Sir,

Optic neuropathy in a patient with chronic inflammatory demyelinating polyneuropathy

Chronic inflammatory demyelinating polyneuropathy (CIDP) is a widespread, often patchy, demyelination of the peripheral nervous system (PNS). Optic neuropathy is a rare manifestation of CIDP. Some sporadic reports have mentioned central nervous system involvement in CIDP.[1–4] But the course and prognosis of optic neuropathy related to CIDP are not well documented in the literature. We illustrate the abnormal findings, relapsing course and treatment of optic neuropathy in a patient with CIDP.

Case report

A 32-year-old man sought care because of acute onset of bilateral visual loss. About 1 year before this episode he had suffered from numbness and weakness of all four distal limbs for more than 10 weeks, and his illness was diagnosed as CIDP based on abnormal findings of nerve conduction velocity and sural nerve biopsy. The electrophysiological study showed diffuse slower nerve conduction velocity with conduction block in the proximal part and delayed or absent F response in bilateral peripheral nerves, such as median nerves, ulnar nerves, peroneal nerves and tibial nerves. The sural nerve biopsy revealed a demyelinating process, some active macrophage-mediated demyelination and no perivascular inflammatory cell infiltration. Since then, the symptoms of CIDP had subsided and no relapse occurred under treatment with oral prednisolone. However, after the dosage of oral prednisolone was tapered to 10 mg per day, he suffered from acute onset of bilateral visual loss.

Ophthalmic examination revealed a best-corrected visual acuity of 6/60 in the right eye and counting fingers at 10 cm in the left eye. There was no disturbance of ocular motility. Slit-lamp examination was normal. Ophthalmoscopy revealed mild pallor of the optic discs bilaterally. Intraocular pressure was 9 mmHg in both eyes. Goldmann visual field test showed large central scotomas in both eyes. The electroretinogram and electro-oculogram were normal, but the pattern visual evoked potential (VEP) disclosed delayed p100 latency bilaterally. Fluorescein angiography was performed and no significant lesion was found (Fig. 1).

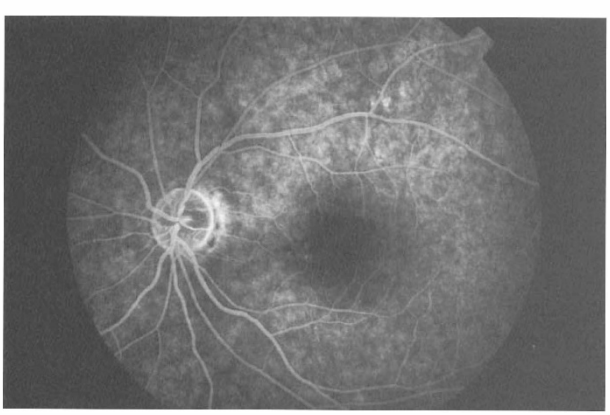

Fig. 1. *Fluorescein angiography showed no significant lesion.*

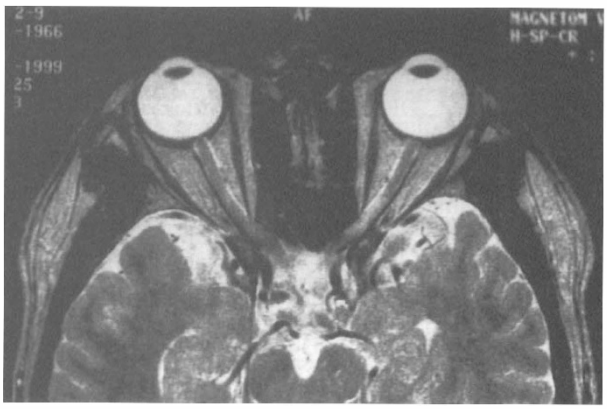

Fig. 2. *T2-weighted MRI scan disclosed enhancement of the left optic nerve.*

The patient did not complain of symptoms of vertigo, focal weakness, sensory abnormalities, urinary incontinence or gait disturbance. Systemic examination revealed the decreased deep tendon reflex over distal limbs. Magnetic resonance imaging (MRI) of brain was performed, but the result was normal. There was no abnormal finding on the chest radiograph. The analysis of cerebrospinal fluid disclosed an elevated protein level (93 mg/dl, normal range 15–45 mg/dl) with a polyclonal pattern and normal cell count. Laboratory investigations of peripheral blood were negative, including complete blood count, immunoglobulin, complement, chronic reactive protein, rheumatic factor, antinuclear antibody and various virus titres.

