

Extra-adrenal phaeochromocytoma

A case report

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Summary

A case of hypertension in a young woman caused by an extra-adrenal phaeochromocytoma (EAP) situated in the hilum of the left kidney is reported. Surgical removal of the EAP resulted in control of the patient's hypertension without further drug treatment. The clinical features, diagnostic work-up and management of EAP are briefly discussed.

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Phaeochromocytoma is a rare tumour with a reported incidence of 1 in 50 000 of the adult population¹ and 1 - 10/1000 of hypertensive patients.² It has been called the '10% tumour' since approximately 10% are malignant, 10% bilateral and 10% extra-adrenal.³ Thus, the calculated incidence of extra-adrenal phaeochromocytoma (EAP) is 1 in 500 000 of the adult population.

Sustained hypertension is found in 50% of patients with phaeochromocytoma and paroxysmal hypertension in the other 50%.⁴ Headache, sweating and palpitations are features of the condition in 60 - 80% of the cases, and less than 50% of patients are thin.⁵

According to Fries *et al.*,⁶ EAP is found in the neck in 2% of cases, the thorax in 12%, the upper abdomen in 43%, the lower abdomen in 29%, the urinary bladder in 12%, and the pelvis in 2%.

The conditions which have been reported to be associated with phaeochromocytoma include the neuro-ectodermal syndromes (von Hippel-Lindau disease, Sturge-Weber syndrome, tuberous sclerosis), renal cell carcinoma, cholelithiasis, congenital heart disease, aganglionic megacolon and mega-ureter.^{4,6-9} There is, however, no association between EAP and the type II multiple endocrine adenopathy syndrome.¹⁰

Malignant change in EAP is reported in one series to be 22% and has a uniformly dismal prognosis, except for cases of vesical neoplasms in which the outlook is better.⁹ At present treatment with iodine-131 meta-iodobenzylguanidine (MIBG) offers some hope of achieving a cure in malignant phaeochromocytomas.¹¹

Case report

A 19-year-old non-pregnant coloured woman complained of headaches, sweating and episodes of syncope. She was of slender build

with a blood pressure of 160/110 mmHg, but the rest of the physical examination was unremarkable. The hypertension was controlled on treatment with labetalol and prazosin.

The patient's full blood count, serum urea, creatinine and electrolyte levels, fasting blood sugar, chest and abdominal radiographs and electrocardiogram were normal.

An ultrasound examination revealed normal kidneys and a 2 x 3 cm well-circumscribed mass with a mixed echoic pattern in the hilum of the left kidney. The excretory urogram was normal. Renal arteriography (Fig. 1) demonstrated a distinct tumour blush in the hilum of the left kidney. The lesion was also clearly delineated on computed tomography (CT) (Fig. 2). Six-hourly urinary vanillylmandelic acid (VMA) estimations revealed values of 3,8, 3,3, 6,5 and 3,2 μmol VMA/mmol urinary creatinine (normal < 3,0). Phenoxybenzamine was added to the patient's treatment regimen, and 1 week later a thoraco-abdominal exploration was carried out. The left kidney and adrenal gland were normal. The tumour in the left renal hilum was enmeshed in a rich vasculature, but with careful dissection was removed completely without compromising the left renal vessels. There was no direct communication between the tumour and the sympathetic trunk or adrenal gland. No other tumours were palpable in the left retroperitoneal region.

The patient's postoperative course was uneventful and at 15 months follow-up she was without symptoms and normotensive. A ¹³¹I-MIBG scan was performed 1 month postoperatively, since this investigation only became available at our institution at that time. No residual foci of phaeochromocytoma could be demonstrated.

Histological study of the tumour confirmed the diagnosis of phaeochromocytoma.

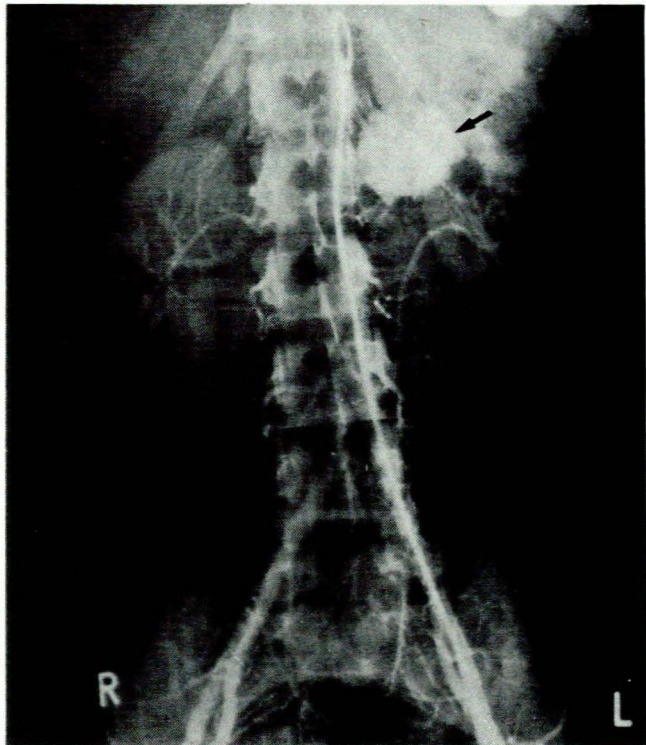


Fig. 1. Arteriogram of the left kidney.

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Fig. 2. CT of the lesion in hilum of the left kidney.

Discussion

Because of its protean manifestations, the diagnosis of pheochromocytoma is easily missed. However, recent data indicate that a complete history alone can result in the diagnosis in nearly 95% of positive cases and can virtually exclude the condition in 99% of negative cases.⁴ The simplest screening test for pheochromocytoma is determination of the urinary metanephrine value, with a 95% sensitivity and 97% specificity. This test is less subject to drug and diet interference than VMA determinations. Fractionated serum catecholamine determination is reported to be virtually 100% diagnostic, but the results need to be interpreted with care because of patients' stress responses.¹²

Localization of a pheochromocytoma can be difficult and hazardous if invasive procedures such as angiography and catheterization of the vena cava are used. CT, however, will localize more than 90% of tumours and has a resolution of approximately 2 cm.¹³

In a series of 8 cases,¹⁴ the MIBG scan was highly accurate, with a specificity of 100%. Tumours as small as 0,2 g were consistently localized but administration of reserpine and tricyclic antidepressants may interfere with the scan.

Although α - and β -adrenergic blocking agents are advocated for the pre-operative control of catecholamine effects, Stewart⁴ has reported a series of 80 cases of pheochromocytomas which were operated on successfully without the use of pre-operative adrenergic blockers. These patients received a transfusion of 2 units of blood on the day before operation and no intra-operative mortality was recorded.

At present the customary laborious and expensive work-up on patients with suspected pheochromocytoma can be obviated, and a high degree of accuracy in diagnosis and localization may be achieved. With CT and MIBG scan on an outpatient protocol the cost of pre-operative studies can be reduced more than tenfold.⁴

The result of surgical extirpation of benign pheochromocytomas is good, provided all the tumour is completely removed. Postoperative normotension is achieved in 67% of patients with sustained and in 95% of patients with paroxysmal hypertension pre-operatively. However, a peri-operative mortality rate of 3,8% has been recorded.¹

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