Corneal deposits after the topical use of ofloxacin in two children with vernal keratoconjunctivitis

Topical fluoroquinolones are broad spectrum, bactericidal agents with activity against both Gram positive and Gram negative corneal pathogens. Single drug administration with topical fluoroquinolones has been effectively used in treatment of bacterial ulcerative keratitis. The drugs are safe, but a white crystalline deposit, that in most cases spontaneously dissolved, has been reported with ciprofloxacin 0.3% and norfloxacin 0.3%. Up to now this side effect has not been described with ofloxacin 0.3%. We present two children with severe vernal conjunctivitis who developed corneal deposits after use of ofloxacin 0.3% eye drops.

Material and methods

Patient 1

A 6 year old boy with vernal keratoconjunctivitis (VKC) developed a painful right eye while on holiday. His ophthalmologist diagnosed a corneal ulcer and prescribed ofloxacin eye drops followed by ofloxacin ointment four times a day for 3 weeks, associated with prednisolone drops. As the inflammation in the right eye persisted, the child was referred to our department. Slit lamp examination revealed mild VKC and a corneal ulcer with a dense, white, sharply demarcated crystalline deposit (Fig 1). All topical treatment was discontinued, but the deposit did not dissolve and inflammation persisted. Twelve days after the discontinuation of the topical treatment a corneal scraping of the deposit was performed in combination with a sub-tarsal injection of corticosteroids in both eyes.

Results

The corneal scraping was analysed by microscopy. To that end, pelloxacin was added as internal standard to a minute tissue sample was extracted with an acid (formic acid, pH 3.0) mixture of water/acetonitrile (60/40). An aliquot was injected onto a 1 mm × 15 cm Versil ODS 5µm column (5 µm), eluted isocratically (40 µl/min) with 90/10 water/acetonitrile pH 3.0. Detection was performed using a Q-TOF mass spectrometer (Micromass, Manchester, UK) in the MS/MS mode. The retention time data and the full scan ion product spectrum (major diagnostic ions m/z: 362.1, 318.1, and 264.1) unequivocally disclosed that ofloxacin was present in the deposit. After removal of the deposit the ulcer epithelialised rapidly.

Comment

Topical ofloxacin 0.3% has been used in the treatment of patients with bacterial keratitis and in an animal model of keratitis and has been tested in an in vitro tear model. So far no reports of corneal deposits have been made. There is only one case report in which deposits are described after the combined use of cyclosporin and ofloxacin; however, the authors attributed the deposit to the use of cyclosporin, although they did not conduct an analysis on the corneal deposits.

We report for the first time two children who demonstrate corneal deposits of ofloxacin adherent to a corneal erosion caused by VKC. Analysis of a corneal deposit proved the presence of ofloxacin while treatment had been discontinued for 7 days, indicative of ofloxacin depot formation by precipitation.

The exact factors contributing to the formation of the fluoroquinolone precipitate are unknown, but pH solubility profiles are of importance. Ofloxacin, like the other fluoroquinolones, exhibits pH dependent solubility. In the in vitro tear model tear drug concentration of ofloxacin remained below solubility at all pH values tested (pH 6.4–7.2). One could argue that VKC associated ulcers occur in a different “microenvironment” compared to bacterial ulcers. Mast cell activation leads to the release of different mediators and inflammatory cells, such as histamine, serotonin, heparin, prostaglandins, protease, tryptase and eosinophils, all of which can contribute to a “microenvironment” potentially promoting ofloxacin precipitation in one way or the other. It would be interesting to find out whether there are significant changes, pH or other, in the tears of children with VKC. In conclusion, we can say that ofloxacin does not seem to form corneal deposits which were used in the treatment of bacterial ulcers, contrary to ciprofloxacin or norfloxacin. However, clinicians should be aware that deposits can occur after the use of ofloxacin in VKC associated ulcers.

Acknowledgements

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References


Chromosomal translocation in a family with ocular abnormalities: indications for karyotype analysis

Hereditary isolated congenital malformations such as congenital cataracts or anterior segment abnormalities are usually considered to be the result of a single gene mutation. The identification of associated abnormalities—dysmorphic features, a malformation in another organ system, or mental handicap, help to identify those who carry more severe genetic defects such as chromosome rearrangements. We describe a family where two healthy men, a father and son, had juvenile cataracts. They both had offspring with more severe ocular abnormalities including anterior segment dysgenesis, as well as mental retardation and dysmorphic features. Recent karyotype analysis revealed a balanced

Figure 1 Slit lamp detail of vernal keratoconjunctivitis associated ulcer with a white sharply demarcated ofloxacin deposit.
translocation in both father and son and a derived unbalanced translocation in their more severely affected children. Analysis of the translocation breakpoints led to the identification of a new human disease gene, MAF, in lens and anterior segment development. Critical, for the family, the finding of the translocation provides an explanation for the severe ocular and other anomalies in the individuals with the unbalanced derivative karyotype. In addition, it allows the option of prenatal diagnosis for future offspring for individuals carrying the balanced translocation.

