A rare manifestation of pediatric CIDP: hypoglossal nerve involvement

Nurşah Yeniay Süt, Miraç Yıldırım, Ömer Bektaş Ayşe Tuğba Kartal, Serap Teber

Department of Pediatric Neurology, Ankara University Faculty of Medicine, Ankara, Turkey

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Chronic inflammatory demyelinating polyradiculoneuropathy (CIDP) is a rare autoimmune peripheral nervous system disease characterized by progressive or relapsing symmetric weakness and sensory dysfunction. It may rarely cause cranial nerve neuropathy. Although cranial nerve involvement is rarer in CIDP than in Guillain-Barré syndrome (GBS), it may occur in rapid-onset and relapsed courses.

INTRODUCTION

Chronic inflammatory demyelinating polyradiculoneuropathy (CIDP) is a rare autoimmune peripheral nervous system disease characterized by progressive or relapsing symmetric weakness and sensory dysfunction, which evolves within at least 8 weeks in the all-age groups. Unlike adults, the clinical manifestation of CIDP in children may occur in less than 8 weeks. It is difficult to distinguish from Guillain-Barré syndrome (GBS), especially when the symptoms develop in less than 4 weeks (acute CIDP). In rapidly progressive pediatric cases, the probability of cranial nerve involvement and sensory deficit increases.

Contrary to GBS, cranial nerve involvement is rare in children with CIDP, particularly hypoglossal nerve involvement is unusual. To date, a few adult patients with CIDP presenting hypoglossal involvement have been reported. Herein, we report the first pediatric case with CIDP presenting with unilateral hypoglossal nerve involvement who responded well to pharmacological therapy.

CASE PRESENTATION

The 8-year-old boy, who was followed up with CIDP for about 7 years, was first admitted to our outpatient clinic at the age of 4 months old with the inability to move his legs. He was misdiagnosed with Guillain-Barre syndrome because of a rapidly progressive course. He had absence of deep tendon reflex and weakness on examination, increased protein in cerebrospinal fluid analysis, and T2 hyperintensity in spinal MRI. He was diagnosed with CIDP at the age of 12 months after the relapse of similar symptoms. The attacks similarly recurred several times and were treated with intravenous immunoglobulin (IVIG), methylprednisolone, azathioprine, and rituximab, respectively. His symptoms improved significantly after rituximab treatment. Two years after his last attack, he presented with dysphagia and asymmetrical face. He had also unilateral tongue deviation, mild atrophy, and fasciculation, indicating hypoglossal nerve involvement. He was administered monthly IVIG (2gr/kg) and then weekly rituximab. Cranial nerve involvement completely resolved in the 3rd month of treatment.

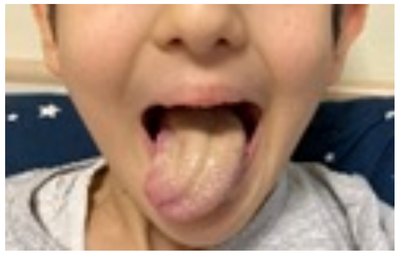

Figure 1. Tongue deviation due to hypoglossal nerve involvement

Figure 2. Significant improvement in hypoglossal nerve involvement in the 3rd month of treatment

CONCLUSION

According to guidelines of the European Academy of Neurology/Peripheral Nerve Society, clinical and laboratory findings such as progressive distal weakness, involvement of more than two sensory and motor nerves in nerve conduction studies, increased protein in CSF examination, spinal MRI findings, good response to immunomodulatory treatment in our patient were consistent with CIDP diagnosis. Our patient was misdiagnosed with GBS because the previously mentioned symptoms lasted less than 4 weeks. However, the first presentation of childhood CIDP could be confused with GBS; this condition is called acute CIDP. In a comparative study comparing the clinical findings of rapid and slow-onset CIDP, Cabasson et al. found that the relapsing course, sensory and cranial nerve involvement were significantly more common in the rapid-onset group, similar to our patient. Although cranial nerve involvement is less common in CIDP than in GBS, it often involves the facial nerve, similar to GBS. Moreover, hypoglossal nerve involvement is very rare in both diseases.

To our knowledge, this case is the first report describing the hypoglossal nerve involvement in a child with CIDP. It should be kept in mind that cranial nerve involvement may occur during relapse in children with CIDP.

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