



Case report

Bilateral optic neuritis related to chronic inflammatory demyelinating polyneuropathy



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ARTICLE INFO

Article history:

Received 26 December 2013

Received in revised form

9 April 2014

Accepted 10 April 2014

Available online 23 June 2014

Keywords:

optic neuritis

bitemporal visual field defect

chronic inflammatory demyelinating
polyneuropathy

ABSTRACT

Chronic inflammatory demyelinating polyneuropathy (CIDP) is a condition that mainly affects the peripheral nervous system; however, the central nervous system has also been involved in rare cases. Herein, we describe the case of a 33-year-old man with CIDP who presented with progressively blurred vision and pain with eye movement in both eyes for 1 month. Ocular examination revealed reduced visual acuities of 0.15 (oculus unitas or OU) and unremarkable fundi (OU). Furthermore, bitemporal visual field defects and prolonged visually evoked potentials were evident. Brain magnetic resonance imaging revealed nothing remarkable along the optic nerve and chiasm. These findings were compatible with the diagnosis of bilateral optic neuritis. The patient's symptoms and visual acuity improved after 5 days of intravenous (IV) corticosteroid pulse therapy, which was subsequently replaced by oral prednisolone therapy with a tapering schedule. The patient's visual acuity returned to 1.0 (OU) 6 months after treatment. However, bilateral optic neuritis recurred in 7 months while the patient was on oral prednisolone and azathioprine. IV corticosteroid pulse therapy was subsequently reinitiated and the patient's visual acuity returned gradually to 1.0 (OU). Bilateral optic neuritis is a rare manifestation of CIDP. It responded well to IV corticosteroid therapy in our case.

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1. Introduction

Chronic inflammatory demyelinating polyneuropathy (CIDP) is defined by the presence of symmetric proximal and distal muscle weaknesses with sensory symptoms and signs in all four extremities. In addition, it must also satisfy the electrophysiological features consistent with peripheral demyelinating neuropathy.¹ Although CIDP is a condition affecting mainly the peripheral nervous system, the central nervous system (CNS) has also been involved in rare cases. Five cases of CIDP-related optic neuropathy have previously been reported.^{2–6} Here, we report a case of bilateral optic neuritis related to CIDP.

2. Case report

A 33-year-old man complained of progressive gait disturbance, dizziness, as well as numbness and weakness in the distal end of all four limbs. The patient was first admitted to the hospital 2 months after the initial presentation of his symptoms because of deteriorating signs such as tongue numbness, slurred speech, and dysphagia. The patient denied recent illness, travel history, and family history. Neurological examination revealed reduced deep tendon reflexes in the distal ends of all four extremities. Electrophysiological study demonstrated prolonged distal latencies, reduced nerve conduction velocities, and delayed F responses in the median, ulnar, and tibial nerves bilaterally. Cerebrospinal fluid (CSF) analysis revealed nothing remarkable except for an elevated protein level (195.7 mg/dL; normal range 10–40 mg/dL). Brain magnetic resonance imaging (MRI) was also unrevealing. Subsequently, four regimens of intravenous (IV) immunoglobulin therapy were administered, which improved the patient's gait function. Although no maintenance therapy was subsequently initiated, the

Conflicts of interest: All contributing authors declare no conflicts of interest.

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<http://dx.doi.org/10.1016/j.tjo.2014.04.002>

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only residual symptom was that of a mild peripheral numbness. Nevertheless, the patient developed progressively blurred vision and pain with eye movement in both eyes in the following month. An ophthalmologist was consulted approximately 1 month after the onset of his visual symptoms. Upon presentation, the patient's best-corrected visual acuities (BCVAs) were 0.15 bilaterally. Pain elicited with ocular movement was apparent bilaterally. He could read 7 out of 15 Ishihara color plate with his right eye and 2 out of 15 with his left eye. Pupils were reactive to light bilaterally without relative afferent pupillary defect. Visual field (VF) examination disclosed a bitemporal VF defect (Fig. 1A), while visual evoked potential examination demonstrated prolonged p100 latencies (Fig. 2). Repeat MRI failed to identify any optic nerve lesion or evidence of multiple sclerosis (MS; Fig. 3). These aforementioned findings were compatible with the diagnosis of bilateral optic neuritis. Other laboratory investigations including complete blood count, immunoglobulin, complement, chronic reactive protein, rheumatic factor, antinuclear antibody, tumor markers, and various viral titers were all negative. Finally, serum aquaporin-4 antibody assay was also unrevealing.

