

Pheochromocytoma: A Rare Cause of Childhood Hypertensive Encephalopathy

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ABSTRACT

Pheochromocytomas are rare neuroendocrine tumours of chromaffin tissues. They are catecholamine secreting tumours which cause severe hypertension and other systemic disturbances. Of all the causes of childhood hypertension, pheochromocytoma constitutes less than 1%. We report the case of a 12 years old child who presented with hypertensive encephalopathy, confirmed histologically to be secondary to pheochromocytoma, and cured with meticulous critical care and surgical resection.

Key words: Pheochromocytoma. Hypertension. Neuroendocrine tumour. Encephalopathy.

INTRODUCTION

Pheochromocytoma is a catecholamine secreting tumour that arises from chromaffin cells. The most common site of origin is in adrenal medulla; however, it may arise anywhere in the body along the sympathetic chain.¹

Patients with pheochromocytoma present with severe hypertension and variety of other symptoms like palpitations, sweating and headaches due to excessive secretion of catecholamine (norepinephrine, epinephrine or dopamine) into circulation.²

Majority of pheochromocytomas are benign but approximately 12% of them are malignant in paediatric population. They often occur sporadically but can be inherited as an autosomal dominant trait. Pheochromocytoma may also be associated with other syndromes like von Hippel-Lindau (VHL) syndrome, multiple endocrine neoplasia (MEN) 2A and 2B, familial paraganglioma (PGL) syndrome and rarely neurofibromatosis-1 (NF-1), MEN-1 and tuberous sclerosis.¹

It is a rare neoplasm in children. Among the hypertensive children, the incidence of surgically confirmed disease ranges from 0.8% to 1.7%.³ The prognosis for completely resectable tumour is excellent.¹

This report describes this rare condition in a 12 years old boy.

CASE REPORT

A 12 years old boy was admitted to the Paediatric Intensive Care Unit (PICU) with acute onset of coma and

signs of raised intracranial pressure. He had an ongoing history of headache and vomiting for 6 months, and irritability and altered sensorium for 3 days. Headache was associated with episodes of sweating, palpitation and abdominal pain. However, there was no history of fever, fits, urinary complaint, chest pain, shortness of breath, petechiae, bruises, oedema, rash or blood transfusion. His past history and family history was insignificant with no history of contact with tuberculosis or any other illness.

On admission, he was unconscious and had height and weight on 50th centile for age. His sensorium was 7/15 on Glasgow Coma Scale. Blood pressure was noted to be very high at 240/150 mm of Hg in upper limbs and 250/150 mm of Hg in lower limbs. Pulse rate was 160 beats/minute, regular and with no radial-femoral delay. He had extensor plantar response with normal tone and reflexes. Fundoscopic examination showed grade four papilloedema with macular stars and cotton wool spots suggestive of malignant hypertensive retinopathy. Rest of the general physical and systemic examination was normal.

The patient was shifted to PICU for management of hypertensive encephalopathy. In addition to supportive treatment and monitoring, he was started on intravenous hydralazine. Later other antihypertensive drugs like diuretics, amlodipine and ACE inhibitors were added. Cerebral oedema was treated with meticulous fluid balance and mannitol. Blood pressure was gradually decreased and patient was normotensive on oral antihypertensive medication by day 5.

The routine investigation including complete blood counts, renal profile, liver profile, serum electrolyte, blood glucose, urine complete analysis, CSF examination and lipid profile, were all normal. Chest X-ray was normal. E.C.G showed evidence of left ventricular strain pattern. Echocardiography showed left ventricular

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hypertrophy with normal structure and no evidence of coarctation of aorta. Initial renal and abdominal ultrasound scan was reported to be within normal limits. Renal Doppler study showed no evidence of renal artery stenosis. Further investigations for the cause of hypertension showed serum renin level of more than 10.8 ng/ml/hr (normal: supine 0.15-2.33 ng/ml/hr, erect 0.31-3.95 ng/ml/hour). A 24-hour urinary VMA level was also high at 29 mg/24 hours (normal < 13.6 mg/24 hours). A further detailed ultrasound scan of abdomen showed a right paraspinal mass measuring about 3-4 cm. CT scan of abdomen showed 4 x 2 cm homogeneously enhancing mass at right supra-renal region separate from right kidney, partially compressing the inferior vena cava with no calcification (Figure 1).

