

## CASE REPORT

## POSTERIOR REVERSIBLE ENCEPHALOPATHY SYNDROME IN A CHILD WITH POST STREPTOCOCCAL GLOMERULONEPHRITIS- A CASE REPORT

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**ABSTRACT**

Posterior reversible encephalopathy syndrome is a condition occurring in majority of case of hypertensive encephalopathy mainly affecting white matter of parieto-occipital region in neuroimaging. It is reversible if timely treatment of hypertensive emergency is done. Here, we are reporting a 11-year-old female child with acute post streptococcal glomerulonephritis leading to typical clinical and radiological features of posterior reversible encephalopathy syndrome.

**Keywords:** Hypertension; Posterior reversible encephalopathy syndrome; Post streptococcal glomerulonephritis

**INTRODUCTION**

Posterior reversible encephalopathy syndrome (PRES) is defined as a clinic-radiological condition characterized by signs and symptoms like headache, seizures, visual abnormalities, impairment of consciousness along with hyper intense lesion in parieto-occipital region of posterior cerebral hemisphere. It was first described by Hinchey et al in 1996.<sup>1</sup> The other names given for PRES are reversible posterior leukoencephalopathy syndrome, reversible posterior cerebral edema syndrome, and reversible occipital parietal encephalopathy.<sup>2</sup> The majority of patients with PRES present with impairment in consciousness, headaches, seizures, visual abnormalities, nausea/vomiting and focal neurological signs.<sup>2</sup> The occurrence of signs and symptoms in PRES is variable, can occur as early as within an hour to 3 months after the exposure to the insult.<sup>3</sup>

The common etiologies of PRES are hypertension, systemic infections, organ transplants, autoimmune

diseases (SLE) malignant tumors, chemotherapy and immuno-suppression.<sup>4</sup> The main pathology of PRES is vasogenic edema leading to white matter changes in neuroimaging. Posterior cerebral hemisphere (mainly parieto-occipital region) is most commonly affected region in PRES. The most common cause of acute hypertension in children is renal parenchymal diseases like acute glomerulonephritis in pediatric population.<sup>5</sup>

**CASE PRESENTATION**

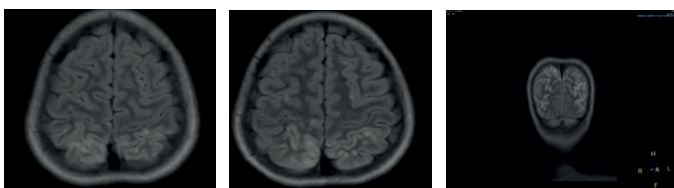
A 11 years old girl presented to Emergency Department with chief complaints of swelling of body starting from face, shortness of breath for 4 days duration. She also presented with headache in temporal and occipital region for 1 day duration. She also complained of sudden blurring of vision for 30 minutes after which she was rushed to hospital by parents. Blurring of vision diminished on the way to the hospital. There was history of red colored urine 1 day back. There was no history of oliguria, drug

intake, head trauma and past history of skin infection.

The patient had no history of surgeries in past, no history of drug allergies. There was no history of hypertension and diabetes or other medical diseases in the past. She was born from a non-consanguineous marriage and her developmental history was age appropriate. She had completed her immunization according to National immunization program of Nepal.

On admission, child was conscious, cooperative, with pediatric Glasgow Coma Scale Score of 15/15(E4M5V5). Her pulse rate was 126bpm, respiratory rate was 30/min and facial puffiness was present. Her blood pressure was (164/124) mm of Hg in right upper arm, (>95<sup>th</sup> centile for age and sex). For blood pressure monitoring, patient was immediately shifted to Pediatric ICU. There was no neuro-cutaneous marker on head to toe examination. Her pupils were equal in size and equally reactive to light. Fundus examination of both eyes was within normal limit. Meningeal signs were absent. Deep tendon jerks were normal. There was no focal neurological deficit. Her other systemic examination was within normal limit.

