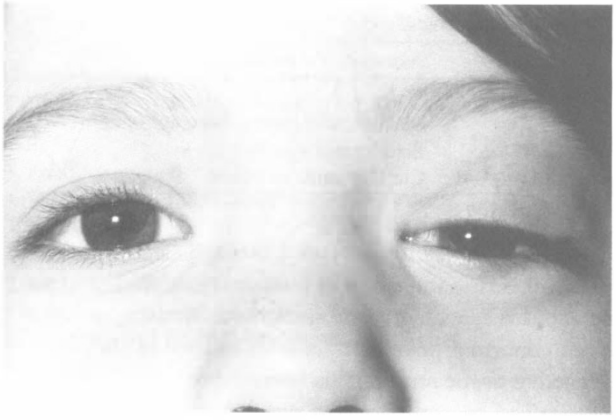

Fig. 2. *The appearance after final treatment.*

Childhood Cancer Study Group (UKCCSG) paediatric pathology panel. There was no evidence of systemic spread.

Treatment comprised sequenced chemotherapy with vincristine, actinomycin D and ifosfamide (IVA) (SIOP/UKCCSG MMT 95 protocol, strategy 953, arm A). She responded until a local recurrence 8 months later, treated with MMT 5 relapse protocol (multiagent chemotherapy including carboplatin, epirubicin, etoposide) and radiotherapy. She has a residual ptosis (Fig. 2) and upper conjunctival fornix redness due to shortening, with no mass palpable, marked mucous keratopathy and retained visual acuity of 20/30.

Comment

We believe this to be the first report of alveolar-type orbital rhabdomyosarcoma associated with NF1. There are 3 previously reported cases of orbital rhabdomyosarcomas in series of NF1 patients, but they were of embryonal or pleomorphic histological type or the histology was not stated, and neither series discussed the ophthalmic presentation.[1,2]

Orbital rhabdomyosarcomas constitute 10% of childhood rhabdomyosarcomas. As with all paediatric malignancies, treatment is according to national/international protocols supervised by regional paediatric oncologists. This girl was treated by the UKCCSG version of the malignant mesenchymal tumour of childhood procotol – MMT 95 study.

Chemotherapy is the primary treatment but radiotherapy is indicated if there is residual tumour after chemotherapy. Long-term sequelae of radiotherapy include visual loss, dry eye, facial asymmetry, and endocrine/growth problems if the hypothalamus or pituitary is involved in the field of radiation.

Prognosis and treatment of alveolar rhabdomyosarcoma differ substantially from that of embryonal rhabdomyosarcoma; it is therefore imperative that cases are discussed before surgery with the regional paediatric oncology team. Histological differentiation between the two types on formalin-fixed tissues is often difficult so increasing reliance is placed on finding 2;13 chromosome translocation (or variants characteristic of alveolar rhabdomyosarcoma).[3–5] These molecular studies require fresh tissue rapidly processed, best done in the regional centre.

References

1. McKeen EA, Bodurtha J, Meadows AT, Douglass EC, Mulvihill JJ. Rhabdomyosarcoma complicating multiple neurofibomatosis. J Pediatr 1978;93:992–3.
2. Mosso ML, Castello M, Bellani FF, *et al.* Neurofibromatosis and malignant childhood cancers: a survey in Italy, 1970–83. Tumori 1987;73:209–12.
3. Kullendorf CM, Donner M, Mertens F, Mandahl N. Chromosomal abnormalities in a consecutive series of childhood rhabdomyosarcoma. Med Pediatr Oncol 1998;30:156–9.
4. Biegel JA, Nycum LM, Valentine V, Barr FG, Shapiro DN. Detection of the t(2;13)(q35;q14) and PAX3-FKHR fusion in alveolar rhabdomyosarcoma by fluorescence *in situ* hybridization. Genes Chromosomes Cancer 1995;12:186–92.
5. Reichmuth C, *et al.* The diagnostic potential of the chromosome translocation (t2;13) in rhabdomyosarcoma: a PCR study of fresh-frozen and paraffin-embedded tumour samples. J Pathol 1996;180:50–7.

