

Figure 2 Axial FLAIR sequence MRI of the brain shows high signal area at the left parieto-occipital boundary. Appearance suggests ischaemia.

occlusive disease with hypertension, also common in PXE acting as an accelerating factor.²⁻⁴ In a study of 100 cases of PXE,⁴ patients who developed ischaemic stroke were found to have only small vessel involvement.

Ischaemic stroke in patients with PXE is usually not seen until the fifth decade of life unlike our patient.^{2,3} Relative risk of developing ischaemic strokes is estimated to be 3.6 in PXE patients less than 65 years compared with the general population.⁴ Other neurological complications quoted in the literature include intracranial aneurysms, subarachnoid and intracerebral hemorrhages, progressive intellectual deterioration, mental disturbances and seizures.⁵ This case reminds the ophthalmologists about the systemic nature of this condition and draws attention to the need for neuroimaging for unexplained visual loss in patients with PXE.

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Sir,

Reattachment of extensive Descemet's membrane detachment following uneventful phaco-emulsification surgery

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Descemet's membrane detachment (DMD) is an uncommon condition with a wide range of possible aetiologies. Probably the commonest cause is a localised detachment occurring at cataract extraction surgery. More extensive detachments give rise to a 'double anterior chamber' after perforation of Descemet's membrane in deep lamellar keratoplasty.¹ Large DMD after cataract extraction is rare. Non-surgical factors that could predispose to DMD are traumatic, congenital glaucoma and corneal ectasias, among others.

DMD detachment can be classified as:

Planar: <1 mm separation from the stroma

- Peripheral detachment only
- Combined peripheral and central detachment

Non-planar: >1 mm separation from the stroma

- Peripheral detachment only
- Combined peripheral and central detachment.²

Case report

An 80-year-old woman was referred by her General Practitioner with a gradual deterioration of vision in her right eye. She had no past ocular history. She had a medical history of chronic renal failure, hypertension and left ventricular failure. On examination the visual acuity was hand movements unaided in the right and 6/12 in the left. There was a dense nuclear cataract in the right eye and mild nuclear cataract in the left eye. There were no other ocular abnormalities. She was listed for right cataract surgery as a day case.

She underwent uncomplicated right phacoemulsification cataract extraction with intraocular lens implant under local anaesthesia. She had clear corneal incision at the 11 o'clock position using a 2.5 mm disposable keratome and a superonasal paracentesis. Divide and conquer phaco-emulsification was performed using a *Millennium Storz* machine (STORZ Ophthalmics Inc, St Louis, MO, USA). The phaco-emulsification power and time were 13% and 2 min and 24 s respectively. The viscoelastic (Provisc) used throughout the procedure was completely removed at the end of the procedure. Balanced salt solution (BSS Alcon) was used as the infusion fluid. Corneal incision was sealed with two 10/0 vicryl sutures. Finally the patient had a subconjunctival injection of Cefuroxime 125 mg and Betnesol 2 mg and the eye was covered with a plastic shield.

The patient was reviewed 4 days following surgery. She was asymptomatic. Her right visual acuity was 6/36 unaided, improving to 6/18 with pinhole. There were Descemet's membrane folds, mild inflammatory reaction in the anterior chamber and the intraocular pressures were 12 mmHg in the right eye and 18 mmHg in the left. She was given guttae Dexamethasone 0.1% to be used four times a day, until the next visit.

She was closely monitored and 7 weeks after surgery, her vision in the right eye had deteriorated to 6/60 unaided, 6/36 with pinhole. The patient was complaining of right epiphora, but the eye was comfortable otherwise. The right corneal sutures had dissolved completely and there were no signs of ocular inflammation. Right intraocular pressure was 16 mmHg. The posterior cornea was noted to have a combined non-planar DMD involving approximately two thirds of the cornea, extending from 5 o'clock around to 12 o'clock (Figure 1).

Following discussion she was scheduled for right anterior chamber tamponade with air under local anaesthesia. The procedure was uneventful. Three paracenteses to drain the sub-descemet fluid were performed. The anterior chamber was filled with

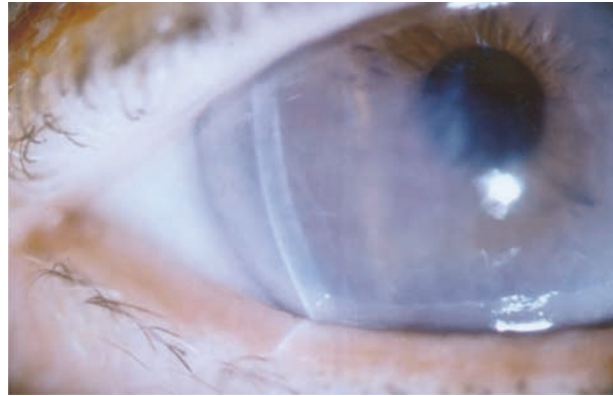


Figure 1 Seven weeks after surgery, a slit lamp view of the right eye shows a localised corneal epithelial oedema and the detachment of the Descemet's membrane.

filtered air in its entire volume and Provisc was used to cover the internal aspect of the corneal incision. The patient was asked to keep a postoperative face-up position for one week and took Acetazolamide sustained release 250 mg stat dose that evening. The following day, her right eye was comfortable, the corneal epithelial oedema had cleared and the Descemet's membrane appeared to be reattached completely. There was a small wrinkle of the Descemet's membrane running from the phacoemulsification corneal wound inferiorly (Figure 2).

One week after the anterior chamber reformation her right visual acuity was 6/24 unaided improving to 6/9 with $-3.00/+1.25 \times 55^\circ$. The Descemet's membrane remained attached and the cornea was clear, her intraocular pressures were 10 mmHg in the right eye and 12 mmHg in the left eye.



Figure 2 One day after surgery, slit lamp view shows a clear cornea. The edge of the Descemet's membrane is still visible running inferiorly.

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.