PAEDIATRIC PHAECHROMOCYTOMA - A CASE REPORT AND LITERATURE REVIEW

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SUMMARY
Phaeochromocytoma continues to be a rare tumour in children. It can be missed if a high index of suspicion is not retained in unusually presenting cases. Early diagnosis, accurate localisation of the tumour, careful pre- and intra-operative management are all key to a successful outcome.

A 12-year-old boy reported to the Korle-Bu Teaching Hospital in Accra with unusual symptoms. A definitive diagnosis was delayed. His course and outcome are described with an emphasis on the maintenance of a high degree of suspicion of the diagnosis of phaeochromocytoma in children of all ages.

Keywords: Paediatric Phaeochromocytoma

INTRODUCTION
Phaeochromocytoma is a chromaffin tumour. It has been described in both adults and children. There are however no large-scale series of children with phaeochromocytoma. Hypertension or symptoms of hypertension continue to be the commonest mode of presentation¹. In most cases, the tumour is unilocular. Occasionally both adrenal and non-adrenal tumours have been found in the same patient, although phaeochromocytomas in children have been found to be more often extra-renal and multifocal than in adults². Phaeochromocytoma has been described as a great mimic due to the wide variety of symptoms and signs with which it presents. Familial associations may occur and should be looked out for.²

CASE REPORT
N.A. a well-nourished and apparently healthy 12-year-old Ghanaian adolescent is a pupil who presented on the 18th July 2000 with a sudden onset of generalised tonic-clonic convulsions, which affected mainly his upper limbs and eyes. There was no previous or family history of epilepsy and no history of drug ingestion. Clinical examination showed a blood pressure of 210/120 mmHg. The Blantyre Coma Scale (BCS) (see appendix) was 2 with conjugate eye deviation to the left¹. There were no abdominal masses nor a bruit.

Differential diagnoses made included idiopathic hypertension, hypoglycaemia or epilepsy.

Investigations done included random blood glucose, a full blood count, blood urea and electrolytes and creatinine, urinalysis, electroencephalogram (EEG) and a blood film for malaria parasites. The results of all of these tests were within normal limits.

His blood pressure was controlled by intravenous (i/v) Hydralazine 8mg then 3mg; his seizures were controlled with intravenous Phenobarbitone 60mg 12hourly and he was discharged home after 5 days on the two drugs.

Two months later he was re-admitted with convulsions and twitching affecting the left side of his face and body, with conjugate left eye deviation and markedly decreased muscle tone.

The initial clinical impression then was of Jacksonian epilepsy. The blood pressure remained elevated at 200/120 mm Hg, so he was admitted as a case of hypertensive encephalopathy of unknown cause.

Investigations done at that time included computerised tomography of the head, intravenous urogram, chest and abdominal X-rays, which were all found to be normal. He was put on plain Nifedipine 20 mg thrice daily, propranolol 40mg daily in addition to oral phenobarbitone, which brought his

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blood pressure down to 120/70 mmHg. He was then discharged home on Nifedipine 10 mg thrice daily and Propranolol 20 mg nocte but was lost to follow-up.

He presented again 2 months later in coma preceded by early morning headache, vomiting, profuse sweating and dizziness. On re-admission, his blood pressure was 190/110 mmHg but fluctuated between 200/120 and 140/100 mmHg irrespective of the maintenance drugs. A presumptive diagnosis of hypertension secondary to pheochromocytoma was made. A 24 hr urinary vanil mandelic acid (VMA) value was 3.5mg, the normal range being 1.9-9.8mg/24hr. An abdominal ultrasound showed no obvious intra-abdominal abnormality. An adrenal Doppler ultrasound was requested but not done. An initial CT scan was reported as normal. His blood pressure was controlled by the addition of Bendrofluazide (2.5 mg daily) to his previous regime (ie Nifedipine and Propranolol) and he was discharged home, the suspicion of pheochromocytoma having been regarded as not proved.

Five months later (a year after the initial presentation), he was rushed in again with tonic-clonic seizures worse in the upper limbs. His blood pressure was 230/140 mmHg. A repeat urinary VMA gave a value of 29.9mg/24hrs and a repeat abdominal renal CT scan showed a left supra-renal mass which was 3cm by 3.5 cm in size this time confirming the diagnosis of pheochromocytoma.

He was prepared for surgery and his blood pressure stabilised with phenoxy benzamine and propranolol at 140/100 mmHg given over a 2-month period. The delay was due to the unavailability of an α-blocker for parenteral use if required during surgery.

He was given Lorazepam 1mg the night before surgery. The tumour was excised in toto under general anaesthesia at laparotomy. The adrenal vein was initially clamped before full mobilisation and excision of the gland. A central venous line inserted did not show any circulatory upsets with tumour handling. His blood pressure remained stable throughout the procedure. The post-operative period was uneventful and his blood pressure remained at 100/60 mmHg until he was discharged home 2 weeks after surgery.

He has been reviewed on several occasions, the latest being 8 months after surgery. His blood pressure is normal at 100/60 mm Hg without any medication. He is back in school.

DISCUSSION

This clinical presentation fits with the usual pattern of presentation of pheochromocytomas. Most are commonly in boys with a peak incidence of 9-13 years. Ciftei et al looking at their 30-year experience with the disease noted a mean age of 10.7+/- 2.9 years and a male: female ratio of 3:1. In Poland, Januszewicz et al found surgically confirmed pheochromocytoma in only 6 out of 668 (0.8%) of children admitted with significant hypertension. Stringel G. et al found in their series that headache, blurring of vision and sweating with hypertension were the most common symptoms on presentation. In all these cases, there was initial sustained hypertension followed by an accelerated phase of malignant hypertension. This fits in with the experience in Korte-Bu where NA initially appeared well controlled on antihypertensives but then represented with out-of control hypertension. The usefulness of iodine 131 metiodobenzylguanidine to locate these tumours is disputed. The Polish series found a large number of false negatives while Caty M.G. et al in Michigan, found it very useful in combination with computed tomography. In our case, the final diagnosis was made by CT scan and elevated VMA.

The delay in diagnosis of this case may have been due to the fact that the tumour was initially small thus releasing small amounts of catecholamines. His loss to follow-up on several occasions also contributed to this delay.

Ligation of the renal vein was not associated in this case with any disturbances in the circulation of the patient unlike the experiences reported by Stringel et al.

Turner et al. in a Los Angeles review determined that the only predictive indicators of an uncomplicated outcome were preoperative resolution of symptoms and normalisation of blood pressure. In this case, the blood pressure was normalised totally before surgery and his outcome was uneventful. He remained normotensive without medication as was the experience of Caty MG et al.

REFERENCES


Appendix
Blantyre Coma Scale
This scale popularised by Molyneux ME et al assesses the central nervous system in a situation where coma arises as a result of an infective process.

The parameters are as follows:
Best motor response:
2: Localises painful stimulus (firm pressure with blunt end of pencil against sternum)
1: Withdraws from painful stimulus (firm pressure with blunt end of pencil against nail bed on finger or toe)
0: Any other motor response or absent motor response to pain.

Best verbal response:
2: Cries adequately in response to painful stimulus.
1: Moans or abnormal cry in response to painful stimulus.
0: No vocal response to painful stimulus.

Eye movements
1: Follows subject (eg. Mother’s face) with eyes
0: Does not follow