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Sir,

Ankyloblepharon filiforme adnatum

Ankyloblepharon is a full-thickness fusion of the eyelids at the lateral canthus (external ankyloblepharon) or the medial canthus (internal ankyloblepharon) producing shortening of the palpebral fissure. Ankyloblepharon filiforme adnatum is a benign congenital anomaly in which single or multiple strands of fine connective tissue join the upper and lower eyelids.

Case report

A 4480 g, white male boy was born at 40 weeks gestation to a 21-year-old third gravida. The pregnancy was normal and the mother denied any drugs, alcohol or smoking. The previous two siblings, aged 4 and 2 years, both males, were healthy. There was no family history of congenital anomalies or consanguinity. At birth the baby

was noted to have fused eyelids of the right eye in addition to bilateral pigmented hydroceles. A detailed paediatric assessment failed to reveal any other congenital abnormalities. There was a single band of tissue connecting the right upper and lower eyelids (Fig. 1), which was not adherent to the underlying eyeball. The band was cut by a no. 15 knife with no bleeding. Ophthalmic examination revealed normal anterior and posterior segments. The left eye was normal. The patient has a follow-up appointment in 6 months time.

Comment

Ankyloblepharon filiforme adnatum was first described by Von Hasner[1] in 1881. It is a rare benign congenital anomaly in which single or multiple strands of fine connective tissue join the upper and lower eyelids anywhere along the lid margin but never at the lateral or medial canthus.[2] These strands arise from the grey line[3] (anterior to meibomian gland orifices and posterior to the cilia) and join the upper and lower lids. The strands are extensile and by forcibly opening the lids their length can be almost doubled.[4] Ankyloblepharon filiforme adnatum usually occurs as a bilateral condition, but can be unilateral.[5] It may be present as an isolated sporadic anomaly, but has primarily been reported in association with cleft lip and cleft palate.[6] It can also be part of a well-defined syndrome: Hay–Wells syndrome,[7] also known as ankyloblepharon – ectodermal dysplasia – clefting syndrome; popliteal – pterygium syndrome[8] characterised by intercrural webbing of fingers; Curly hair – anklyoblepharon – nail dysplasia syndrome (CHANDS);[9] and Edwards syndrome[10] (trisomy 18). Other associated anomalies reported are hydrocephalus, meningomyelocele and imperforate anus,[11] patent ductus arteriorus,[12] ventricular septal defect[13] and bilateral syndactyly.[14] The annual incidence reported is 4.4 per 100 000 births.[15]

The eye itself has been reported to be normal except in one case report by Scott *et al.*[16] where it was found to be associated with infantile glaucoma and iridogoniodysgenesis. The aetiology of this condition is not known but a number of theories have been put forward. The currently accepted theory is that this condition is due interplay of temporary epithelial arrest and rapid mesenchymal proliferation, allowing union of lids at abnormal positions.[17] The histology of the connective tissue strands has been shown to consist of a vascularised central core surrounded by stratified squamous epithelium.[18]

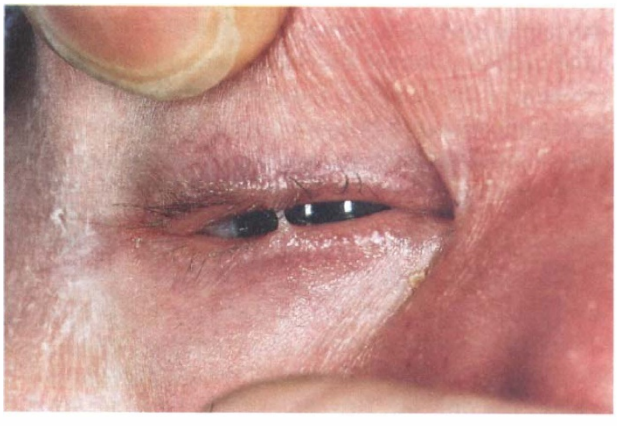

Fig. 1. *Ankyloblepharon filiforme adnatum of the right eye showing a single band of tissue connecting the upper and lower eyelids.*

Table 1. *Classification of ankyloblepharon filiforme adnatum*

Group	Associated abnormalities
I	None
II	Cardiac and central nervous system
III	Ectodermal syndromes
IV	Cleft lip and/or palate

In 1980 Rosenman *et al.*[6] divided ankyloblepharon filiforme adnatum into four subgroups. Subgroups I and II were sporadic, and III and IV were autosomal dominant with variable expressivity (Table 1).

