

Case Report

Posterior reversible encephalopathy syndrome seen in a case of acute post-streptococcal glomerulonephritis: a case report

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ABSTRACT

Posterior reversible encephalopathy syndrome (PRES) is a clinical and radiological condition that involves the acute onset of headache, confusion, optical impairments, and seizures with accompanying vasogenic edema on brain imaging. Here is a 12 years old male patient presented with complaints of swelling in the face and legs, headache and vomitings. He had a history of fever, sore throat and tonsillitis 12 days back. On physical examination weight and height of the patient was between 25-50th percentile, BP was 142/92 mmHg (>99th centile), he had pretibial +2 edema and periorbital edema. At presentation urine analysis showed haematuria, proteinuria at nephritic level, elevated urea and creatinine, increased ASO titre, low C3 and C4 levels. After admission, he had an episode of convulsion when BP was 162/98 mm hg. Magnetic resonance imaging brain showed bilateral occipital, parietal, frontal cortex and subcortical white matter T2/Fluid-attenuated inversion recovery (FLAIR) hyperintensity, suggestive of PRES. EEG was found to be normal. MRI was found to be normal in the first month after starting on antihypertensive and anticonvulsive treatment. In the first year of the follow-up, the blood pressure, neurological examination and urinalysis findings were found to be normal. Thus, it is suggested that early diagnosis of PRES in patients with clinical features and prompt treatment will prevent permanent neurologic sequelae.

Keywords: Acute glomerulonephritis, Childhood, Posterior reversible encephalopathy syndrome

INTRODUCTION

Posterior reversible encephalopathy syndrome (PRES) is a clinical and radiological condition that involves the acute onset of headache, confusion, optical impairments, and seizures with accompanying vasogenic edema on brain imaging.³ The radiological findings most commonly described include transient bilateral grey and white-matter changes compatible with vasogenic edema in the posterior cerebral hemispheres, parieto-occipital areas and cerebellum.¹ Although the exact pathophysiological mechanism is not completely understood and remains controversial, the predominant proposed mechanism is endothelial dysfunction, which is preceded by hypertension, immunosuppressive agents, or cytotoxic

medication.³ Though frequently described in adults, the incidence in pediatric population is being increased.² In children, PRES has been reported in association with Acute post-streptococcal glomerulonephritis (APSGN), Henoch-Schönlein purpura (HSP), Nephrotic syndrome (NS), lupus nephritis and use of calcineurin inhibitor.⁴⁻⁶ Here a pediatric patient who had vomitings, headache and seizure during the course of APSGN, and whose MRI findings were consistent with PRES has been presented.

CASE REPORT

A 12 years old male patient presented with complaints of swelling in the face and legs, headache and vomitings. He had a history of fever, sore throat and tonsillitis 12 days

back and has not been treated for it. The presenting complaints started after 12 days with swelling in face, legs, reduced urine output. Headache which is more in occipital and temporal regions and vomitings. On physical examination weight and height of the patient was between 25-50th percentile, BP was 142/92 mmHg (>99th centile), he had pretibial +2 edema and periorbital edema which was more in the morning. At presentation urine analysis results are: pH: 5, density: 1020, protein 2+, erythrocyte 3+, leukocytes 1+, urinary microscopy showed plenty of erythrocytes, 4-5 leukocytes. Other laboratory tests were: hemoglobin: 9.5 g/dl, hematocrit: 29.6%, white blood cells: 16460/mm³, platelets: 3,28,000/mm³, urea: 68 mg/dl, serum creatinine: 1.6 mg/dl, sodium: 134 mmol/l, potassium: 4.2 mmol/l, calcium: 7.4 mg/dl, phosphorous: 7.8 mg/dl, total protein: 6.2 g/dl, albumin: 3.2 g/dl, triglyceride: 130 mg/dl, total cholesterol: 190 mg/dl, urine protein/creatinine ratio: 1.2, anti-streptolysin O antibody: 356 IU/l, C3-32 mg/dl (N: 83-177), C4-26 (N: 15-45).

Acute post streptococcal glomerulonephritis was considered in the patient. The patient was advised fluid restriction and salt restriction. Diuretics (furosemide) and CCB (amlodipine) were started. After half an hour of admission, he had seizure activity in the form of upward deviation of eyes and tonic contraction of the whole body. Seizure activity controlled after giving midazolam. loading dose of phenytoin 20 mg/kg was given and treatment continued with maintenance doses. At the time of seizure, his bp was 169/90 mmHg. Sodium nitroprusside was used to control BP. Since he had seizures, headache and vomiting, MRI was ordered considering PRES. Magnetic resonance imaging brain showed bilateral occipital, parietal, frontal cortex and subcortical white matter T2/Fluid-attenuated inversion recovery (FLAIR) hyperintensities, suggestive of PRES. EEG was found to be normal. Follow up MRI after one month of hypertensive therapy was found to be normal. Antihypertensives were gradually tapered and discontinued. On follow up, neurological examination is normal, BP is within normal limits and urinary findings are within normal ranges over 6 months of follow up. Consent has been taken from the relatives of the patient to publish the case.

