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.

References

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Sir,

Bilateral familial inferotemporal retinal dialyses

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