demonstrated either within the orbit or in the region of the pituitary fossa. Full blood count and blood film showed evidence of recurrence of iron deficiency anaemia with haemoglobin of 6.1 g/dl, MCV of 62.9 fl, red cell anisocytosis, hypochromasia, polychromasia and microcytosis.

All symptoms improved following transfusion with 4 units of blood, with the return of haemoglobin level to 11 g/dl. Oral iron replacement therapy continued for several weeks following transfusion and, because of the rapid improvement on treatment, a planned MRI examination was cancelled. The optic disc oedema gradually resolved over the following 10 weeks (Fig. 2), with no recurrence of optic disc oedema over a 6 month follow-up period.

Comment

Optic disc oedema secondary to iron deficiency anaemia is very rare and the cause is unknown. Capriles[2] suggested that iron deficiency could cause cerebral and optic disc oedema by a dual mechanism of anaemic anoxia due to low haemoglobin levels and histotoxic anoxia due to failure of the cytochrome system. Jacobs[3] found that some patients with severe iron deficiency anaemia have no detectable level of cytochrome oxidase (an iron-containing enzyme) in the buccal mucosa; however, a return to normal levels occurred within 24 h following iron therapy. Stoebner *et al.*[4] observed that iron replacement therapy causes resolution of optic disc oedema before the occurrence of a significant rise of haemoglobin concentration. This may be the result of regeneration of the deficient enzymes. These observations suggest that depletion of iron-containing enzymes may be an important factor in causing the optic disc oedema. However, the reason why only a minority of patients with iron deficiency anaemia develop the condition remains unknown.

Although iron deficiency is common, only a few cases with associated optic disc oedema have been reported.[2,4–8] The patients were usually female with an age range of 14–52 years, and most presented in the second or third decade of life.[2] Intracranial pressure was raised in most of the cases, but cases with normal intracranial pressure were also reported.[5,6] The majority of the patients had headaches, including those with normal intracranial pressure, and in some patients there was significant nausea and vomiting. Some patients had asymmetrical optic disc oedema, and two patients had only one eye affected.[2] As in our patient, treatment with blood transfusion or iron supplements led to resolution or improvement of the optic disc oedema.[2,4–8] Patients in whom iron therapy was delayed developed optic atrophy and blindness.[2]

Patients with optic disc oedema secondary to iron deficiency anaemia usually present to physicians with systemic symptoms, but our patient presented to the ophthalmologist with headaches and visual obscurations mimicking benign intracranial hypertension or raised intracranial pressure due to a space-occupying lesion.

Bilateral optic disc oedema usually requires urgent imaging of brain and orbits. If this is normal, iron deficiency anaemia should be included in the differential diagnosis regardless of intracranial pressure.

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Sir,

Partial unilateral third nerve palsy and bilateral internuclear ophthalmoplegia: an unusual presentation of multiple sclerosis

Internuclear ophthalmoplegia (INO) is perhaps the most characteristic and common motility lesion associated with multiple sclerosis (MS). Between 35% and 50% of patients with MS will develop an INO, often bilateral.[1] Lesions of the medial longitudinal fasciculus (MLF) cause INO. In some instances adjacent structures can be involved resulting in additional clinical signs. Partial third nerve palsies have rarely been reported in association with MS.[2] We present a patient with partial unilateral third nerve palsy and bilateral INO.

Case report

A 50-year-old woman presented with sudden-onset binocular diplopia and drooping of the left eye upper lid. She had noticed reduced vision in her right eye 2 years previously, which spontaneously resolved.

On examination, she had a 4 mm ptosis of her left upper lid, alternating exotropia and slight left hypotropia. There was limitation of adduction in both

