A Rare Presentation of Posterior Reversible Encephalopathy Syndrome in Recovery Phase of Guillain–Barré Syndrome

Abstract

Posterior reversible encephalopathy syndrome (PRES) is a clinical and radiological syndrome, in which a patient presents with neurological symptoms, including headache, seizures, altered sensorium, and loss of vision, and accompanied with characteristic magnetic resonance imaging findings which are likely to be reversible. Guillain–Barré syndrome (GBS) is an acute demyelinating polyneuropathy presumably related to immunological mechanisms. Here, we describe a patient who had PRES in recovery phase of GBS while he was neither on any immunomodulator nor was having hypertension. He recovered completely clinically as well as radiologically.

Keywords: Guillain–Barré syndrome, posterior reversible encephalopathy syndrome, IVIg

Introduction

Posterior reversible encephalopathy syndrome (PRES) is a clinical and radiological syndrome, which was first described by Hinchey et al.[1] in 1996. A patient presents with neurological symptoms, including headache, seizures, altered sensorium, and loss of vision, and accompanied with characteristic magnetic resonance imaging (MRI) findings which are usually reversible. This condition has been described by various names including reversible posterior leukoencephalopathy, reversible posterior cerebral edema, reversible occipito-parietal encephalopathy, and hypertensive encephalopathy.[2] The main causes include a sudden increase in blood pressure, renal failure, fluid retention, immunosuppressive drugs, eclampsia, sickle cell disease, and intravenous immunoglobulin (IVIg) treatment.[1,3] MRI findings are typically reversible and appear as vasogenic edema in the parieto-occipital lobes.[4,5]

Guillain–Barré syndrome (GBS) is an acute demyelinating polyneuropathy presumably related to immunological mechanisms. Patients with GBS are preceded by an acute infection within 1–3 weeks, which are mainly respiratory or gastrointestinal. After the role of autoimmunity became clear, plasmapheresis and IVIg were used for the treatment. IVIg is commonly used in pediatric GBS treatment. The commonly seen side effects of IVIg are myalgia, headache, nausea, vomiting, fever, anaphylaxis, thromboembolism, and aseptic meningitis. PRES is a rare and potentially severe adverse event following IVIg therapy. Occurrence of GBS with PRES is a rare entity.[6,7] Dysautonoma is a proposed mechanism for such occurrence.

Case Report

An 8-year-old boy was brought with chief complaints of weakness over the lower limbs progressing to the upper limbs for 4 days and difficulty in breathing for 2 days. On admission, the patient’s pulse and blood pressure were normal, but the patient had difficulty in breathing due to involvement of respiratory muscles. On central nervous system examination, the patient was conscious and oriented, there was hypotonia in all four limbs, the power was 2/5 on both shoulder muscles and was 0/5 in all other muscles acting on other joints, deep tendon reflexes were absent, and plantar reflex was not elicitable.

The patient had symmetrical ascending paralysis involving respiratory muscle, so clinical diagnosis of GBS was made and was corroborated by nerve conduction velocity test which was suggestive of sensory-motor polyneuropathy.
The patient was put on a ventilator and given IVIg 2 g/kg over 5 days. In view of no improvement, a repeat dose of IVIg 2 g/kg over 5 days was given. The patient required prolonged ventilation; therefore, tracheostomy was done. After 14 days of the second course of IVIg, no improvement was observed, so seven cycles of plasmapheresis were done. Following which, the child showed improvement in the power of respiratory muscle, and slowly, the patient was weaned off from a ventilator. There was an improvement in power of the upper limb muscle, followed by lower limb muscle. Gradually, the patient continued to improve; hence, the patient was weaned off from oxygen, and tracheostomy tube was removed.

After 1½ months of admission and almost 1 month after IVIg therapy and 15 days after the last plasmapheresis cycle, the child had five episodes of convulsions generalized in nature. Following which, the patient had altered sensorium, inappropriate talks, and complete loss of vision. Blood pressure was normal. Clinical diagnosis of acute disseminated encephalomyelitis and PRES was made. MRI brain showed a bilateral asymmetrical area of abnormal enhancement in the parieto-occipital lobe with patchy areas of restricted diffusion at places suggestive of PRES [Figure 1]. The patient was started on anticonvulsants, following which, on the 2nd day, improvement in sensorium and vision was noted. Physiotherapy was continued, and as there was no new episode of convulsions, anticonvulsant was tapered off. Gradually, there was an improvement in power over a period of time; hence, the patient was discharged. After 3 months, MRI brain was repeated which was suggestive of normal study [Figure 2].

