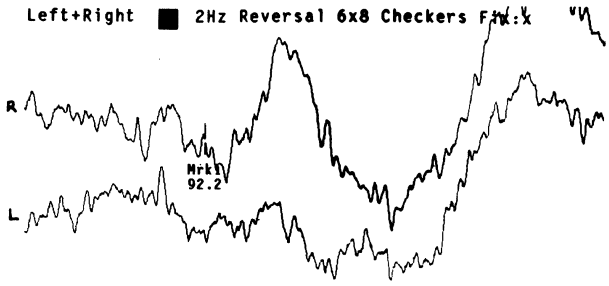

Fig. 2. *Pattern-reversal visual evoked potentials show normal P100 latencies but diminished amplitudes in the right eye (upper tracing) and especially in the left eye (lower tracing).*

involvement are strong support for a generalised mechanism of injury. This possibility resulted from a systemic autoimmune demyelination, rather than random, viral invasion of both optic nerves. In the present case, the interval of 11 days between the onset of the measles rash and the development of bilateral visual loss is enough for host immune mechanisms to become active. Possible mechanisms for the secondary autoimmune process include molecular mimicry between viral and neural antigens, or incorporation of virally coded antigens into neural tissue such as oligodendrocyte membrane or myelin sheath. Another factor may be inherited susceptibility to the production of an autoimmune response. All possible mechanisms are likely to result in demyelination. Recovery of good vision may also be consistent with primary demyelination. However, why the optic nerves were singled out in our patient remains unclear.

Visual evoked potentials 8 months later revealed low amplitudes but normal latency bilaterally. This is because some optic nerve axons might have been structurally disrupted as a sequel of acute inflammation.

It is not suggested that the present case is a variant of multiple sclerosis. Studies of simultaneous bilateral optic neuritis in adults have shown that subsequent development of multiple sclerosis is uncommon.[9] Moreover, our patient had neither a family history nor a previous or subsequent attack to suggest multiple sclerosis and oligoclonal bands were absent from her cerebrospinal fluid.

The visual improvement following high-dose corticosteroid therapy was good in our patient, although spontaneous recovery without treatment has been reported.[10,11] The natural course of acute bilateral optic neuritis following viral infections tends to be rather benign, with good prognosis for return of vision to near-normal levels. There are also no controlled studies convincingly demonstrating that any form of therapy significantly alters the course of optic neuritis. However, corticosteroids are of theoretical value in suppressing the presumptive immunological reaction and thus may shorten the recovery time.[12] Therefore, unless contraindicated, high-dose corticosteroid therapy may be justified in cases of bilateral retrobulbar optic neuritis following viral infections such as measles, or when one eye has previously defective vision.

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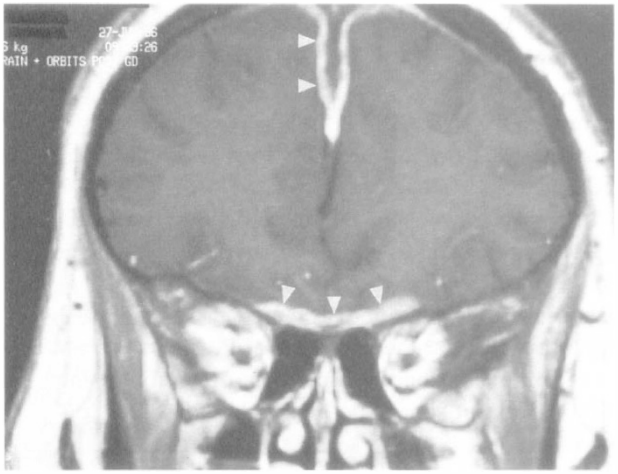
Sir,

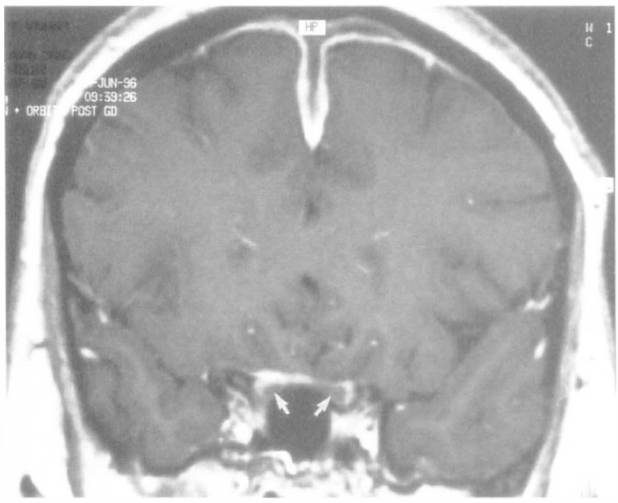
Idiopathic hypertrophic chronic pachymeningitis presenting with acute visual loss

