Conn’s syndrome due to an ectopic adrenal adenoma

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Summary: A 31 year old woman presented with resistant hypertension. Investigations revealed that she had Conn’s syndrome. Computed tomography showed both adrenals to be normal but an ectopic adenoma was identified posterior to the stomach. Surgical excision of the tumour confirmed benign aldosteronoma and cured her hypertension. We believe this to be the first report of Conn’s syndrome caused by an ectopic adrenal adenoma.

Introduction

Conn1 first described a new clinical syndrome of primary aldosteronism in 1955. Most cases of Conn’s syndrome are due to solitary adrenal adenomas.2-4 We report an unusual case of Conn’s syndrome caused by an adrenal adenoma (aldosteronoma) found outside the left adrenal gland where both adrenals themselves were normal.

Case report

A 31 year old woman was seen in the medical outpatient clinic for difficult control of hypertension. Five years previously she was prescribed the contraceptive pill and two months later her general practitioner found the blood pressure to be raised. Blood pressure remained elevated in spite of discontinuing the contraceptive pill. She was being treated with cyclophentiazide and atenolol at the time of presentation to the hospital. She did not complain of muscle weakness, polyuria, polydipsia, paraesthesiae or tetany. She had not ingested liquorice preparations. There was no family history of hypertension. On examination her blood pressure was 190/110 mmHg. There was no radiofemoral delay of pulse. The kidneys were not enlarged and there was no renal artery bruit. The muscle power was normal. Fundi showed arteriovenous nipping.

On investigation the full blood count and blood glucose were normal. Electrocardiogram showed left ventricular hypertrophy. Chest X-ray showed cardiomegaly. Serum urea was 4.2 mmol/l, serum creatinine 76 μmol/l and creatinine clearance 110 ml/min. Her intravenous pyelogram was normal. Serum electrolytes (mmol/l) on presentation were sodium 143, potassium 2.2, chloride 100, bicarbonate 35, and 2 weeks after stopping the diuretic the potassium level was 2.8. The 24-hour urinary excretions of electrolytes were sodium 135 mmol and potassium 92 mmol in a volume of 1880 ml. The 24-hour urinary-free cortisol was 126 nmol (normal range 100–300 nmol). Her plasma aldosterone level was 1040 pmol/l (100–600 pmol/l). Plasma renin activity in pmol per hour per ml was 1.15 (1.17–2.39) recumbent and 1.84 (2.99–4.3) ambulant. Radio-immunoassays were used to measure both plasma aldosterone7 and plasma renin activity.8 Computed tomographic scan showed a 1 cm diameter low attenuation cystic mass identified above the left adrenal but both adrenals themselves were normal (Figure 1).

**Figure 1** CT scan of abdomen demonstrating the ectopic adrenal adenoma marked by arrow.
She became normotensive on treatment with oral spironolactone 400 mg daily and at surgery a 15 mm x 7 mm nodule found 3 cm above the left adrenal gland was excised. The cut surface of the nodule was golden yellow and the histological features were of an adrenal adenoma (Figure 2). There was no evidence of malignant change. Adrenalectomy was not required as the tumour was an ectopic one. Her blood pressure and serum electrolytes have been normal since surgery without drug therapy.

Discussion

Conn et al.\(^1\) found muscle weakness (75%) nocturnal polyuria (74%), headache and polydipsia to be the four most common symptoms in their analysis of 103 patients with primary aldosteronism. Our patient did not report any symptoms at all.

Computed tomography (CT) is a valuable method of locating adrenal adenomas.\(^5\)\(^-6\) Our patient had normal adrenal glands on CT scan but a small mass was noted posteromedial to the stomach above the left adrenal (Figure 1).

Normally adrenal adenomas form in the zona fasciculata of the adrenal cortex and are found within the adrenal gland. We believe this to be the first case report of Conn’s syndrome caused by an aldosteronoma found outside the adrenal glands.

References

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