He received pulse therapy of methylprednisolone (250 mg q6h for 3 days) followed by oral prednisolone (30 mg b.i.d.), his visual acuity recovering rapidly to 6/7.5 in the right eye and 6/6 in the left eye and no visual field defect being observed on Goldmann perimetry. Thereafter, the dosage of oral prednisolone was tapered gradually.

Unfortunately, blurred vision in the left eye recurred 4 months later. The visual acuity was 6/8.6 in the right eye and 6/20 in the left eye. There was no complaint of weakness or numbness over the extremities. Goldmann visual field examination showed cecocentral scotoma in the left eye. Dilated fundus examination revealed only mild pallor of the optic discs bilaterally. MRI disclosed enhancement of the left optic nerve on the T2-weighted image, so optic neuropathy on the left side was confirmed (Fig. 2).

Increasing the dosage of oral prednisolone (30 mg b.i.d.) improved the visual acuity rapidly to 6/7.5 in both eyes. The visual field defect also diminished. Thereafter, the rate of tapering of steroid was slowed down. After 6 months of follow-up the dosage of oral prednisolone was tapered to 30 mg per day gradually, and there has been no relapsing episode to date.

Comment

Central nervous system (CNS) involvement is a rare manifestation of CIDP, and its precise incidence is unclear. Mendell *et al.*,[1] Pakanlis *et al.*,[2] Uncini *et al.*[3] and Ohtake *et al.*[4] reported a total of 11 of 55 consecutive patients with clinical signs or symptoms of CNS involvement. Optic neuropathy was found in 8 of these 11 patients. In general, these clinical features are indistinguishable from those of multiple sclerosis. In addition to clinical CNS findings, subclinical involvement can be demonstrated by VEP, brainstem auditory evoked response (BAER), electroencephalography (EEG) and MRI. In these four studies there were 17 patients with abnormal results of evoked potentials or MRI but no CNS symptoms or signs noted. In these 17 patients, VEPs were abnormal in 11, BAERs in 2 and EEGs in 1. MRI showed changes suggestive of CNS demyelination in 7 patients. According to the VEP findings of these reports, the optic nerves seem to be most frequently affected in CIDP patients.

The mainstay of CIDP treatment is corticosteroid, for instance prednisolone 60–100 mg per day for 2–4 weeks, then gradually tapered to 5–20 mg every other day. The beneficial effect of corticosteroid is to speed recovery and to prevent relapse. This disease is also responsive to plasma exchange or intravenous infusion of high dose of immunoglobulin.

Optic neuropathy is one of the clinical manifestations in CIDP with CNS involvement, but its course, therapy and prognosis are not well documented in the literature. Our patient initially suffered from PNS demyelination, which was followed by optic neuropathy about 1 year later. Under treatment with corticosteroid, his PNS symptoms and signs subsided significantly and there has been no relapse to date. However, recurrent episodes of optic neuropathy occurred within 4 months in spite of pulse therapy with methylprednisolone. The different courses and responses to corticosteroid between the myelinopathy in the PNS and CNS suggest a possible difference in their immunopathogenetic mechanisms.

In our report, intravenous corticosteroid accelerated the recovery of visual function but did not prevent the subsequent attack of optic neuropathy. It needs to be further investigated whether CIDP-related CNS demyelination is also responsive to other therapies of CIDP, such as plasmapheresis or intravenous infusion of immunoglobulin.

Although CIDP is classified as a demyelinating disorder of the PNS, it does cause clinical or subclinical CNS problems, especially optic neuropathy, and needs cooperation between neurologists and ophthalmologists for more effective treatment.

References

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Sir,

Extensive globe rupture from blunt trauma: a case with a hope

Ocular trauma is a commonly encountered presentation to eye casualty. The majority of the injuries are minor, but over 10% of patients will lose useful vision in the injured eye.[1] Various aspects of serious ocular trauma, including the demography, prognostic variables, histopathological characteristics and role of vitrectomy, have been studied to identify features predictive of outcome and establish useful guidelines for management. We report a patient with extensive globe rupture who, despite the presence of uveal prolapse at scleral wound and an incomplete scleral wound closure, made a good visual recovery, which was against that predicted by outcome features.

Case report

A 28-year-old man presented to the eye casualty with his left eye having been hit by a metal bar at work. His visual acuity was perception of light on the left. Ocular examination revealed a full-thickness corneal laceration starting at the 9 o'clock hour position, extending downwards to the 7 o'clock hour position of the cornea and into the sclera, where it was obscured by extensive