Clinical details

Three days before presentation to another hospital, a 56-year-old woman awoke and noted almost total loss of vision in her right eye without any preceding visual disturbance, pain or systemic ill-health. She was known to have non-insulin-dependent diabetes mellitus and to smoke 20 cigarettes per day. On examination, her right visual acuity was reduced to, at best, the perception of hand movements in the superior nasal quadrant. There was a marked right relative afferent pupillary defect but otherwise full ophthalmological examination of both eyes was normal. General examination and her plasma viscosity were also normal and a presumptive diagnosis of non-arteritic posterior ischaemic optic neuropathy (PION) was made and conservative management instituted.



(a)



(b)

Fig. 1. *Post-gadolinium coronal T1-weighted MRI scan of the intracranial optic pathways showing (a) undulating thickening and enhancement of the falx, and of the dura of the anterior cranial fossa floor (arrowheads), and (b) dural thickening extending posteriorly, encasing the optic nerves in the optic foramina (white arrows).*

Seven weeks later, she developed painless blurring of vision in the left eye which progressed over 48 h. Examination at this point revealed an acuity on the left of 6/24, Ishihara 1/17, a full visual field to confrontation, but no abnormal ocular findings. The right eye was unchanged. A contrast-enhanced CT scan of the brain and orbits was normal but her ESR was now 52 mm/h and the diagnosis was changed to arteritic PION. She was commenced on intravenous dexamethasone and the vision in her left eye improved to 6/6 within a few hours.

She was then transferred to this institution for further management. Sequential bilateral temporal artery biopsies showed no evidence of arteritis and her chest radiograph, full blood count and biochemical tests were all normal. Serological tests for syphilis and *Brucella* and an autoantibody screen, including ANCA, were also normal. Cerebrospinal fluid (CSF) analysis revealed a mild lymphocytic pleocytosis (19 cells/mm^3), but protein, glucose and cytology were normal. CSF staining and culture, including that for acid-alcohol fast bacilli

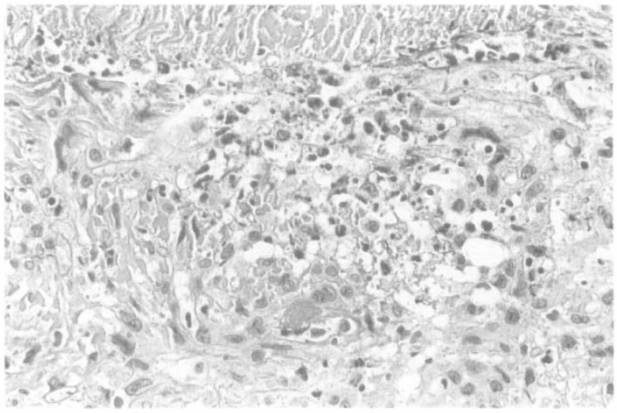


Fig. 2. *A focal collection of chronic inflammatory cells including lymphocytes and histiocytes. There is no confluent or caseous necrosis, but numerous nuclear fragments from individual cell necrosis are present. (Haematoxylin and eosin; original magnification ×430.)*

and *Cryptococcus neoformans*, were negative. However, magnetic resonance imaging (MRI) showed marked thickening of the dura mater over the cerebral hemispheres (Fig. 1a) and the floor of the anterior cranial fossa, with encroachment on the optic nerves at the optic canals bilaterally (Fig. 1b). The brain itself appeared normal.

Meningeal biopsy revealed multiple foci of inflammation containing lymphocytes, multinucleated giant cells and non-caseating microgranulomas though without any more specific features (Figs. 2, 3). Immunocytochemical analysis showed the lymphocytes to be both B and T cells. Subsequent CT scans of her thorax, abdomen and pelvis were normal. A working diagnosis of idiopathic hypertrophic cranial pachymeningitis was made and she was continued on oral prednisolone.

