ANCA has been reported as being both sensitive and specific.[3] However, histological investigation, including that of the enucleated eye, did not show granulomas or vasculitis – only evidence of chronic inflammation. Limited presentations of Wegener's granulomatosis occur in which no other system is involved. ANCA testing has been felt to be very helpful in such cases in making the diagnosis.[4] In this case a positive pANCA was found, but not initially. A moderately raised vWF and ESR were found, indicating endothelial damage and suggesting a vasculitic process (Table I). ANCA titres have been shown to vary with disease state but not always in step with the disease.[5]

Positive ANCA serology is found in a spectrum of vasculitic diseases including polyarteritis nodosa, microscopic polyarteritis and Wegener's granulomatosis. Two main types are recognised: cANCA, which has a high positive predictive value for Wegener's granulomatosis; and pANCA, which is less specific but does suggest a vasculitis, as occurred in this case.[5] This case is interesting as it is an ANCA-positive anterior segment vasculitis which does not appear to be Wegener's granulomatosis. The term Wegener's vasculitis has been used in pulmonary conditions in which granulomas are not found.[5] Perhaps this case could be labelled as limited Wegener's vasculitis. In such cases serial serological titres may help elucidate the diagnosis.

'Pulsed' immunosuppression has been shown to be effective in treating anterior segment disease.[6] A confident diagnosis of the underlying condition is preferred before the initiation of long-term immunosuppressive therapy but, as our case demonstrates, treatment may need to be started in its absence.[2] Relapses of Wegener's granulomatosis have been associated with infections.[7] In each relapse, in our case, an infective organism was found (Table I). This has not been previously documented in ophthalmic cases and suggests there may be a role for maintenance antibiotics.

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Sir,

Toxic Retinopathy Secondary to Repeat Intravitreal Amikacin and Vancomycin

Controversy exists concerning the indications and safety of single and repeat intravitreal antibiotic injections in the management of bacterial endophthalmitis. We report an unusual toxic reaction following repeat intravitreal injection of amikacin and vancomycin in a child with endophthalmitis following a penetrating eye injury.

Case Report

An 8-year-old white boy was injured in the right eye while playing darts. On presentation to the Accident & Emergency Department 1 hour later, he had a small nasal scleral perforation 2.5 mm from the limbus. There was minimal uveal and vitreous herniation but no injury to the lens or the retina. Visual acuity was 6/4 unaided. There was no relative afferent pupillary defect and the other eye was normal. Intravenous cephazolin and topical gentamicin and steroids were commenced. A few hours later, increased vitreous activity led to the diagnosis of early endophthalmitis. Primary wound repair under general anaesthesia was combined with sampling from the injury site for microbiological analysis and pars plana intravitreal injection of amikacin 0.4 mg (in 0.1 ml saline) and vancomycin 1.0 mg (in 0.1 ml saline) and subconjunctival injection of vancomycin 25 mg, ceftazidime 100 mg and 0.5 ml of Mydricaine No. 2.

Marked anterior chamber and vitreous activity 24 hours after primary repair signified deterioration. Endophthalmitis was clinically confirmed during examination under anaesthesia when a white opacity was observed at the vitreous base extending inferiorly from the injury site. Uncomplicated wide diagnostic core vitrectomy was performed 32 hours after the initial repair and a repeat intravitreal injection of amikacin 0 4 mg and vancomycin 1.0 mg and a subconjunctival injection of vancomycin 25 mg and ceftazidime 100 mg were given. The optic nerve head and macula were normal.

The child was distressed and photophobic 12 hours following vitrectomy. Examination was difficult and visual acuity unobtainable. Nevertheless, intraocular pressure was normal and anterior chamber and vitreal activity were

