by other tumour types, such as melan A and calretinin.Common epithelial markers such as cytokeratin, EMA, CEA are generally negative.

Although ACC could be positive for synaptophysin, it is negative for chromogranin.

Prognosis of the conventional ACC depends on the cancer stage but overall the prognosis is poor.'

The 5-year cancer specific survival rate of patients with ACC was 38% as noted from USA SEER database [1]. It is worth noting that surgery on theprimary site even in metastatic ACC significantly improved overall and cancer-specific survival of patients with ACC [17].

Conclusion

ACC continues to be a rare malignancy worldwide with an annual incidence of 1 case per million populations. However, most cases continue to be diagnosed only in advanced stages and are associated with poor survival. These findings underline the need for specific diagnostics tools with new and more effective treatment for adrenocortical cancers.

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