Clinics in Diagnostic Imaging (39)

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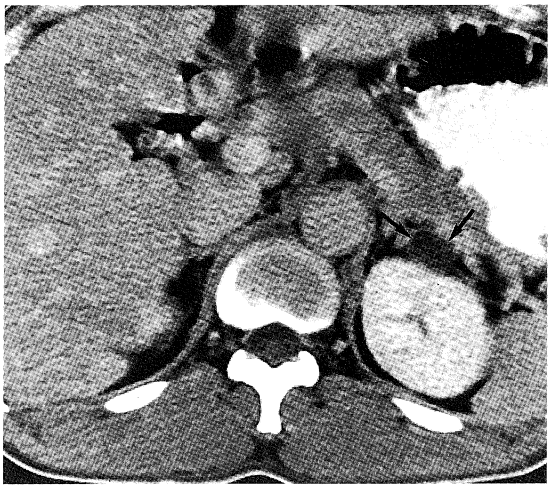

Fig 1a

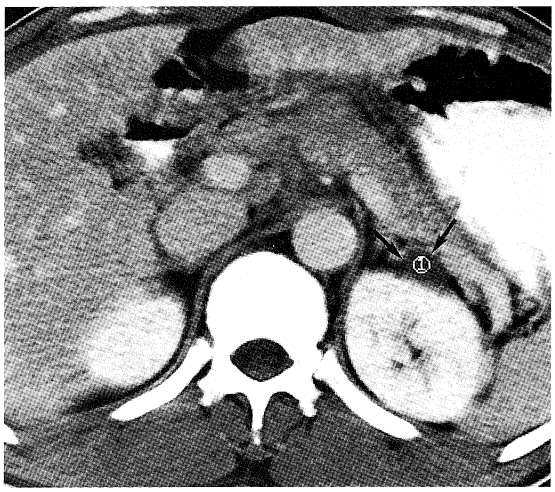

Fig 1b

Fig 1a & b – Contiguous enhanced CT scans taken (a) at the level of the upper poles of the kidneys, and (b) 10 mm more caudally. CT number in the area indicated by the cursor (1) measures 28.4 HU.

CASE PRESENTATION

A 33-year-old man presented with generalised, progressive muscle weakness of two years duration. He had no past history of note. On examination, his general condition was good. His blood pressure was elevated, being 160/100 mm Hg. Results of his relevant blood investigations were urea 18 mg/dL (normal 8 – 20 mg/dL), creatinine 1.2 mg/dL (normal 0.7 – 1.5 mg/dL), sodium 146 mmol/L (normal 135 – 150 mmol/L), potassium 2.2 mmol/L (normal 3.5 – 5.5 mmol/L), chloride 96 mmol/L (normal 98 – 108 mmol/L) and bicarbonate 36 mmol/L (normal 20 – 30 mmol/L). Full blood count and urine analysis were normal. What do the computed tomography (CT) scans (Figs 1a & b) show? What is the diagnosis?

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IMAGE INTERPRETATION

CT scans showed an oval mass in the left adrenal gland, with an attenuation number of 28.4 HU, indicating that it was solid. It was well-defined, measured 1.9 cm across and did not enhance. The mass was homogeneous with no evidence of calcification (Figs 1a & b).

DIAGNOSIS

Conn's syndrome due to adrenocortical adenoma (Conn's tumour)

CLINICAL COURSE

The patient underwent a left partial adrenalectomy. Pathological examination of the excised tumour confirmed the diagnosis of an adrenocortical adenoma. The patient recovered uneventfully. The post-operative blood pressure normalised (120/70 mm Hg) and his blood electrolyte levels also returned to normal on follow-up.

