sheath decompression on each side. This suggests that optociliary shunts are dynamic structures which can appear and disappear with changes in central retinal venous pressure.

CASE REPORT

A 21-year-old man presented in 1981 with a history of right earache with discharge, headache, and obscurations of vision in both eyes. Examination showed bilateral papilloedema but no other neurological abnormality. Cerebrospinal fluid (CSF) had normal constituents but pressure was raised at greater than 400 mm H₂O. Computed tomography scan showed petrous osteomyelitis and small cerebral ventricles. He was treated by right mastoidectomy and during this procedure the lateral dural venous sinus was visibly thrombosed. The headache and obscurations did not resolve so he underwent lumboperitoneal CSF shunting in 1982. He required numerous shunt revisions to maintain CSF pressure control over the next 4 years. The shunt was removed at the time of a lumbar 4/5 laminectomy in 1986. Headache and visual obscurations recurred and the shunt was reinserted. However, during the following year, the shunt became infected and was removed again. His headache worsened and visual obscurations were occurring 15 times a day in each eye.

In April 1988 he underwent right optic nerve sheath decompression via a medial orbital approach under general anaesthesia. The procedure was without complication and the frequency of obscurations immediately reduced to roughly twice per week. One week after surgery the optociliary shunt that had been evident on the right disc before surgery was much reduced in calibre (Fig 1).

The left eye continued to suffer frequent obscurations and by later in the year, these were occurring 20–30 times a day. In January 1989 the left optic nerve was decompressed using the same technique. Again surgery went smoothly and by 4 days postoperatively the optociliary shunt vessel on this disc was much reduced in diameter (Fig 2). Both optociliary shunts continued to undergo further involution with time and, late in 1992, there was no swelling of either disc. Both discs were flat and pale with no conspicuous optociliary shunts.

At last follow up in early 1993, vision was preserved at 6/5 N5 in both eyes and, although visual fields were smaller than a normal individual of his age, his vision was functionally normal. Repeated visual field assessment showed residual enlargement of the blind spots although the optic discs were clinically not swollen.

COMMENT

Optociliary shunts may be congenital or acquired.^{1,2} Acquired optociliary shunts are classically associated with optic nerve sheath meningioma³ although central retinal vein occlusion is the most common cause in clinical practice. Dowhan *et al*¹ give a list of causes including glaucomatous optic atrophy, chronic papilloedema,² optic nerve glioma, arachnoid cyst of optic nerve, neurofibromatosis, benign intracranial hypertension (pseudotumour cerebri), optic disc drusen, optic nerve coloboma, and osteosclerosis.

Optociliary shunt vessels develop when dilatation of collateral channels connecting retinal and choroidal venous systems occurs as central retinal venous pressure (CRVP)

rises.^{2,4} The increase in CRVP may follow obstruction in the vessel lumen (central retinal vein occlusion), nerve itself (glioma), subarachnoid space (raised CSF pressure), or by disease of the nerve sheath (meningioma).

Previous reports of resolution of optociliary shunts are scarce and we have only been able to locate one case in which resolution followed optic nerve sheath decompression. Perlmutter *et al*⁵ described a patient with benign intracranial hypertension (pseudotumour cerebri) where optociliary shunt vessel calibre was reduced after bilateral optic nerve sheath decompression.

Other procedures modifying CSFP or intracranial pressure have rarely been reported to cause resolution of optociliary shunts. Dowhan *et al*¹ reported two patients with neonatal hydrocephalus where optociliary shunts disappeared following CSF shunting procedures. Tyson and Lessell⁶ described optociliary shunt vessels associated with chronic atrophic papilloedema which resolved when a massive meningioma was removed.

The case reported here and the others quoted demonstrate that optociliary shunts due to raised CSFP with chronic papilloedema are reversible when CSFP is reduced. The reduced CSFP reduces CRVP and permits more venous drainage by the central retinal vein, reducing the calibre of the blood column in the optociliary shunt vessels. This regulation of calibre appears to occur rapidly with normalisation of CRVP and study of optociliary shunt vessel calibres may provide longitudinal information regarding CSFP in the optic nerve sheath in patients with chronic papilloedema.

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Paroxysmal superior rectus and levator palpebrae spasm: a unique presentation of multiple sclerosis

EDITOR,—Paroxysmal diplopia due to spontaneous ocular motor nerve discharge has not been described in multiple sclerosis, although it may occur in other conditions such as superior oblique myokymia¹ and ocular neuromyotonia.² We describe a patient who

Fig 1A

Fig 1B

Figure 1 (A) Right optic disc immediately before nerve sheath decompression. A 'Y' shaped optociliary shunt at the 8 o'clock position on the optic disc connects the inferior central retinal vein tributary with the disc margin. (B) Same optic disc 7 days after nerve sheath decompression. The optociliary shunt is reduced in calibre and barely visible.