The patient received IV methylprednisolone for the first 3 days (125 mg q6h), which was then tapered to 80 mg q8h on Day 4 and reduced again to 40 mg q8h on Day 5. Thereafter, the patient was maintained on oral prednisolone (60 mg q.d.) and tapered off of it gradually. The patient's peripheral numbness and visual acuity improved soon after the corticosteroid therapy. Eventually, the patient's BCVAs returned to 1.0 bilaterally with normal color test. When the steroid was discontinued after 4 months, unsteady gait and limb numbness recurred. Consequently, prednisolone (25 mg

q.o.d.) and azathioprine (25 mg q.d.) were administered for maintenance.

Unfortunately, blurred vision (oculus unitus or OU) recurred following 3 months of maintenance therapy. This time, the BCVAs were 0.3 (OU) but the patient did not complain of weakness or numbness of the extremities. VF examination showed generalized depression with paracentral scotoma in both eyes (Fig. 1B). Consequently, corticosteroid pulse therapy was reinitiated and the patient's BCVAs returned gradually to 0.6 (OU) in 3 weeks. In addition, azathioprine 25 mg q.d. was added to the treatment regimen in the following month. Prednisolone was slowly tapered to 15 mg q.d. to prevent recurrence. Nevertheless, CIDP recurred with unsteady gait and hand weakness bilaterally 10 months later. This time, there were no visual symptoms and the patient's BCVAs were 1.0 (OU). Corticosteroid pulse therapy was administered for 3 days, followed by plasma exchange. Those symptoms soon improved after completing the treatment course. The patient is currently on oral prednisolone (60 mg q.d.) and azathioprine (50 mg q.d.).

3. Discussion

CIDP typically presents as a progressive or relapsing, symmetric motor neuropathy with proximal and distal muscle weaknesses developing over 2 or more months. Deep tendon reflexes are often reduced or absent.⁷ Traditionally, CIDP has been considered a heterogeneous disorder with a broad spectrum of clinical phenotypes. The European Federation of Neurological Societies/Peripheral Nerve Society's (EFNS/PNS) CIDP treatment guideline has defined

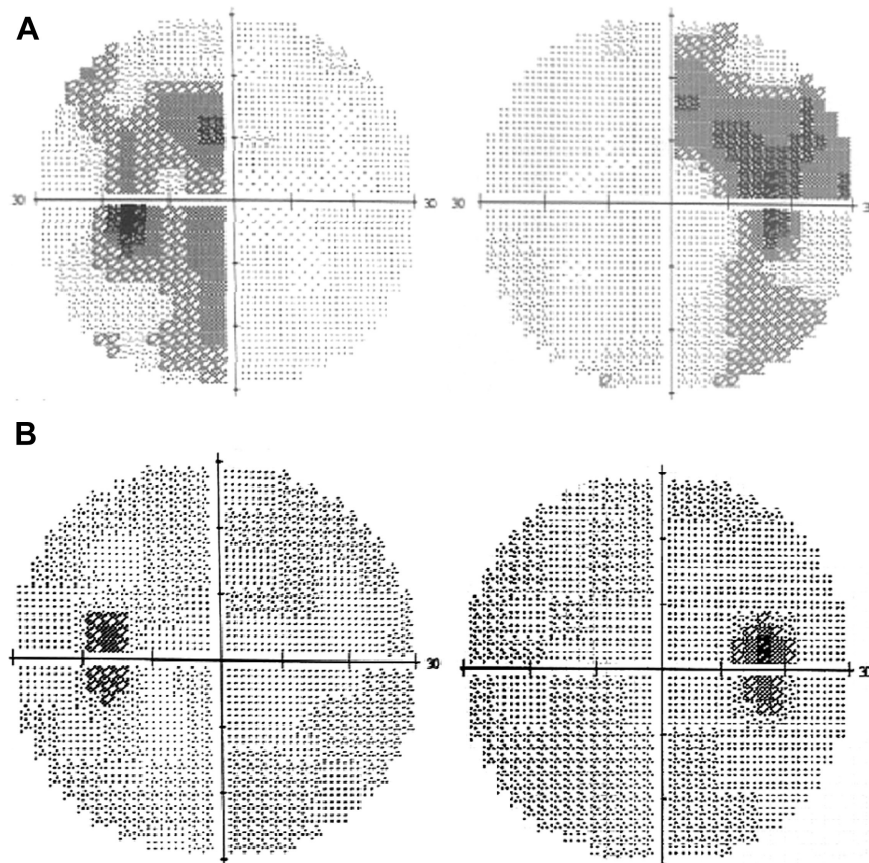


Fig. 1. (A) Visual field (VF) showed bitemporal visual field defect. (B) VF showed bilateral general depression with paracentral scotoma.

