Posterior reversible encephalopathy syndrome a rare presentation of post-streptococcal glomerulonephritis

Sai Chandar Dudipala[1], M Prashanthi[2], Laxman Kumar Ch[3]

From [1]Assistant Professor, [2]Resident, [3]Professor, Department of Pediatrics, Prathima Institute of Medical Sciences, Karim Nagar, Telangana, India

Correspondence to: Sai Chandar Dudiapala, Department of Pediatrics, Prathima Institute of Medical Sciences, Karim Nagar, Telangana, India. E-mail: drsaichander@gmail.com

Received - 15 January 2020 Initial Review - 28 January 2020 Accepted - 11 February 2020

ABSTRACT

Acute post-streptococcal glomerulonephritis (PSGN) is a classic example of acute nephritic syndrome in children. It is typically characterized by gross hematuria, edema, hypertension, and acute kidney injury (AKI). Patients show diverse clinical profiles from being asymptomatic to mild syndrome or significant complications such as AKI, cardiac failure, or encephalopathy. Hypertension is found in up to 90% of patients and only 10% may have neurological symptoms. Only a few present with posterior reversible encephalopathy syndrome (PRES). Although PRES is a rare, but severe complication, there is a good outcome with appropriate treatment. Here, we report a case of PSGN in an 11-year-old female child who presented with altered sensorium, seizures, and vision loss. She was diagnosed as PRES on neuroimaging, which recovered with appropriate treatment.

Key words: *Glomerulonephritis, Posterior reversible encephalopathy syndrome, Vision loss*

Acute post-streptococcal glomerulonephritis (PSGN) is a classic example of acute nephritic syndrome in children. It is typically characterized by gross hematuria, edema, hypertension, and acute kidney injury [1]. PSGN is most common in children aged 5–12 years. Posterior reversible encephalopathy syndrome (PRES) is a clinicoradiological syndrome, presenting with headache, altered sensorium, convulsions, visual disturbances, and radiological findings of bilateral hyperintensity in T2-weighted images in the posterior part of cerebral hemispheres [2]. In the literature, various atypical presentations of PSGN were reported. We report a case of atypical presentation of PSGN presenting as PRES with status epilepticus and transient vision loss.

CASE REPORT

An 11-year-old girl presented to the emergency room with generalized tonic-clonic seizures lasting for 40 min. The child was stabilized with oxygen, intravenous midazolam, and levetiracetam. The child had headaches, vomiting, and blurring of vision for 24 h before admission. There was no history of fever, trauma, hematuria, and toxin ingestion. On inquiry, the child had a sore throat and fever 3 weeks back and was treated symptomatically.

On examination, the child had a Glasgow coma scale (GCS) of 8/15, pulse rate 120/min, blood pressure 198/130 mmHg, and oxygen saturation of 96%. Pupils were mid-dilated with a sluggish reaction to light, and there were no signs of meningeal irritation. Hence, the child was diagnosed as hypertensive emergency and started on intravenous labetalol infusion, nifedipine, and mannitol.

On laboratory evaluation, urine analysis showed plenty of red blood cells with blood. Creatinine was 1.7 mg/dL. Liver function tests, hemogram, serum electrolytes, and C-reactive protein were normal. C3 level was 18 mg/dL (normal 90–180 mg/dL) and anti-streptolysin O (ASO) titer was high (1600 IU/mL). The antinuclear antibody profile was negative. The electroencephalogram showed encephalopathic features. Magnetic resonance imaging (MRI) brain showed multifocal T2 FLAIR hyperintensities in the cortex, subcortical white matter in the frontal, and parieto-occipital lobes bilaterally (Fig. 1a).

GCS was improved 6 h after admission; however, the patient was unable to see. The fundus examination was normal. The blood pressure was controlled with nifedipine and labetalol infusion. After 3 days, the child regained her vision. Creatinine was normal after 4 days. The child was discharged 7 days after admission with levetiracetam and nifedipine.

