

Multiple phaeochromocytoma (adrenal and extra-adrenal)

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Case presentation

A 17-year-old-girl was referred from a rural hospital with problems of hypertension secondary to suspected coarctation of the aorta, and aortic incompetence. Her main complaints were headaches and palpitations.

Clinically she had mild to moderate hypertension, tachycardia, left ventricular enlargement and a systolic murmur. There were no abdominal masses. Investigations performed excluded coarctation of the aorta and aortic incompetence. ECG confirmed sinus tachycardia with left ventricular hypertrophy. Renal artery stenosis was also excluded by a normal renogram. She was then investigated for other causes of hypertension including phaeochromocytoma.

Serum urea and electrolytes were normal. Urine vanillylmandelic acid (VMA) and metanephrines were also normal.

Imaging

CXR

There were no positive findings despite clinical early cardiomegaly.

MIBG scan

This showed no features of phaeochromocytoma.

Ultrasound

A homogeneous soft-tissue mass measuring 4.5 x 4.2 cm, slightly lobulated and well defined was visualised on the left side of the aorta just above its bifurcation. No calcifications were noted. The aorta was not compressed.

Another similar mass measuring 2.9 x 2.9 cm, triangular, and more regular in outline, was visualised at the upper pole of the left kidney.

There were no other positive findings (Fig. 1).

CT scan of the abdomen

The patient was allergic to iodine and had to be premedicated with prednisone, and alpha-blockers were also given to avert a hypertension crisis (Fig. 2).

A homogeneous retroperitoneal tumour, isodense to skeletal muscle, was found to the left of the aorta just above its bifurcation. It was lateral to the aorta, and anterolateral to the spinal column and left psoas muscle. It measured 6.0 x 4.2 x 5.8 cm. The



Fig. 1. Transverse sonar mid-abdomen showing left para-aortic mass — organ of Zuckerkindl.

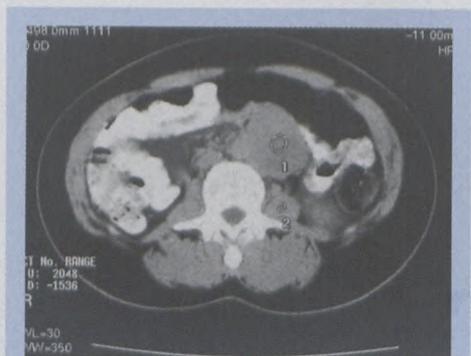


Fig. 2. Pre-contrast CT scan of the abdomen — axial showing a left para-aortic tumour isodense to skeletal muscle.

margins were lobulated and clear. There was a marked homogeneous enhancement post contrast except for a small hypodense area at the inferior pole thought to be cystic necrosis. There was no infiltration or compression of surrounding structures (Fig. 3.)

A second tumour was found in the left adrenal gland measured 3.8 x 2.3 x



Fig. 3. Reconstructed CT image — coronal showing left para-aortic mass just above the aortic bifurcation — organ of Zuckerkindl.

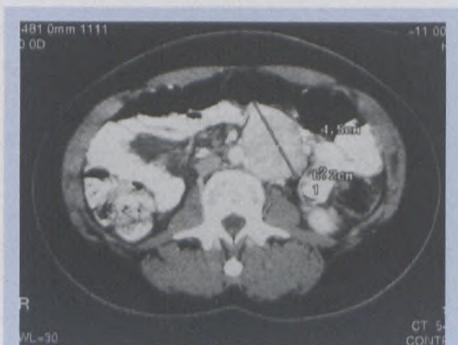


Fig. 4. Contrast-enhanced CT — axial showing markedly enhancing extra-adrenal phaeochromocytoma, left para-aortic region.

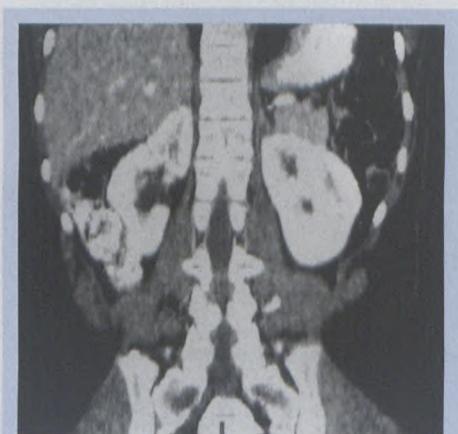


Fig. 5. Contrast-enhanced CT of the abdomen. Coronal reconstruction shows left suprarenal tumour (adrenal phaeochromocytoma).

36.6 cm. It was smooth and enhanced homogeneously and markedly post contrast (Fig. 4).