Discussion

PRES is seen in both adults and children and is characterized by a reversible, predominantly posterior (classically parieto-occipital) leukoencephalopathy with clinical features on a spectrum ranging from headaches and visual changes to confusion/altered mental status and seizures. Seizures are the most common neurologic symptoms. The seizures are usually generalized tonic-clonic type and multiple, as was in our case. Temporary restlessness and agitation may be present. Lethargy, stupor, and coma may develop. The patients are usually confused, and there may be some abnormalities of vision such as hemianopia, blurred vision, and cortical blindness. Our patient had complete loss of vision. The treatment of PRES is mainly symptomatic with withdrawal of offending immunosuppressant. The prognosis is generally good, with complete recovery within a few days. Our patient also showed recovery. Only a few cases suffer from sequelae, such as sequelae, and rarely, death may occur. PRES is thought to be caused by a breakdown of circulatory autoregulation in the circumstances of acute hypertension. The posterior circulation of the brain is particularly susceptible due to sparse sympathetic innervation. GBS is occasionally associated with hypertension. Almost two-thirds of patients who have GBS have transient hypertension. GBS is an inflammatory polyradiculoneuropathy. This inflammatory process may have a role in PRES by changing capillary permeability. The high percentage of patients with PRES who have autoimmune disorders may support the theory that PRES is in part caused by endothelial dysfunction, a process in which the host autoimmune response is essential.

In a case report, the patient complained of severe headache on day 28 after the initial administration of IVIg, and MRI of the brain indicated vasogenic lesions which resolved 17 days later. Another report of PRES associated with IVIg of a 14-year-old girl with GBS described the development of PRES only 3 days after the initial administration of IVIg, much sooner than our patient.

Unusual features of our case were development of PRES after 1½ months of admission when the patient...
was in recovery, had normal BP, and was not on immunosuppressant. This case also raises a question whether dysautonomia persists till very late in GBS as in our case.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

References

1. 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.
2. Girişgen I, Tosun A, Sömmez F, Ozsunar Y. Recurrent and atypical posterior reversible encephalopathy syndrome in a child with peritoneal dialysis. Turk J Pediatr 2010;52:416-9.
3. McCoy B, King M, Gill D, Twomey E. Childhood posterior reversible encephalopathy syndrome. Eur J Paediatr Neurol 2011;15:91-4.
4. Onder AM, Lopez R, Teomete U, Francoeur D, Bhatia R, Knowbi O, et al. Posterior reversible encephalopathy syndrome in the pediatric renal population. Pediatr Nephrol 2007;22:1921-9.
5. Inceçik F, Hergüner MO, Altunbasak S, Erbey F, Leblebisatan G. Evaluation of nine children with reversible posterior encephalopathy syndrome. Neurol India 2009;57:475-8.
6. van Diest D, van Goethem JW, Vercruyssen A, Jadoul C, Cras P. Posterior reversible encephalopathy and Guillain-Barré syndrome in a single patient: Coincidence or causative relation? Clin Neurol Neurosurg 2007;109:58-62.
7. Lamy C, Oppenheim C, Méder JF, Mas JL. Neuroimaging in posterior reversible encephalopathy syndrome. J Neuroimaging 2004;14:89-96.
8. Kwon S, Koo J, Lee S. Clinical spectrum of reversible posterior leukoencephalopathy syndrome. Pediatr Neurol 2001;24:361-4.
9. Garg RK. Posterior leukoencephalopathy syndrome. Postgrad Med J 2001;77:24-8.
10. Skiba V, Etienne M, Miller JA. Development of chronic epilepsy after recurrent episodes of posterior reversible encephalopathy syndrome associated with periodic lateralized epileptiform discharges. Seizure 2011;20:93-5.
11. Fugate JE, Rabinstein AA. Posterior reversible encephalopathy syndrome: Clinical and radiological manifestations, pathophysiology, and outstanding questions. Lancet Neurol 2015;14:914-25.
12. Roth C, Ferbert A. The posterior reversible encephalopathy syndrome: What’s certain, what’s new? Pract Neurol 2011;11:136-44.
13. Okada T, Hiyoshi K, Noto N, Fujita Y, Fuchigami T, Okubo O, et al. A case of Guillain-Barré syndrome accompanied by sympathetic overactivity and hypertensive encephalopathy. Acta Paediatr 1996;85:1006-8.
14. Kieseier BC, Tani M, Mahad D, Oka N, Ho T, Woodroofe N, et al. Chemokines and chemokine receptors in inflammatory demyelinating neuropathies: A central role for IP-10. Brain 2002;125:823-34.
15. Aird WC. The role of the endothelium in severe sepsis and multiple organ dysfunction syndrome. Blood 2003;101:3765-77.
16. Zx Li, Jd Gao, Hm Yu, Jiang K. Posterior reversible encephalopathy syndrome associated with IVIg in a child with Guillain-Barré syndrome: Case Report and review of the literature. HK J Paediatr (new series) 2016;21:211-3.
17. Koichihara R, Hamano S, Yamashita S, Tanaka M. Posterior reversible encephalopathy syndrome associated with IVIG in a patient with Guillain-Barré syndrome. Pediatr Neurol 2008;39:123-5.