At 6 month review she remained systemically well and continued to have excellent vision in the left eye. Though the vision of the right eye had failed to improve, a repeat MRI scan showed partial resolution of the falcine meningeal thickening.

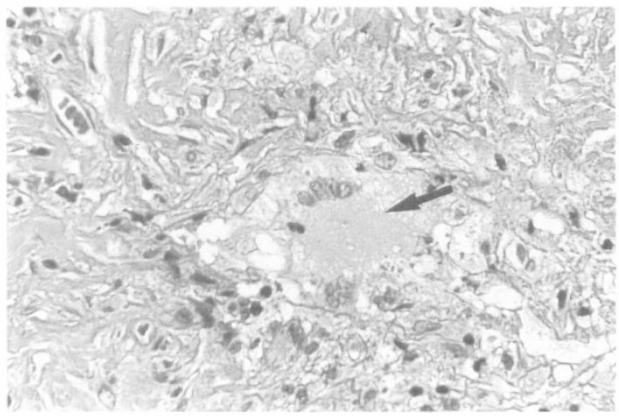


Fig. 3. *Part of a separate non-caseating granuloma containing a multinucleate giant cell (arrow). (Haematoxylin and eosin; original magnification ×430.)*

Discussion

In summary, a 56-year-old diabetic smoker presented with sudden unilateral visual loss of the right eye followed 7 weeks later by involvement of the other eye. The initial diagnosis of non-arteritic PION was changed to arteritic PION when the second eye became involved. In retrospect, both diagnoses were erroneous, and further investigation showed her to be suffering from idiopathic hypertrophic cranial pachymeningitis.

PION (ischaemia of the retrobulbar portion of the optic nerve) is a well-recognised entity and is characterised by sudden loss of vision, usually unilaterally, associated with visual field loss and an afferent pupillary defect, but no initial changes in the retina or optic disc.[1–4] It is distinguished from the much more common anterior ischaemic optic neuropathy (AION) by the absence of swelling of the optic disc. Like AION, it may result from both arteritic and non-arteritic vascular mechanisms.[5–9] However, the clinical features of PION may also occur with optic nerve compression, inflammation, infiltration, trauma or toxic damage. It is therefore only reasonable to make an initial diagnosis of PION in the presence of sudden onset of unilateral visual loss accompanied by the clinical and laboratory features of temporal arteritis.[10] In all other situations, PION must be regarded as a diagnosis of exclusion.[10–12]

In the case presented, it is likely that a diagnosis of PION was made when the patient first presented because she was a known diabetic who smoked, and therefore at high risk of a vascular event. However, the exact timing of the onset of the visual loss was not clear and there were no features of temporal arteritis at presentation to support the diagnosis. Initial correct diagnosis might have resulted in earlier treatment with steroids and might have saved the vision in her right eye and prevented subsequent problems with her left.

Hypertrophic pachymeningitis was described as early as 1876 (see Michel *et al*.[13] for a review). The condition is characterised by chronic inflammatory hypertrophy of the dura mater (though usually the pia and arachnoid are also involved), and typically occurs in the posterior fossa and falco-tentorial regions.[14–17] Although most early cases were ascribed to syphilis or tuberculosis, many other causes have since been described, including bacterial and fungal infection and non-infectious inflammatory conditions such as temporal arteritis, sarcoidosis and Wegener's granulomatosis.[18] In many cases, however, no cause can be identified, and the condition is then known as idiopathic hypertrophic cranial pachymeningitis (IHCP).[13–17,19–24] In the case presented, the non-caseating microgranulomas might have suggested sarcoidosis, but there were no other features to support this diagnosis.

IHCP often presents with multiple cranial neuropathies,[15,17,18] and ocular involvement includes blurring of vision,[15] progressive blindness,[14,20,21] papilloedema,[19] nystagmus,[19] ophthalmoplegia[14–16,22] and proptosis.[16] Cranial CT imaging commonly fails to reveal any abnormality in IHCP[14,15,19] and this case reinforces the superiority of MRI in the investigation of intracranial causes of visual loss.[25] Steroids (with or without azathioprine) have been reported to be of benefit in some cases of IHCP,[14,17,20,23] but the effects may be transient,[14,15,17,20,24] or they may be ineffective.[19] Radiotherapy has been tried without benefit.[24]

In conclusion, we believe this is the first reported case of IHCP presenting as sudden visual loss, and suggest that it should be added to the list of differential diagnoses in this situation. The case also highlights how important it is that the diagnosis of PION should not be made without further investigation, preferably using MRI, and that it should be treated as a diagnosis of exclusion.

