Unusual large functional adrenocortical carcinoma in a young woman

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DESCRIPTION

A 27-year-old woman presented with an abdominal lump and features of Cushing syndrome with virilisation, namely, weight gain, acne, hyperpigmentation, hirsutism, central obesity, rounded face, stretch marks, osteoporosis, dyslipidaemia, hypertension and amenorrhoea (figure 1). Serum testosterone and cortisol levels were elevated. Oestradiol

and 24 h urinary metanephrine were within normal range. contrast-enhanced CT of the abdomen showed a right supra renal mass measuring $9.5 \times 8.3 \times 5.5$ cm (figure 2). The clinical diagnosis was adrenocortical carcinoma (ACC). The left adrenal gland was normal. Per operatively, the tumour was free from the kidney and entire tumour was removed intact (figure 3).

Figure 1 (A) Contrast-enhanced axial CT image showing a large right suprarenal mass with central area of necrosis and haemorrhage. (B) Contrast-enhanced coronal (CT) image showing right suprarenal mass measuring $9.5 \times 8.3 \times 5.5$ cm displacing the right kidney inferiorly and liver superiorly with irregular enhancement, necrotic areas with cystic degeneration and peripheral rim of calcification. (C) Contrast-enhanced coronal CT with angiogram showing multiple feeding vessels from the right renal artery and aorta.

Figure 2 (A–D) The patient with round face, central obesity, acne, hirsutism, hyperpigmentation and stretch marks.

Figure 3 (A and B) Clinical photograph of adrenal gland after excision (gross specimen).

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Histopathology showed a tumour with nests and trabeculae. A few bizarre cells with hyperchromatic nuclei, calcification, mitoses (>25 per high power fields) and necrosis were present. A focal capsular breach was identified. Immunohistochemistry was positive for vimentin, melan-A and inhibin, and negative for cytokeratin and chromogranin, consistent with ACC. The patient was doing well 6 months after surgery, with no features of recurrent/residual disease.

ACC is an uncommon aggressive neoplasm having an incidence of about 1–2 cases per million population.¹ It has bimodal age distribution with peak in the first decade of life in children and in fourth to fifth decades in adults. However, ACC most commonly occurs in adults and is slightly more common

in females than in males.¹ About 60% of these produce hormones that can lead to Cushing's syndrome (45%), mixed Cushing's and virilisation syndrome (25%), or virilisation alone (<10%).² Virilisation occurs more frequently in children and indicates malignancy in 70% of cases. Surgery is the best treatment modality. The overall 5-year survival rate varies from 16% to 38%.³ Recurrence is common even after complete surgical removal, occurring in 23–85% of patients.³ Death usually occurs within the first 2 years.

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Learning points

- ▶ Increasing adrenal tumour size correlates significantly with risk of malignancy.
- ▶ At the time of presentation, the majority of adrenocortical carcinomas will be functional.
- ▶ Adrenocortical carcinoma should be managed aggressively, as it has a high recurrence rate and poor prognosis.

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