DISCUSSION

Primary aldosteronism (or Conn's syndrome) results from autonomous hypersecretion of aldosterone from the adrenal cortex. It is characterised clinically by hypertension, hypokalaemia, metabolic alkalosis, and periodic muscle weakness and paralysis. In the majority of cases (79%), primary aldosteronism is due to a benign aldosterone-secreting adrenocortical adenoma. Bilateral adrenal hyperplasia is present in 20% while adrenocortical carcinoma is a very rare cause[1-3]. Imaging has an important role in distinguishing between adenoma and bilateral hyperplasia, as the former condition should be treated surgically while the latter condition is best treated medically.

Most adrenocortical adenomas are small, averaging less than 2cm in size, while 15% – 20% of aldosteronomas are micronodules measuring less than 1 cm across[3,4]. The availability of thin-section (5 mm or thinner), high-resolution CT scans make this modality highly effective in the detection of adenomas, with a sensitivity of 70% – 90%[3,5]. Adenomas are seen on CT scans as low-density masses, usually less than 10 HU, due to their high cytoplasmic lipid content[6] (Fig 2). They rarely calcify and do not enhance significantly after contrast administration. One weakness of CT is that it cannot reliably distinguish between aldosteronomas and non-functioning adenomas, particularly when a lesion is solitary[7]. But if the appropriate clinical and biochemical information is taken into consideration, the chances of a solitary mass in normal-sized adrenal glands being something other than an aldosteronoma is extremely low[8].

On magnetic resonance (MR) imaging, adenomas usually appear slightly hypointense or isointense relative to the liver on T1-weighted images, and are slightly hyperintense or isointense on T2-weighted images[8]. MR scans appear to be equal to CT in sensitivity and may be more specific for the detection of adrenal adenomas[9]. Venous sampling is the most accurate means for localising aldosteronomas but may be technically difficult to perform. In successfully-catheterised cases, the sensitivity approaches 100%[5]. A cholesterol-based radiopharmaceutical, iodomethyl 19-norcholesterol (NP-59), has been used to distinguish unilateral adenoma from bilateral adrenocortical hyperplasia. However, NP-59 scintigraphy is time-consuming, expensive and less accurate compared to CT. Therefore its use may be limited to cases where CT is equivocal[8].

Other adrenal disorders which may also give rise to hypertension in young adults include Cushing's syndrome, which may in turn be due to a variety of lesions, and phaeochromocytoma. Cushing's syndrome results from excessive production of glucocorticosteroids, with the diagnosis usually being made on clinical and biochemical grounds. The most common cause of Cushing's syndrome is iatrogenic steroid administration, while endogeneous causes include excessive adrenocorticotrophic hormone (ACTH) production in 75% of cases and excessive cortisol from adrenal tumours in the other 25%[8]. The majority of ACTH-dependent Cushing's syndrome are due to a pituitary abnormality (Cushing's disease), with the adrenals showing hyperplastic changes in 71% of these cases[10]. These hyperplastic changes may be smooth (Fig 3) or nodular, the latter being the less common variety[11]. Hyperplastic glands usually cause overall enlargement of both glands, although occasionally, a single dominant hyperplastic micronodule may simulate an unilateral adenoma.

Functioning adrenocortical adenomas account for about 10% – 20% of Cushing's syndrome. These adenomas are unilateral, usually larger than 2 cm in size, and have a variable CT appearance with contrast enhancement being seen[8] (Fig 4). Adrenal carcinomas comprise about 10% – 15% of Cushing's syndrome, with the majority being larger than 6 cm in diameter

