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## CASE REPORT

# Giant adrenal tumor presenting as Cushing's syndrome and pheochromocytoma: A case report



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### KEYWORDS

Giant adrenal tumor;  
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**Abstract** We report a case of a 35-year-old lady who presented with Cushingoid features and associated raised urinary metanephrine. The patient underwent open adrenalectomy. Histopathological examination revealed adreno-cortical carcinoma with microscopic lymphovascular invasion. Postoperative period was uneventful and is on follow-up for the last one year and is doing well.

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## 1. Introduction

Adrenal carcinoma is an aggressive malignant neoplasia arising from the adrenal cortex with poor prognosis. It represents 0.02% of all neoplasia. Global incidence is 0.5–2 per every 1,000,000 inhabitants [1]. The age distribution is reported as bimodal with a first peak in childhood and a second higher peak in the fourth and fifth decade [2,3]. Women are more often affected than men in the ratio of 1.5 [3–5]. Tumors are classified as functioning when they

are associated with endocrine manifestations or elevated hormone levels. Cushing's syndrome is due to hypercortisolism while pheochromocytoma is a catecholamine secreting tumor of the adrenal medulla or extra adrenal sites. Adrenal carcinoma accounts for approximately 33%–53% of cases of Cushing's syndrome [6,7]. However, it is rare for adrenocortical carcinoma to present clinically as pheochromocytoma. We report a case of a 35-year-old lady who presented with Cushingoid features and associated raised urinary metanephrine.

## 2. Description of case

A 35 years old female presented with the chief complaint of altered menstrual symptoms for the last 10 months. She

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also complained of dull aching pain in the left flank without any radiation or shifting and frequent episodes of generalized headache, palpitation and anxiety for the last 5 months. The palpitation was abrupt in onset and lasts for about 30 min to 1 h, occurs 4–5 times per week, and was associated with day-to-day household activities. The patient gradually developed swelling of both her lower limbs and she had great difficulty in getting up from the squatting position. On clinical examination, she was found to have chemosis with swelling of eye lids, flushing of face, increased facial hair distribution, hypertension, centripetal obesity, and bilateral pedal edema. On examination of her abdomen, a firm mass at left hypochondriac region approx 10 cm × 7 cm in size was felt with smooth surface and well defined margins, and fingers could not be insinuated below the costal margin. Laboratory work-ups including full blood count, renal function tests, serum electrolytes, and liver function tests were within normal limit. Serum cortisol [morning – 31.79 µg/dL (normal: 4.30–22.40), evening – 32.73 µg/dL (normal: 3.09–16.66)] and 24 h-urine norepinephrine – 117.21 µg per 24 h (normal: 12.10–85.50), dopamine – 592.82 µg per 24 h (normal: 52.00–480.00) levels were raised. CT scan revealed a 18.3 cm × 12 cm × 16 cm left sided hypervascular retroperitoneal mass without any invasion of the adjacent organs and showing focus of calcification and microscopic fat component (Fig. 1). Preoperatively, the patient was placed on  $\alpha$  blockers (Tab. Prazosin 5 mg at bedtime) for the effective management of blood pressure. After effective stabilization of blood pressure, she was taken up for exploration. On exploration of abdomen, a left adrenal mass of approx 21 cm × 12 cm × 8 cm in size, fixed to left kidney with evidence of local invasion was found (Fig. 2). The left adrenal vein was isolated and suture ligated before attempts were made to dissect out the adrenal mass. The mass weighed 380 g (Fig. 3). The capsule of the mass was intact with hemorrhagic and necrotic areas seen at places on cross section. Intraoperatively there were fluctuations of blood pressure, which was managed effectively with Nitroglycerine infusion. Histopathological examination revealed

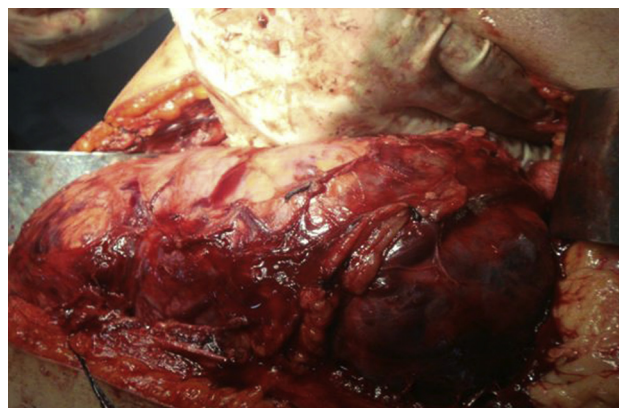


Figure 2 Intraoperative view.

adreno-cortical carcinoma (Mitotic rate 60/50 high power field) with microscopic lymphovascular invasion and invasion limited to the capsule and small vessel. There was extensive tumor necrosis with hemorrhage and calcification (Fig. 4). However, no microscopic features suggestive of pheochromocytoma seen on histopathology of adrenal medulla. Postoperative period was uneventful and is on follow-up for the last one year and is doing well.