Family report
A man with juvenile onset cataracts, and a family history of this, sought genetic advice with his partner. This man (II.3, Fig 1) was the father of three children with ocular anomalies and developmental delay. He had juvenile onset, progressive cataracts which were removed at the age of 24 years. The cataracts were a result of acquired, widespread dot opacities, with anterior and posterior sutural densities. General and other ocular examination was normal. This couple's only living child, a daughter age 5 years (III.1, Fig 1), had been noted at birth to have opaque corneas, small lens remnants in each eye, and very flat anterior chambers with rims of abnormal iris tissue. Bilateral microphthalmia was present with axial lengths of 12 mm. Fundal examination and an ERG were normal. Bilateral corneal grafts were attempted, but rejection occurred. This child had significant developmental delay with learning and motor difficulties, saying approximately five single words and only just walking at the age of 5 years. She had microcephaly and dysmorphic features (Fig 2A and B).

Two previous siblings, now dead, had also had ocular and other anomalies. The elder of these (III.1, Fig 1) had unilateral cataract, global developmental delay, and similar facial features to III.3 (Fig 2C). She died age 6 years as a result of aspiration pneumonia (Fig 2D). Her sister (III.2, Fig 1) had cloudy corneas at birth. She died soon after birth as a result of laryngeal stenosis (Fig 2D).

Analysis of the family history revealed that the patient's father (I.1, Fig 1), had also had juvenile onset cataracts but was otherwise well. He had two other children with ocular anomalies and mental impairment. The elder daughter (III.1, Fig 1), had juvenile onset cataracts and died age 29 years because of cardiac arrhythmia. The second daughter (III.2, Fig 1), developed cataracts in early childhood and had bilateral dense cataracts at the age of 30 years. Both these individuals had limited speech and required assistance with personal care. These women have dysmorphic facial features similar to those seen in their developmentally delayed nieces.

Karyotype analyses were requested on surviving family members. These demonstrated that the two otherwise normal men (I.1 and II.3), with juvenile onset cataracts, were carriers of the same balanced translocation 46,XX,der(5);t(5;16)(p15.3;q23.2) (Fig 1). Their two surviving offspring (II.2 and III.3) with more severe ocular anomalies, mental retardation, and dysmorphic facial features both had the same unbalanced, derived derivative karyotypes, 46,XX,der(5);t(5;16)(p15.3;q23.2) (Fig 1). This had been taken at necropsy for a karyotype analysis on III.2 which had also shown additional material on chromosome 5. This would be consistent with the unbalanced karyotype seen in her sibling, but unfortunately contact with the family about this at the time did not proceed. Karyotype analyses were not performed in II.1 or III.1 before their deaths and had not been performed in III.3 before the genetic referral.

Comment
The diagnosis of the chromosomal translocation has significant implications for management and options for individuals in this family. The indications for karyotype analysis are not specifically emphasised to ophthalmologists as this is generally not part of the ophthalmologist's role in patient care. Nevertheless, chromosomal anomalies are frequently reported in the ophthalmic literature and contributions to identification of several genes in human eye disease, including PAX6, FOXC1, PITX2, and MAF from this family, have been made through analysis of chromosomal abnormalities.

In assessing the child with a congenital ocular anomaly, the presence of at least one other anomaly including another malformation or mental retardation, with or without dysmorphic features, is an indication for karyotype analysis. For the child, the finding of a chromosomal abnormality identifies the likely underlying cause, gives information regarding prognosis and directs management options. For the family, the origin of the chromosomal abnormality is critical in determining the likelihood of recurrence in a future pregnancy.

The parents of a child with a chromosome abnormality should be offered karyotype analysis. If one of the parents is a carrier of a balanced form of the translocation, this carries a risk of recurrence of up to 20% or sometimes higher of having a malformed and mentally retarded child as a result of transmission of an unbalanced chromosomal complement. Couples where one partner is a balanced translocation carrier can be offered prenatal diagnosis in a future pregnancy. If a parent is a balanced translocation carrier, one of his or her parents may also be a carrier so that siblings and other more distant relatives may also carry the translocation with its attendant risks for a future pregnancy. If the parental karyotypes are normal, the chromosomal abnormality has probably arisen as a de novo event in the egg or sperm that went on to create that child, and the likelihood of recurrence in a future pregnancy is low. Such risk analyses vary between families depending on family history and the particular chromosomal rearrangement: referral to a clinical genetics unit for assessment is recommended.