Fig 2A

Fig 2B

Figure 2 (A) Left optic disc immediately before nerve sheath decompression. The optociliary shunt vessel is at the 10.30 o'clock position on the optic disc connecting the superior central retinal vein tributary with the disc margin. (B) Same optic disc 4 days after nerve sheath decompression. The optociliary shunt is considerably reduced in calibre.

presented with paroxysmal vertical diplopia due to superior rectus/levator complex spasm as the presenting feature of multiple sclerosis.

CASE REPORT

A 34-year-old man presented with a 3 week history of paroxysmal vertical diplopia without torsion or oscillopsia. Short attacks, lasting 3 to 4 seconds, would occur at approximately 5 minute intervals, continuously throughout the day. During attacks, he had not noticed any ocular deviation or lid position abnormality. His previous ocular and medical history was unremarkable.

Examination revealed normal ocular motility initially, although several short lived episodes lasting only 2 to 4 seconds occurred during the examination (Fig 1). Hyper-deviation of the right eye, marked right upper lid retraction, and restriction of right down-gaze were observed during episodes, indicating spasm of the right superior rectus/levator complex. Upgaze, adduction, and abduction were full and pupillary responses normal throughout and neither torsion nor micro-tremor could be detected. Left ocular motility and lid position remained normal throughout, and general and neurological examinations were normal.

Episodes occurred in a random fashion and were not associated with precipitating factors, including sustained voluntary extraocular muscle activity. During a period of 3½ hours, 54 episodes lasting only a few seconds each were recorded. Intervals between episodes typically lasted 2 to 4 minutes.

Magnetic resonance imaging (MRI) revealed multiple lesions characteristic of multiple sclerosis, including one in the mid-brain in the region of the third nerve fascicle (Fig 2). Cerebrospinal fluid analysis revealed

oligoclonal bands which were not present in serum and a diagnosis of multiple sclerosis was made.

Carbamazepine 200 mg twice daily was commenced and within 24 hours all symptoms and attacks had ceased. Two months and 6 months later, the carbamazepine was discontinued and on each occasion symptoms recurred within 48 hours. Treatment was therefore continued. One year after initial presentation he remains symptom free on carbamazepine.

COMMENT

Intranuclear ophthalmoplegia, gaze paresis, ocular motor nerve palsies, and nystagmus commonly occur as a result of brainstem and cerebellar involvement in multiple sclerosis.³ Although paroxysmal symptoms due to spontaneous discharge in peripheral nerves are well described and include paraesthesia evoked by neck flexion⁴ (Lhermitte's symptom) and tonic motor spasms evoked by movement,⁵ paroxysmal diplopia due to

spontaneous ocular motor nerve discharge has not been described in multiple sclerosis.

In superior oblique myokymia¹ and ocular neuromyotonia,² paroxysmal ocular motor nerve discharge is an intrinsic feature. In neuromyotonia, discharges are usually triggered by sustained voluntary extraocular muscle activity and ephaptic transmission has been postulated to account for co-firing of different muscles.²

Co-firing of the superior rectus/levator complex also occurs in third nerve synkinesis because of 'aberrant regeneration' of fibres and is only evident with voluntary neuromuscular activity. Ephaptic transmission may also play a role in this condition.

In our patient, two points should be considered. Firstly, spontaneous firing as a result of ectopic action potentials has been shown to occur in injured demyelinated axons in animal models⁶ and in traumatic and post irradiation neuropathy in humans.^{7,8} The underlying mechanism is thought to be impaired neural ionic buffering and elevated extracellular potassium concentration.⁹ In fact, elevated extracellular potassium has been demonstrated around demyelinated mammalian axons which exhibit ectopic action potentials.¹⁰ Secondly, isolated superior rectus/levator complex involvement indicates a fascicular rather than a nuclear lesion and this was consistent with the MRI findings.

All patients with demyelinating disease reporting paroxysmal diplopia should be examined carefully for spontaneous neural activity and, if this is demonstrated, anticonvulsant therapy should be considered.

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Figure 1 Normal ocular motility during asymptomatic periods (first, second, third row). During attacks, spasm of the right superior rectus/levator complex results in upper lid retraction and restricted downgaze of the right eye (bottom row). Attacks were self limiting, lasting typically between 2 and 4 seconds.

Fig 2A

Fig 2B

Figure 2 T1 weighted magnetic resonance imaging scan showing (A) periventricular demyelinating lesions and (B) a lesion in the midbrain in the region of the right third nerve fascicle (arrow).

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