The bands frequently resolve spontaneously after a few months, or if obstructing the visual axis they may be released by a simple operative procedure.

The practical importance of this anomaly is that when it occurs it should alert the clinician to the possible presence of other congenital problems.

References

1. Von Hasner. Ankyloblepharon filiforme adnatum. Z Heilkol 1881;2:429.
2. Duke-Elder S. System of ophthalmology, vol. 3. London: Kimpton, 1964:869–71.
3. Khanna VN. Ankyloblepharon filiforme adnatum. Am J Ophthalmol 1957;43:774–7.
4. Weill S. Arch Aukenheilkd 1908;59:278. Cited by Duke-Elder S. System of ophthalmology, vol 3. London: Kimpton, 1964:869.
5. Kapoor MS, Sood GC, Aurora AL, Kapoor S. Ankyloblepharon filiforme adnatum. Indian J Ophthalmol 1977;25:43–4.
6. Rosenman Y, Ronen S, Eidelman AL, Schimmel MS. Ankyloblepharon filiforme adnatum. Am J Dis Child 1980;134:751–3.
7. Schwayder TA, Lane AT, Miller ME. Hay–Wells syndrome. Pediatr Dermatol 1986;3:399–402.
8. Kind HP. Popliteal pterygium syndrome. Helv Paediatr Acta (Basel) 1970;25:508–15.
9. Baughman FA. CHANDS: the curly hair – ankyloblepharon – nail dysplasia syndrome. Birth Defects 1971;7:100–2.
10. Clark DI, Patterson A. Ankyloblepharon filiforme adnatum in trisomy 18 (Edwards syndrome). Br J Ophthalmol 1985;69:471–3.
11. Kazarian EL, Goldstein P. Ankyloblepharon filiforme adnatum with hydrocephalus, meningomyelocele and imperforate anus. Am J Ophthalmol 1977;84:355–7.
12. Lobestein A, Haarscher A. Anklyoblepharon filiforme congenital et malformation cardiaque associée. Bull Soc Ophtalmol Fr 1953;2:128–31.
13. Rogers JW. Anklyoblepharon filiforme adnatum. Arch Ophthalmol 1961;65:114–7.
14. Akkermans C, Stern LM. Anklyoblepharon filiforme adnatum. Br J Ophthalmol 1979;63:129–31.
15. Jain S, Atkinson AJ, Hopkinson B. Ankyloblepharon filiforme adnatum. Br J Ophthalmol 1997;81:708.
16. Scott MH, Richard JM, Farris BK. Anklyoblepharon filiforme adnatum associated with infantile glaucoma and iridogoniodysgenesis. J Pediatr Ophthalmol Strabismus 1994;31:93–5.

17. Judge H, Mott W, Gabriels J. Ankyloblepharon filiforme adnatum. Arch Ophthalmol 1929;2:702–8.
18. Wintersteiner H. Ein neurer Fall von Ankyloblepharon filiforme. Arch Augenheilkd 1908;59:196.

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Sir,

Epstein–Barr virus dacryoadenitis as a complication of bone marrow transplant in a child with combined immunodeficiency

Dacryoadenitis may result from bacterial or viral infection or granulomatous disease such as sarcoidosis and may affect up to 1 in 300 people with acute Epstein–Barr virus (EBV) infection.[1] The clinical course in the immunocompetent patient is usually mild and self-limiting.

We present a case of severe acute bilateral dacryoadenitis in a child who had recently undergone a bone marrow transplant. Concurrent EBV infection was demonstrated in blood and tears by polymerase chain reaction (PCR).

