Hypertensive emergency due to vasculitis: a case report

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ABSTRACT

Hypertensive emergencies commonly occur due to an underlying renal or reno-vascular disease. Vasculitis is an uncommon cause. We report a 5 year old boy who presented with hypertension, heart failure and posterior reversible encephalopathy syndrome (PRES). Subsequent evaluation led to the diagnosis of polyarteritis nodosa (PAN). PAN is an uncommon but well known cause of hypertension. However, hypertensive encephalopathy and PRES have been rarely reported in childhood PAN.

Keywords: hypertensive emergency, posterior reversible encephalopathy syndrome, vasculitis, polyarteritis nodosa, CT angiography

Introduction

Hypertensive emergency (HE) is an uncommon but potentially life-threatening condition in children.¹ The commonest cause for HE in infants and children is a renal or reno-vascular abnormality whereas in older children and adolescents, primary hypertension is more common.²,³ Extensive evaluation may become necessary when renal and reno-vascular diseases have been ruled out. We describe a young child presenting with hypertensive encephalopathy who was subsequently diagnosed with polyarteritis nodosa (PAN).

Case report

A 5 year-old-boy presented with a 2-day history of irritability followed by progressive drowsiness and jerky movements of the lower limbs. After an Electroencephalography (EEG) and magnetic resonance imaging (MRI) of the brain had been performed, he was referred to our hospital.

On arrival, he had a Glasgow Coma Scale (GCS) of 3/15, shallow respirations, marked sinus tachycardia (150/min), feeble pulses, mottled extremities and blood pressure (BP) of 150/100 (110) mm Hg. His pupils were equal in size and reacting normally. Heart sounds were normal. There were no focal neurological deficits or signs of meningeal irritation. Functional echocardiography showed a dilated and dysfunctional left ventricle (LV) with hypertrophy. A provisional diagnosis of hypertensive encephalopathy with heart failure was made.

The child was intubated and ventilated in view of his low GCS. A 10 ml/kg fluid bolus was given slowly and he was started on a nitroglycerine infusion (starting at 0.5 μg/kg/min). Labetalol infusion was added subsequently (beginning at 0.5μg/kg/min). Due to poor cardiac contractility, milrinone was started at 0.5 μg/kg/min. He was empirically started on ceftriaxone and acyclovir for possible central nervous system (CNS) infections.

On detailed enquiry, the child had a 6-month history of intermittent fever and lethargy, with weight loss since 3 months. He had been extensively evaluated for an infectious etiology. Complete blood counts, blood and urine cultures, smear for malarial parasite, serology for dengue, typhoid fever and scrub typhus were negative. He had anorexia and intermittent abdominal pain for one week prior to admission. On detailed clinical examination, the child weighed 11 kg, and was thinly built with appropriate height for age. He had no organomegaly or lymphadenopathy. His fundus examination was normal.

Investigations showed a total leucocyte count of 31300/ mm³ with 18% stabs, 4% metamyelocytes, 1% myelocytes. Erythrocyte sedimentation rate was 54 mm/hr. C-reactive protein was 57.2 mg/L. Renal function, electrolytes, liver function, Creatine Kinase,
Troponin I and Serum ammonia were normal. Serology for hepatitis B, hepatitis C, HIV I and II were negative. MRI brain were suggestive of posterior reversible encephalopathy syndrome (PRES). His EEG was normal.

**Figure 1:** MRI Brain suggestive of posterior reversible encephalopathy syndrome

An evaluation for the cause of hypertension was undertaken. Echocardiography confirmed the presence of LV dysfunction and hypertrophy. The ejection fraction (EF) was 40%. The coronary arteries were normal. Abdominal ultrasonography showed a pseudoaneurysm arising from a mesenteric vessel most likely the right gastroduodenal artery. The kidneys and adrenals appeared normal. No other masses were noted. Computerised tomography (CT) angiography of the chest and abdomen confirmed the presence of a partially thrombosed pseudoaneurysm of the right gastroepiploic artery with an aneurysm of the right hepatic artery. The aortic arch, its branches and other thoracic and abdominal vessels were normal. Both kidneys appeared large and showed a striated nephrogram. His 24-hour urinary vanillyl mandelic acid (VMA) was normal. A diagnosis of polyarteritis nodosa (PAN) was made.

