International Journal of Contemporary Pediatrics
Thomas M et al. Int J Contemp Pediatr. 2019 Sep;6(5):2225-2227
http://www.ijpediatrics.com
pISSN 2349-3283 | eISSN 2349-3291

DOI: http://dx.doi.org/10.18203/2349-3291.ijcp20193758

Case Report

Paroxysmal hypertension with reversible posterior leukoencephalopathy syndrome in a child: an unresolved riddle

Mili Thomas[1]*, Veena V. Nair[2], Lekha Hrishikesan[3], Kamala Swarnam[1]

[1]Department of Pediatrics and Neonatology, [2]Department of Pediatric Endocrinology, [3]Department of Nephrology, Ananthapuri Hospitals and Research Institute, Chackai, Thiruvananthapuram, Kerala, India

Received: 18 May 2019
Revised: 30 June 2019
Accepted: 02 July 2019

***Correspondence:**
Dr. Mili Thomas,
E-mail: militreesa@gmail.com

ABSTRACT

Nine year old girl was presented with paroxysmal episodes of hypertensive emergency. She was asymptomatic with normal blood pressure without antihypertensives in between the episodes. MRI brain was suggestive of reversible posterior leukoencephalopathy. Acute episodes were managed with IV labetalol infusion and amlodipine. She was evaluated extensively to find out the etiology of hypertension. Cardiac and renal causes were ruled out. Work up for pheochromocytoma, hyperaldosteronism, porphyria and vasculitis were negative. The case is reported in view of the rare presentation and the leukoencephalopathy noted in this case.

Keywords: High blood pressure, Hypertensive emergency, Paroxysmal hypertension, Pseudopheochromocytoma children, Reversible posterior leukoencephalopathy

INTRODUCTION

Hypertensive emergency is characterized by acute severe elevation of blood pressure along with end organ damage like hypertensive encephalopathy, cardiac failure or renal disease. The evaluation of children with hypertensive emergency includes recognition of target organ damage and identifying the underlying etiology.[1,2] Paroxysmal hypertension warrants evaluation for pheochromocytoma.[3] Sometimes the evaluation for etiology may be negative making the diagnosis and management difficult. A nine year old girl who presented with paroxysmal hypertension and posterior reversible encephalopathy is reported.

CASE REPORT

Nine year old non obese girl referred to the emergency department with history of vomiting and upper abdominal pain for 6 days, one episode of transient loss of vision and high blood pressure. No history of headache, seizures, cola colored urine, edema, sweating, palpitation, flushing, rashes or diarrhea. No history of drug intake. There was no stress at home or school. At admission she was conscious, dehydrated. She had tachycardia and BP was160/100mm Hg, There was no pallor/edema. All peripheral pulses were palpable. There was no cardiac murmur, bruit over major vessels or focal neurological deficits. Fundus examination was normal. No family history of hypertension. Blood pressure got controlled with oral amlodipine.

One year back she was admitted with vomiting, abdominal pain and one episode of abnormal gaze. Blood pressure at admission was 140/90mm Hg. Further BP measurements were normal. EEG was normal. MRI brain was suggestive of reversible posterior leukoencephalopthy syndrome/ encephalitis. CSF study

was normal. She was treated symptomatically. Repeat MRI done 3 months later was normal. Blood pressure measurements on follow up were within normal limits.

Observation

MRI brain showed symmetrical T2W/FLAIR cortical and subcortical hyperintensities in parietal, occipital and middle frontal gyrus of frontal lobe which was suggestive of reversible posterior leukoencephalopthy syndrome (RPLS) (Figure 1).

