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Sir,

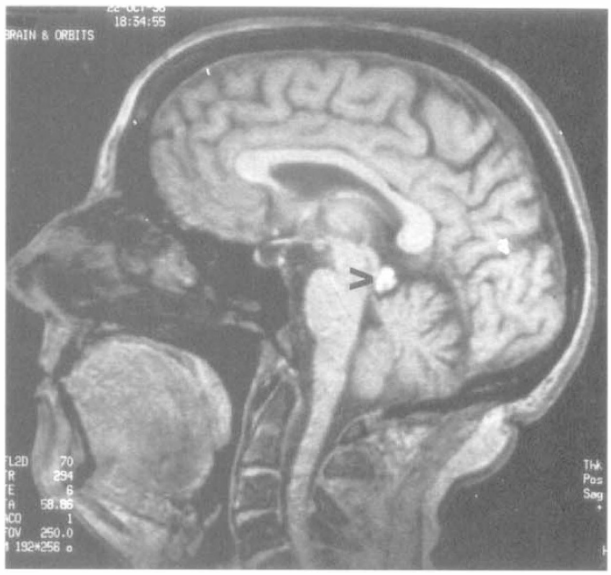
An unusual cause of intermittent vertical diplopia

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.

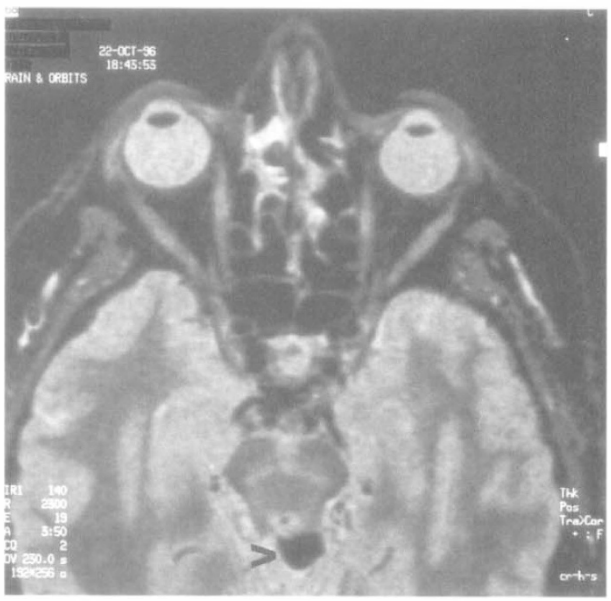
Case report

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.*

Magnetic resonance imaging (MRI) of the patient's brain demonstrated a small lesion, hyperintense on T1-weighted images, overlying the quadrigeminal plate (Fig. 1). This was felt to be consistent with a congenital lipoma. No other abnormality was seen.

In view of the fact that the lesion was located over the site at which the fourth nerve exits, and that there was no other explanation for a 15-year history of intermittent vertical diplopia, it was felt that the probable cause of the exertional diplopia was fourth nerve paresis related to the lipoma. No active treatment was instigated, though the patient was advised to avoid strenuous exercise in the future.

Comment

Many conditions are well recognised to give rise to intermittent symptoms of binocular diplopia. The most obvious of these is myasthenia gravis, whose symptomatic hallmark is variability/fatiguability. Other diseases produce worsening of symptoms in certain situations, such as Uhthoff's phenomenon in multiple sclerosis, or the exacerbation of Graves' ophthalmopathy that occurs secondary to venous congestion first thing in the morning. Likewise, decompensating strabismus can result in intermittent diplopia, as can superior oblique myokymia or some drugs (e.g. anticonvulsants). Rare oculomotor causes include convergence spasm (spasm of the near reflex), convergence-retraction nystagmus and ocular neuromyotonia. The visual phenomenon of hemifield slip resulting from chiasmal lesions has also been reported to cause diagnostic confusion.[1]

Our patient had had identical symptoms for 15 years with no other neurological or ophthalmological symptoms or signs. There were no features of congenital strabismus (NB: normal vertical fusion range), and no clinical features of myasthenia gravis, Graves' ophthalmopathy or any of the rare causes of binocular diplopia listed above.

The lipoma demonstrated by MRI scanning lay over the quadrigeminal plate, at the point where the fourth nerve exits the brain. While such lipomas have been well described,[2,3] they have not been associated with this particular symptom complex. Nevertheless, it is quite conceivable that under conditions of exertion, the change in cerebrospinal fluid dynamics may have resulted in a slight distortion of the intracranial anatomy, causing transient compression of the fourth nerve, which was then relieved by rest (and the resultant return of intracranial contents to normal).

In the absence of any alternative explanation for this patient's symptoms, we believe the lipoma to have been responsible, and would therefore suggest that structural lesions affecting the tectal plate should be included in the differential diagnosis of intermittent vertical diplopia.

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Sir,

Spontaneous resolution of iris and cutaneous haemangiomata in diffuse neonatal haemangiomatosis

Diffuse neonatal haemangiomatosis (DNH) is a rare disorder, characterised by the presence of multiple cutaneous haemangiomata and associated visceral haemangiomata involving predominantly the liver, but also the central nervous system, gastrointestinal tract and lungs.[1,2] When diffuse, multiple, cutaneous haemangiomata occur without involvement of visceral organs, a good prognosis is the rule (benign neonatal haemangiomatosis[3]). In DNH, however, the outcome is predictably poor if untreated, with mortality and morbidity related to complications associated with visceral involvement. Intracerebral and gastrointestinal haemorrhage or, more usually, high-output cardiac failure as a result of arteriovenous shunting in the liver, often prove fatal within the first few months of life.[1,2] Abnormal ocular findings are common,[4] and although they only rarely lead to ocular complications, ophthalmologists should be aware of the systemic associations. We report a surviving, untreated case of DNH with ocular haemangiomata.

Case report

A female, Caucasian infant weighing 3460 g was born at term. She was a first child and there was no family history of haemangiomata or consanguinity. The child was noted to have multiple, red cutaneous lesions, which blanched on pressure (Fig. 1), and a diagnosis of multiple cutaneous haemangiomata was made. Haematological investigation revealed a normal full blood count and coagulation screen (including platelets).

The cutaneous capillary haemangiomata, including lesions on the lips and gums, subsequently enlarged and became more numerous. The child was noted to have enlarging vascular lesions of both irides and an ophthalmic opinion was sought. Ophthalmic