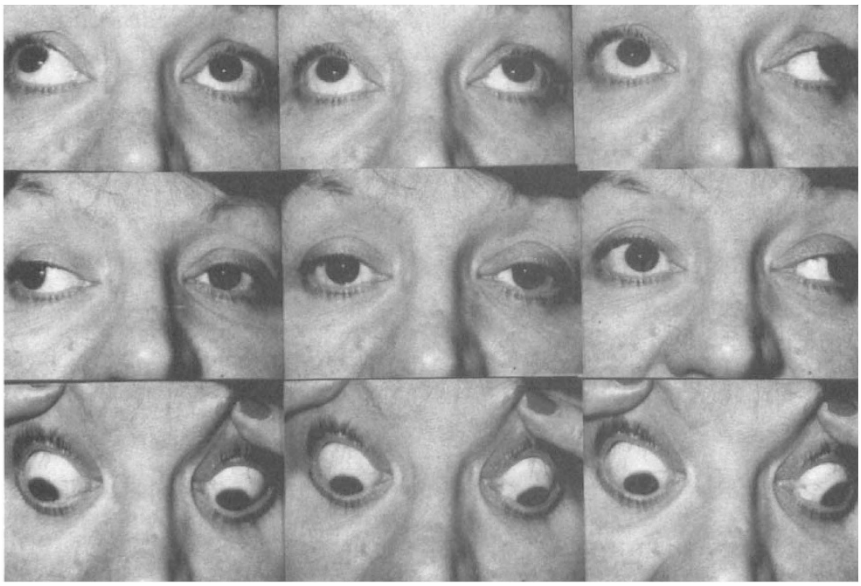

Fig. 1. *Bilateral internuclear ophthalmoplegia (note the limitation of adduction of both eyes), ptosis and some limitation of elevation in the left eye.*

eyes with abduction nystagmus and slight restriction of elevation of her left eye (Fig. 1). Her visual acuity, colour vision, anterior segment, pupillary responses, fundoscopic examination and neurological evaluation were normal. A provisional diagnosis of bilateral INO with left partial oculomotor nerve palsy was made. An urgent magnetic resonance imaging (MRI) examination and referral to a neurologist were organised.

MRI showed multiple small foci of high signal intensity on proton density and T2-weighted sequences within the deep white matter of the cerebral hemispheres, involving the periventricular, subcortical regions and the corpus callosum (Fig. 2). There was relative sparing of the brain stem. The appearances were consistent with primary demyelination.

During the neurologists' review a month later, she described an episode of sensory disturbance in her lower limbs 25 years ago and similar symptoms 12 months ago. On evaluation during the same visit, her oculomotor symptoms had completely resolved and ocular movements were full with normal saccades. Neurological examination showed an asymmetrical clonus at the right ankle with pathological brisk reflexes in both lower limbs and extensor plantar response.

Routine haematological, biochemical, microbiological and immunological investigations were unremarkable. Lumbar puncture revealed a normal cerebrospinal fluid protein, glucose, cell count and electrophoresis.

Comment

Multiple sclerosis usually presents in young adults with focal neurological signs. The ophthalmic presentations include optic neuritis, INO, cranial nerve palsies (III, IV and VI) and nystagmus.

INO can be isolated or associated with eye movement disorders such as conjugate hypermetric saccades, rebound nystagmus, conjugate gaze-evoked nystagmus and marked disorder or vestibulo-ocular response (VOR) suppression in the horizontal plane.

Bilateral INO in a young patient is most commonly caused by multiple sclerosis. The other causes include basilar artery disease,[3,4] vasculitides, syphilis, fourth ventricular brain stem tumours, Chiari malformation and trauma.

Many patients with INO have no visual symptoms. In others, the presenting symptom may be diplopia due to limitation of adduction or skew deviation and oscillopsia

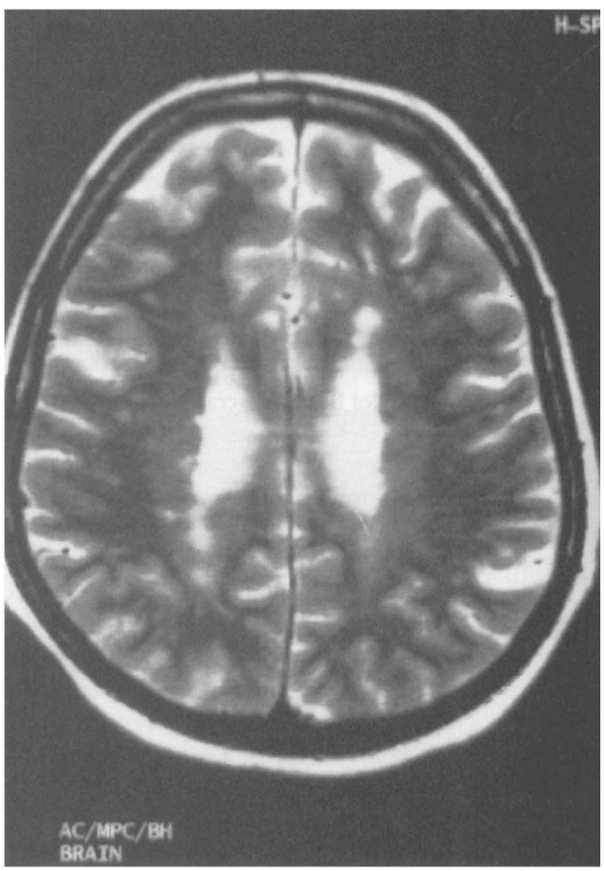

Fig. 2. *Axial T2-weighted MR sequence shows multiple small foci of high signal intensity involving the periventricular region.*

due to adduction lag, abduction nystagmus or deficient VOR response.