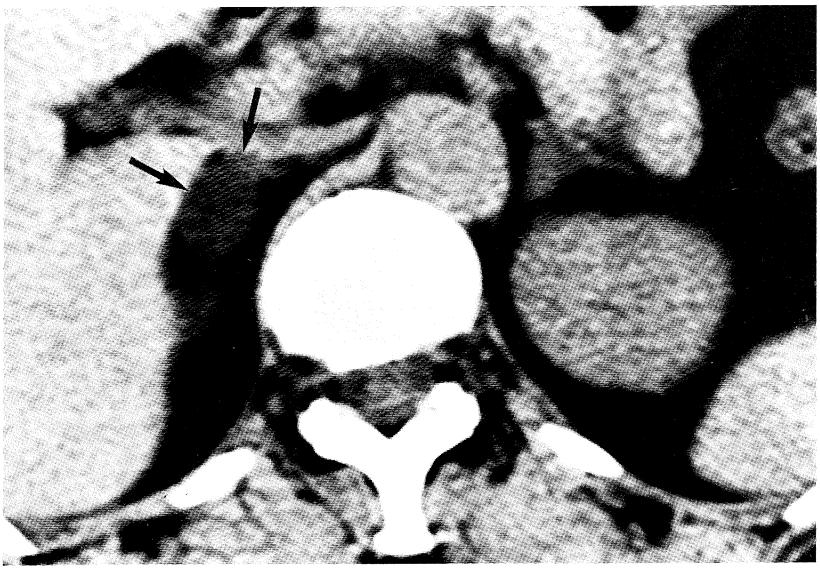

Fig 2 – 49-year-old woman with adrenocortical adenoma. She presented with muscle weakness for one year and was hypertensive (blood pressure 210/110mm Hg). Unenhanced CT scan shows a right adrenal mass (arrows), with a CT number of minus 1.9 HU, indicative of lipid content.

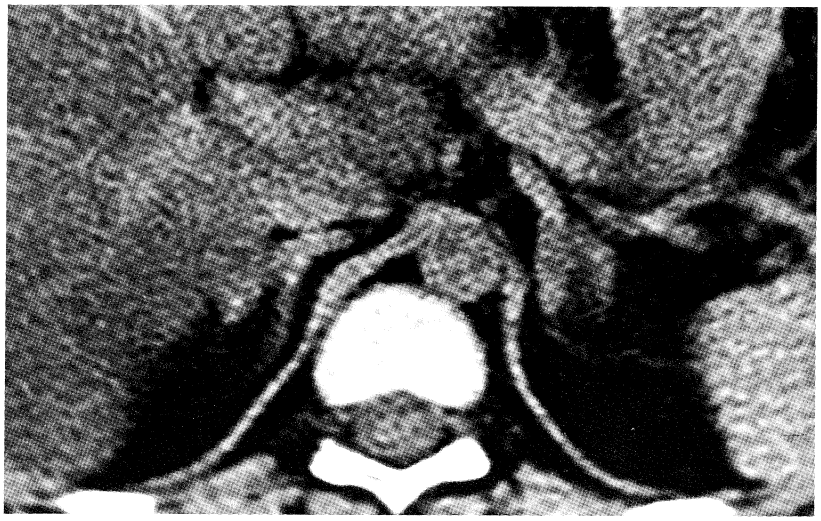

Fig 3 – 9-year-old boy with adrenal hyperplasia secondary to Cushing's disease. Unenhanced CT scan shows smooth enlargement of both adrenal glands.

