Sir,

Acute onset bilateral cataracts in an infant with vertically transmitted HIV with CMV retinitis despite treatment

Cytomegalovirus (CMV) retinitis occurs in 14–50% of AIDS patients,[1] but CMV retinitis in children with vertically transmitted HIV remains rare.[2] We are not aware of any cases of acute bilateral cataracts in such patients while on medical treatment for CMV.

Case report

This patient was a known case of vertically transmitted HIV. He was born at full term (birth weight 2.7 kg), postnatal examination was normal and at 2 months of age he was commenced on co-trimoxazole prophylaxis. Subsequently he showed neurodevelopmental delay. At 10 months he developed CMV retinitis and HIV encephalopathy. He had a diffuse haemorrhage retinitis on the right eye involving the macula; the left eye showed only atrophic changes at the mid-periphery. The right eye showed very poor fixation but the left eye was able to follow large toys. The CD4 count at this time was 203 cells/mm^3. Electrophysiological testing showed poor electroretinographic and visual evoked responses from both eyes. The CT scan showed diffuse cerebral atrophy with calcification consistent with CMV infection. The patient was started on intravenous ganciclovir 5 mg/kg twice a day.

The retinitis in the right eye responded to the treatment. Three months later a focus of active inflammation was noted in the periphery of the left eye, with formation of vitreous haemorrhage. The child exhibited roving eye movements implying a significant deterioration of vision.

Ten months after treatment, the patient's mother suddenly noticed that the patient's pupils had become white. Bilateral dense cataracts were seen; there was no anterior segment inflammation and there was no view of the posterior segments. Tonometry revealed intraocular pressure of 8 mmHg in both eyes. B-scan ultrasonography revealed flat retinae. The patient was treated conservatively.

Discussion

CMV retinitis is described in adult patients with $CD4^+$ counts below 50 cells/ml.[3] Screening is therefore recommended when the CD4 count drops to 60 cells/ml. The CD4 count at the time of our patient developing the CMV retinitis was 203 cells/ml. However, if reference is made to a childhood CD4 chart[4] it is seen that the normal level at this age is above 3000 cells/ml. It therefore needs to be emphasised that when interpreting CD4 counts as an assessment of risk of CMV retinitis in children, childhood white cell count values are normally higher than adult values and the reference values from the appropriate laboratory should be consulted whenever childhood CD4 counts are being interpreted.

In our case the reason for the rapidity of onset of severe bilateral cataracts is uncertain, especially as both retinae were flat. It may be ascribed to the HIV infection, CMV or the combination. In one report, 1 of 39 eyes with CMV retinitis treated with ganciclovir and foscarnet subsequently developed cataract,[5] although this was not in a child. In another study involving 101 HIV-positive patients without CMV retinitis, opacities were seen in the lens cortex of 52% of the eyes.[6] Lens opacities have also been described in mice infected with HIV virus[7] and it has been suggested that HIV protease gene expression alone can cause cataract formation.[8] This may occur through the action of the HIV-1 protease enzyme itself or activation of enzymes such as calpain (intracellular cysteine proteases) leading to the fragmentation of crystallins. It is the organisation of crystallins that maintains the transparency of the lens in normal circumstances.

In summary, we describe acute onset bilateral cataracts in a child with vertically transmitted HIV infection and CMV retinitis. The pathogenesis is uncertain, with no evidence of retinal detachment or anterior segment inflammation. Mouse studies have suggested it may be related to direct viral invasion of the lens and enzymatic lens protein degradation.

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Sir,

Bilateral familial inferotemporal retinal dialyses

Retinal detachment associated with retinal dialysis occurs in 10% of all rhegmatogenous retinal

detachments. Controversy exists in the literature as to the roles that genetics and environment play in the aetiology of bilateral dialyses.[1–10] We report a family in which bilateral inferotemporal dialyses were found in siblings, without prior history of ocular trauma, indicating a possible genetic predisposition to this condition.

Case reports

Case 1 An 11-year-old girl was referred complaining of progressive blurring of left eye (LE) vision for 1 month. Visual acuity was 6/9 right eye (RE) and hand movements LE with low myopic correction. Examination showed extensive bilateral inferotemporal retinal dialyses with detachments. The macula was attached in the RE and detached in the LE. She underwent bilateral scleral buckling procedures 2 months apart. Five years later visual acuity with correction is 6/5RE and 6/24LE and the retinae are attached.

