evaluated carefully if there is a member showing the characteristic clinical findings of the disease, especially in consanguineous marriages.

References

1. Urbach E, Wiethe C. Lipidosis cutis et mucosae. Virchows Archiv Pathol Anat Physiol Klin Med 1929;273:285–319.
2. Blodi FC, Whiney RD, Hendricks CA. Lipid-proteinosis (Urbach Wiethe) involving the lids. Trans Am Ophthalmol Soc 1960;58:155–66.
3. İrkeç M, Orhan M, Orhan D, Durgun B, Can Ç. Dry eye syndrome associated with Urbach-Wiethe disease. J Pediatr Ophthalmol Strabismus 1996;33:265–8.
4. Harper JI, Duance VC, Sims TJ, Light ND. Lipoid proteinosis: an inherited disorder of collagen metabolism? Br J Dermatol 1985;113:145–51.
5. Newton JA, Rasbridge S, Temple A, Pope FM, Black MM, Mckee P. Lipoid proteinosis: new immunopathological observations. Clin Exp Dermatol 1991;16:350–4.
6. Feiler-Ofry V, Lewy A, Regenbogen L, Hanau D, Bat-Miriam Katznelson M, Godel V. Lipoid proteinosis (Urbach-Wiethe syndrome). Br J Ophthalmol 1979;63:694–8.

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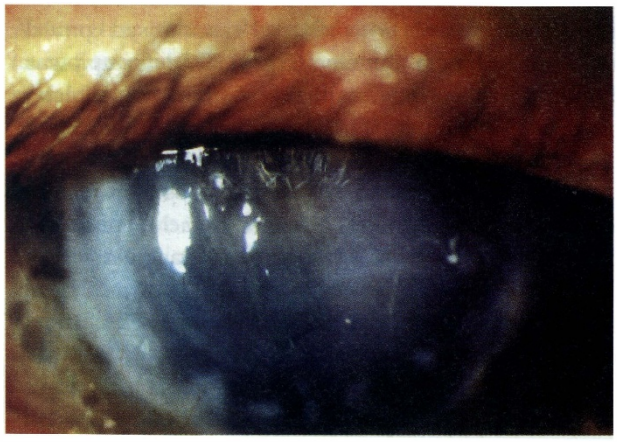
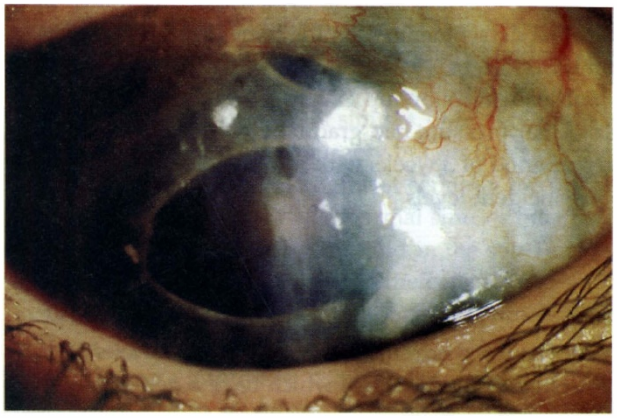


(a)



(b)

Fig. 1. *(a), (b) Anterior segment photographs showing bilateral thin vascularised cornea.*

Sir,

Spontaneous perforation of the globe in Ehlers Danlos syndrome

Ehlers Danlos syndrome is a congenital disease associated with abnormal collagen formation. We present a case of spontaneous rupture of the globe in this condition.

Case report

A 50-year-old Caucasian man with a history of Ehlers Danlos syndrome presented to eye casualty with a 1 day history of blurring of vision in his left eye and noticing that his left eyeball felt very soft while washing his face that morning. There was no history of trauma. His past ocular history included two penetrating keratoplasties in each eye for keratoconus and a right retinal detachment 17 years previously. The last graft performed in the left eye was 5 years ago. His ocular medications included 4% flourometholone eye drops twice a day to both his eyes and saline drops to the left eye.