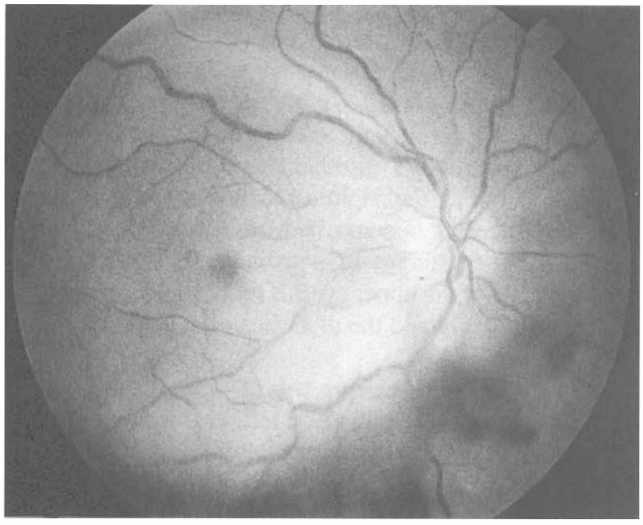

Fig. 1. *Red-free fundus photograph showing 'cherry-red spot' at the macula.*

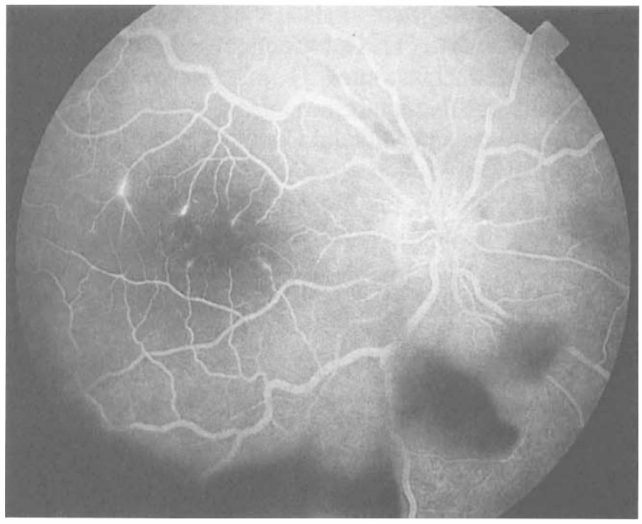

Fig. 2. *Fluorescein angiogram. Note the macular ischaemia and leakage from small vessels.*

reduced. At 36 hours after vitrectomy there was no evidence of vitreous activity but the patient's visual acuity was hand movements with a relative afferent pupillary defect. Despite prompt control of the infection, severe macular damage was apparent on funduscopy in the form of a 'cherry-red spot' surrounded by oedematous retina (Fig. 1).

The scotopic electroretinogram revealed a selective reduction of the *b* wave (normal *a* wave), and absent oscillatory potentials. The 30 Hz (cone) responses were grossly reduced and delayed. The electro-oculogram index was recorded at 110%, which was grossly subnormal. Fluorescein angiography showed underperfusion of the retina in the macular area and leakage of dye from surrounding small vessels (Fig. 2). Complete blood count, clotting screen, biochemical profile and computed tomography of the head and orbits were normal. No growth was obtained from the vitreous samples. A trial of systemic steroids did not improve the visual acuity or electroretinogram.

Discussion

Dart-induced perforating injuries are often associated with poor visual outcome directly from the injury and from endophthalmitis.[1] Prompt management of bacterial endophthalmitis necessitates administration of intravitreal antibiotics prior to culture results. Broad-spectrum cover is required. Increasing bacterial resistance has prompted the current use of amikacin and vancomycin in combination.[2]

Repeated intravitreal antibiotics injections in the management of endophthalmitis have been advocated by several authors.[3] Studies have documented an enhanced turnover of intravitreally injected drugs after vitrectomy, especially in inflamed eyes. Despite the attraction of maintaining a therapeutic level of antibiotics, it is possible that retinal toxicity may occur with repeated injections.[4] It has been suggested that the more rapid decrease in vitreous levels of these antibiotics in vitrectomised eyes may not protect the retina from cumulative toxicity.

A previous report described a retinal toxic reaction following inadvertent intravitreal injection of a high dose of tobramycin during sub-Tenon injection.[5] Other reports described similar reactions following intravitreal injection of gentamicin, clindamycin[6] and amikacin.[7] The retinopathy was characterised by marked capillary occlusion at the posterior pole with oedema and intraretinal haemorrhages and was associated with poor visual prognosis. The proposed mechanism is either a direct retinal toxicity or secondary retinal infarction, possibly due to leucocytic plugging. In the case presented here, prompt control of the infection, the characteristic appearance of the angiogram and the electrodiagnostic changes[8] were suggestive of a similar retinal toxic reaction.

This devastating toxic reaction, occasionally encountered after a single exposure to intravitreal antibiotics, may reflect a variable individual susceptibility to the antibiotics used. This report shows toxic damage to human retina after repeated intravitreal injection of amikacin and vancomycin. The short interval of 32 hours between the two injections may have increased the intravitreal level dangerously close to the threshold for toxicity. It appears that vitrectomy did not protect the retina from cumulative toxicity. This highlights the dangers inherent in this mode of treatment and ophthalmic surgeons should be aware of this complication. The timing of re-injection of intravitreal antibiotics should be carefully considered to avoid potentially toxic intravitreal levels.

