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Spontaneous intracerebral haemorrhage caused by extra-adrenal phaeochromocytoma

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Abstract

We report an 18-year-old man who presented with a sudden onset of headache followed by left hemianopia. A brain CT scan showed intracerebral haemorrhage in the left frontoparietal area, but a cerebral angiogram and MRI revealed no vascular anomaly. The patient was managed conservatively and his headache and visual loss improved over time. Hypertension in the form of paroxysmal attacks led us to suspect phaeochromocytoma. Subsequently, the patient was diagnosed with an extra-adrenal phaeochromocytoma in the left para-aortic area following endocrinological evaluation, abdominal CT scan and ¹³¹I-meta-iodobenzylguanidine (MIBG) scintigraphy. The patient presented here illustrates the importance of a careful search for a remediable cause of hypertension in children and young adults with spontaneous intracerebral haemorrhage.

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1. Introduction

Phaeochromocytomas commonly arise from the adrenal medulla, but approximately 10% of these tumours have been reported to develop at extra-adrenal sites, including the paraspinal and prevertebral sympathetic ganglia and the organ of Zuckerkandl. Phaeochromocytomas account for only 0.1–0.5% of patients with hypertension,¹ but it is imperative that the condition be diagnosed early to avoid its devastating complications. Among these complications, spontaneous intracerebral haemorrhage is an unusual, but potentially fatal, complication and cerebral haemorrhage caused by extra-adrenal phaeochromocytoma is rare. Herein, we report a rare patient with an extra-adrenal phaeochromocytoma presenting with spontaneous intracerebral haematoma.

2. Case report

An 18-year-old man, who had been taking antihypertensive medication for 1 month on advice received from a local clinic, presented with a sudden onset of headache followed by left hemianopia. He had experienced palpitations and chest discomfort during physical exertion for 2 years prior to his admission but, unfortunately, this was ignored. CT scan of the brain revealed a left frontoparietal haematoma (Fig. 1). The patient was managed conservatively and his headache and visual loss improved over time. His blood pressure, which was in the range 160–190/100–120 mmHg, was controlled with alpha, beta and calcium channel blockers. A cerebral angiogram and MRI did not demonstrate a vascular anomaly. Paroxysmal attacks of hypertension led us to suspect phaeochromocytoma. Evaluation of a 24-h urine specimen showed vanillylmandelic acid (VMA) 12.6 mg/day (normal < 8 mg/day) and metanephrine 5.4 mg/day (normal < 1.2 mg/day). Abdominal CT scan

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Fig. 1. Brain CT scan demonstrating an intracerebral haemorrhage in the left posterior frontal and parietal region.

demonstrated a well-defined enhancing mass in the left para-aortic area with no abnormality in the adrenal glands; ^{131}I -meta-iodobenzylguanidine (MIBG) scintigraphy showed high uptake in the same area (Fig. 2). Total resection of the tumour was achieved using a retroperitoneal approach through a left subcostal incision. Macroscopically, the mass was well encapsulated, the cut surface being dark with haemorrhagic areas and areas of necrosis. Microscopic examination revealed a well-defined tumour cell nest (Zellballen) bound by a delicate fibrovascular stroma. The cells varied considerably in size and shape, with a finely granular basophilic cytoplasm and round to ovoid nuclei with prominent nucleoli. Immunohistochemical staining showed strong immunoreactivity to chromogranin and synaptophysin. These histopathological findings confirmed the diagnosis of pheochromocytoma (Fig. 3). Postoperatively, the patient improved rapidly and was discharged on the 7th postoperative day. One month after abdominal surgery, the patient was free of symptoms and normotensive without medication; in addition, urinary VMA (3.3 mg/day) and metanephrine (0.2 mg/day) levels had returned to normal.

3. Discussion

Pheochromocytomas are functionally active tumours of sympathetic nerve cells that occur most commonly in the adrenal medulla, but can also develop in extra-adrenal sites. These tumours produce adrenaline and noradrenaline, and thus cause sustained or paroxysmal hypertension. Pheochromocytoma is often accompanied by complications in other organs; although cerebrovascular disease is serious, its incidence is comparatively low. Cerebral haemorrhage