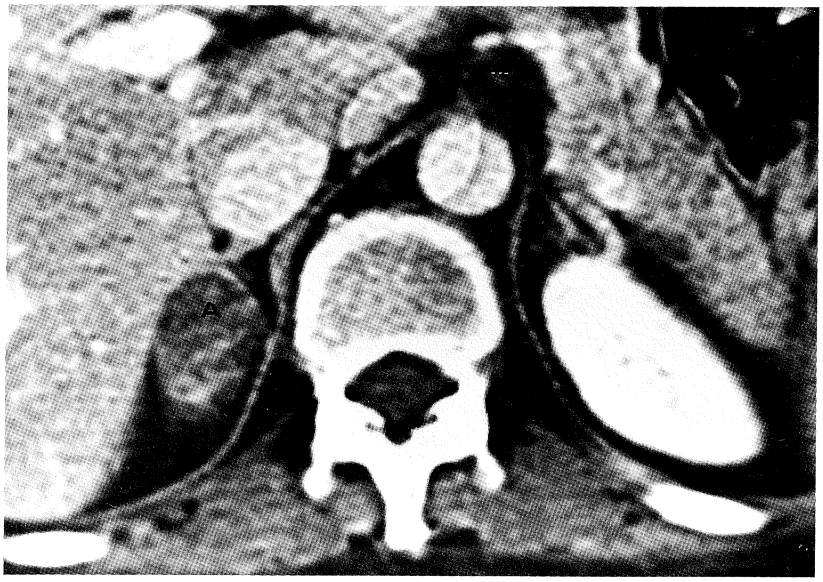

Fig 4 – 29-year-old woman with Cushing's syndrome caused by an adrenocortical adenoma. Enhanced CT scan shows a mildly-enhancing right adrenal mass (A).

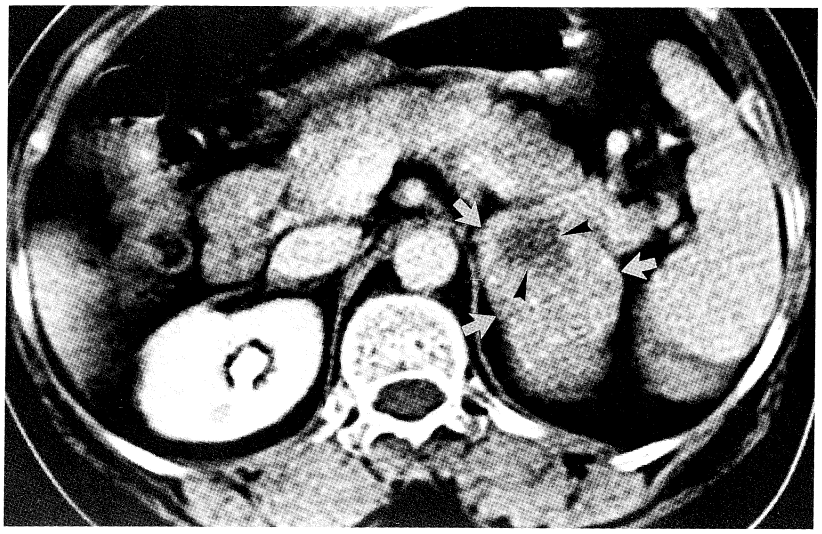

Fig 5 – 54-year-old woman with Cushing's syndrome caused by an adrenal carcinoma. Enhanced CT scan shows a large heterogeneously-enhancing left adrenal mass (arrows), with a central necrotic area (arrowheads).

at time of presentation. Because of its large size, these tumours can often be detected on ultrasound. On CT scans, adrenal tumours are seen as large heterogeneous masses, with areas of necrosis and calcification (Fig 5)[12]. Small carcinomas may resemble adenomas. It is important to search for evidence of possible carcinomatous involvement of the adjacent organs such as the inferior vena cava and regional lymph nodes, and metastases to the liver, lung and bone. CT and MR imaging are equally effective in this respect[8]. Adrenal carcinoma does usually not show uptake on cholesterol based scintigraphy[11].

Phaeochromocytomas arise from paraganglion cells anywhere in the autonomous nervous system, and produce catecholamines which account for the patients' clinical features and biochemical findings. 90% of phaeochromocytomas are found in the medulla of the adrenal gland. The less common varieties of these tumours can be remembered using the "rule of tens": 10% are extra-adrenal, 10% are associated with neuroectodermal disorders, 10% are multiple, and 10% are malignant. Only about 10% of adrenal phaeochromocytomas are non-functional[13]. Diagnosis of phaeochromocytomas is best established by 24 hour collection of excreted catecholamines, metanephrines and vanillymandelic acid (VMA). The diagnosis of phaeochromocytoma should be followed by studies to localise the tumour. For most patients with sporadic non-familial phaeochromocytomas, CT scans are most appropriate for identifying adrenal gland lesions.

