# Steroid Sensitive Nephrotic Syndrome Presented as Posterior Reversible Encephalopathy Syndrome - A Rare Case Report

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## ABSTRACT

Posterior reversible encephalopathy syndrome (PRES) is a rare serious syndrome of central nervous system that can develop in both adults and children. It is characterised by acute onset of headache, confusion, seizures, or focal neurological deficits along with radiological abnormalities in the parietal and occipital lobes. In the past, this syndrome has been mainly described in adults but rare in children. However, it is not uncommon in paediatric nephrology. Hypertension, renal disease, immunosuppression, and chemotherapy of malignancies are triggers for PRES. Here, we report a case of 12 years old boy with steroid-sensitive nephrotic syndrome presenting as PRES.

**Keywords**: Hypertension; Posterior reversible encephalopathy syndrome; Steroid-sensitive nephrotic syndrome

## INTRODUCTION

Posterior reversible encephalopathy syndrome (PRES), a rare disorder of central nervous system, was first described by Hinchey as a variable combination of impairment of consciousness, seizure activity, headaches, vomiting, visual abnormalities or cortical blindness and focal neurological signs. Radiological findings closely resemble hypertensive encephalopathy, which includes symmetric distribution of patchy or globe-like lesions, partially or completely reversible subcortical vasogenic edema in the posterior white matter.[1-3] PRES can develop in various clinical conditions like autoimmune diseases, systemic infections, pre-eclampsia, hypertension, organ transplantation, malignancies, chemotherapy and immunosuppression especially with calcineurin inhibitors.[2,3] Children on prolonged steroid therapy or calcineurin inhibitor therapy in nephrotic syndrome and chronic kidney disease are at risk of developing PRES.[4] Reports of PRES are scarce in the paediatric population. Here, we report a case of a 12 year child with steroid-sensitive nephrotic syndrome presenting as hypertensive encephalopathy in emergency.

## CASE REPORT

A 12 year old boy, presented with chief complaints of generalized body swelling for 10 days, decreased urine output for two days and headache, vomiting, abnormal body movements and diminution of vision for one day. There was no history of fever, cough, pain abdomen, hematuria, loose stool, jaundice, rash, loss of consciousness or altered sensorium. Also, there was no history of head trauma, drug intake or any similar history in the past. On examination child was lethargic, afebrile with vitals PR - 112/min, RR - 26/min, BP - 152/96 mm Hg (> 99$^{th}$ centile) and all peripheral pulses were well palpable. GCS was E3V4M5 = 12/15 and he was not able to count finger or recognise parents, there was no ptosis or rectus muscle palsy, pupil was normal size and reactive to light. Rest of the examination was within normal limits.

Laboratory examination revealed proteinuria of 4+, low serum albumin of 2.0 g/dl, high serum cholesterol of 524.5 mg/dl and urinary protein to creatinine ratio of 9.93. All of these findings were suggestive of nephrotic syndrome. Haemoglobin was 11.9 gm/dl, total leucocyte count- 16719/mm$^3$ (N 75% and L 16%), Blood urea nitrogen - 49.5 mg/dl, serum creatinine - 0.6 mg/dl, aspartate aminotransferase / Alanine aminotransferase 19/26 units/litre, serum calcium - 8.9 gm/dl, ESR - 9 mm/ 1$^{st}$hr. Mantoux was negative, HIV test, Hepatitis B surface antigen and antibody to hepatitis C virus were non-reactive, fundus examination was within normal limits, ultrasonography of abdomen including kidney, ureter and bladder was suggestive of bilateral renal parenchymal disease. Urine culture and blood culture was sterile and urinary electrolytes was within normal limit. Collagen profile showed - C3 - 137 mg/dl (79 - 152 mg/dl), C4 - 45.4 mg/dl (16-38 mg/dl), antinuclear antibody 0.3 IU/ml (< 1.0 IU/ml) and anti-double stranded DNA 13 IU/ml (< 20 IU/ml). Subsequently Contrast Enhanced Computed Tomography head was done which showed ill-defined hypo dense areas in right frontal, bilateral parietal lobes suggestive of non-specific edema. This CT imaging features were suggestive of PRES. (figure 1)