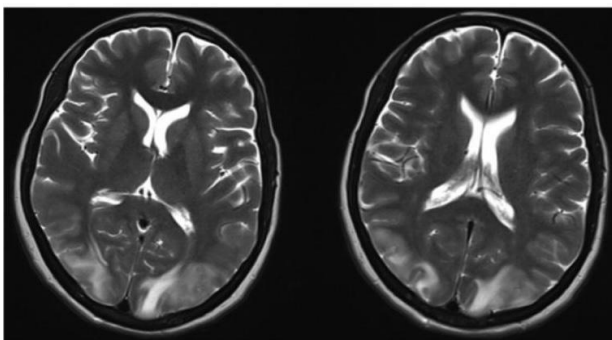


Figure 1: Axial view showing bilateral occipital, parietal frontal cortex and subcortical white matter T2/FLAIR hyperintensities.

DISCUSSION

Posterior reversible encephalopathy syndrome was first described by Hinchey et al in 1996 for the first time and it is gradually being more frequently recognized in children.^{1,7} PRES is a clinical and radiological condition that involves the acute onset of headache, confusion, optical impairments, and seizures with accompanying vasogenic edema on brain imaging. PRES is a complex disorder with many causative factors, including underlying conditions such as hypertensive encephalopathy, eclampsia, collagen disease, and severe infection. Although the exact pathophysiological mechanism is not completely understood and remains controversial, the predominant proposed mechanism is endothelial dysfunction, which is preceded by hypertension, immunosuppressive agents, or cytotoxic medication. Magnetic resonance imaging (MRI) facilitates prompt diagnosis and treatment and leads to good outcomes. Findings are characterized by the following: hyperintensity on fluid-attenuated inversion recovery images and apparent diffusion coefficient mapping, and isointensity on diffusion weighted images involving the parieto-occipital or posterior frontal cortical-subcortical regions that are recognized in >90% of patients, and reversibility of neuroimaging abnormalities, with the latter being the most important.³

PRES is generally observed with increased blood pressure. The blood pressure values were found to be above the 99th percentile in all 17 pediatric patients with PRES presented by Prasad et al.⁷ It was suggested that PRES is related with sudden increases in blood pressure.¹⁰ In our patient, the blood pressure value measured at the time of severe headache, vomiting and subsequent convulsion 162/98 mmHg, compared to BP at the time of presentation which was 142/92 mmHg. This suggested that PRES findings in our patient were related with the sudden increase in blood pressure. It is thought that functional vasomotor response that occurs because of exceeding the autoregulator capacity in PRES is transient. It is stated that the findings rapidly reverse with improvement of blood pressure.⁷ However, in contrast to the phrase 'reversible' in its name, PRES may not always be reversible.⁹ Permanent neurological damage can be observed due to prolonged hypertension and convulsions. Hence, permanent findings were observed in three of 17 cases of PRES presented by Prasad et al.⁷ Also in a series including 11 paediatric patients presented by Yamada et al chronic epilepsy was observed in two patients and abnormal EEG findings were observed in one patient.⁵ In our patient, the imaging findings were found to be normalized on MRI repeated in the first month of the follow-up.

Delayed in the diagnosis of PRES may lead to permanent neurological damage.^{8,9} The diagnosis should be promptly made with typical MRI findings along with clinical findings. The most common MRI finding is edema which is more prominent in the parieto occipital region.⁷ Involvement of the brainstem, cerebellum, basal ganglia

and even frontal lobes can also be seen.⁷ In our patient, lesions were observed in the right temporooccipital lobe.

The lesions in posterior reversible encephalopathy syndrome were observed to be hypointense on T1 weighted images, hyperintense on T2 weighted images and compatible with increased diffusion on diffusion weighted images. Vasogenic edema is the main finding of posterior reversible encephalopathy syndrome, in the presence of which water molecules surrounding the cells can move freely and increase in diffusion is seen. In conditions causing cytotoxic damage like infarction, a decrease in the movement of water molecules and thus a limitation in diffusion is observed due to decrease in the Na/K ATPase enzyme activity. Therefore, diffusion MRI is helpful to differentiate between PRES and ischemic conditions.⁹ It can be helpful in deciding treatment also. Hence, recent guidelines do not recommend reducing blood pressure in mild-moderate hypertension in ischemic stroke, whereas in PRES reducing blood pressure is the absolute treatment in order to reduce edema.⁴ Thus controlling blood pressure is main treatment recommended for posterior reversible encephalopathy. Thus, it is recommended to reduce blood pressure below the 99th percentile in the first hour or reduce the mean blood pressure by 25% during the initial 8 hours of treatment.⁹ Prolonged seizures due to lack of appropriate treatment and/or prolonged hypertension can lead to permanent neurological damage or brain infarction.^{5,7}

MRI findings observed in posterior reversible encephalopathy syndrome can be confused with gliomatosis cerebri, progressive multi-focal leukoencephalopathy, demyelinating conditions and infarcts. This can lead to unnecessary tests and treatment which can necessitate biopsy.^{1,7} Since early recognition of PRES results in prevention of severe neurological sequelae, keeping PRES in mind when it accompanies clinical findings is important.⁵ In our patient, clinical findings of severe vomiting, headache and seizures lead to suspicion of PRES and MRI showed findings consistent with PRES. Immediate measures to control seizures and reduction of BP were started. Thus, it has been possible to avoid permanent neurologic sequelae in our patient.

CONCLUSION

Conclusively, PRES must be primarily considered in the differential diagnosis when patients present with severe headache, seizure, visual disturbance or loss of consciousness and history is suggestive of acute glomerulonephritis, lupus and nephrotic syndrome and treatment should be directed to reduce blood pressure as soon as possible along with imaging studies including diffusion MRI. The present case developed PRES in the course of acute post-streptococcal glomerulonephritis when systolic BP was increased thus necessitating

considering PRES in pediatric patients who present with hypertension and other clinical signs.

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