As his BP stabilised over the next 72 hours, his pulses and perfusion improved. He was weaned off nitroglycerine (maximum dose 10 μg/kg/min), labetalol (maximum dose 4 μg/kg/min) and milrenone. Enalapril was started. His consciousness improved and he was successfully extubated on day 4 of admission. Carvedilol and hydralazine were subsequently added to his antihypertensive regime. Ceftriaxone and acyclovir were stopped.

In the high dependency area, he developed right partial seizures which were treated with levetiracetam. MR angiography of the brain showed white matter changes consistent with a vasculitic disease but no infarcts. The angiography was normal. He was treated with methylprednisolone at 30mg/kg for 3 days followed by oral prednisolone. The antihypertensives were continued and he was discharged after 14 days of hospitalisation.

Two weeks after discharge, he was noted to have reticulate rash over his feet (livedo reticularis) and right sided foot drop both consistent with the diagnosis of PAN. He was started on monthly doses of cyclophosphamide at 500 mg/m². At six weeks, his echocardiography showed a normal myocardial function with an EF of 60%. His BP was below the 90th centile.

**Discussion**

The initial evaluation of hypertension in children includes a detailed history and clinical examination (including anthropometry with calculation of Body Mass Index) and basic investigations (complete hemogram, renal function tests, urinalysis, abdominal ultrasonography and chest radiography). Subsequent evaluation, especially when the renal status is normal, may vary based on initial findings. An abdominal ultrasonography with a doppler study would identify problems related to the renal parenchyma, renal vasculature, adrenals, any suspected masses and visceral artery abnormalities such as aneurysms, and as such appears to be the single most useful investigation in such patients.

Systemic PAN is a medium-vessel vasculitis estimated to have an incidence of one per one million children, comprising about 3% of all vasculitides in children. Rheumatological and vasculitic disorders constitute a very small percentage of children presenting with hypertensive crises. Nonetheless, hypertension is a common feature of PAN and is...
considered to be secondary to increased plasma renin. A prospective study in children in East India found that 93% of children with PAN were hypertensive though none of them had abnormal renal function or renal vasculature.

The most common organs/vessels involved in PAN are the skin, muscles, kidneys and gastro-intestinal tract while cardiac and neurological involvement are less common. Constitutional symptoms such as fever, weight loss and muscle or joint pains are seen at presentation. Depending on the organs involved, skin changes (livedo reticularis, purpura, necrosis, painful subcutaneous nodules), hypertension, hematuria, proteinuria, ischemic heart disease, abdominal pain, testicular pain or mononeuritis multiplex (sensory or motor) may be present. While the disease generally evolves insidiously, acute presentations including hypertensive crises, stroke, digital gangrene and even myocardial infarction have been described in children. PRES has been reported rarely in childhood PAN.

The diagnosis of PAN (European League against Rheumatism/Pediatric Rheumatology European Society criteria) requires the presence of either histopathological or angiographic abnormalities along with two out of seven clinical criteria. Though data are limited, childhood PAN seems to have a better outcome compared to adults. Reported mortality varies from 1.1 to 4% while end-stage renal disease has been reported in 1.8% of children. The long term management protocol involves a prolonged course of steroids with cyclophosphamide or azathioprine as adjuvants.

Various case reports have described the management of hypertensive crises in PAN with sodium nitroprusside and labetalol in the acute phase. The use of angiotensin converting enzyme inhibitors (enalapril and lisinopril), thiazides (hydrochlorothiazide) and labetalol has been described for long term management in adults.

The management of heart failure in the presence of hypertensive encephalopathy is challenging. A rapid normalisation of blood pressure to improve cardiac output by afterload reduction could not be achieved due to the risk of cerebrovascular compromise. Milrenone was therefore added as a bridge to augment cardiac output during the interim 48-72 hours.

In conclusion, vasculitis should be considered as a differential diagnosis in a child with a hypertensive emergency especially when the renal function is normal and no other cause is apparent. The presence of visceral artery aneurysms on the background of prolonged fever, weight loss and abdominal pain are pointers towards PAN. Abdominal ultrasonography and CT/ MR angiography are useful when the etiology is obscure.

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References
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