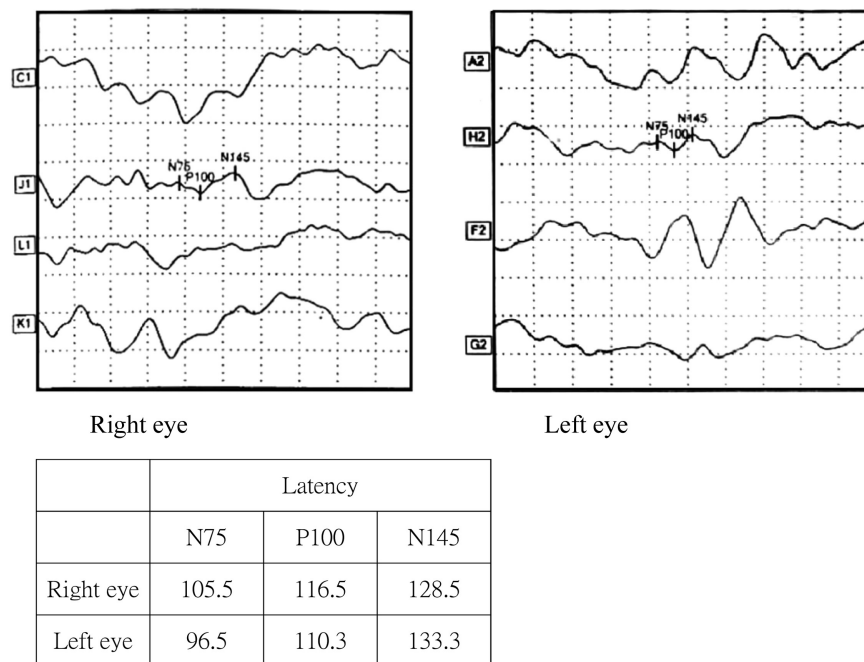


Fig. 2. Prolonged latency of pattern visual evoked potentials in both eyes.

several variants of CIDP, including pure sensory, pure motor, predominantly distal weakness, and CIDP with focal presentation.¹ Although CIDP is considered to be an autoimmune disorder of the peripheral nervous system, in two large series, CNS involvements were observed in 5% and 8% of the patients, respectively.^{8,9}

Our patient's symptoms of bilateral and symmetric muscle weakness, numbness in the extremities, decreased deep tendon reflexes are compatible with the diagnosis of CIDP. The patient's

nerve conduction velocity (NCV) examination demonstrated prolonged distal latency of medial and ulnar nerve bilaterally, which also satisfies the CIDP's electrodiagnostic criteria from the EFNS/PNS guideline.¹ In addition, elevated protein levels with normal white cell counts were also demonstrated repeatedly in our patient's CSF. This is also in accordance with the guideline. Unfortunately, nerve biopsy was not performed due to the patient's refusal. However, the diagnostic utility of nerve biopsy for suspected CIDP is still controversial.¹⁰ Nerve biopsy is used mainly when other studies fail to establish the diagnosis of CIDP clearly, particularly when electrophysiological criteria for demyelination are not met.

In our patient, the diagnosis of chiasmal optic neuritis should be considered due to bilateral temporal VF defect. Chiasmal optic neuritis is a clinical syndrome consisting of acute visual loss with a chiasmal VF defect pattern and/or radiographic demonstration of chiasmal inflammation. The incidence of chiasmal optic neuritis is rare worldwide, and most are associated with inflammatory demyelinating diseases, most commonly, MS. There were three cases of MS-related chiasmal optic neuritis¹¹ and one idiopathic chiasmal optic neuritis¹² reported in 2012 in Taiwan. In our case, the MRI did not reveal any evidence of MS, chiasmal lesion, or other pathology. Furthermore, chiasmal optic neuritis related to CIDP has never been reported, and our case was the first one.

Including our patient, there have only been six patients, totaling 11 eyes, with CIDP-related optic neuropathies reported in literature.^{2–6} The clinical features of these patients are summarized in Table 1. In these six patients, there were four males and two females. The patient's age at the time of diagnosis ranged from 18 to 57 years. Of the five patients who received MRI with contrast, two demonstrated optic nerve enhancements, whereas the other three demonstrated nothing remarkable. The initial visual acuity (VA)s ranged from counting fingers at 10 to 1 cm. Furthermore, 7 of 11 eyes had initial VAs of 0.15 or worse. All patients had final VAs greater than 0.6 except for one, who had final VAs of light perception (OU) only. However, in comparison with other patients, this patient with poor final VAs was diagnosed with CIDP for more than 2 decades and suffered several recurrences.