In some instances in which MLF are damaged, adjacent structures, particularly the paramedian pontine reticular formation, abducens nuclei, abducens fascicle and oculomotor nuclear complex, are also involved. Ptosis, partial nuclear oculomotor nerve palsy and trochlear nerve palsy are some of the clinical signs described with INO.[3–5] Isolated pupil-sharing third nerve palsy has also been reported as a presenting sign in a patient with MS.[2]

Our patient presented with horizontal diplopia and unilateral ptosis. The clinical signs suggested bilateral INO with left partial oculomotor nerve palsy. Though the MRI of the midbrain did not show any foci of demyelination, it is likely that both the MLF and a part of the oculomotor fascicles were involved. The presence of ptosis suggests that the fibres controlling the levator palpebrae superioris muscle are located caudally in the oculomotor fascicles.

A clinical diagnosis of MS was made on the basis of the patient's past history and her most recent problems. MRI helped to confirm the diagnosis. In most patients, visual symptoms resolve completely, because of either recovery of function of MLF axons or central adaptive mechanisms. Our patient had complete resolution of symptoms in less than 2 weeks.

We are not aware of any reports of bilateral INO and partial unilateral oculomotor nerve palsy in MS. This is an example of the diverse ocular presentations of this demyelinating disorder.

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Sir,

A family with Rieger's syndrome and aniridia

Rieger's syndrome is an autosomal dominant disorder that affects the anterior segment of the eye bilaterally, causing glaucoma in 50% of those affected. It is associated with dental hypoplasia, craniofacial dysmorphism and umbilical stump abnormalities. The responsible gene, *RIEG/PIX2*, is on chromosome 4q25.[1] There are also other genes associated with Rieger's syndrome, such as *RIEG2*[2] and *FKHL7*.[3]

Aniridia encompasses a group of congenital ocular disorders with iris hypoplasia in common. It may be inherited in an autosomal dominant or recessive pattern. There is also a sporadic form associated with Wilms' tumour. Aniridia is caused by mutations in the transcriptional regulator, the *PAX6* gene on chromosome 11p13.[4]

We report here the rare occurrence of both Rieger's syndrome and aniridia in a family and the molecular study of their *PAX6* status.

Case report

We examined a 39-year-old Chinese woman who had a phthisical right eye with only hand-movement visual acuity, and a left eye with visual acuity of 20/20. She had a history of total retinal detachment of her right eye after complicated combined trabeculectomy and cataract surgery 5 years earlier. Her left eye had open-angle glaucoma, controlled with four topical medications. On examining her left eye, there were peripheral iris strands attached to a posterior embryotoxon, iris stromal hypoplasia, corectopia and ectropion uveae (Fig. 1). The cup: to disc ratio of her left eye was 0.6, which was compatible with her visual field loss. On general examination she was noted to have a broad flat nasal bridge, maxilla hypoplasia, telecanthus, hypertelorism and microdontia, compatible with Rieger's syndrome. Her husband had normal vision and no ocular abnormalities were detected.

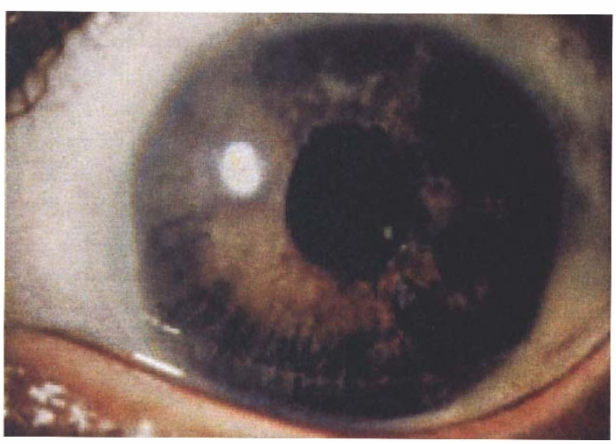

Fig. 1. *Slit-lamp photograph of the patient's left eye, showing typical Rieger's features including posterior embryotoxon, iris stromal hypoplasia, corectopia and ectropion uveae.*