Case 2 The 12-year-old brother of case 1 was undergoing routine refraction/ocular examination when he was found to have bilateral (asymptomatic) inferotemporal retinal dialyses, extending almost 180° (Figs. 1, 2). Visual acuities with low myopic correction were 6/9RE and 6/9LE and both maculae were attached. He underwent bilateral scleral buckling procedures 2 months apart. One year later, both retinae remain attached with stable visual acuities at 6/9.

An older brother and both parents were examined by one of us (P.K.L.) and no dialyses were found in any of these family members.

Comment

Retinal dialyses comprise 10% of all rhegmatogenous retinal detachments.[1] In his manuscript on dialysis aetiology, Scott[10] has defined a retinal dialysis as a tear in the retina whose anterior edge is at the ora serrata and whose posterior edge is attached to the vitreous base. Thus a dialysis is different from a more posteriorly located retinal tear in that the vitreous traction in a dialysis is applied to the posterior rather than the anterior retinal edge. Blunt trauma has long been postulated to be a major predisposing factor in the development of retinal dialysis, possibly due to

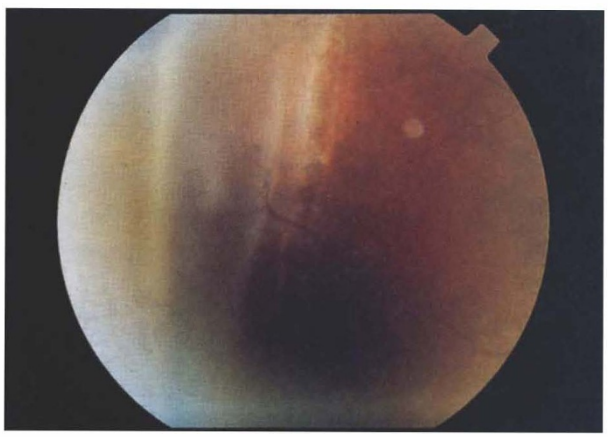

Fig. 1. *Case 2. Right eye.*

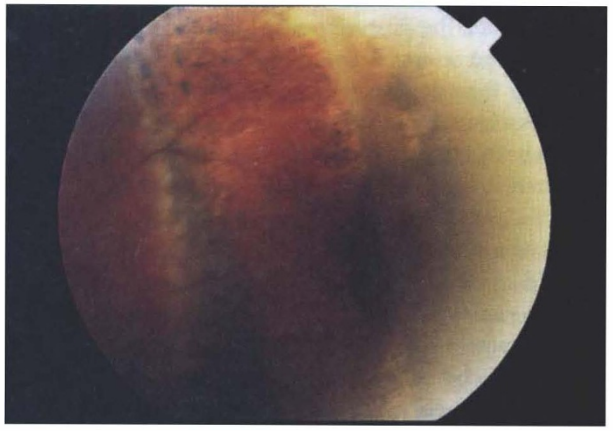

Fig. 2. *Case 2. Left eye.*

distortional injury to the globe, with vitreous traction on the retina in the region of the ora serrata.

Prior studies on retinal dialysis have shown that the majority of traumatic dialyses are located superonasally.[1–3,9,10] Heredity has also been implicated in the aetiology of retinal dialysis and studies have indicated that these dialyses are more likely to be located inferotemporally.[1–4,10] Some investigators have reported large series of dialyses without evidence of genetic predisposition,[2] but there are many other reports that propose multifactorial causation with a genetic predisposition.[3–7,10]

We report two siblings who were found to have extensive bilateral retinal dialyses in similar locations, without evidence of previous ocular trauma. Spontaneous giant retinal tears of 180° are more common than giant retinal dialyses, but are commonly associated with congenital vitreous anomaly and/or detachment of the pars plana, as in some cases of Stickler's syndrome. In both of our cases the vitreous was normal, there was no sign of detachment of the pars plana and the posterior vitreous was attached throughout the fundus. We could find no evidence for any hereditary genetic predisposition to retinal detachment, such as Stickler syndrome, and there was no family history of retinal detachment. These children had previously documented normal eye examinations, apart from mild myopia, and had no evidence of congenital eye disease. The presence of bilateral inferotemporal dialyses in these siblings suggests that genetic factors can play a role in the development of retinal dialysis. Previous reports have suggested that foregoing trauma, long since forgotten by the patient (and/or relatives), is the likely aetiology of retinal dialyses.[2] This aetiology is unlikely in these children, given that each would have had to suffer trauma to both eyes and there was not a shred of evidence to suggest either accidental or non-accidental injury.

