A rare form of Guillain-Barré syndrome: pharyngeal-cervical-brachial variant

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Guillain-Barré syndrome is clinically characterized by acute onset of generalized, symmetrical, and ascending muscle weakness and areflexia from peripheral nerve involvement. In Guillain-Barré syndrome variants, however, some patients have unusual distribution of muscle involvement. Pharyngeal-cervical-brachial variant of Guillain-Barré syndrome is characterized by oropharyngeal, neck, and upper limb muscle involvement. Although Guillain-Barré syndrome is one of several post-infectious diseases that cause limb muscle weakness, the incidence of pharyngeal-cervical-brachial variant is relatively low. Here we report the case of a 16-month-old boy who developed a rare form of Guillain-Barré syndrome, the pharyngeal-cervical-brachial variant of the disease. We concluded that taking all the other etiologic reasons into consideration, pharyngeal-cervical-brachial variant of Guillain-Barré syndrome should be remembered in patients with symptoms of bulbar and upper extremity weakness not only for early diagnosis but also to plan the treatment early and follow up the potential complications.

Key words: Guillain-Barré syndrome, pharyngeal-cervical-brachial variant.

Guillain-Barré syndrome (GBS) includes a spectrum of acquired, immune-mediated disorders causing dysfunction or degeneration in peripheral nerves, spinal sensory and motor nerve roots and, occasionally, cranial nerves. The incidence of the disease has been estimated to range from 0.5 to 1 in 100,000, in individuals younger than 18[1].

Guillain-Barré syndrome is clinically characterized by acute onset of generalized, symmetrical, and ascending muscle weakness and areflexia from peripheral nerve involvement. In GBS variants, however, some patients have unusual distribution of muscle involvement. Miller-Fisher syndrome is characterized by the triad of ataxia, areflexia, and external ophthalmoplegia[2]. In polyneuritis cranialis, bilateral facial neuropathy is the most common manifestation, which more typically occurs in the evolution of a syndrome of ascending paralysis with other clinical features of acute inflammatory demyelinating polyneuropathy. Rarely, facial and bulbar weakness present with dysphagia and dysphonia and only minimal, if any, appendicular weakness. Pharyngeal-cervical-brachial (PCB) variant of GBS is characterized by oropharyngeal, neck, and upper limb muscle involvement. Although GBS is one of several post-infectious diseases that cause limb muscle weakness, the incidence of PBC is relatively low[1-3].

The diagnosis of GBS is relatively easy in patients with typical constellation of clinical features, with laboratory examination of the cerebrospinal fluid and electrodiagnostic studies. Recognition of atypical cases is important because it permits anticipatory monitoring for complications of the disease and identifies therapeutic options for affected children.

Here we report the case of a 16-month-old boy who developed a rare form of GBS, the PCB variant of the disease.

Case Report

A 16-month-old boy was admitted to our hospital with difficult respiration and bilateral arm weakness. He had an acute gastroenteritis two weeks before. He had experienced difficulty

in swallowing and bilateral arm weakness for the last three days. His past medical history and family history were unremarkable. His mental examination was normal. He had cyanosis and difficulty in respiration. In neurological examination, he was alert. He had dysphagia, but his eye movements and pupillary light reflex were normal. His upper extremities were flask and he had loss of head control. Strengths in distal and proximal muscles in upper extremities were 1/5 bilaterally. Strength in the legs was normal except for trace weakness of the hip flexors. The deep tendon reflexes were absent in both arms but mildly depressed in the legs. The plantar responses were flexor, and sensation was intact. There was no ataxia or tremor. Autonomic functions were normal.

Laboratory examination revealed serum electrolytes, complete blood count, and liver function tests to be normal. Viral panel (cytomegalovirus, Epstein-Barr virus, rubella, rubeola and herpes viruses), cultures for salmonella, shigella and campylobacter, and serological examination for mycoplasma were also normal. Cervical magnetic resonance imaging (MRI) was normal. The cerebrospinal fluid examination on day 8 indicated cytoalbuminologic dissociation.

In addition, nerve conduction studies indicated acute motor axonal injury and sparing of sensory responses, especially in the arms.

He was diagnosed with PCB variant of GBS. He was treated with intravenous immunoglobulin 2 g/kg administered in 3 doses. Over the following 15 days, muscle strength in his arms was 3/5 bilaterally and lower extremity strength was normal. His respiration became nearly normal, and dysphagia was disappearing.