On follow-up, after 2 months, the child becomes normotensive and stopped nifedipine. After 3 months, the ASO titer and C3 levels were normal. MRI brain was normal after 6 months (Fig. 1b). After 1 year of follow-up, the child still had microscopic hematuria. Finally, the diagnosis of PSGN with PRES was made based on clinical features, hypertension, low C3 levels, and reversible MRI brain findings.

DISCUSSION

Patients with PSGN sometimes manifest with atypical or unusual clinical presentations, which may lead to delay in the diagnosis or misdiagnosis of the disease [3]. Atypical manifestations of PSGN can be classified as (1) co-occurrence of immune-mediated

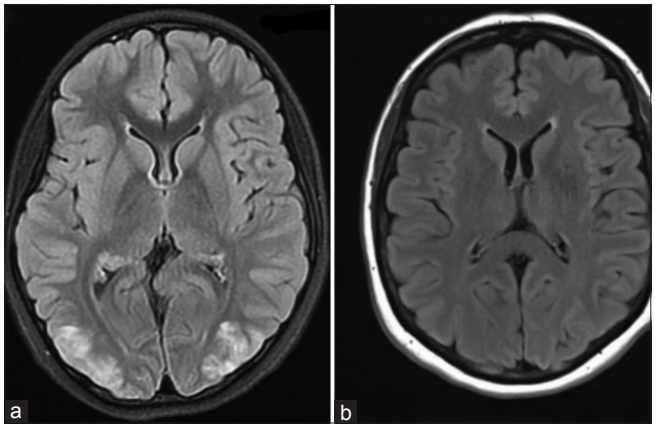

Figure 1: (a) Magnetic resonance imaging (MRI) brain showing bilateral areas of abnormal signal intensity in cortical and subcortical areas of the parieto-occipital region, which is hyperintense on T2 FLAIR weighted image and resolution of abnormal intensity on follow-up MRI brain (b)

diseases, (2) nonimmune-mediated complications, and (3) unusual clinical presentations or courses.

PRES is considered as nonimmune-mediated complication of PSGN [4]. PRES is described as clinicoradiological syndrome, presenting with headache, altered sensorium, convulsions, visual disturbances, and radiological findings of bilateral hyperintensity in T2-weighted images in the posterior part of cerebral hemispheres [2,3]. PRES has mainly been described in adults but has also been reported in children [3,5]. PRES in children has been shown to be associated with Henoch-Schönlein purpura, acute lymphoblastic leukemia, steroids, hemolytic uremic syndrome, intra-abdominal neurogenic tumors, porphyria, and bone marrow transplant [6,7]. In literature, PRES associated with PSGN has also been reported [8,9].

Although the underlying pathophysiology of PRES remains elusive, various theories have been proposed, namely, hypertension-induced breakdown in cerebral autoregulation; cerebrovascular endothelial dysfunction; and vasoconstriction and hypoperfusion with subsequent ischemia and vasogenic edema [10]. The preferential involvement of the posterior brain in PRES may be caused by its relative paucity of sympathetic innervation in comparison to the anterior circulation.

The diagnosis of PRES is mainly based on neuroimaging findings along with clinical features. Bartynski described three types of patterns on MRI brain. Almost 95% patients showed vasogenic edema with 75% patients in the parieto-occipital regions. Sometimes infraction (10–25%) or an intracranial bleed (15%) can be found on imaging [11,12]. In the present case, the child had encephalopathy, status epilepticus, and vision loss with classical MRI brain findings. An important feature of PRES is the reversibility of imaging abnormalities, which was seen in our case with the MRI brain after 6 months of being normal.

Although the association between PSGN and PRES has been reported [8,9,13], acute glomerulonephritis presenting with symptoms of PRES is extremely rare in children [2,14]. A total of 5–10% of children had PRES who were admitted to hospital with acute nephritic syndrome with various causes, but an exact association with PSGN was not known.