On the basis of clinical features, raised urinary VMA, and right suprarenal mass diagnosis of pheochromocytoma was made. Patient was subsequently taken to theatre and the mass was resected by the paediatric surgical team, with close peri-operative and postoperative monitoring. Histopathology of the mass confirmed the diagnosis of pheochromocytoma of adrenal origin. Patient was discharged on 8th postoperative day without any antihypertensive medicine. He has been normotensive since and is under follow-up.

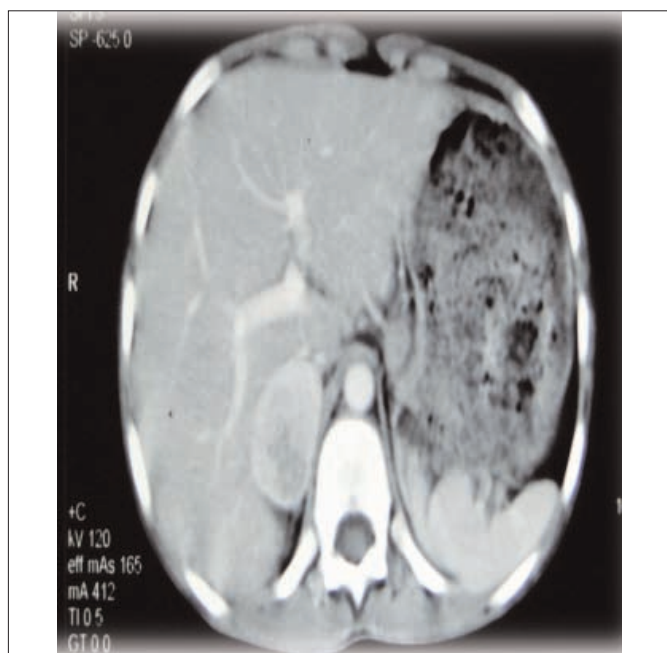


Figure 1: A rounded 4 x 2 cm enhancing mass in right suprarenal area, partially compressing inferior vena cava..

DISCUSSION

Pheochromocytoma is a neuroendocrine tumour of adrenal medulla or extra-adrenal chromaffin tissue.⁴ It secretes excessive amounts of catecholamines, usually norepinephrine and to lesser extent epinephrine.⁵ A tumour that arises from adrenal medulla is termed as pheochromocytoma, and those with extra-adrenal origin

are called paraganglioma. Both of these tumours are rare in children.⁶

Pheochromocytoma is more common in fourth and fifth decade of life with slight female predisposition.⁷ About 90% of cases occur sporadically and are benign.⁷ About 10-12% of cases occur in children at an average age of 11 years.¹

Clinical features of pheochromocytoma consist of classic triad of episodic headache, sweating, tachycardia and usually accompanied by hypertension.² Symptoms of mass effect such as abdominal pain, abdominal distension and backache are reported in less than 1/3rd cases in children.⁶ In contrast to adult, most children present with sustained hypertension.^{3,6} If not detected early, children may present with malignant hypertension and its complications like increased intracranial pressure and encephalopathy.³ Other less frequent features are pallor, orthostatic hypotension, constipation, psychiatric disorder, increased ESR, hyperglycemia and dilated cardiomyopathy.^{3,8}

Increased urinary excretion of VMA (major metabolite of epinephrine and norepinephrine) helps in diagnosing pheochromocytoma.⁵ Tumour is localized by USG, CT, or MRI and small tumours can be localized by MIBG scan - if available.⁹ As has been seen in this case, small tumours may be missed on routine ultrasound scan especially if performed by less experienced radiologist.

Approximately 10% of all pheochromocytoma are malignant but fortunately these tumours are rare in children and mostly extra-adrenal in origin. Surgical removal of these tumours is curative, but require meticulous pre-operative, intra-operative and post-operative care. Pre-operatively alpha and beta adrenergic blockade and fluid loading are required. Appropriate anaesthesia and expansion of blood volume is required to prevent fall in blood pressure intra-operatively and within 48 hours postoperatively. Long-term follow-up is indicated because functional tissue at other site may be manifested many years after initial surgery.^{8,10}

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