When a patient is seen with an ocular abnormality associated with mental retardation, dysmorphic features, or another malformation, a request or referral for karyotype analysis is indicated because of the significant risk of undetected chromosomal abnormalities.

Table 1

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Figure 1 Pedigree and clinical features of translocation family. Individuals with the balanced translocation (B) have cataract alone, while those with the unbalanced derivative karyotype (U) have additional features including other ocular anomalies, mental retardation, dysmorphic features, and other malformations. In individuals with the balanced translocation (B), 46,XY,t(5;16)(p15.3;q23.2), telomeric portions of 5p and 16q are translocated with no obvious loss of chromosomal material. Individuals with the unbalanced translocation (U), 46,XX,der(5);t(5;16)(p15.3;q23.2), have inherited the derived chromosome 5 (der(5)) from their father as well as his normal chromosome 16 and their mother’s normal 16, so they are trisomic for the telomeric portion of 16q containing MAF and monosomic for a small telomeric portion of 5p. NK = not known.
impact that the diagnosis of a chromosomal abnormality has for the individual and family. In this family the diagnosis of a familial chromosomal abnormality was made only recently. Before this five children were born over a period of 35 years with ocular abnormalities and associated features including mental retardation, dysmorphic features, and other non-ocular malformation, all caused by a chromosome abnormality derived from the balanced translocation in their parent.

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maximal medications, laser iridotomy was performed in her right eye. She had stopped topiramate 1 day before presentation. On the second day, pressures were normal and B scan confirmed uveal effusions. By day 3, her vision was 20/20 in both eyes, and her pressures were normal on brimonidine 0.2% in both eyes. Gonioscopy revealed posterior trabecular meshwork right eye and no angle structures left eye (anterior chamber depths 2.69 mm right eye, 2.20 mm left eye). After 1.5 weeks, pressures were normal without medications. Angles were open to scleral spur in both eyes. Anterior chamber depths were 2.87 mm in both eyes. No uveal effusions were seen.

**Comment**

Drug induced myopia has been associated with sulfa drugs such as acetazolamide, sulfamethoxazole/trimethoprim, indapamide, promethazine, spironolactone, isosorbide dinitrate, and bromocriptine. Other drugs include tetracycline, corticosteroids, hydrochlorothiazide, penicillamine, quinine, metronidazole, isoretinoin, and aspirin. Recently, bilateral angle closure glaucoma with uveal effusions have been associated with topiramate.

Although the mechanism for topiramate induced myopia is unknown, it may partly be from topiramate’s weak carbonic anhydrase inhibitor activity or a prostaglandin mediated effect. Since rechallenging at lower doses does not cause recurrence of myopia, allergic hypersensitivity is unlikely. Uveal effusions with ciliary body swelling cause forward rotation of the lens-iris diaphragm, causing myopia and angle closure glaucoma.

Including the three other cases in the ophthalmic literature which reported angle closure glaucoma, all were women between ages 34 and 53. The onset of high pressures varied from 1 day to 2.5 weeks. Angle closure glaucoma has been noted in a 5 year old (Clark T, personal communication, 19 April 2002).

Although peripheral iridotomy may relieve some pupillary block (as suggested by case 2), most glaucoma cases resolve without miotics or iridotomy. Patients who develop blurred vision should promptly discontinue topiramate to prevent progression to angle closure glaucoma. Pediatric and developmentally delayed patients on topiramate should be closely monitored during the first 2 weeks of treatment.

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**References**


**LASIK induced choroidal infarcts**

Laser in situ keratomileusis (LASIK) is now commonly used for correction of refractive error. Anterior segment complications relating to flap occurs occasionally. However, posterior segment complications are rare. Choroidal infarct following LASIK, to our knowledge, has never been reported. Here we report a case of bilateral choroidal infarct following a routine LASIK procedure.

**Case report**

A 23 years old woman underwent a LASIK excimer procedure to both eyes for correction of hypermetropia. Her preoperative refractive errors were right eye +5.00 and left eye +3.75, improving vision to 6/6 in the right eye and 6/5 in the left eye. However, the axial length was not measured. She had no other ocular pathology. The anterior segments and the fundi were normal. She was generally healthy and her blood pressure was 120/76 mm Hg. Her routine blood tests and chest x ray were within normal limits. She did not have any history of thromboembolic phenomenon, therefore a routine thrombophilia screening test was not performed.