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Sir,

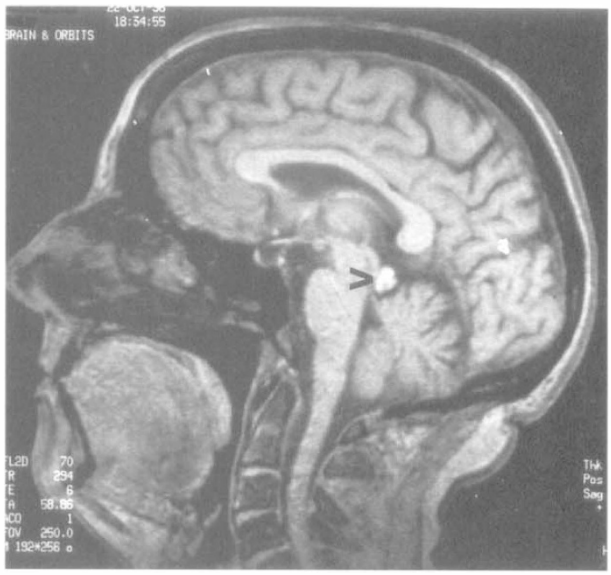
An unusual cause of intermittent vertical diplopia

Common causes of palsy of the superior oblique include closed head trauma, vascular disease, decompensated congenital paresis, localised orbital processes and herpes zoster infection. Patients usually present with vertical diplopia, which may be associated with a head tilt. We present an interesting case of intermittent vertical diplopia, presumed to be due to intermittent fourth nerve paresis induced by physical exertion.

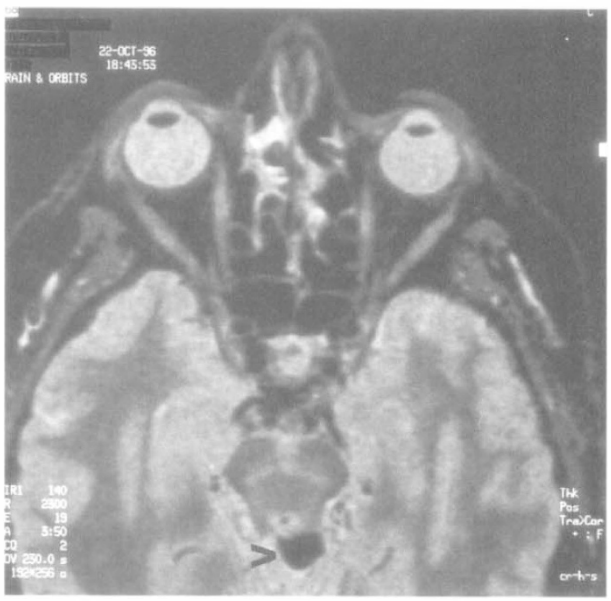
Case report

A 66-year-old, right-handed woman was referred to the neuro-ophthalmology clinic with a 15 year history of intermittent vertical diplopia. Episodes lasted about 10 min, and were precipitated by physical exertion, usually hill walking, which was a keen pastime of hers. There was no associated torsional element to the diplopia. The attacks occurred every few months, though they had recently increased in frequency just prior to presentation. She was asymptomatic between attacks, was otherwise fit and well, a non-smoker, and had no significant past medical or ocular history.

On examination, her visual acuities were 6/6 bilaterally, with normal ocular and neurological examination. In particular, ocular motor testing showed no evidence of fatiguability, and although there was a slight clinical suggestion of a positive Bielschowsky on head tilt to the left, a Hess chart showed no abnormality. The vertical fusional amplitude was 4 prism dioptres (normal). Routine screening blood tests were normal, and anti-acetylcholine receptor antibodies were not detected.



(a)



(b)

Fig. 1. *MRI scan of brain showing a small lipoma, approximately 0.8 cm in diameter (indicated by the arrow), situated in the quadrigeminal cistern, close to but not encroaching upon the colliculi. (a) Sagittal section, T1-weighted. (b) Axial section, fat suppression.*