In our case, transient cortical blindness is described, which is one of the manifestations of PRES. Overall 33% patients with PRES had visual disturbances. Gupta *et al*. reported that PSGN with PRES presented with hypertension, transient vision loss with computed tomography of brain findings, which was reversed after hypertension control [13]. Kaarthigeyan and Vijayalakshmi reported a case with typical PSGN features and transient blindness [15].

CONCLUSION

PSGN can present atypically as PRES, which is a rare and severe entity. MRI brain is necessary. By early recognition and appropriate treatment, neurological sequelae and possible death can be prevented. Pediatricians should be aware of this unusual severe neurological complication, as early recognition may improve prognosis.

REFERENCES

1. Pan CG, Avner ED. Nelson Text Book of Paediatrics. 20th ed. Philadelphia, PA: Elsevier; 2015. p. 2498-501.
2. Hinchey J, Chaves C, Appignani B, Breen J, Pao L, Wang A, *et al*. A reversible posterior leukoencephalopathy syndrome. N Engl J Med 1996;334:494-500.
3. Fischer M, Schmutzhard E. Posterior reversible encephalopathy syndrome. J Neurol 2017;264:1608-16.
4. Eison TM, Ault BH, Jones DP, Chesney RW, Wyatt RJ. Post-streptococcal acute glomerulonephritis in children: Clinical features and pathogenesis. Pediatr Nephrol 2011;26:165-80.
5. Prasad N, Gulati S, Gupta RK, Sharma K, Gulati K, Sharma RK, *et al*. Spectrum of radiological changes in hypertensive children with reversible posterior leucoencephalopathy. Br J Radiol 2007;80:422-9.
6. Singhi P, Subramanian C, Jain V, Singhi S, Ray M. Reversible brain lesions in childhood hypertension. Acta Paediatr 2002;91:1005-7.
7. Khan SJ, Arshad AA, Fayyaz MB, Ud Din Mirza I. Posterior reversible encephalopathy syndrome in pediatric cancer: Clinical and radiologic findings. J Glob Oncol 2018;4:1-8.
8. Dasgupta MK, Sarkar S, Patra C. Posterior reversible encephalopathy syndrome in a case of post-streptococcal glomerulonephritis. J Pediatr Sci 2013;5:e1192.
9. Wirrell EC, Hamiwka LD, Hamiwka LA, Grisaru S, Wei X. Acute glomerulonephritis presenting with PRES: A report of 4 cases. Can J Neurol Sci 2007;34:316-21.
10. Bartynski WS. Posterior reversible encephalopathy syndrome, part 2: Controversies surrounding pathophysiology of vasogenic edema. AJNR Am J Nueroradiol 2008;29:1043-9.
11. Grioni D, Rovelli A, Pavan F, Prunotto G. The diagnosis of posterior reversible encephalopathy syndrome. Lancet Neurol 2015;14,1073-4.
12. Bartynski W. Posterior reversible encephalopathy syndrome, part 1: Fundamental imaging and clinical features. AJNR Am J Nueroradiol 2008;29:1036-42.
13. Gupta S, Goyal VK, Talukdar B. Reversible posterior leucoencephalopathy syndrome in post streptococcal glomerulonephritis. Indian Pediatr 2010;47:274-6.
14. Aeby A, David P, Fricx C, Jissendi P, Blecic S, Van Bogaert P. Posterior reversible encephalopathy syndrome revealing acute post-streptococcal glomerulonephritis. J Child Neurol 2006;21:250-1.
15. Kaarthigeyan K, Vijayalakshmi AM. Cortical blindness in a child with acute glomerulonephritis. Indian J Nephrol 2012;22:42-4.

Funding: None; Conflicts of Interest: None Stated.

How to cite this article: Dudipala SC, Prashanthi M, Ch LK. Posterior reversible encephalopathy syndrome a rare presentation of post-streptococcal glomerulonephritis. Indian J Child Health. 2020; 7(2):96-97.

Doi: 10.32677/IJCH.2020.v07.i02.015