Posterior Reversible Encephalopathy Syndrome in Post-Streptococcal Glomerulonephritis

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Abstract
Posterior reversible encephalopathy syndrome (PRES) is a recognized brain disorder most commonly associated with hypertension, toxemia of pregnancy, or the use of immunosuppressive agents. Its clinical features include headache, decreased alertness, confusion, diminished spontaneity of speech, seizures, vomiting, and abnormalities of visual perception like cortical blindness. Magnetic resonance imaging shows edematous lesions primarily involving the posterior supratentorial white matter. We describe a 14-year-old boy who developed neurological symptoms of PRES during the course of acute post-streptococcal glomerulonephritis (APSGN).

Introduction
Posterior reversible encephalopathy syndrome (PRES) is a clinico-radiological syndrome manifesting with headache, confusion, seizures, visual disturbances, bilateral grey and white matter abnormalities suggestive of oedema in the posterior regions of cerebral hemisphere.

Case Report
14 year old boy presented with fever, headache associated with vomiting and passing high-coloured urine. Three hours prior to admission patient developed seizures involving left lower limb which became generalised later. Patient had five such episodes and he was brought to our hospital in a state of post-ictal confusion. On examination, patient had puffiness of face and swelling all over the body and confused, responded to simple oral commands, Temp 104° F, PR 110/min regular, BP 160/ 90 mmHg, RR 22/min, perritorbial puffiness of face (+); CVS/RS/ABD – NAD; CNS - optic fundi : normal; pupils: central, circular, 3 mm reacting to light and accommodation, no other focal neurological deficit. Urine analysis revealed plenty of erythrocytes, hyaline casts and RBC casts urine albumin 3+; Hb-11.2 g/dl; HCT was 29.6%; ESR-24 mm/hr; platelets - 301x10⁹, blood urea serial values (35,32,30,27) mg/dl; creatinine (1.5,1.3,1.0,1.0) mg/dl; serum albumin-2.6gm/dl; antistreptolysin O titre - at admission (77 IU/ml), after 1 week (207 IU/ml), 2 weeks after admission 497 IU/ml (normal range <200 IU/ml); complement C3 on admission 27 mg/dl, after 1 week 96 mg/dl (normal 90-120 mg/dl); 24 hr urine protein 900 mgs/day. Ultrasound abdomen was normal; EEG showed background delta rhythm suggestive of cerebral dysfunction; MRI contrast (Figure 1) showed bilateral cortical and subcortical white and grey matter involvement over occipital and parietal region suggestive of PRES. He was diagnosed as having acute post-streptococcal glomerulonephritis (APSGN) with PRES. He was put on salt-restricted diet, fluid restriction, diuretics, anticonvulsants and antibiotics. With treatment blood pressure came down, there were no further convulsions, facial puffiness improved and the patient was discharged with Tab. Amlodipine 2.5 mg OD and was reviewed after 2 weeks with repeat complement C3, urine routine, and other routine blood investigations. Urine analysis showed Albumin trace, no casts, Antistreptolysin O and C3 showed increasing titres. Repeat MRI showed no abnormality and the diagnosis of PRES was established by characteristic MRI findings which reversed to normal after 2 weeks (Figure 2).

Discussion
Posterior reversible encephalopathy syndrome (PRES), also known as reversible posterior leukoencephalopathy syndrome (RPLS), is a recently described brain disorder associated with findings on neuroimaging that suggest white-matter edema, mostly in the posterior parieto-temporo-occipital regions of the brain. However, radiological lesions in PRES are rarely isolated to these areas, and often involve the cortex, frontal lobes, basal ganglia and brainstem.

PRES has been shown to be associated with, ganglioneuroma, Henoch Schonlein purpura, acute lymphoblastic leukaemia, chemotherapy, hemolytic uremic syndrome, Addison’s disease, hypertension, intraabdominal neurogenic tumors, porphyria and bone marrow transplant. It is estimated that PRES occurs in 5% to 10% of children hospitalized

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with acute glomerulonephritis of all etiologies, the prevalence of PRES associated with APSGN is unknown. PRES caused by hypertension has been reported in seven children from 7 to 15 years of age with APSGN. All patients exhibited abnormal findings of the brain MRI or CT in the white matter of the parietal and occipital lobes, and recovered without any neurological sequelae following adequate treatment of the associated hypertension.

One patient with APSGN without severe hypertension but had features of PRES. The most important factor in development of paediatric hypertensive PRES is the rapidity of blood pressure elevation and the degree of elevation relative to the patient’s baseline pressure. It has been suggested that blood pressures more than 30% above normal for age should alert clinicians to the possibility of hypertensive PRES. Although the underlying pathophysiology of PRES remains elusive, three theories have been proposed: (1) hypertension-induced breakdown in cerebral auto-regulation, (2) cerebrovascular endothelial dysfunction (3) vasoconstriction and hypoperfusion with subsequent ischemia and vasogenic edema.

The preferential involvement of the posterior brain in PRES may be caused by its relative paucity of sympathetic innervations in comparison to the anterior circulation. The outcome of PRES is generally favourable, but delay in initiating the appropriate treatment may result in permanent damage to the brain.

In our patient the diagnosis of PRES was established by (1) presence of hypertension, (2) characteristic MRI contrast picture, (3) paramedian occipital lobe and calcarine were spared that distinguish from bilateral infarction of posterior cerebral artery territory. Important characteristic feature is reversibility of the imaging abnormalities after 2 weeks.

In conclusion, patients with PSGN sometimes exhibit atypical or unusual clinical manifestations such as PRES which may lead to diagnostic delays or misdiagnosis of the disorder. Recognition of this is important in order to ensure that the patient receives prompt treatment.

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**References**