Discussion

In nations with widespread immunization programs in place, where poliomyelitis has been nearly eliminated, GBS remains the most common cause of acute paralysis in childhood. Typical GBS is characterized by bilaterally symmetrical ascending paralysis, absent of deep tendon reflexes, sensory loss, cytoalbuminologic dissociation in cerebrospinal fluid and typical findings in nerve conduction studies. In up to 45% of affected children, facial weakness and ophthalmoplegia were reported associated with extremity weakness. A number of variant forms of GBS are recognized. Miller-Fisher syndrome is characterized with ataxia, areflexia, and ophthalmoplegia. Some of them are associated with facial weakness, dysarthria, dysphagia, abnormal pupil reactions, and weakness of the extremities. Electrophysiological studies usually reveal axonal degeneration. Polyneuritis cranialis is defined as acute, multiple, symmetrical cranial neuropathies, with the most frequently involved cranial nerves being III and IV[1-3].

In 1986, Ropper[4] described the first patients with PCB variant of GBS. The disease was characterized with muscle weakness predominantly in oropharyngeal, neck, shoulder and arm muscles and dysphagia. In their cases, the lower limbs were spared, and this was initially thought to be a diagnostic hallmark for this variant. However, some patients were later described to have mild lower limb weakness, similar to our patient. The cerebrospinal fluid examination may be normal or protein may be mildly elevated. Nerve conduction studies often revealed motor axonal injury and sparing of sensory responses, as was found in our case. However, in some cases, electrodiagnostic findings may be normal or with evidence of a primary demyelinating process[5].

Pharyngeal-cervical-brachial variant of GBS is a rare disorder in childhood. Furiya et al.[5] reported a 15-year-old boy with acute onset ataxia, diplopia, dysarthria, and dysphagia, followed by muscle weakness in neck and arm muscles. That patient had symptoms of both Miller-Fisher syndrome and PCB variant of GBS. Mogale et al.[6] reported two children (10 and 3 years old) with acute demyelinating PCB variant. Murakami et al.[7] reported a 15-year-old girl who developed bulbar palsy and upper limb dominant muscle weakness two weeks after a cytomegalovirus infection. In PCB variant of GBS, bulbar palsy is noticed from the early stage of the disease and improves in a few months.

Our patient had dysphagia at presentation, followed by muscle weakness in neck and arms. There was a history of antecedent infection as gastroenteritis. Examination of the cerebrospinal fluid and electrodiagnostic studies supported this diagnosis. Spinal lesions were excluded with his MRI findings. Other acute neuropathies with upper extremity predominant weakness, such as Tangier disease, tyrosinemia,

and porphyria, were excluded on biochemical testing. Poliomyelitis was excluded with his clinical findings and nerve conduction studies. Our patient was younger than those reported previously in the literature.

Guillain-Barré syndrome is one of several autoimmune disorders and is often preceded by an infectious disease. Anti-ganglioside antibodies are detected in the sera of patients with GBS and its variants. It is thought to be a useful marker for supporting the diagnosis. Specific nerve antigens have been reported in various types of GBS. In Miller-Fisher syndrome and polyneuritis cranialis, anti-ganglioside GQ1b antibodies have been shown[1,5,8,9]. Murakami et al.[7] reported a girl who developed PCB variant of GBS after cytomegalovirus infection and who had isolated elevation of anti-GT1a IgG antibody. It is interesting, because Koga et al.[10] reported that all GBS variants with GT1a-specific IgG antibody had positive serology for Campylobacter jejuni infection. However, in our case, there was an antecedent infection as gastroenteritis, we did not show any pathological agent, and assays for these antibodies were not available.

Adult patients with PCB variant of GBS commonly have prolonged illness. Electrophysiological studies are not good predictors of the outcome. Treatment strategies in GBS could theoretically employ interventions at multiple stages in the process of autosensitization and targeted tissue inflammation. Studies suggest that plasmapheresis and intravenous immunoglobulins are safe and effective treatment for children with GBS, especially in more severely affected children[1]. In our case as well, early results were satisfactory after intravenous immunoglobulin.

In conclusion, PCB variant of GBS can be seen in childhood, though not often. Taking all the other etiologic reasons into consideration, PCB variant of GBS should be remembered in patients with symptoms of bulbar and upper extremity weakness not only for early diagnosis but also to plan the treatment early and follow up the potential complications.

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