# Adrenocortical carcinoma with metastasis in abdominal wall: A rare case report

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#### **ABSTRACT**

Carcinoma of the adrenal cortex is a rare malignancy and account for only 0.05–0.2% of all cancers. It has a slight predilection for female patients. It has a poor prognosis due to aggressive behavior. Here, we report a case of a male patient who suffered from adrenocortical carcinoma with metastasis in the abdominal wall. To the best of our knowledge, this is the first case where we have noticed metastasis in the abdominal wall. This case is presented for of its rarity.

**Key words:** Abdominal wall, adrenal cortex, adrenocortical carcinoma, metastasis

#### INTRODUCTION

Adrenocortical carcinoma is a rare malignancy with an estimated incidence of 0.5–2 cases/million persons yearly.<sup>[1]</sup> There is a bimodal age distribution with peaks in the first and fourth to fifth decades of life.<sup>[2,3]</sup> About 60% are functional tumors that secretes hormones and present with features with Cushing's syndrome due to cortisone, virilizing tumors due to androgens or feminizing tumors due to estrogens.<sup>[4]</sup> At the time of diagnosis about one-fifth of the adrenocortical carcinoma have already spread locally and about half of the patients present with metastasis.<sup>[4]</sup> The most common sites for distant metastasis are the lungs, liver, lymph nodes, and bones. In our case, the patient of adrenal carcinoma had metastasis in the abdominal wall, which is not yet mentioned in the literature. The prognosis of adrenocortical carcinoma is variable but generally poor.

#### CASE REPORT

A 65-year-old male came with complaints of pain in the abdomen in the right lumbar region since 6 months on

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examination the abdomen was soft, tender, and right suprarenal mass was vaguely palpable. There was no organomegaly. The vanillylmandelic acid level was 11.3 mg/24 h (normal 1.6–13.2), and metanephrine levels were 173  $\mu$ g/dl (0–350) which were within normal limits.

Chest X-ray was normal. Ultrasonography abdomen showed right suprarenal large solid hypoechoic slightly lobulated mass lesion. Inferior vena cava is slightly compressed computed tomography (CT) scan of abdomen revealed a large heterogeneously space occupying lesion in the right suprarenal area showing peripheral enhancement with internal nonenhancing areas causing compression of inferior vena cava and displacing the right kidney inferiorly [Figure 1].

Gross: Received a globular circumscribed tissue mass of  $10 \text{ cm} \times 7 \text{ cm} \times 4 \text{ cm}$ . Cut surface of the tumor was soft with a yellowish tan appearance. We also received another single tissue piece of  $0.5 \text{ cm} \times 0.4 \text{ cm}$ , which was labeled as abdominal wall nodule [Figure 2].

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Microscopic examination showed tumor tissue composed of large polygonal cells with hyperchromatic nuclei and eosinophilic cytoplasm at places showing clear cytoplasm. The cells are arranged in nests sheets and trabeculae. Mitotic activity, tumor giant cells, and areas of necrosis seen. The capsular invasion also seen. Stroma is scanty and delicate [Figure 3]. The specimen labeled as abdominal wall nodule also showed similar tissue [Figure 4]. A diagnosis of adrenocortical carcinoma with metastatic deposits in the abdominal wall was made.

### **DISCUSSION**

Adrenocortical carcinomas are rare and lethal neoplasm with the incidence of 0.5–2 cases/million person's yearly.<sup>[1]</sup> With a prevalence of about 3% in the population aged over 50 years.<sup>[2,3]</sup> An adrenocortical carcinomas more often occurring woman than in men (ratio 1.5:1).<sup>[5,6]</sup> This tumor is associated with several genetic syndromes such as

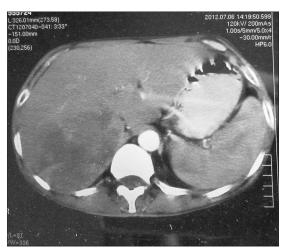


Figure 1: Computed tomography abdomen revealed a large heterogeneously space occupying lesion in the right suprarenal area

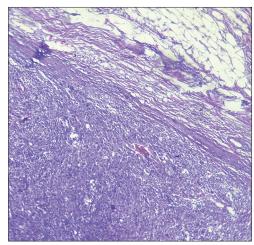


Figure 3: Tumor tissue composed of large polygonal cells with hyperchromatic nuclei and eosinophilic cytoplasm at places showing clear cytoplasm. The cells are arranged in nests sheets and trabeculae

multiple endocrine neoplasia type I, Beckwith–Wiedemann syndrome, Li–Fraumeni syndrome.<sup>[5]</sup> Adrenocortical carcinomas are usually diagnosed late with about half of the patients revealing metastasis when they are diagnosed<sup>[4]</sup> and the average size of the tumor is more than 12 cm.<sup>[7]</sup> About 62% of adrenocortical carcinomas are functioning and 38% are nonfunctional hormonal syndrome including feminization or mineralocorticoid excess syndrome will occur with functioning Adrenocortical carcinoma.

The diagnosis of adrenocortical carcinoma depends on clinical suspicion and diagnostic imaging studies. CT scan is considered the diagnostic study of choice for evaluating the adrenal masses. The diagnosis between benign and malignant adrenal masses is difficult. Macroscopically a weight of more than 500 g, a grossly lobulated cut surfaces, necrotic areas calcifications, and intratumoral hemorrhage have been used to predict the presence of malignancy. [8,9] The microscopic diagnostic Weiss score is the most widely used tool, features include a high nuclear grade, mitotic rate more than 5/50 HPF, atypical mitotic figures, necrosis,



Figure 2: A globular circumscribed tissue mass of 10 cm × 7 cm × 4 cm. Cut surface soft with a yellowish tan appearance

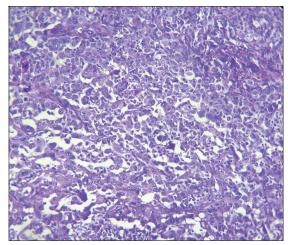


Figure 4: Abdominal wall nodule shows same tumor tissue

invasion of venous structures, sinusoidal structures, and capsular invasion. [9,10] In this case, five criteria of Weiss score was fulfilled, and diagnosis of metastasis adrenal carcinoma to the abdominal wall was made.

The most common site for distant metastasis is the liver (48–85%), lungs (39–60%), lymph nodes (7–29%), and bones (7–13%).<sup>[7]</sup> In this case, patient had metastasis in the abdominal wall.

Surgery is the treatment of choice and provides the only chance for cure. However, aggressive behavior of this cancer limit the cure rate and medical therapy with mitotane plays an important role in cases in which surgery is not suitable, and it is effective in treating patients with metastatic adrenocortical carcinoma.<sup>[7]</sup> Adrenocortical carcinomas are relatively radioresistant therefore radiotherapy is usually reserved for palliating bone metastasis and for unresectable local recurrence.[11] After curative resection two-third of patients experience recurrence within on to 2 years, but because some patients have recurrence many years later, long-term follow-up is needed. [4] The prognosis of adrenocortical carcinomas is poor. With an overall 5 years survival rate of 15-47%. The 5 years survival rate decrease from 30% to 45% in stage I, 12.5-57% in stage II, 5-18% in stage III, and 100% in the stage IV tumor.[11] When the metastatic disease is present at initial presentation death usually occurs within 1 year.[12]

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#### **Conflict of interest**

There are no conflict of interest.

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