Orbital plexiform neurofibroma

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Plexiform neurofibromas are peripheral nerve sheath tumours considered to be pathognomonic of von Recklinghausen's disease (neurofibromatosis type I). It is an autosomal dominant condition with almost 100% penetrance but with variable expression of the phenotype. Nodular and plexiform neurofibromas may be found at any anatomical site, café au lait spots are seen on the skin, and Lisch nodules on the iris. Signs of the syndrome are usually present within the first year of life.

We report the case of a girl with an orbital plexiform neurofibroma with no other signs of neurofibromatosis.

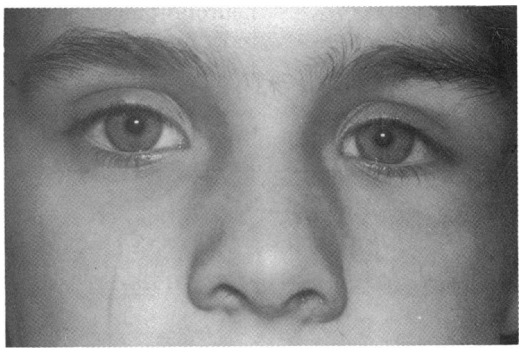

Figure 1 Smooth swelling at the superotemporal margin of the left orbit.

Case report

An 8-year-old girl presented with a swelling on the temporal aspect of the left orbital margin. It had been present for 1 year as a bluish lump which had fluctuated in size but was not associated with diplopia, proptosis, or reduced visual acuity (Fig 1). She was otherwise well and there was no family history of neurofibromatosis. On examination there was a firm diffuse nodular mass at the superolateral orbital margin. On funduscopy, there were no retinal folds and the optic discs were normal. The computed tomography scan showed a mass extending from the orbital margin into the temporal fossa (Fig 2). The swelling became painful during the following year so an excision biopsy was performed. The lesion was located subcutaneously extending deep to temporalis fascia. It was debulked but complete clearance of the inferonasal part of the lesion was not possible because there was no capsule and it was not limited by tissue planes. Histopathological examination showed a plexiform neurofibroma (a benign tumour).

Her postoperative recovery was marked by a subcutaneous haematoma at the operation site 1 month after the original surgery; this was drained under general anaesthesia. During the following year there was a recurrent swelling extending into the left eyelid causing a ptosis. Further surgery was performed to excise the diffusely spreading vascular lesion, histology was as before. There has been no recurrence after 1 year of follow up.

Comment

This girl had a solitary plexiform neurofibroma with no other signs of von Recklinghausen's neurofibromatosis. Many cases occur as a sporadic mutation which could explain the lack of family history.

There are two histological types of primary neurofibroma: either the 'isolated' nodular tumours which compress surrounding tissues and have histological features of perineurial cells or 'plexiform' neurofibromas which are diffuse, infiltrating, and non-encapsulated with histological features of proliferating Schwann cells.

Orbital plexiform neurofibromas may be massive, extending intracranially where they may be associated with arachnoid cysts and intracranial tumours.[1] In this case there was no such extension although the tumour infiltrated through the temporalis fascia and across the eyelid, through the levator palpebrae superioris threatening normal lid movement and causing pain.

There have been previous reports of patients with plexiform neurofibromas at other anatomical sites without any other evidence of neurofibromatosis; however, orbital neurofibromas which are uncommon (three in a previous series of 214 children with orbital tumours) are reported in association with von Recklinghausen's disease.[2]

The presence of abnormal feeding vessels[3] has been demonstrated by angiography and there may be two types of plexiform neurofibroma: one with normal vasculature and the other with

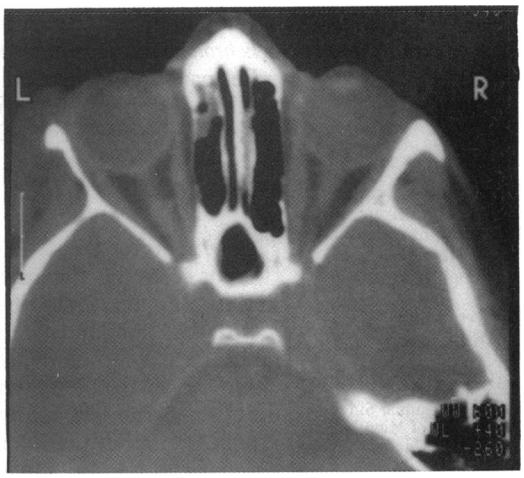

Figure 2 Axial computed tomography scan shows a soft tissue lesion at the left orbital margin, extending into the temporal fossa without any bony erosion or extension into the orbit.

