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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R.W.D Bell[1]
G.G. McIlwaine[1,2]
C.J. Lueck[2]
[1]The Princess Alexandra Eye Pavilion
Royal Infirmary of Edinburgh
Edinburgh EH3 9HA, UK
[2]Western General Hospital
Edinburgh EH4 2XU, UK

Dr R.W.D. Bell ✉
The Princess Alexandra Eye Pavilion
Royal Infirmary of Edinburgh
Chalmers Street
Edinburgh EH3 9HA, UK

Tel: +44 (0)131 5361000
Fax: +44 (0)131 5363897

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

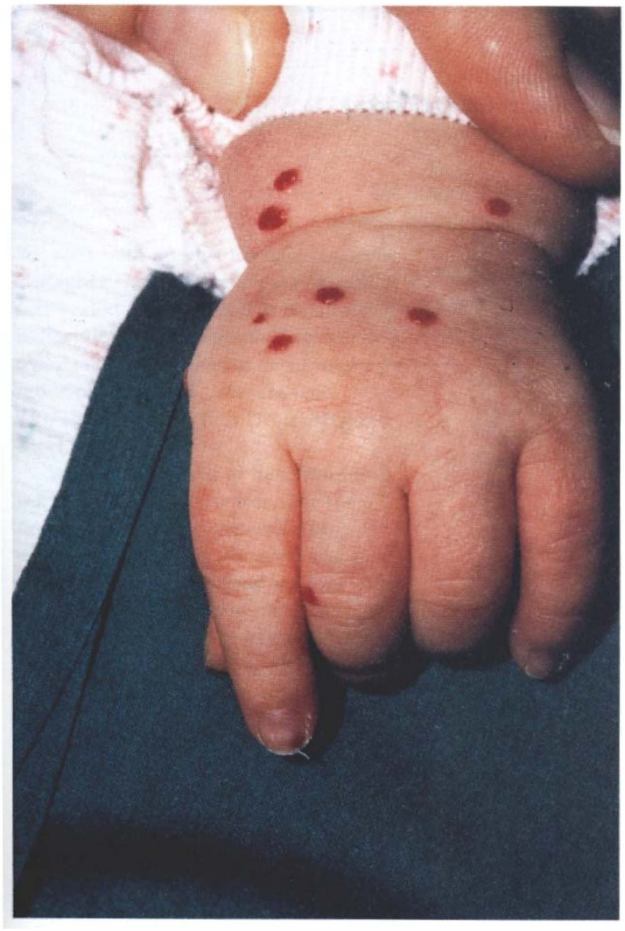

Fig. 1. *Multiple cutaneous haemangiomata.*

examination at 12 days of age found haemangiomata on the eyelid margins and, in addition, a single haemangioma on the right iris, and two on the left (Fig. 2). Ocular examination was otherwise unremarkable; in particular there were no conjunctival or retinal vascular malformations.

Further investigation by computed tomography, echocardiography and abdominal ultrasound, revealed multiple liver haemangiomata, the largest measuring 3 cm, but none in the lungs, brain, or heart. The child remained well with normal platelet counts, blood biochemistry and no evidence of cardiac failure. In view of this, and the possible side effects of systemic treatment, it was decided not to treat actively using either corticosteroids or interferon alpha.

The child was reviewed on a number of occasions and remained well. The visible haemangiomata began spontaneously to reduce in size at 5 months. At 10 months, repeat liver ultrasonography showed a solitary, much smaller hepatic lesion 1.6 cm in diameter, and it is presumed that the others have involuted. At 2 years 6 months the child remains well, the cutaneous haemangiomata are much smaller, with most having resolved completely, and the iris lesions have disappeared.

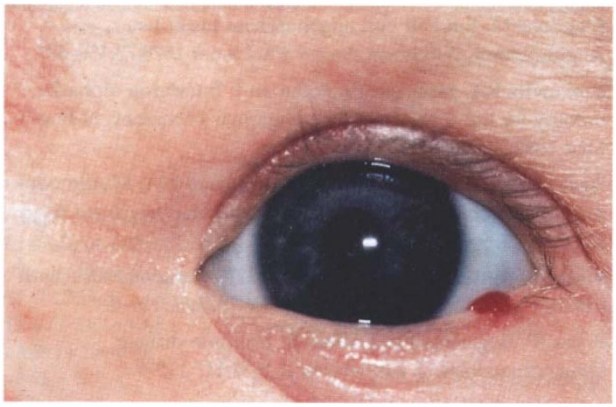

Fig. 2. *Iris and lid haemangiomata of the left eye.*

Comment

DNH was first described in the German literature in 1878.[5] Untreated, the condition was almost universally fatal and, despite the advent of systemic prednisolone, the mortality rate remained high.[1,2] Subsequently, the effects of interferon alpha-2a on immature vascular tissue have been exploited in the treatment of life-threatening childhood haemangiomata including DNH, and substantially improved the prognosis in many.[6] Interferon alpha-2b has recently been used successfully in combination with therapeutic hepatic embolisation and systemic corticosteroids.[7]

Ocular involvement may include iris, conjunctival and chorioretinal haemangiomata and is noted in between 32% and 50% of both surviving and fatal cases reported. Weiss and Ernest[8] reported a case associated with development of glaucoma requiring treatment with cyclocryotherapy. Our case is similar to that described by Fryns *et al.*,[9] where involution occurred without any treatment.

In the majority of cases of DNH, liver lesions predominate and result in the majority of morbidity. Why some cases survive and others do not is uncertain but may be related to the number, size and location of visceral lesions, such that if initially the child survives treated or untreated without major complications, the natural history of the haemangiomatous lesions favours resolution.

In conclusion, clinical evaluation of infants with multiple cutaneous haemangiomata should determine the presence and extent of visceral involvement, and in view of the apparent improvement in prognosis using corticosteroids and interferon, an informed choice may be made regarding treatment.

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I.G. Bryce
V. Pai
J.A. Bradbury
Department of Ophthalmology
Bradford Royal Infirmary
Bradford, UK

J.A. Bradbury, FRCOphth ✉
Bradford Royal Infirmary
Duckworth Lane
Bradford BD9 6RJ, UK
Tel: +44 (0)1274 364806
Fax: +44 (0)1274 364895