Her laboratory investigations revealed hemoglobin of 11 gm/dl, white blood cell count 15,300/cumm and platelet counts was 2lakh/cumm. Anti Streptolysin O (ASO) titre was positive (>200IU/l) and serum C3 level was very low (0.02gm/l). Her urine microscopic examination showed hematuria along with proteinuria. Ultrasound of abdomen and pelvis and renal Doppler studies were found to be normal. Her renal function tests, liver function tests were within normal limit. The patient was admitted with provisional diagnosis of acute post streptococcal glomerulonephritis with hypertensive emergency on basis of clinical and laboratory findings. For the treatment of hypertensive emergency Labetalol infusion was started for 48hrs along with oral amlodipine and furosemide. Her blood pressure gradually came to normal within 48hrs of admission. Meanwhile her MRI of brain showed hyper intense lesion in bilateral parieto-occipital region suggestive of PRES. She was also given Inj. Mannitol in between to release cerebral edema.



**Figure: T2 and FLAIR hyperintense area involving bilateral parieto-occipital cortex and sub-cortical white matter showing no restriction diffusion**

She had one episode of generalized tonic clonic seizures on the day of admission. She received Inj.

diazepam for this episode and seizure was controlled. She was discharged with the final diagnosis of Acute Post Streptococcal Glomerulonephritis with Posterior Reversible Encephalopathy Syndrome after five days of hospital stay on oral Amlodipine. Repeated c3 level and MRI could not be done due to financial constraints.

## DISCUSSION

There are many theories associated with pathogenesis of PRES. Hyper perfusion theory is one of them which states that when systemic arterial pressure becomes greater than the cerebral auto regulation, it leads to hyper perfusion resulting in increase in permeability of blood brain barrier causing vasogenic edema. Usually posterior circulation is more affected than anterior because of less availability of sympathetic innervations. The another theory states that PRES is a result of cytotoxic edema occurring due to endothelial damage caused by circulating or direct toxic agents.<sup>6</sup>

Most common cause of hypertension in children is renal parenchymal diseases, among them 10% of cases are complicated with hypertensive encephalopathy.<sup>7</sup> Hypertension encephalopathy with renal disease was first described in 1928.<sup>8</sup> Only 70% to 80% of children with PRES have systemic hypertension.<sup>9</sup> Seizures is most common symptom in PRES, followed by encephalopathy, headache, visual disturbances and focal neurological deficits. Initially seizures are localized gradually progress to become generalized, ultimately leading to status epilepticus in most of the cases.<sup>10</sup> There may be drowsiness, confusion, restlessness, agitation, stupor and frank coma. The visual symptom of PRES may vary from blurring of vision, homonymous hemianopsia to even cortical blindness.<sup>6</sup>

The gold standard for diagnosis of PRES is T2weighted MRI, fluid-attenuated inversion recovery (FLAIR). T2 weighted MRI shows region of high signal intensities suggesting of cerebral edema and FLAIR helps in detection of subcortical and cortical lesions in PRES. The four main MRI pattern of PRES are holo-hemispheric watershed pattern, superior frontal sulcus pattern, dominant parietal-occipital pattern and partial expression of the three primary patterns. MRI is considered superior to CT scan for the diagnosis of PRES.<sup>2</sup> Diffusion weighted MRI helps to differentiate between vasogenic and cytotoxic edema.<sup>6</sup> Though PRES is reversible, may lead to complications like cerebral ischemia, cerebral hemorrhage and cerebral herniation.<sup>2</sup>

Early diagnosis and prompt treatment of cause along with hypertension remains the main objective of the case management. Immediate correction of high blood pressure along with discontinuation of offending agents provides early recovery of patients. MAP should

be decreased by 20-25% within first 2 hours. Rapid reduction of blood pressure can lead to alterations in cerebral perfusion pressure resulting in cerebral ischemia.<sup>11</sup> In our case blood pressure of the patient was gradually reduced and came to normal range after 48hrs of admission.

## CONCLUSION

PRES should be suspected in any case of acute glomerulonephritis which may be presented with hypertensive emergency, new onset of seizures and blurring of vision. Early diagnosis and treatment are the main modalities of treatment. PRES is reversible with good prognosis if early intervention is done.

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