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Sir,

Congenital Tarsal Kink: A Rare Cause of Neonatal Corneal Ulcers

Congenital tarsal kink is a rare condition of unknown origin, in which a fold is present in the upper tarsal plate at birth. The bent edge of the tarsus may then traumatise the cornea causing ulceration.[1–3]

Case Report

A 1-week-old baby girl presented to the Eye Department with a history of sticky eyes and failure of eye opening since birth. Perinatal conjunctival swabs were negative.

The mother's obstetric history was unremarkable, and maternal microbiological investigations proved negative. Ocular examination revealed bilateral upper lid oedema, absent lid crease and conjunctival chemosis in the upper fornices (Fig. 1). Bilateral central corneal ulceration was also present, with the right eye more severely affected (Fig. 2).

The baby was treated with intensive topical penicillin and tetracycline and oral erythromycin, which resulted in slight improvement in the clinical signs. Further extensive microbiological investigations failed to detect a causative organism. The possibility of non-accidental injury was excluded.

The left ulcer slowly epithelialised following 2 weeks of treatment, but the right eye showed only minimal improvement. Lid oedema had now settled and it was apparent that the tarsus of both upper lids was rotated. Manipulation of the upper eyelids resulted in correction of the tarsal anomaly and the ulcer healed with residual corneal scarring.

Although corneal scarring was asymmetrical, the right eye being more affected than the left, vision as assessed by preferential looking following resolution of the ulcers was equal in both eyes and within normal limits for the stage of development.

Discussion

Congenital entropion of the upper eyelids is extremely rare.[4] Congenital tarsal kink represents a severe form of entropion whereby the tarsal plate is kinked along its horizontal length causing inversion of the eyelid and lashes.[1–3] It is characterised by blepharospasm and absence of the upper lid fold. Severe corneal ulceration may result from the inturned lids and lashes abrading the cornea. It is thought that mechanical trauma to the cornea occurs *in utero*.[2] The aetiology of the condition is unknown; a primary defect of the tarsus has been proposed as a possible underlying abnormality. Alternatively, overaction or malpositioning of orbicularis muscle fibres may result in an infolding of the tarsus *in utero*.[5]

Primary management of congenital tarsal kink requires manual unfolding of the tarsal plate. The lid may then be taped shut and a pressure dressing applied for 24–48 hours.[6] A bandage contact lens may aid corneal healing.

More severe cases require surgical management. A variety of surgical procedures have been described, including tarsal split,[1] tarsal wedge resection,[7] lamellar tarsoplasty[2] and repositioning of the anterior lamella.[5,8] As the condition may be corrected by altering the position of the orbicularis and creating a skin crease with a simple

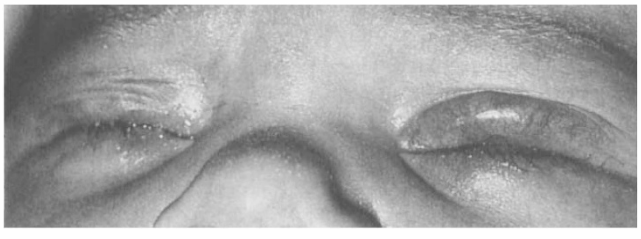

Fig. 1. *Eyelids showing tarsal plate anomaly.*

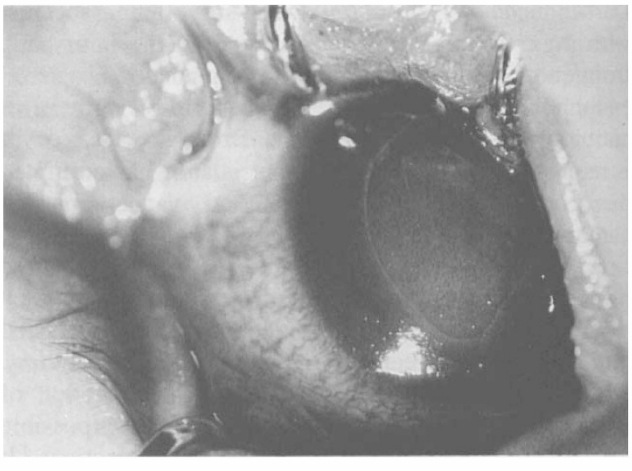

Fig. 2. *Right cornea showing central ulceration.*