Optic Neuritis in a Patient with Bickerstaff's Brainstem Encephalitis

Jae Gun Park
Chang Hun Bin
Min Su Park

Department of Neurology,
Yeungnam University
College of Medicine, Daegu, Korea

Dear Editor,

Bickerstaff's brainstem encephalitis (BBE) is characterized by progressive ophthalmoplegia, ataxia, impaired consciousness, and hyperreflexia. BBE is one of the spectrum diseases of anti-GQ1b antibody syndrome affecting both the central nervous system (CNS) and the peripheral nervous system (PNS).¹ We report a patient with optic neuritis (ON) and BBE who was effectively treated with steroid and intravenous immunoglobulin (IVIg) therapy.

A 34-year-old man presented with confusion combined with lateral and downward gaze limitation of the left eye. A cerebrospinal fluid study showed white blood cells at 54 cells/mm³, 92.12 mg/dL protein, and a normal glucose level. Brain magnetic resonance imaging (MRI) revealed diffuse leptomeningeal enhancement (Fig. 1A). He was diagnosed as BBE and started on steroid pulse therapy (1 g/day for 5 days). However, weakness in the extremities and right facial palsy developed on the day after starting steroid pulse therapy, and his deep tendon reflexes were decreased in general. His serum was positive for anti-GQ1b antibody. A nerve conduction study showed a motor-dominant sensorimotor polyneuropathy that was not consistent with the criteria for demyelinating disease. We diagnosed the patient as BBE with Guillain-Barré Syndrome (GBS).

We started IVIg therapy at 0.4 g/kg for 5 days. Left-eye blindness appeared when his mentality was fully recovered on the third day of IVIg treatment. A thorough review of his initial MRI findings revealed enhancement in the left optic nerve (Fig. 1A). Follow-up MRI showed improvement of the leptomeningeal and left-optic-nerve enhancement, but new multiple hyperintense lesions had developed in the brainstem (Fig. 1B). The visual evoked potential (VEP) indicated the presence of a prechiasmal lesion on the left side (Fig. 1C). Additional steroid pulse therapy (1 g/day for 5 days) resulted in a small improvement of his visual acuity (from no light perception to finger counting at 30 cm). A third session of steroid pulse therapy (0.5 g/day for 5 days) was started 11 days after finishing his second pulse therapy. He was discharged at 35 days after his first admission, when he was able to walk by himself with improvement of his left visual acuity to 0.1. At 3 months after discharge his left eye visual acuity had fully recovered and VEP had improved markedly compared to the earlier measurements (Fig. 1D).

The pathogenesis of anti-GQ1b antibody syndrome involves anti-GQ1b antibody binding with nerve GQ1b gangliosides that are distributed throughout the CNS and PNS.¹ The third, fourth, and sixth cranial nerves are those with the highest percentage of GQ1b gangliosides, followed by the second cranial nerve. Thus, although GQ1b gangliosides are less abundant in the CNS than other cranial nerves,¹⁻³ ON which is CNS could be accompanied with BBE since they have the same underlying pathologic mechanism. The present patient, who had both ON and brainstem lesions, needed to be differentiated from neuromyelitis optica (NMO). NMO was ruled out from his differential diagnosis because the clinical features and nerve conduction study and MRI findings differed from findings that are typical of NMO.

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Correspondence

Min Su Park, MD
Department of Neurology,
Yeungnam University
College of Medicine,
170 Hyeonchung-ro, Nam-gu,
Daegu 42415, Korea
Tel +82-53-620-3685
Fax +82-53-627-1688
E-mail minsupark@ynu.ac.kr

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Fig. 1. Initial and follow-up MRI images and visual evoked potential (VEP) waveforms. A: Initial MRI showed left-optic-nerve gadolinium enhancement (arrow) and leptomenigeal enhancement in T1-weighted contrast-enhanced images without brainstem signal changes. B: Follow-up MRI showed multiple high signal changes in the brainstem in T2-weighted images (arrows). Optic-nerve and leptomenigeal enhancement had disappeared. C: Initial VEP indicated a prechiasmal lesion on the left side. D: Follow-up VEP showed improvement.

BBE is a rare immune disorder itself, and a case of ON with superimposed BBE and GBS fully cured by steroid and IVIg therapy would be extremely rare. There has been only one reported case of ON with BBE that was not effectively treated.⁴ The scarcity of the disease and the poor mentality of patients makes it difficult to identify ON in BBE patients, but we recommend physicians to still attempt to confirm whether ON is present in any patient with BBE.

There have been no previous reports on the application of additional steroid pulse therapy or other types of steroid therapy for ON in anti-GQ1b antibody syndrome, but there have been some reported cases of refractory ON treated with steroid pulse therapy and IVIg therapy.⁵ If a patient with ON and BBE shows a minimal response to a single steroid or IVIg therapy, additional pulse therapy may be helpful.

Conflicts of Interest

The authors have no financial conflicts of interest.

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