It is possible that a genetic predisposition is present in all instances of retinal dialysis and that in those patients who develop spontaneous dialysis, predisposing factors are strongest. In his description of the mechanism of dialysis formation, Scott postulates an abnormal adhesion between the vitreous base and its attachment to

the ora serrata, the posterior edge of the vitreous base being seen, on scleral depression, to extend more posteriorly than normal. The oral frill is a degenerative change associated with this posterior extension of the vitreous base. He goes on to postulate that the oral frill is a precursor to dialysis, attentuation of the retina in the area of the oral frill occurring as a result of dynamic traction or minor trauma. A break is then formed in the attenuated area as further retinal degeneration occurs and fluid passes behind the retina. Some eyes that develop dialyses have an inherited weakness or oral degenerative change and are predisposed to the development of dialyses with normal eye movements or minimal blunt trauma. Other eyes, with lesser degenerative change, require much higher levels of trauma to lead to dialysis formation.[10]

Our cases serve to emphasise the importance of examining both eyes in all patients with inferotemporal non-traumatic retinal dialysis, as they may have a genetic predisposition, due to inherited abnormalities, associated with degenerative changes at the ora serrata.

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Sir,

Spontaneous anterior capsular rupture in Alport syndrome

Alport syndrome is most commonly an X-linked disease affecting basement membrane collagen. It is characterised by progressive nephritis and associated with high-tone sensorineural deafness; more variably certain ocular changes occur.[1] Anterior lenticonus is an integral part of the disease.[2] The lens capsule has been observed to have marked anterior thinning associated with a decrease in the number of epithelial cells.[3]

In this report, a patient with Alport's syndrome who developed spontaneous anterior capsular lens rupture is described.

Case report

An 18-year-old man with Alport syndrome was first examined in the eye department 1 year after successful renal transplantation. He complained of rapidly progressive visual loss in his left eye. Visual acuity in the right eye was 20/30 with −2.75 cylinder axis 180° and 20/400 in the left eye. Slit-lamp examination revealed mild anterior lenticonus in the right eye, and irregular protrusion of the opacified anterior lens surface with capsular folds and fibrosis in the left eye. Anterior chamber reaction with 2+ cells was present. No retinal or corneal changes were found. The patient was lost to follow-up, during which time he underwent planned extracapsular lens extraction and intraocular lens implantation for the left eye elsewhere. He presented 4 months later with visual deterioration in his right eye of 2 weeks' duration. Right visual acuity was 20/70 with −1.00 cylinder axis 180° and 20/25 in the left eye with posterior chamber intraocular lens. In the right eye, the anterior lens capsule had protruded forming a localised bulla at the anterior pole of the right lens (Fig. 1). Two weeks later another blister appeared above the first one (Fig. 2); the anterior capsule burst 4 days later through

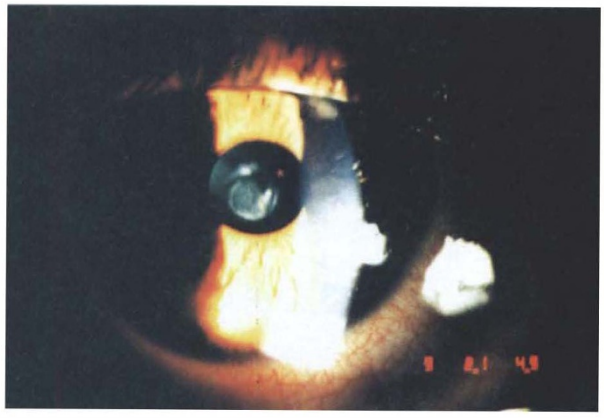

Fig. 1. *Blister formation at the anterior pole of the lens with underlying localized opacity.*