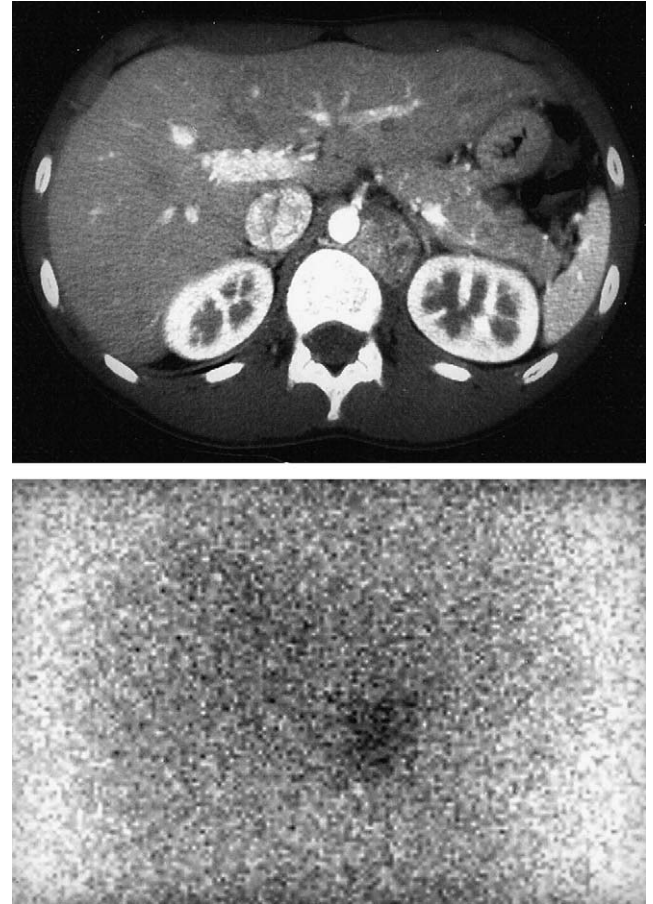


Fig. 2. Contrast-enhanced abdominal CT scan (upper) showing a well-defined, heterogeneously enhancing mass in the left para-aortic area. ^{131}I -Meta-iodobenzylguanidine (MIBG) scintigraphy (lower) showing an abnormally high tracer uptake in the same area.

is an unusual complication^{2,3} and cerebral haemorrhage caused by extra-adrenal pheochromocytoma is rare. To our knowledge, only one other patient has been reported previously in whom pheochromocytoma of the urinary bladder caused cerebral haemorrhage.⁴

The most important diagnostic studies are the biochemical evaluation of the urine and blood, which reflect catecholamine production by the tumor and radiological studies for localization of a mass. The biochemical tests for pheochromocytoma are based on the detection of abnormally increased levels of several free catecholamines or metabolites in the urine or blood and are routinely used in the evaluation of hypertensive patients. These studies generally involve measurement of the 24-h urinary excretion of free catecholamines and metabolites, including total metanephrines and VMA. Plasma and urinary metanephrine and constant monitoring of blood pressure are useful when screening for pheochromocytoma. Accurate tumor localization is essential and CT, MRI and ^{131}I -MIBG scintigraphy are the imaging techniques of choice. ^{131}I -MIBG scintigraphy is highly sensitive, especially where the CT and MRI findings are negative or equivocal, and is particularly useful for extra-adrenal tumors and metastases.⁵ Complete

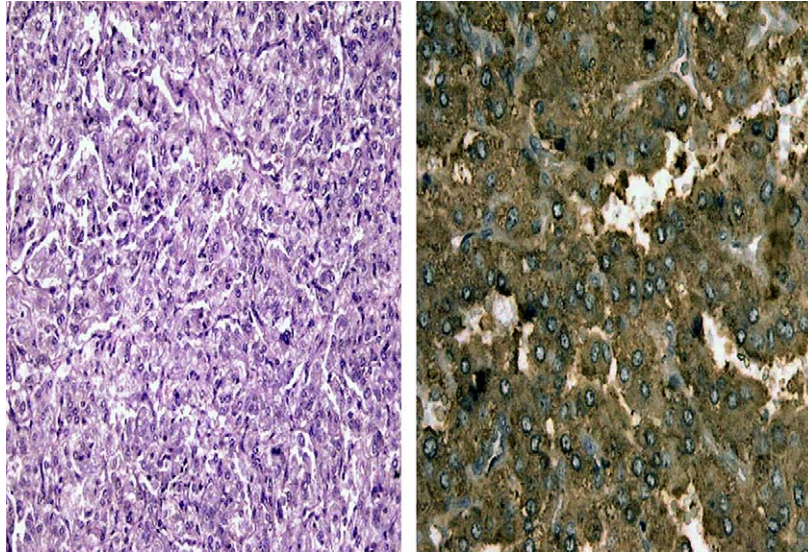


Fig. 3. Photomicrographs of the tumour specimen showing characteristic small clusters (Zellballen) separated by delicate fibrovascular stroma. The cells had a finely granular basophilic cytoplasm (left; haematoxylin and eosin stain; original magnification $\times 100$) and were positive for chromogranin staining (right; chromogranin stain; original magnification $\times 200$). This figure is available in colour on Science Direct.

surgical excision is the only curative therapy for pheochromocytoma, but should be performed as an elective procedure after careful pre-operative preparation.

Pheochromocytoma is a rare cause of secondary hypertension. However, its diagnosis is important, as the hypertension is usually cured with resection of the tumor, whereas it is potentially lethal if left untreated and is associated with long-term morbidity. Pheochromocytoma must be considered in the differential diagnosis of hypertensive patients with signs of sympathetic hyperactivity. The patient presented here highlights the importance of careful investigation to determine the remediable causes of hypertension in children and young adults before initiating antihypertensive therapy.

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Giant unruptured anterior communicating artery aneurysm presenting with seizure

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