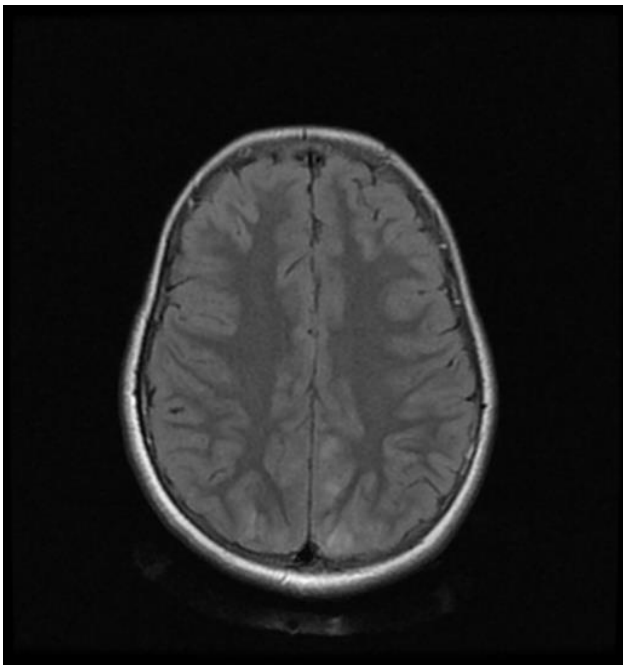

Figure 1: MRI brain T2W image showing symmetrical cortical and subcortical hyperintensities in the occipital lobe.

She was extensively worked up for hypertension. Renal, renovascular and cardiac causes were ruled out. Serum calcium, Thyroid function, Plasma renin activity and aldosterone concentration were within normal limits. Work up for pheochromocytoma including MRI abdomen, urine 24 hour metanephrine, normatanephrines and plasma free metanephrine, normatanephrines was normal. Vasculitis work up was negative. Urine porphobilinogen was negative.

Amlodipine was stopped as blood pressure was consistently low. Blood pressure was monitored daily and was 90/60mm Hg without antihypertensives for 2 weeks. Again she presented with similar symptoms, blood pressure was 160/ 110mm Hg. As she was not tolerating orally IV Labetalol infusion was started at 0.25mg/kg / hour and then increased to 0.4mg/kg/hour and blood pressure got controlled.

As the child had paroxysmal hypertension with RPLS she was advised for further evaluation like MIBG scan for pheochromocytoma, psychological counseling to find out stress factor for pseudopheochromocytoma, repeat plasma metanephrine and normetanephrine during hypertensive crisis and 5 hydroxy indole acetic acid for carcinoid syndrome and 24 hour urine porphobilinogen for acute intermittent porphyria. Due to financial constrains it was not done. Amlodipine was started and then tapered and stopped. No definite diagnosis has been made till now. She is on follow up with normal blood pressure without medications for last 6 months.

DISCUSSION

Severe and symptomatic hypertension in children is usually due to secondary hypertension. Intermittent hypertension can be seen in renal diseases, drugs and poisons (cocaine, sympathomimetics, heavy metals, vitamin D) central and autonomic nervous system disorders, pheochromocytoma, hypercalcemia, porphyria etc. Children with pheochromocytoma have sustained rather than intermittent hypertension.[4] Other causes of paroxysmal hypertension includes pseudopheochromocyoma, labile hypertension and panic attacks.

Pseudopheochromocytoma is due to activation of the sympathetic nervous system due to unrecognized emotional factors that have been kept from conscious awareness.[5,6] Labile hypertension manifests with transient blood pressure elevation along with a stress that the patient is aware of. Panic attack can also be associated with elevation in blood pressure.[7]

Reversible posterior leukoencephalopathy is due to disordered cerebral autoregulation and endothelial dysfunction that occurs in hypertensive encephalopathy, eclampsia, and the use of cytotoxic / immunosuppressant drugs. Subcortical white matter in the posterior cerebral hemispheres is characteristically affected. Prompt recognition and treatment is important in preventing the permanent damage that can occur in this otherwise typically reversible condition.[8]

Our patient had 3 episodes of hypertensive emergency with reversible posterior leukoencephalopathy syndrome. She was asymptomatic with normal blood pressure in between the episodes. All major etiologies were ruled out. We couldn't find out the exact etiology of her paroxysmal hypertension and she needs further work up for the same. The case is reported in view of the rare presentation and the leukoencephalopathy noted in this case.

Funding: No funding sources
Conflict of interest: None declared
Ethical approval: Not required

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Cite this article as: Thomas M, Nair VV, Hrishikesan L, Swarnam K. Paroxysmal hypertension with reversible posterior leukoencephalopathy syndrome in a child: an unresolved riddle. Int J Contemp Pediatr 2019;6:2225-7.