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Case report

Multifocal motor neuropathy and visual pathway impairment: A case report

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ABSTRACT

Background: Multifocal motor neuropathy (MMN) occasionally presents with cranial nerve involvement. However, no MMN cases with visual pathway impairment demonstrated by visual evoked potential (VEP) have been reported.

Case report: A 36-year-old man was admitted to our hospital with progressive muscular weakness. On admission, neurological findings revealed bilateral muscle weakness and atrophy of the distal upper limbs. The blood tests were positive for GM-1 ganglioside antibodies. Nerve conduction studies revealed bilateral conduction block in the median nerve. He was diagnosed with MMN. Intravenous immunoglobulin treatment improved muscle weakness and blurred vision, which was not a complaint when he was first seen. Moreover, VEP showed a post-treatment shortening of P100 latency. These treatment effects were consistently observed for 3.5 years.

Significance: Our findings suggested that MMN could affect the visual pathway through autoimmune mechanisms.

1. Introduction

Multifocal motor neuropathy (MMN) is among the immune-mediated neuropathies associated with anti-GM1 antibodies. MMN presents as a chronic progressive motor neuropathy, which is characterized by asymmetric muscle weakness and atrophy in the distal limbs with partial motor conduction block (Yeh et al., 2020). Rarely, some patients with MMN present with cranial nerve involvement other than limb muscle weakness (e.g., hypoglossal and abducens nerve palsies) (Kaji et al., 1992; Galassi et al., 2012). Here, we describe the case of a patient with MMN who presented with visual pathway impairment.

2. Case report

A 36-year-old man was admitted to our hospital with bilateral progressive muscular weakness in the upper limbs (day 0). He was diagnosed with MMN in another hospital and treated using intravenous immunoglobulin (IVIg) 11 years prior. However, he had discontinued visiting the hospital on his judgment. Upon neurological examination, visual field, pupils, light reflex, and ocular movement were normal. No other cranial nerve disorders were detected. There were bilateral muscle atrophy and weakness of the distal upper limb, mainly on the right side (Medical Research Council Scale for Muscle Strength: 1–3). There were no deep tendon reflexes of the right triceps brachii and bilateral brachioradialis; there were no evoked pathological reflexes. The grasping power was 13.5 and 22.0 kg in the right and in the left hand, respectively. There were no findings of sensory disorders, ataxia, and autonomic dysfunction. Additionally, the complete blood count and biochemical examinations, including liver and kidney

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function, revealed normal findings. Autoantibody tests revealed positivity for anti-GM1 IgM antibodies. Cerebrospinal fluid analysis results were normal. Contrast-enhanced magnetic resonance imaging (MRI) did not reveal abnormalities in the brain, optic nerve, and spine. Nerve conduction studies revealed bilateral conduction block in the median nerve between the wrist and elbow segment (Fig. 1). The sensory nerve action potentials and sensory conduction velocities were normal. He was diagnosed with MMN recurrence and treated using IVIg from day 4. There was gradual post-treatment improvement in the muscle weakness of the upper limbs. Additionally, although he did not present pre-treatment complaints of visual symptoms, he said, "I could see more clearly, and I felt like fatigue of my eyes was removed" after treatment. He was discharged on day 18.

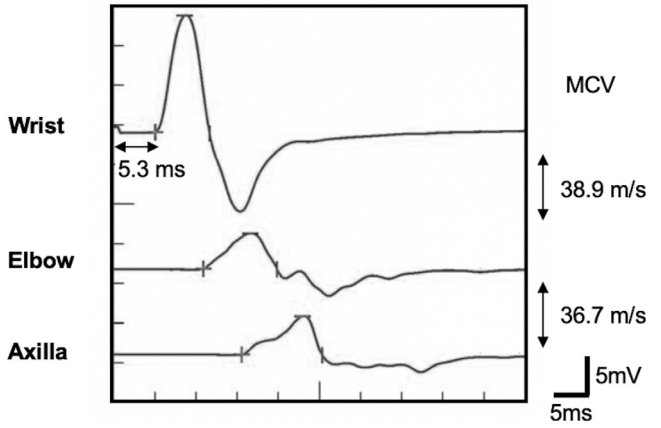

Fig. 1. Motor-nerve conduction studies focused on the right median nerve on admission. Prolonged distal latency, slowed conduction velocities, and conduction block between the wrist and elbow are observed. MCV, motor-nerve conduction velocity.

At 5 months after being discharged, he developed recurrent upper limb muscle weakness. IVIg administration improved muscle weakness and bilateral blurred vision. At 9 months after discharge, ophthalmologic examination was unremarkable (eyesight [right, 0.7; left, 0.6], intraocular pressure [right, 12 mmHg; left, 9 mmHg], and Fricker value [right, 39.33; left, 37.33]). The post-treatment improvement in muscle weakness and blurred vision was observed repeatedly for 3.5 years after the discharge. Therefore, in patients with MMN, there might be visual pathway involvement in addition to motor nerve involvement. Fig. 2 presents the results of the P100 latencies of visual-evoked potential (VEP) and grasping power obtained within 1 week before and after the treatment throughout the last 2.5-year follow-up examinations. P100 latencies were within the normal range (<121.0 ms) before IVIg treatment (Rt, 109.9 ± 2.4 ms; Lt, 108.0 ± 2.2 ms [mean ± standard deviation]). There was significant post-treatment shortening in the latencies (Rt, 103.4 ± 2.4 ms; Lt, 103.4 ± 0.9 ms; p = 0.03, Wilcoxon's signed-rank sum test) (Fig. 2, Supplementary Figure) (Tobimatsu et al., 1993).

