Relapse with Dysphagia in a Case of Chronic Inflammatory Demyelinating Polyradiculoneuropathy

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Abstract

Glossopharyngeal and/or vagus nerve involvement is infrequent in patients with chronic inflammatory demyelinating polyradiculoneuropathy (CIDP). We herein report the case of a 69-year-old Japanese woman who presented with muscle weakness and numbness of the extremities with dysphagia. The serum anti-ganglioside GM1 immunoglobulin IgM antibody levels were elevated, and treatment with intravenous immunoglobulin (IVIg) resulted in a dramatic improvement; the weakness, numbness and dysphagia all resolved. However, relapse comprising dysphagia alone occurred on hospital day 26, and treatment with IVIg again proved extremely effective. IVIg therapy can be effective against cranial nerve involvement in cases of CIDP.

Key words: chronic inflammatory demyelinating polyradiculoneuropathy, dysphagia, glossopharyngeal nerve, vagus nerve, modified barium swallow, intravenous immunoglobulin

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Introduction

Chronic inflammatory demyelinating polyradiculoneuropathy (CIDP) is a remitting or chronic progressive autoimmune disease causing progressive or relapsing and remitting weakness and numbness. Involvement of the cranial nerves (CNs), particularly CN IX (glossopharyngeal nerve) and/or CN X (vagus nerve), is infrequent in patients with CIDP (1). Rotta et al. reported cases showing distinct features outside the usual clinical phenotype of CIDP, with 5% of patients (4 of 87 cases) showing predominant CN involvement. In that report, CNs III, IV and VI were affected in three patients, CN VII in two patients and CNs IX, X and XI in one patient each (1). However, the details of the therapeutic response to treatment with intravenous immunoglobulin (IVIg) and/or corticosteroids remain unclear. We herein report the case of an elderly woman with CIDP who presented glossopharyngeal and/or vagus nerve involvement.

Case Report

A healthy 69-year-old Japanese woman noticed limbs weakness that disappeared spontaneously after three months. After an additional two months, quadriplegia and slight dysphagia developed, which subsequently slowly progressed over four months, leading to hospital admission. On admission, the patient demonstrated symmetrical proximal and distal muscle weakness with diminished tendon reflexes, numbness in the fingers and slight dysphagia. The results of hematological, serum biochemistry and a urinalysis were within the normal limits, with the exception of hyperlipidemia. Neither monoclonal gammopathy in the serum nor Bence-Jones proteins in the urine were detected, and there was no evidence of abnormal glucose tolerance or infectious diseases, including hepatitis C, hepatitis B or acquired immune deficiency syndrome. Screening tests for vitamin and collagen vascular diseases were within the normal limits. We subsequently examined the levels of serum antibodies against several types of glycolipids, including GM1, GM2, GM3, GD1a, GD1b, GD3, GT1b, GQ1b and galactocere-
A small number of cases of CN involvement in patients with CIDP have been described (3). CIDP with involvement of CNs IX and/or X is infrequent, with clinical features reported in only three cases. McCann et al. reported a CIDP patient with muscle weakness, sensory ataxia and multiple CN abnormalities, including dysphagia. In that case, neither steroid therapy nor plasmapheresis had an effect. An autopsy revealed widespread onion bulb formation in CNs III, IV, V, VI, X, XI and XII (3). In addition, Rotta et al. reported a case of steroid- and IVIg-responsive CIDP with dysphagia. In that case, the onset of dysgeusia, dysphagia, bilateral facial numbness and weakness preceded that of typical symptoms, such as muscle weakness of the limbs (1). Mazzucco et al. reported a case of ataxic CIDP in which the patient exhibited recurrent tetraparesis, bilateral facial drooping, dysphonia, dysphagia and dyspnea. Negative results were obtained for anti-GM1, GD1a, asialo-GM1, GD1b antibodies and GQ1b IgG, and treatment with steroids and IVIg was effective (4). In both previous cases of CIDP with dysphagia as well as the current case, the administration of IVIg proved effective. These reports show that IVIg is effective for CIDP with dysphagia. Our patient is distinctive in that the recurrence occurred in the form of dysphagia alone. This shows that CIDP can recur with only CN dysfunction. In addition, the effect of IVIg was dramatic for the CN IX and/or X involvement. Therefore, IVIg may be an effective treatment for CIDP with CN palsy.