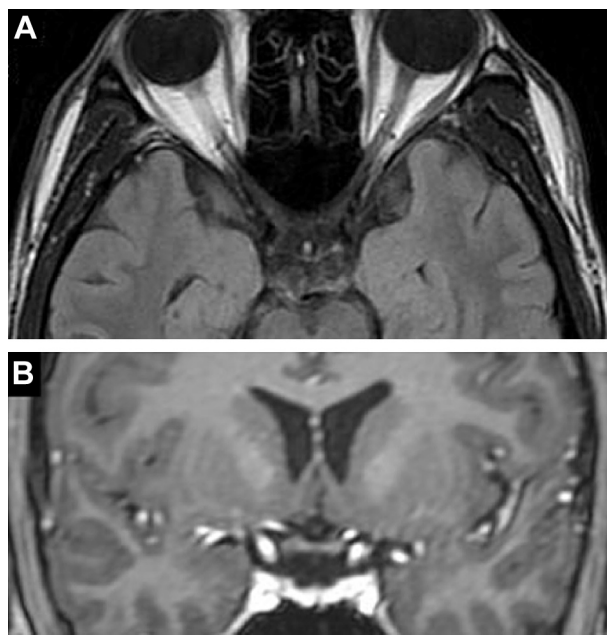


Fig. 3. Magnetic resonance imaging of optic nerve and chiasm. (A) Axial FLAIR image did not reveal any optic nerve or intraorbital lesion. (B) Coronal T1-weighted image after gadolinium injection did not show abnormal enhancement or enlargement of chiasm.

Table 1

Clinical features of six patients with CIDP-related optic neuritis.

	Age/Sex	Eye	Disc	Ocular pain	Initial VA	Visual field	VEP	MRI	Treatment	Follow-up period/Prognosis
Imamura et al (1994) Japan ²	35/M	OU	Swelling	+	OD 0.01 OS 0.02	Central scotoma	WNL	Negative	Oral prednisolone 60 mg q.o.d.	2 mo// OU 1.0
Lee et al (1999) America ³	57/F	OU	WNL	–	OD 1.0 OS 0.8	Superior arcuate defect + paracentral OD Inferior nasal + paracentral OS	Delayed latency	Right optic nerve enhancement	Steroid pulse therapy	6 mo/OD 0.8 OS 0.6
Tsai et al (2000) Taiwan ⁴	32/M	OU	Mild pallor	–	OD 0.1 OS counting finger (CF)/ 10 cm	Central scotoma	Delayed latency	Left optic nerve enhancement	Steroid pulse therapy	10 mo, recured once/OU 0.8
Holtkamp et al (2001) Germany ⁵	41/ M	OU	Disc atrophy	–	OD 0.6 OS 0.25	N/A	N/A	N/A	Oral azathioprine + prednisolone for CIDP	7 mo/ OU Light perception
Watanabe (2013), Japan ⁶	18/F	OS	Mild pallor	+	OD 1.0 OS 0.1	N/A	WNL	Negative	Immunoadsorption	1 y OU 1.0
Our case, (2013) Taiwan	33/M	OU	WNL	+	OD 0.15 OS 0.15	Bitemporal VF defect	Delayed latency	Negative	Steroid pulse therapy	2 y, Recurred once/OU 1.0

CIDP = chronic inflammatory demyelinating polyneuropathy; MRI = magnetic resonance imaging; N/A = not available; OD = oculus dexter; OS = oculus sinister; OU = oculus unitas; VEP = visual evoked potential; VF = visual field; WNL = within normal limits.

Currently, there are no formal guidelines for the treatment of CIDP-related optic neuritis. According to the optic neuritis treatment trial (ONTT), IV methylprednisolone therapy may lower the recurrence rate of optic neuritis in comparison with oral prednisolone (25% vs. 41%). In addition, high-dose IV methylprednisolone therapy was found to be more effective in improving short-term VA, particularly, VF and contrast sensitivity recovery.¹³ By contrast, some studies suggest that long-term remissions can be achieved with pulse high-dose corticosteroids in approximately 25–40% of CIDP patients.¹⁴ Therefore, the use of IV steroid was reasonable in our case. Like previous case reports of CIDP-related optic neuritis, our experience also confirmed IV steroid's effectiveness.

CIDP with optic neuritis may potentially be a new category of its own. Unfortunately, the number of currently documented patients is insufficient to make such a distinction. There were a few common traits noted among those with CIDP-related optic neuritis. For example, it was shown that those with optic nerve involvement had a milder CIDP progression. In addition, the patients were mostly middle-aged Asian males. Furthermore, they tend to respond favorably to steroid therapy.

Optic neuritis is a rare manifestation of CIDP, which involves mainly the peripheral nervous system. Clinicians need to recognize CIDP as a possible cause of optic neuritis and know that corticosteroids can hasten the recovery of visual symptoms. Furthermore, the maintenance therapy should be tapered off carefully to avoid CIDP and optic neuritis recurrence.

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