3. Discussion

To the best of our knowledge, this is the first reported case of repeated improvements in visual symptoms with IVIg treatment for MMN. Visual symptom improvement was associated with the P100 latency shortening of VEP.

Chronic inflammatory demyelinating polyradiculoneuropathy (CIDP), which is another demyelinating neuropathy with motor conduction block, is often accompanied by visual symptoms (Imamura et al., 1994; Holtkamp et al., 2001). The mechanism for visual impairment in CIDP remains under debate. The immunolog-

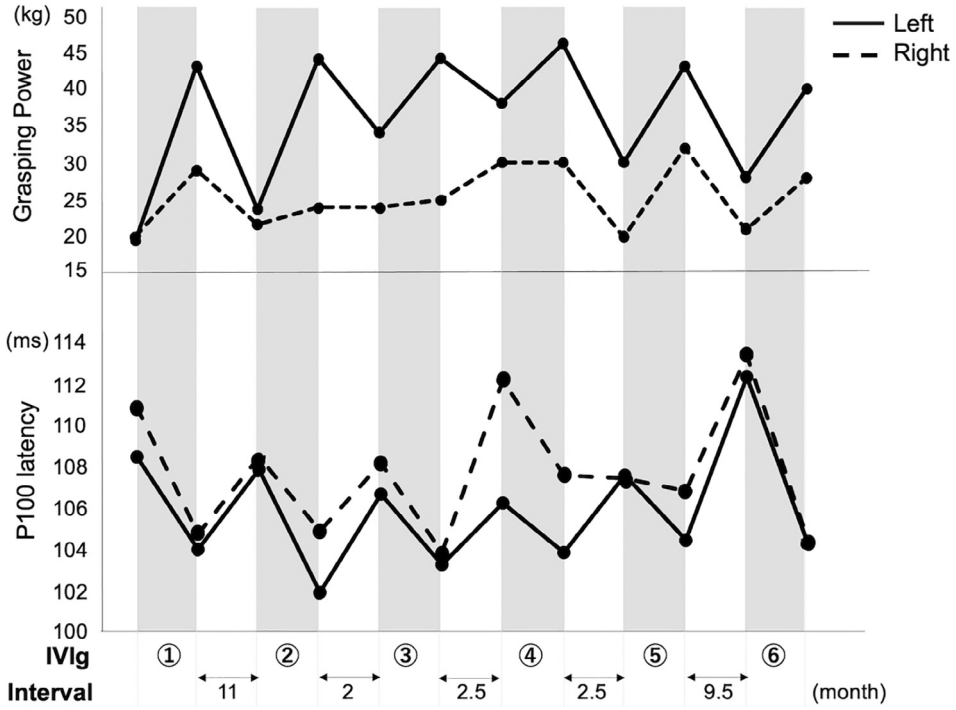

Fig. 2. Clinical course. We examined the grasping power and VEP within 1 week before and after IVIg. VEP to checkerboard pattern reversal stimulation was recorded at midline occipital portion (MO). Before IVIg treatment, there is bilateral deterioration of grasping power in the hands, although P100 latencies are within the normal range (<121.0 ms). After every IVIg treatment, an improvement in the grasping power is observed. In addition, P100 latencies have shortened significantly (p = 0.03; Wilcoxon's signed rank-sum test). VEP, visual evoked potential; IVIg, intravenous immunoglobulin.

ical mechanism affecting the glial cells may be associated with visual pathway impairment; however, myelin in the central nervous system comprises oligodendrocytes, rather than the Schwann cells in the peripheral nerves (Prasad and Galetta, 2011). Regarding MMN, there was only one reported case with optic neuritis, which was indicated by contrast enhancement on optic nerve MRI (Lee et al., 1999). The mechanism underlying visual pathway impairment in our patient remains unclear. However, several immunological processes have been considered. For example, GM1 antibodies can impair the visual pathway since GM1 ganglioside, which is one of the target antigens in patients with MMN, is expressed in both the peripheral and central nervous systems (Marconi et al., 2005).

Some patients with CIDP present with prolonged P100 latency of VEP; however, the utility of VEP in evaluating the treatment effect remains unclear (Stojkovic et al., 2000; Pakalnis et al., 1988; Gigli et al., 1989; Takeda et al., 2010; Knopp et al., 2014). We did not detect prolonged P100 latency; however, the post-treatment shortening of P100 latency may indicate the presence of visual pathway impairment. However, VEP abnormalities result from disorders in various regions, including the retina, optic nerve, optic tract, optic radiations, or visual cortex (Donnel, 2019). In our case, there were no obvious abnormalities on ophthalmologic examination and contrast MRI findings. Therefore, we could not precisely localize the lesion responsible for visual symptoms.

Taken together, our findings indicated that some patients with MMN might present with visual pathway impairment. Further evaluation of visual pathway involvement is needed in patients with MMN, even in the absence of conscious complaints of visual symptoms. Additionally, VEP may be a useful tool for monitoring the effect of IVIg treatment on the visual pathway.

Declaration of Competing Interest

The authors declare that they have no known competing financial interests or personal relationships that could have appeared to influence the work reported in this paper.

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Ethics approval

This study was approved by the institutional review board of Kansai Electric Power Medical Research Institute.

Author contributions

K.K. and S.W. contributed to data acquisition. K.K. primarily analyzed the data and performed the statistical analyses. The first draft of the manuscript was written by K.K., S.W., Y.O., Y.T., and M.I.

M.I. and T.H. revised the manuscript for important intellectual content. All authors have read and approved the final manuscript.

Appendix A. Supplementary data

Supplementary data to this article can be found online at https://doi.org/10.1016/j.cnp.2021.05.003.

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