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abnormal vessels which form an integral part of the lesion forming a 'haemangioneurofibroma'. Such pathology could explain the profuse bleeding encountered when excising this lesion.

Malignant conversion to a sarcoma has been described in plexiform neurofibromas but these have been in children with the other signs of neurofibromatosis.[4] Our patient will be reviewed indefinitely because the neurofibroma may recur, although the full syndrome of von Recklinghausen's neurofibromatosis is unlikely to develop at this age.

1 Boltshauser B, Stocker H, Sailer H, Valavanis A. Intracranial abnormalities associated with facial plexiform neurofibromas in neurofibromatosis type I. *Neurofibromatosis* 1989; 2: 274–7.
2 Porterfield JF. Orbital tumours in children: a report on 214 cases. *Int Ophthalmol Clin* 1962; 2: 319–35.
3 Littlewood AH, Stilwell JH. The vascular features of plexiform neurofibroma with some observations on the importance of pre-operative angiography and the value of pre-operative intra-arterial embolisation. *Br J Plast Surg* 1983; 36: 501–6.
4 Chaudhuri B, Ronan SG, Manaligod JR. Angiosarcoma arising in a plexiform neurofibroma: a case report. *Cancer* 1980; 46: 605–10.

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Thelaziasis: report of two cases

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We report here two cases of thelaziasis, a rare parasitic infestation of the eye in humans caused by a spiruroid nematode of the genus *Thelazia*. Although the total number of cases in the world is not available 28 cases, 17 from Japan, seven from China, two from Korea, and one each from Russia and India, have been reported.[1,2] The complete life cycle of the nematode is not yet fully understood; however, a snail is considered to be a probable intermediate host.[3] Burnett *et al* found a species of *Fania* to be a naturally and experimentally susceptible intermediate host for the larval development of *Thelazia californiensis*.[4] A case of unilateral conjunctivitis caused by the same species was described by Knierin and Jack and it was thought that a fly or gnat was the possible mode of transmission in the Siskiou mountains of North California.[5] Generally, the clinical manifestations of human thelaziasis comprised conjunctival congestion, pain, excessive lacrimation, and irritation. In Australia it is called 'bung' or 'blue eye' because of congestion and chemosis; there it is caused by *Filaria hebronema*, another species of *Thelazia*.[6] Permanent scarification and fibrous opacities of the conjunctiva and cornea may develop due to the presence and repeated movement of the worms.

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Case reports

Two cases of thelaziasis have been seen in Manipur: an 18-month-old boy who presented with the complaints of excessive tears, irritation, and redness of the left eye of 6 days' duration; and a 3-year-old boy who suffered from a blunt injury to the left eye and presented with mild pain, irritation, and conjunctival congestion of 7 days' duration. On casual examination, the parents of both children had noticed thread-like moving worms on the affected eye. Clinical findings were similar and comprised conjunctival congestion, local oedema, and excessive lacrimation. No gross ulceration and follicles were seen; instead, there were whitish worms seen moving in the conjunctiva across the cornea. Routine examination of blood, urine, and stool revealed no abnormal findings. Two adult worms from each patient were removed from the conjunctival sac after instillation of 4% lignocaine hydrochloride. The worms were later identified as adult *Thelazia callipaeda* on the basis of the hexagonal mouth, absence of alae and cuticular ornamentations except for conspicuous transverse striations near the anterior end which are considered to aid movement across the smooth surface of the cornea. The infection in both cases might have occurred from close association with infected dogs or through house flies, *Musca domestica*.

Comment

Diagnosis depended on the recognition of creamy white worms coiled in the conjunctival sac or migratory worms over the cornea. Removal of the worms and morphological study under a dissecting microscope were needed for species identification. Complete removal of the worms with eye forceps provides a cure as no drug is yet known to be effective against the nematode. Because of the rarity of reports on human thelaziasis clinicians, apparently unaware of the condition in which spiruroid nematode infestation can give rise to such a clinical manifestation, might think of other causes of the conjunctivitis. Thorough and careful examination of the affected eye, bearing in mind the possibility of such a condition, would justify the recognition of more cases and the right treatment.