Eye examination revealed a visual acuity of hand movements close to face in the right eye and counting finger 1/2 m in the left eye. There was marked scleral

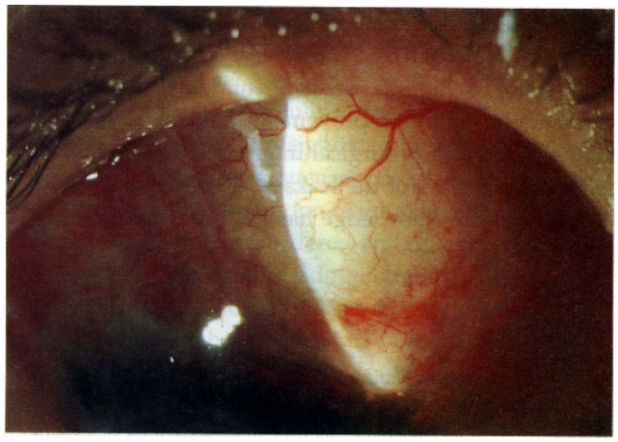


Fig. 2. *Anterior segment photograph showing a superior diffuse conjunctival bleb.*

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 scleromalacia[13] 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.

References

1. Cameron JA. Corneal abnormalities in Ehlers Danlos syndrome type VI. Cornea 1993;12:54–9.
2. Cordella M, Vinciguerra E. Ocular manifestations of Ehlers Danlos syndrome. Minerva Ophthalmol 1966;8:103–7.
3. Bitgood JJ, Whitley RD. Pop eyed: an inherited z-linked keratoglobus in the chicken. J Hered 1986;77:123–5.
4. Traboulsi E, Kattan H. Skeletal and connective tissue disorders with anterior segment manifestations. In: Krachmer JH, Mannis MJ, Holland EJ, eds. Cornea. St Louis: CV Mosby, 1997:925–42.
5. Lahoud S, Brownstein S, Laflamme MY, Poleski SA. Keratoconus with spontaneous perforation of the cornea. Can J Ophthalmol 1987;22:230–3.
6. Hyans SW, Dar H, Newmann E. Blue sclera and keratoglobus. Br J Ophthalmol 1969;53:54–7.
7. Nelson ME, Talbot JF. Keratoglobus in Rubenstein Taybi syndrome. Br J Ophthalmol 1989;73:385–7.
8. Byers PH. Ehlers Danlos syndrome: recent advances and current understanding of the clinical and genetic heterogeneity. J Invest Dermatol 1994;103:475–525.
9. May MA, Beauchamp GR. Collagen maturation defects in Ehlers Danlos keratopathy. J Pediatr Ophthalmol Strabismus 1987;24:78–82.
10. Newlin A, Sugar J. Corneal and external eye manifestations of systemic disease. In: Yanoff M, Duker JS, eds. Ophthalmology, vol 5. St Louis: CV Mosby, 1999:15.2.
11. Forman AR. Situs inversus of the disc in the Ehlers Danlos syndrome, type III. Ophthalmology 1979;86:844–6.
12. Green WR, Friedman Kien A, Banfield WG. Angioid streaks in Ehlers Danlos syndrome. Arch Ophthalmol 1966;76:197–204.
13. Parc C, Legeais JM, Renard G. Post-traumatic scleromalacia in an Ehlers Danlos syndrome. J Fr Ophtalmol 1998;21:761–3.
14. Bodanowitz S, Hesse L, Postgens H, Kroll P. Retinal detachment in Ehlers Danlos syndrome: treatment by pars plana vitrectomy. Ophthalmology 1997;94:634–7.
15. Syndrome of blue sclera and keratoglobus (ocular type of Ehlers Danlos syndrome). Graefes Arch Klin Exp Ophthalmol 1977;204:235–46.

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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