On CT scans, phaeochromocytomas are seen as rounded masses, with speckled calcifications in about 12% of tumours. The tumours may undergo marked necrosis and typically display marked enhancement (Fig 6)[8]. As intravenous administration of an ionic contrast agent may precipitate a hypertensive crisis, α_1 – adrenergic blockade is advised prior to CT scanning. This complication does not appear with non-ionic contrast agents, therefore usage of this newer agent is recommended[14]. MR imaging is as accurate as CT for identifying adrenal phaeochromocytomas although MR scans are slightly better for detecting extraadrenal lesions[15]. ^{123}I or ^{131}I meta-iodo-benzylguanidine (MIBG) scintigraphy is particularly useful for demonstrating sites of ectopic phaeochromocytomas, and metastatic and local recurrent disease. MIBG, a nonepinephric analogue, labels catecholamine precursors and is concentrated in adrenergic storage vesicles. A positive MIBG scintiscan, however, always requires correlation with CT or MR imaging[8].

In the work-up of adrenal lesions, whether presenting in a symptomatic patient or incidentally-discovered, the initial step remains a careful clinical evaluation and detailed biochemical assessment. The nature of the biochemical abnormality then dictates further management and investigation, including imaging. The adrenal glands are usually imaged to localise and characterise pathology which has been indicated biochemically. CT is currently the single

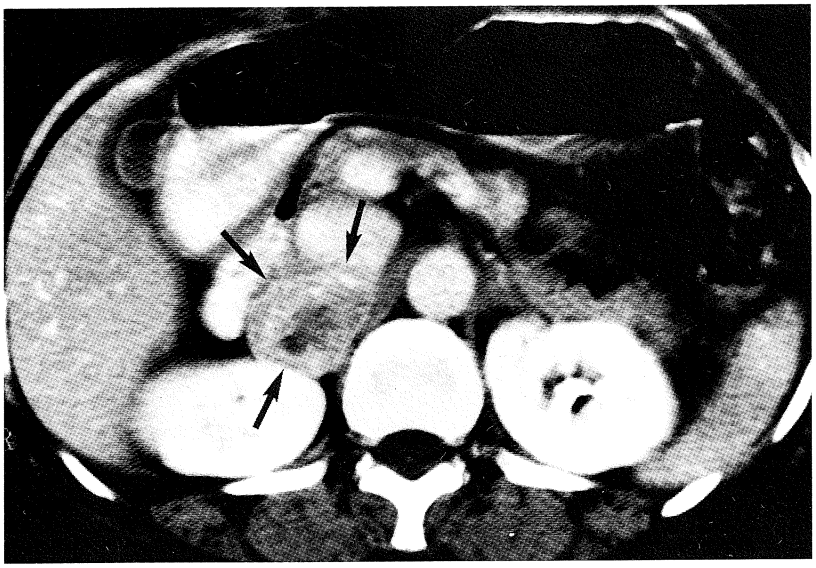

Fig 6 – 42-year-old woman with phaeochromocytoma. Her VMA levels were elevated. Enhanced CT scan shows marked heterogeneous enhancement of a right adrenal mass (arrows). It has a central area of necrosis.

most important imaging method for identifying adrenal masses, although MR scans, especially chemical-shift imaging, are being increasingly used[16,17]. Adrenocortical and adrenomedullary scintigraphy have a role in selected situations. A strategic approach to imaging adrenal lesions should reduce the need for percutaneous biopsy in diagnosis[18-20].

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ABSTRACT

A 33-year-old man presenting with muscle weakness and hypertension was found to have hypokalaemia and metabolic alkalosis. Computed tomography (CT) showed an adrenal mass. Conn's syndrome due to an adrenocortical adenoma was confirmed at surgery. Hypertension-related adrenal disorders such as Cushing's syndrome, carcinoma and phaeochromocytoma are discussed. CT is currently the single most useful imaging method for identifying adrenal masses.

Keywords: adrenal tumour; computed tomography (CT); Conn's syndrome; Cushing's syndrome; hyperaldosteronism; phaeochromocytoma