

A case of adrenocortical carcinoma causing mineralocorticoid and glucocorticoid excess

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Initial presentation and investigations

A 68 year old farmer presented to a local hospital with hypertension and hypokalaemia. He had a past medical history of non ST elevation myocardial infarction following which he was treated with antihypertensive agents. These were withdrawn and supine and ambulant aldosterone levels were raised (supine: 1900pmol/l (100 – 400), ambulant: 2000pmol/l (100 – 800)). Plasma renin was normal. CT scan of abdomen showed a 5.9 x 5.3 cm left adrenal mass.

Further investigation

The patient was referred to our tertiary hospital. He was Cushingoid with plethora, proximal myopathy and dyspnoea on minimal exertion. Blood pressure was 183/93mmHg. He had hypokalaemia (K⁺ 3.0mmol/l). Other electrolytes, renal function and liver function tests were normal. Fasting blood glucose was 9.5mmol/l and HbA1c 8.2%. 24 hour urinary free cortisol (UFC) measurements were 1384 and 759nmol/24 hours (65-275). 24 hour urinary catecholamines were normal.

There was loss of diurnal variation of cortisol levels and with no suppression after dexamthasone (see table 1) suggesting an autonomous source of cortisol excess not dependent on ACTH. Dihydroepiandrosterone sulphate (DHAS) was low (0.7µmol/l (2.0-15.2)).

Day	Dexamethasone dose (mg)	Cortisol (nmol/l)		ACTH (ng/l)		UFC (nmol/24h)	Aldosterone (µmol/l)
		0900h	2400h	0900h	2400h		
1	0	517	581	8	7	-----	-----
2	0	533	557	7	<5	727	2400
3	8	-----	-----	-----	-----	-----	-----
4	0	480	-----	-----	-----	-----	1800

Table 1

Management

He was treated with Metyrapone and underwent open laparotomy. An 8 – 10cm mass was found in the left adrenal gland. Pathology of the lesion showed an adrenal cortical neoplasm with malignant potential which was incompletely excised (pT3). Post operative cortisol levels were 176 – 200nmol/l but blood pressure was 110/70mmHg (off antihypertensives) and blood glucose 5.3mmol/l. He was commenced on Mitotane.

Discussion

This case shows the importance of fully investigating adrenal masses. Adrenocortical adenocarcinoma is rare and has an incidence of 0.5/million population per year¹. A literature review by Ng and Libertino found that 62% of tumours were functional with almost 40% of these presenting with Cushing’s syndrome². Mineralocorticoid excess affected 2.5%. CT scanning is the best imaging modality and can help identify which lesions are likely to be malignant (larger lesions >5cm, blurred margins, heterogeneous contrast enhancement) and can identify distant metastasis². MRI can be used to assess thrombosis in blood vessels. Pre-operative investigations help plan the operation and, if cortisol excess is detected, hypoadrenalism post-operatively period can be treated. The main prognostic indicators are early tumour stage and complete surgical resection of the tumour^{2,3}.

References

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