### 3. Discussion

Adrenocortical carcinoma is itself a rare disease, of which functional adrenocortical carcinoma accounts for 50%–79% of cases [8]. Rapidly progressing Cushing's syndrome with or without virilization is the most frequent presentation [9]. Adrenocortical carcinoma can present with dysfunctional uterine bleeding in women due to increased amounts of androstenedione and estrogens [10]. However, adrenocortical carcinoma presenting with features of pheochromocytoma alone is a rare entity and it is rarest to have features of both Cushing's syndrome as well as pheochromocytoma in the same patient with adrenal tumor. Despite extensive PubMed search, no reports of the existence of an adrenal tumor presenting with both features of Cushing's syndrome as well as pheochromocytoma has been found till date.

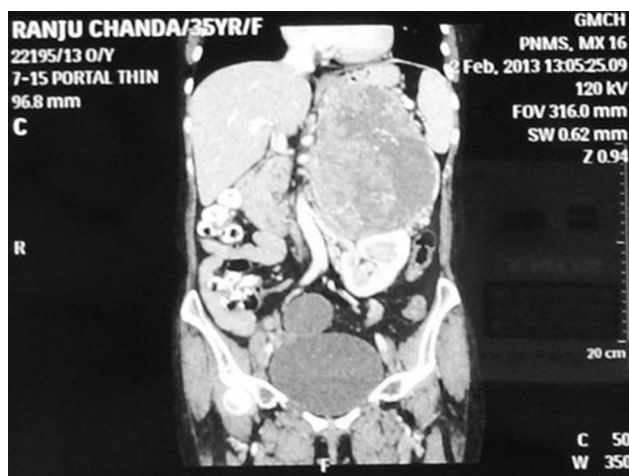


Figure 1 CT scan abdomen (coronal section) showing the adrenal gland.

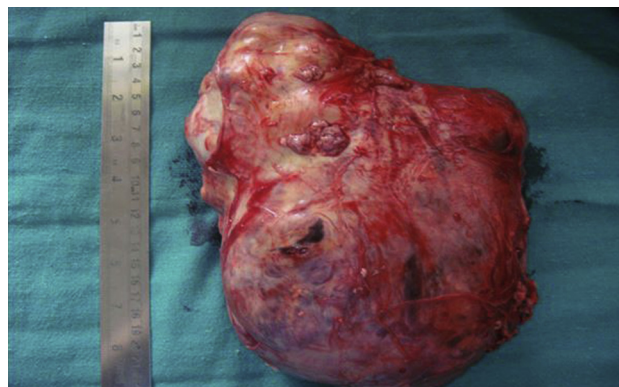
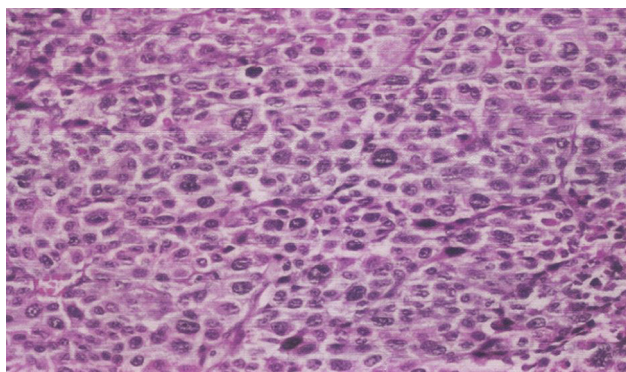


Figure 3 Specimen.



**Figure 4** Histopathological report of the specimen (40× magnification; Haematoxylin and Eosin stain).

#### 4. Conclusion

A functional giant adrenocortical carcinoma with features of both Cushing's syndrome and pheochromocytoma is a rare entity. Surgical extirpation is a good management option for giant, resectable adrenocortical carcinoma. Precise preoperative work-up and cautious pre-, peri- and postoperative management for the functional component is of utmost importance. A further long stringent follow-up will throw light into the behavior of this entity.

#### Conflicts of interest

The authors declare no conflict of interest.

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