The right adrenal gland was normal. Both renal arteries were normal. No lymph node enlargement was noted. There were no liver or bone metastases. Coronal and sagittal multiplanar reconstruction (MPR) aided in localisation and visualisation of tumours (Fig. 5).

Biopsy

An excision biopsy was performed and two phaeochromocytomas removed. No postoperative complications were reported.

Histology confirmed the diagnosis of phaeochromocytoma.

Discussion

Phaeochromocytoma is a subtype of the paragangliomas, a neuroendocrine tumour that arises from paraganglionic tissue.¹

It is responsible for 0.1% of hypertension cases.

Location

It occurs anywhere in the sympathetic nervous system from the neck to the sacrum: (i) adrenal medulla (85 - 90%); (ii) extra-adrenal (10 - 15%), with the majority adjacent to the kidneys; and (iii) other sites including the organ of Zuckerkandl, the gonads, bladder and thoracic para-vertebral region.

Our patient's tumour was in the left adrenal gland and the organ of Zuckerkandl (extra-adrenal site adjacent to aortic bifurcation).

Clinical findings

Symptoms are secondary to excess catecholamine production.

Classical presentation is: (i) paroxysmal (50%) or sustained (50%) hypertension accompanied by headaches, tachycardia, diaphoresis, palpitations and anxiety, with 9% of patients asymptomatic;² (ii) elevated vanillylmandelic acid in 24-hour urine and metanephrines (these were normal in our patient); and (iii) elevated serum catecholamines.¹⁻⁴

Rule of 10

This postulates that 10% of cases are:¹⁻³ (i) familial; (ii) children; (iii) bilateral adrenal; (iv) extra-adrenal; (v) multiple; and (vi) malignant.

Associations

1. Multiple endocrine neoplasia (phaeochromocytoma, usually bilateral and almost always intra-adrenal).¹

• Sipple syndrome: Phaeochromocytoma, medullary carcinoma of the thyroid (MCT) and parathyroid adenoma (multiple endocrine neoplasia (MEN) type II).

• Mucosal neuroma syndrome (MEN type III) — MCT plus intestinal ganglioneuromatosis plus phaeochromocytoma.

2. Neuroectodermal disorders: Tuberous sclerosis, neurofibromatosis, Von Hippel Lindau syndrome.²

3. Familial phaeochromocytosis.

4. Carney's syndrome: Paraganglioma plus gastric epithelioid leiomyosarcoma plus pulmonary chondroma.²

Imaging

Plain X-rays or IVU with tomograms may reveal a mass above the kidney with renal involvement but in our case it did not help. Both are low in sensitivity and specificity.³

Ultrasound is helpful in children where it has proved to be highly successful.⁴ A well-margined solid/complex cystic tumour, homo or heterogeneous is found in the adrenal or para-aortic areas.¹⁻³

CT scan. Localisation is accurate in 91% of cases with tumours > 2 cm.² Sensitivity is 93 - 100%. It is the method of choice. Detection rates are high.

The small number of failures tend to occur in children with poorly developed fat planes and in cases of extra-adrenal phaeochromocytoma. CT shows adrenal/extra-adrenal mass with strong contrast enhancement. Low density areas are due to necrosis or haemorrhage.² Calcifications are rare.

NB: IV contrast injection may precipitate hypertensive crisis in patients not on alpha-adrenergic blockers.⁴ In

our case the patient was already on treatment.

MRI

The detection rate is also high with MRI and it may eventually replace CT as the investigation of choice since it involves no radiation.² Its use is limited by cost and availability.²

Phaeochromocytomas are extremely hyperintense on T2W1 and show marked enhancement post contrast. Intensity is considerably higher when compared with adenoma and metastases.²

Nuclear medicine studies

Scintigraphy with MIBG is 80 - 90% sensitive, and 98% specific.²

It is useful in detection of extra-adrenal tumours or when adrenals are normal on CT and MRI. False-positive and false-negative results have both been recorded.³

In our patient MIBG showed no evidence of phaeochromocytoma; false-negative result.

Arteriography and venous sampling

These are occasionally used in the search for ectopic tumours.³

Arteriography: Localisation is useful in more than 90%. Usually hypervascular lesion with intense tumour blush enlarged feeding arteries and neovascularisation.²

Complications

Malignancy in 10% of lymph nodes, liver, lungs, and bone. Metastases may be hormonally active.²

References

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An information pack is available from the Senior Personnel Manager, Medical Personnel Department, Wishaw General Hospital, 50 Netherton Street, Wishaw ML2 ODP. Telephone +44 (0)1698 366551. Fax +44 (0)1698 366554. Please quote ref MW276. Closing date 27 September 2002.