thinning in both eyes with bilateral vascularised corneal grafts and peripheral thinning (Fig. 1). The left pupil was irregular and the eye was aphakic. A conjunctival bleb was present superiorly (Fig. 2). The anterior chamber was formed and there was no evidence of leakage of the aqueous (Seidl's negative). A limited fundus examination did not show any evidence of retinal detachment or choroidal effusion in the left eye. The intraocular pressure was 10 mmHg in the right eye and 3 mmHg in the left. A diagnosis of spontaneous perforation of the globe under the conjunctival bleb was made. In view of the underlying pathology no surgical intervention was planned and the patient was started on prednisolone 1% eye drops every 2 h together with atropine eye drops three times a day to his left eye. The eye remained stable over the next 2 days with no evidence of any retinal or choroidal detachment when the patient returned to his country of origin.

Subsequent follow-up locally revealed that his intraocular pressure had gradually built up to 13 mmHg over a period of 3 weeks with improvement in his vision to pre-incident levels.

Comment

Ehlers Danlos syndrome is a collection of various connective tissue disorders which have a genetically determined abnormality of collagen. It is associated with skin laxity, hyperextensibility and hypermobility of joints, short stature, skin bruising and visceral abnormalities.¹⁻⁷ There are ten different subtypes depending on the type of collagen deficiencies, with type VI having ocular associations.^{8,9} Thin sclera, keratoconus, keratoglobus, lens subluxation, myopia, and ocular fragility to minor trauma are the usual ocular manifestations.¹⁰ Other ocular associations include situs inversus of the disc,¹¹ angioid streaks,¹² traumatic scleromalacia¹³ and retinal detachment.¹⁴ Perforation of the globe following minor ocular trauma is a known association of Ehlers Danlos syndrome;^{14,15} however, spontaneous perforation of the eyeball as in the present report is exceedingly rare.¹

Knowledge of the underlying pathology, extensive thinning of the sclera and the diffuse conjunctival bleb with absence of choroidal detachment made conservative treatment our choice. Improvement in intraocular pressure without any surgical intervention suggests spontaneous sealing of the leakage, an observation that to the best of our knowledge has not been reported before in literature.

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P. Puri
M. Gupta
J. Chan
Department of Ophthalmology
Royal Hallamshire Hospital
Sheffield, UK
Mr P. Puri ✉
Department of Ophthalmology
Royal Hallamshire Hospital
Glossop Road
Sheffield S10 2JF, UK

Sir,

Orbital cellulitis following corneal gluing under sub-Tenon's local anaesthesia

Orbital cellulitis following ophthalmic surgery is very rare, but has occurred after strabismus surgery, blepharoplasty or retinal surgery.¹ Other sources for orbital infection include post-operative endophthalmitis or coincidental sinusitis,^{2,3} and it has been reported following surgery with retrobulbar or peribulbar local anaesthesia.⁴

We present a case of orbital cellulitis due to group A, beta-haemolytic *Streptococcus*, the virulent causative organism for necrotising fasciitis, arising after sub-Tenon's local anaesthesia in an immunocompromised patient.

Case report

An 81-year-old man with a 60 year history of rheumatoid arthritis, controlled on methotrexate and non-steroidal anti-inflammatory drugs, presented with a sudden onset of watering and reduced visual acuity of his

Fig. 1. CT showing proptosis, scleral thickening and diffuse opacity within left orbital fat.

pseudophakic left eye. An infero-nasal peripheral corneal perforation, due to a rheumatoid corneal melt, was apparent and there was a slow leak of aqueous humour and iris plugging of the perforation. A therapeutic soft contact lens was fitted and topical ofloxacin and dexamethasone administered, together with high-dose oral prednisolone.

Two days later, corneal gluing was performed because of persistent aqueous leak. After preparation of the skin and conjunctival sac with aqueous povidone-iodine, a sub-Tenon's local anaesthetic was administered through a small conjunctival incision in the infero-nasal fornix. Cyanoacrylate glue was applied to the corneal perforation, a therapeutic contact lens was inserted and the topical medications continued.

Proptosis developed rapidly during the third post-operative night, with reduced ocular movements, a left relative afferent pupillary defect and mild purulent discharge from the site of the sub-Tenon's infiltration. The contact lens and glue remained in place and the ocular media remained clear. B-mode ultrasonography revealed scleral thickening, with fluid in the sub-Tenon's space, and CT demonstrated scleral thickening, proptosis and patchy opacity within the left orbital fat (Fig. 1).

Fig. 2. Left proptosis due to severe orbital cellulitis after sub-Tenon's anaesthesia, with purulent discharge from the injection site, ischaemic sclera and an infero-nasal corneal melting.

Although afebrile, the patient had a neutrophil leucocytosis and he was commenced on high-dose systemic cefuroxime and metronidazole.

The patient's condition had deteriorated by the following day, with pyrexia, increased proptosis, an absence of ocular ductions and an area of ischaemic temporal sclera (Fig. 2). As large numbers of group A, beta-haemolytic streptococci had been isolated from earlier conjunctival swabs, the treatment was changed to very high dose intravenous penicillin G (2.4 g four times daily) with gradual improvement, apart from a persistent mild optic neuropathy.