The child was managed supportively and symptomatically with antihypertensive medications, initially started with amlodipine and later shifted on ACE inhibitor, anticonvulsants and diuretic. Prednisolone was started as per ISPN Guidelines for first episode nephrotic syndrome.[5] Child regained vision and full consciousness within two days. No further episode of seizure was noted during the hospital stay. At discharge his blood pressure was 106/70 mm Hg (between 50$^{th}$ - 90$^{th}$ centile), urinary protein was absent and edema subsided. Repeat CT cranium was done which was normal and child is doing well in follow up

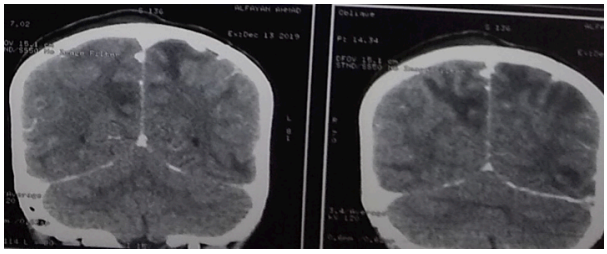

**Figure 1.** Ill-defined hypo densities areas, noted in right frontal, bilateral parietal lobes suggestive of non-specific edema.

probably due to prompt initiation of antihypertensive therapy.

## DISCUSSION

PRES can be diagnosed according to typical clinical manifestations and brain imaging. Patients generally present with headache, vomiting, visual perception abnormalities and seizures. Other studies also state that PRES should be one of the differential diagnosis in a child with nephrotic syndrome, if presented with headache and visual disturbance.[6,7] The exact pathophysiological mechanism of PRES is still not clear. Three hypotheses have been suggested, which include: first, after exposure to causative agent (severe hypertension), auto regulation mechanism of intracranial pressure fails, leading to vasogenic edema; second, after exposure to causative agent (mild-to-moderate hypertension), cerebral vasoconstriction and hypo perfusion cause vasogenic brain edema and ischaemia; third, endothelial injury with disruption of the blood-brain barrier leads to fluid and protein transudation in the brain.[2] It can also result from the failure of the cerebrovascular auto-regulation mechanisms due to sudden elevations in blood pressure and secondary hypertension in children is commonly related to renal changes, suggesting that these are the main etiologies of PRES.[8]

Ishikura et al. showed that PRES may occur during moderate to severe nephrotic state in most of the paediatric patients.[4] Several additive factors predispose to the development of PRES in these patients, namely low serum albumin level, generalised edema, increase in vascular permeability, unstable fluid status and renal insufficiency along with child receiving immunosuppressive drugs like cyclosporin.[4] Gera et al also stated that PRES is not uncommon in paediatric kidney disease. Hypertension, renal disease and immunosuppression are triggers for PRES.[9] However, in our case most unusual part was that child developed PRES at initial presentation and he was not taking any immunosuppressive drugs and calcineurin inhibitor.

Prompt management of PRES leads to favourable outcome. It has been reported that outcome of PRES is satisfactory after withdrawal of the offending drugs or improvement of the etiological factors like hypertension.[10] PRES is mostly fully reversible in a day to weeks. However, prolonged seizures and hypertension may result in death or permanent neurological disability. Early recognition and timely management are important to prevent irreversible neurological damage.

PRES with nephrotic syndrome has mostly been associated with steroid dependent [6,7] and resistant nephrotic syndrome or it may develop during the course of illness. We reported this case as this was a very rare presentation of very common disease nephrotic syndrome presenting as PRES. High degree of suspicion is required in all cases of nephrotic syndrome presenting with neurological manifestations even in the first time with seizures, visual disturbances, headache, and altered mentation to diagnose PRES.

## CONCLUSIONS

PRES should be considered one of the possibilities in children with nephrotic syndrome even at first diagnosis if, presenting with seizures, visual disturbances, headache, and altered mentation. Early recognition is crucial for timely diagnosis and treatment resulting minimal permanent neurologic damage.

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