Case report

An 8-year-old girl presented with a 1 week history of pain and redness around both eyes. Ocular examination revealed reduced aqueous tear secretion and mild conjunctival injection with no follicular or papillary changes. Visual acuities, anterior and posterior segments were all normal.

Two months previously she had undergone a 1DQ mismatched, T-cell replete, unrelated donor bone marrow transplant (using low-intensity conditioning[2]) to treat combined immunodeficiency (with an autoimmune component which included poliosis). Her post-transplant course was not smooth and she remained neutro- and lymphopenic. She had also developed anorexia and a mild skin rash suggestive of mild graft-versus-host disease (GVHD). Her ocular findings were thought to be consistent with GVHD. She was commenced on tear film supplements and mild topical steroid drops.

Two days later she had increased pain and upper lid oedema. Examination showed bilateral palpable tender swollen lacrimal glands but no pre-auricular or submandibular lymphadenopathy (Fig. 1). There were bilateral temporal small perilimbal subconjunctival haemorrhages with mild punctate epithelial keratitis and minimal conjunctivitis. Samples taken over the previous 2 weeks for plasma PCR detected a rapid rise in EBV viral load from none to 165 000 copies/ml. She was commenced on foscarnet and the anti-CD20 monoclonal

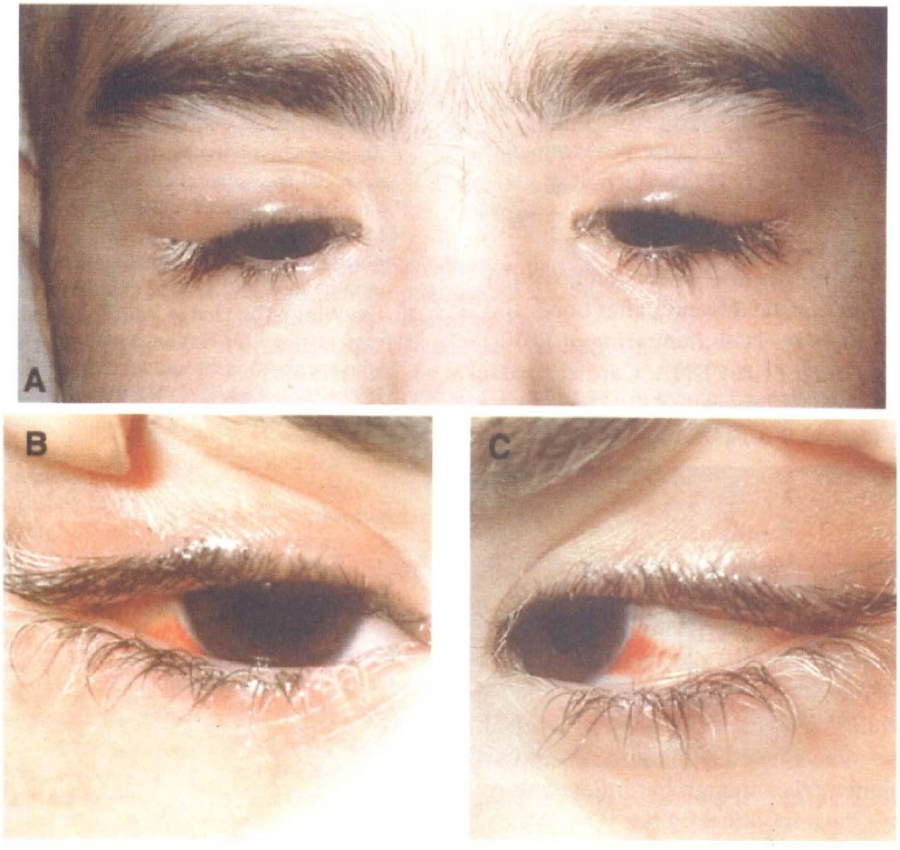

Fig. 1. *(A) Face showing bilateral periorbital oedema, especially in the region of the lacrimal glands. (B) Right eye showing temporal subconjunctival haemorrhage. (C) Left eye showing temporal subconjunctival haemorrhage.*