Comment

Sinusitis is the most common cause of orbital cellulitis, accounting for about 70% of cases in adults,⁵ and about a quarter may be secondary to trauma, with entry of organisms through the skin or sinuses. Orbital cellulitis is very rare after ophthalmic surgery but, in this case, the sub-Tenon's anaesthetic was almost certainly the portal for infection: imaging showed normal sinuses but marked scleral thickening with fluid in the sub-Tenon's space; purulence was observed at the conjunctival incision; the operation was a minor corneal procedure, with no subsequent endophthalmitis.

Sub-Tenon's infiltration using a blunt cannula has been advocated as a safe and effective technique for administration of ocular local anaesthesia;⁶ compared with needle injection techniques, however, it involves a significant breach of the conjunctiva, with an open portal for entry of pathogenic organisms.

Group A streptococci are the putative cause for the very serious necrotising fasciitis.^{7,8} Necrotising fasciitis may be associated with very minor superficial trauma,⁷ and occurs more commonly where there is concurrent medical disease, such as diabetes and alcoholism, or systemic immunosuppression – as in this case.⁹ This patient's conjunctival flora may also have been altered by the presence of other local factors such as topical antibiotic, the therapeutic contact lens and the cyanoacrylate glue. Progression of group A *Streptococcus* infection may be very rapid and prompt treatment with high-dose penicillin is essential,⁷ but surgical debridement may be required if necrosis progresses.

The risk of orbital cellulitis after sub-Tenon's anaesthesia is small, but significant in view of the potentially severe consequences. Prophylaxis (such as povidone-iodine preparation of the skin and conjunctiva) should therefore be considered and, given the rapid onset of anaesthesia after administration, sub-Tenon's anaesthesia could be given after the sterile preparation and immediately before the start of surgery.

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Brian Redmill
Charles Sandy
Geoffrey E. Rose
Orbital Service
Moorfields Eye Hospital
London EC1V 2PD, UK

Mr B. Redmill ✉
Department of Ophthalmology
Lincoln County Hospital
Greetwell Road
Lincoln LN2 5QY, UK

Tel: +44 (0)1522 573949
e-mail: brianredmill@mac.com

Sir,

Keratoconus associated with a chromosome 7, 11 translocation

We present a case of keratoconus with a balanced chromosome translocation between chromosomes 7 and 11. The keratoconus is demonstrated with corneal topography. Chromosomal rearrangements should be considered in cases of keratoconus presenting in childhood.

Keratoconus is an ectatic corneal disorder with a prevalence of about 50 in 100 000.¹ The hereditary pattern of keratoconus is not predictable, and family histories have been reported in 6–8% of cases. In some families, the suggested mode of inheritance is autosomal dominant with variable penetrance and expression.² Keratoconus has been described with a number of systemic conditions, including Marfan's and Ehlers Danlos syndromes. Keratoconus is also recognised to be more frequent in two chromosomal conditions: Down's syndrome (trisomy 21) and Turner's syndrome.² Recently, an association with a chromosome 13 ring abnormality has been described.³

Case report

A 13-year-old girl with short stature was referred from her optician to the eye clinic with myopic astigmatism as she was found to have frequent changes in her refraction. She wore gas-permeable contact lenses.

Snellen visual acuity was 6/24 unaided right (6/9 with contact lens) and counting fingers left (6/9 with contact lens). Refraction was right +1.50/–5.00×80, left –9.00/–2.50×80. Examination of the left cornea showed it to be steep with central apical mid-stromal scars and Vogt's striae. The mid-stromal scars were due to an episode of left acute hydrops which settled on conservative treatment. The right cornea was normal in appearance. Corneal topography of the right eye demonstrated the astigmatism and inferior corneal steepening typical of keratoconus (Fig. 1).⁴ The left corneal topography showed over 17 dioptres of astigmatism and corneal steepening inferiorly.

The patient had been investigated at the age of 12 years for short stature. She was not dysmorphic and systemic examination was unremarkable. As part of the investigative process chromosome analysis was done. A blood sample revealed a balanced translocation between the long arms of chromosome 7 and 11. The karyotype was 46, XX, t(7,11)(q34;q13.3), inv (9)(p11q12). Chromosome analysis of the mother's blood showed a pericentric inversion of chromosome 7q22.1–q34, which was also present in the mother's sister; neither the mother nor the maternal aunt had keratoconus. The father had normal eyes: his chromosomes showed a pericentric inversion of chromosome 9.

A radiograph of the proband's wrist showed that bone age was 2–3 years behind actual age. All other investigations, including pelvic ultrasound and hormonal profile (total T₄, estradiol, luteinising hormone, follicle stimulating hormone, prolactin and progesterone), were normal. The short stature was attributed to delayed puberty, with menarche commencing at age 15 years, and the patient is now normal height and weight.

Comment

This girl with delayed puberty and keratoconus had no family history of keratoconus and two normal older female siblings. She has a *de novo* balanced translocation between the long arms of chromosome 7 and 11 as well as a familial pericentric inversion of chromosome 9. Both parents have pericentric inversions: the father of chromosome 9, as seen in the proband, and the mother of chromosome 7.

Pericentric inversion of chromosome 9 is a normal variant and is not associated with keratoconus. Such inversions are relatively common and are not known to predispose to *de novo* translocations in offspring. However, since the maternal inversion and translocation in the proband have a common breakpoint on