In this study, the MBS was useful for evaluating the patient’s swallowing impairment and the efficacy of treatment. Pharyngeal retention is defined as the presence of a material remaining in the pharynx (valleculae and/or pyriform sinuses) after swallowing is completed (5). Pharyngeal retention is caused by poor pharyngeal clearance with CN impairment, including impairment of CNs IX and/or X. In the current patient, the dysphagia was mild, and MBS was useful for identifying impaired swallowing and evaluating the effect of therapy. CN IX and/or X involvement is infrequent in cases of CIDP, and MBS may be used to prevent missing the presence of mild dysphagia in patients in whom dysphagia is suspected.

Caudie et al. reported finding IgG or IgM antibodies to GM1 in 43.8% of patients with Guillain-Barré syndrome, 15% of patients with amyotrophic lateral sclerosis, 19% of patients with chronic inflammatory demyelinating polyneuropathy and 9% of healthy controls (6). Anti-ganglioside GM1 IgM antibodies were detected in our patient. Ryo et al. reported a 19-year-old woman with CIDP who presented with CN involvement and anti-ganglioside GM1 IgM anti-

Discussion

pearence of the patient’s symptoms. No pharyngeal retention was shown on MBS, and her swallowing normalized (Figure B). As of one additional year after the second cycle of IVIg, no signs of recurrence had been detected while continuing treatment with oral prednisolone.

Figure. Modified barium swallow on hospital day 29 shows barium in the pyriform sinuses (arrow) after swallow completion (A). Four months after the second course of intravenous immunoglobulin (IVIg), no pharyngeal retention is evident (arrow) (B).

Broside, using an enzyme-linked immunosorbent assay. IgM anti-GM1 antibodies were positive, while IgM antibodies to other antigens were negative and no IgG antibodies were positive. A cerebrospinal fluid (CSF) analysis revealed an elevated protein level (81 mg/dL) and normal cell count (1 cell/mm³), with albuminocytologic dissociation. CSF cytology yielded negative results; however, electrophysiological studies identified conduction block in the right ulnar nerve. Prolongation of distal latency was also observed in the right median, bilateral peroneal and right tibial nerves. Meanwhile, F-waves in the bilateral median nerves were absent, whereas cervical magnetic resonance imaging revealed cervical nerve root hypertrophy. The results of the laboratory studies fulfilled the European Federation of Neurological Societies and Peripheral Nerve Society criteria for definitive CIDP (2).

The patient was treated with IVIg at a dose of 0.4 g/kg for five days starting on hospital day 2. The quadriplegia and dysphagia subsequently began to improve, and her symptoms completely resolved by hospital day 15. However, the dysphagia reappeared on hospital day 26, and a neurological examination showed poor soft palate elevation. Although no weakness of the limbs was evident at the time of relapse, impaired swallowing was identified on a modified barium swallow (MBS) study performed on hospital day 29, with pharyngeal retention remaining in the pyriform sinuses after swallow completion (Figure A). Corticosteroid therapy with methylprednisolone was initiated (1,000 mg/day for three days starting on hospital day 27), followed by oral prednisolone; however, the dysphagia remained essentially unchanged for 10 days after the start of corticosteroid treatment. The administration of a second course of IVIg at a dose of 0.4 g/kg for five days was started on hospital day 38, and the dysphagia improved dramatically, with disapp
bodies in a Japanese article (7). In that case, recurrent distal limb weakness and numbness of the extremities with ophthalmoplegia were noted. The pathology of CIDP remains unknown, and the further accumulation of cases is required to clarify the effects of anti-ganglioside antibodies in CIDP patients with CN involvement.

The authors state that they have no Conflict of Interest (COI).

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