Seizure in a Child with Guillain-Barré Syndrome: Association or coincidence!

A 6-year-old normally developing girl presented with progressive gait instability for the past three days followed by loss of ambulation along with paresthesia and pain in bilateral lower limbs. There was a history of probable viral upper respiratory infection two weeks prior to the presentation. On examination, she had flaccid weakness (power was MRC grade 2 in all limbs) and areflexia, without respiratory or bulbar involvement. Rest of the central nervous system (CNS) examination was unremarkable. Nerve conduction study showed motor-sensory axonal polyneuropathy. With a clinical diagnosis of Guillain Barré syndrome (AMSAN variant), she was started on IVIg (2 g/kg over 5 days). On third day of illness, she had an episode of generalized tonic-clonic seizure, lasting for two minutes. At that time, she did not have any fever. Her heart rate and respiratory rate, blood pressure, serum electrolytes and infection workup were normal. Magnetic resonance imaging of the brain and inter-ictal electroencephalogram were unremarkable. cerebrospinal examination revealed albumino-cytological dissociation (5 cells, protein: 74mg/dL), normal sugar, and sterile culture. Stool culture yielded no growth. She regained ambulation within two weeks and there was no seizure recurrence.

Guillain-Barré syndrome (GBS) is an immune-mediated polyradiculoneuropathy that primarily affects peripheral nervous system; however, rarely it can involve CNS [1]. The reported manifestations of GBS are encephalopathy, seizures, dystonia, myoclonus, visual disturbance, and nystagmus [2]. The pathogenetic mechanism of these CNS features are posterior reversible encephalopathy syndrome secondary to autonomic instability, watershed infarction, demyelination, adverse effects of immunoglobulin and hypoxic brain injury due to respiratory complications [3]. Isolated seizures without encephalopathy or aseptic meningitis are rarely reported in GBS.

Koul, *et al*. [4] reported a 10-year-old girl with Fischer variant of GBS who had recurrent myoclonic seizures during the course of the illness. The exact mechanism of seizures in GBS is unknown. Koul and colleagues [4] suggested a brain stem origin of the myoclonus. Seizures are a common manifestation in various autoimmune neurological and systemic disorders. GM1 is expressed both in central and peripheral nervous system; however, the pathological changes are evident only in peripheral nervous system as blood brain barrier is less permeable for autoantibodies [5]. The possible pathophysiological mechanism in the index case may be immune-mediated neuronal damage as other postulated mechanisms are unlikely to explain seizures in our case, or it can be merely a coincidence due relatively high prevalence of unprovoked seizures and GBS in the community.

Prateek Kumar Panda and Indar Kumar Sharawat*
Pediatric Neurology Division, Department of Pediatrics, All India Institute of Medical Sciences, Rishikesh, Uttarakhand. India.
**sherawatdrindar@gmail.com*

References

1. Koichihara R, Hamano S-I, Yamashita S, Tanaka M. Posterior reversible encephalopathy syndrome associated with IVIG in a patient with Guillain-Barré syndrome. Pediatr Neurol. 2008;39:123-5.
2. Sutter R, Mengiardi B, Lyrer P, Czaplinski A. Posterior reversible encephalopathy as the initial manifestation of a Guillain-Barré syndrome. Neuromuscul Disord. 2009;19:709-10.
3. Van Diest D, Van Goethem JWM, 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.
4. Koul RL, Nair PM, Chacko A, Venugopalan P. Myclonic seizures in a young girl with Fishers variant of Guillain-Barre syndrome. Neurosciences Journal of Riyadh Saudi Arab. 2002;7:188-90.
5. Yuki N. Guillain-Barré syndrome and anti-ganglioside antibodies: a clinician-scientist's journey. Proceedings of the Japan Academy Ser B Physical and Biological Sciences. 2012;88:299-326.