Comment

DMD is not an uncommon complication after cataract surgery.³ Vastine *et al* suggest surgical intervention for large planar and scrolled detachments.⁴ Walland *et al* also recommend repair of large DMD should it be recognised intraoperatively, and that an expanding gas should be used.⁵

In this case the patient underwent routine, small incision phaco-emulsification cataract surgery with foldable lens implant and developed persistent corneal oedema. The DMD was not noted until the seventh week postoperative visit and appeared to have been progressive. The signs of DMD were very subtle and the examination was made more difficult by the extensive corneal oedema giving poor details of the anterior chamber. This patient responded well and promptly to anterior chamber reformation with air, with an almost immediate reattachment of the Descemet's membrane. We used air to fill the anterior chamber after a three port paracentesis to drain the sub-Descemet's membrane fluid. Once the Descemet's membrane was attached to the corneal stroma the incision was covered internally with *Provisc*.

In summary, DMD can be a cause of corneal oedema and should be considered as a differential diagnosis in cases of corneal oedema following cataract surgery, especially if the procedure has been uneventful. One will need to examine carefully, as the signs of DMD can be subtle especially if masked by extensive corneal oedema. DMD can be reattached successfully using only air tamponade with good visual outcome.

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Sir,

Susac Syndrome

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We present a case of Susac syndrome, which is rare, but more common than thought. It is characterised by the triad of retinal arterial occlusion, sensorineural deafness and encephalopathy.^{1,2} This condition is often under diagnosed or misdiagnosed as multiple sclerosis³ or systemic lupus erythematosus. We also discuss the differential diagnosis especially in the absence of early diagnosis of the disease.

Case report

A 24-year-old Caucasian female was referred by the neurologist, for ophthalmic examination. She gave a history of migraine type of left-sided headache and fixed small scotoma in her left eye. She also complained of decreased hearing. She was previously fit and healthy and denies any past history of eye problems. She was on the oral contraceptive pill. The visual acuity in either eye was 6/6. Fundi showed two cotton wool spots in the left eye with signs of branch retinal arteriolar occlusion (Figure 1). The fundus fluorescein angiography showed leakage from the vessel walls of the small arteries. The 24–2 automated perimetry showed an area of scotoma corresponding to the cotton wool spot.

The patient developed encephalopathy the following week and was investigated with an urgent MRI scan. The scan showed T2 weighted high signal discrete areas around the periventricular region (Figure 2), in the white and grey matter and corpus callosum.

The lumbar puncture showed a marginally raised cerebrospinal fluid, a pressure of 26 cm, with no increase in cell count. No oligoclonal antibodies were found in the cerebrospinal fluid. The cerebral angiogram was normal. All the immunological tests were normal. Erythrocyte sedimentation rate was 12 mmHg. Antinuclear antibodies (ANA) titre was negative; Florescent treponemal antibodies (FTA-ABS) and C-reactive protein were normal. An audiogram showed sensorineural deafness.

She was treated with intravenous prednisolone 1 g/kg for 3 days. The response to steroid was remarkable with the patient's mental status much improved, though her visual complaints were unchanged. The symptoms of encephalopathy subsided within 48 h. She was started on oral steroid of

Figure 1 Left fundus photograph showing areas of retinal ischemia due to small arteriolar occlusion.

Figure 2 Coronal T2 weighted image with FLAIR, showing high signal lesions around the periventricular region.

60 mg/day after that, which was tapered gradually. The MRI was repeated after 2 weeks and the hyper intense areas had almost disappeared (Figure 3). There has been recurrence twice since with symptoms of milder degree during which period the symptoms were treated with an increase in dosage of oral steroid. She is now asymptomatic with 10 mg of oral steroid. At 10 month follow-up after initial presentation, her visual acuity is 6/6 in either eye with small scotoma in the left eye. Left fundus shows old branch retinal arterial occlusion.

Comment

Susac syndrome was first described independently by Susac in 1976 and since then called by different names like RED-M (retinopathy, encephalopathy, deafness associated with microangiopathy), SICRET (small infarction of cochlear, retinal and encephalitic tissue) or retinocochlear vasculopathy.^{1,5} This syndrome almost commonly affects women in the 20–40 years age group and is characterised by the clinical triad of encephalopathy, branch retinal artery occlusion and sensorineural hearing loss due to microangiopathy of retina, brain and cochlea. Headache is the usual prodromal symptom. The condition has a chronic

Figure 3 Almost complete disappearance of the lesions after treatment with steroids.

relapsing course punctuated by frequent remissions and exacerbations. It is a self-limiting disorder, extending over 1–2 years with remission. The underlying process is believed to be an immunological reaction, leading to small vessel vasculitis⁴ causing micro infarcts in the retina, brain and the apical turn of the cochlea. The important differentials would be multiple sclerosis, aseptic meningitis, systemic lupus erythematosus, Bechet's and complicated migraines. The other vasculitides like sarcoidosis, tuberculosis, syphilis and lymphomas have to be ruled out.

A high index of suspicion, leading to early recognition of this syndrome is important because treatment with immunosuppression may minimise permanent cognitive, audiologic and visual sequelae. This syndrome has a good prognosis when treated early. In patients whom early diagnosis has led to early administration of immunosuppressive therapy, recovery can be almost complete.

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Sir,

Cocaine-associated central retinal artery occlusion in a young man

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Retinal vascular occlusions in young adults are seen very infrequently and are generally associated with systemic disorders.¹ With the increasing abuse of cocaine in Western countries, reports of medical complications are increasing. Besides the more common vascular complications described, such as myocardial infarction² and cerebrovascular events,³ only one case of central retinal artery occlusion (CRAO) in an intravenous cocaine abuser, had until recently been reported.⁴ There have now also been two case reports of CRAO and central retinal vein occlusion in intranasal cocaine abusers.^{5,6}

Case report

A 42-year-old man presented to casualty with a sudden painless loss of vision in his right eye 9 hours previously. He was not on any medication and was otherwise entirely fit and well with no history of migraine. He was a smoker of ten cigarettes a day for the past 20 years. He admitted to smoking 'crack' cocaine (more potent alkaloid form of cocaine) twice a week for the last 4 years, although he denied use of intranasal or intravenous abuse. He had smoked cocaine the previous evening. Visual acuity in the right eye was counting fingers at one metre, left eye 6/4. There was a right relative afferent pupillary defect. Examination of the anterior segments was unremarkable. Examination of the right fundus revealed diffuse retinal whitening and oedema in the posterior pole, with a foveal cherry-red spot and vascular attenuation, with sludging and segmentation of the blood column, consistent with a CRAO (Figure 1). No arterial emboli were seen. The left fundus was unremarkable (Figure 2).

Immediate treatment was instituted comprising 500 mg of acetazolamide intravenously, intermittent ocular massage and re-breathing into a paper bag for half an hour. Cardiovascular examination was normal with no carotid bruits, a regular pulse and no murmur audible.

Blood pressure, echocardiography, carotid ultrasonography, full blood count, erythrocyte sedimentation rate, fasting lipids and glucose, autoantibody screen including anti-cardiolipin antibody, protein S and C levels, antithrombin III level, Factor V Leiden, and blood homocysteine levels, were all normal. He was found to be sickle cell trait positive.