An uncomplicated routine LASIK procedure was performed on 31 August 2001 in both eyes. The keratectomy used was Hansatome. Postoperatively, she complained of reduced and wavy vision, with floats in both eyes. However, 2 months later (3 November, 2001) she had been referred to one us (RJB). On examination her vision was 6/18 with +1.0 D sphi.+1.75D cyl @ 75° and 6/18, with −1.25 D sph.+1.50 D cyl @ 70° in right and left eye respectively. There were normal LASIK scars and no corneal staining. The intraocular pressure (IOP) was 20 mm Hg in both eyes. The anterior chambers were quiet and pupils reacted normally. The retinal examination showed multiple yellow-grey deep retinal lesions at posterior poles in both eyes. In each eye, there was at least one larger triangular shaped lesion measuring approximately 2×3 disc diameters in size temporal to the macula. There were several smaller satellite lesions around the macula, which were somewhat polygonal in shape. These lesions had sharp, well defined borders with a mild degree of retinal oedema. It should be noted that the lesions were almost symmetric in each eye. The discs and blood vessels appeared normal. Fluorescein angiography showed normal transit of dye through the discs and blood vessels. The arteriovenous phase in each eye showed two typical triangular hyperfluorescent areas with apex pointing posteriorly. The smaller polygonal lesions showed punctate hyperfluorescence. The intensity of hyperfluorescence increased with the concentration of circulating dye during the transit and faded away towards the later phases of the angiography. The lesions were situated at a deeper level in comparison with the retinal blood vessels. The late phase in both eyes showed generalised staining of all lesions (Fig 1).

**Figure 1** (A) The arteriovenous phase of the fluorescein angiogram of the right eye shows two punctate hyperfluorescent triangular lesions with apex pointing towards the posterior pole on the temporal side of the macula. There are several smaller lesions above and below the macula. (B) The late venous phase of the fluorescein angiogram of the left eye done at the same session shows a large palquoid lesion on the temporal side of the macula with increased hyperfluorescence due to staining of the lesion.
The patient was treated with 60 mg of oral prednisolone in two divided dosage for 3 weeks which was then reduced to 20 mg twice daily. She was reviewed fortnightly. She was also given flurbiprofen sodium 0.03% as local anti-inflammatory drops to both eyes three times daily for 4 weeks.

After 6 weeks the retinal oedema settled with visible fine pepy pegmentation over the lesions. Fluorescein angiography was repeated which showed increasing hyperfluorescence of all lesions during the transit with minimal staining towards the late phase. On her final review her visual acuities were right eye 6/9 with +1.0 D sph/+0.50 D cyl @ 160° and in left eye 6/6 with +1.0 D sph/-0.50 D cyl @ 180°. The IOP was 18 mm Hg in each eye. The anterior chambers were quiet. The fundus showed well defined fine pigmented lesions with well defined borders. There were no fresh lesions or change in size of earlier lesions.

Comment

Flap related complications following LASIK surgery are well documented. Posterior segment complications such as bilateral macular haemorrhage,3 premacular subhyaloid haemorrhage,4 and macular ischaemic optic neuropathy,5 retinal detachment,6 field defects,7 and macular hole8 have also been reported but to the best of our knowledge this is first case report of choroidal infarcts occurring after LASIK.

During the flap making stage of LASIK, application of suction may increase the IOP to as much as 60 mm Hg or even more. This increase in IOP can result in poor perfusion leading to circulatory imbalance of the retina and the choroid. Alteration of perfusion pressure in susceptible individuals may precipitate clinical features of AION or choroidal infarct. In our case the increase in IOP might have reached much higher or be sustained for a much longer period. This might have been the basis of infarction of choriocapillaris lobules. The smaller lobules coalesce to form larger lesions. Subsequent pigment changes seen were due to changes in the overlying retinal pigment epithelium.

Bilateral almost symmetrical lesions indicate that it could not have occurred exclusively as a result of raised IOP during LASIK. Therefore it is possible that this patient must have had other risk factors and she was susceptible to the effect of raised IOP. The refractive surgeons must be aware of the general physical status of a patient's health while considering keratorefractive surgery where a rise of IOP may be an associated factor.

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A case of Boerhaave’s syndrome associated with raised intraocular pressure

We would like to report a fatal case of post-ematic oesophageal perforation (Boerhaave's syndrome). The condition was attributed to nausea and retching caused by raised intraocular pressure (IOP). We believe the case highlights the importance of early diagnosis and management of such rare cases.

Case report

An 81 year old man presented with a painful right eye. He felt nauseated and was retching. On examination he was found to have 6/18 right eye and 6/12 pinhole 6/5 left eye. The right eye was mildly inflamed and pseudophakic as a result of recent surgery. Right IOP was 58 mm Hg. The left eye showed no abnormality. Past ocular history included complicated right cataract surgery requiring pars plana vitrectomy and removal of lens pseudophakic as a result of recent surgery. Right IOP was 58 mm Hg. The left eye showed no abnormality. Past ocular history included complicated right cataract surgery requiring pars plana vitrectomy and removal of lens fragment 6 weeks previously. Five weeks later he developed an inferior retinal detachment which was repaired by further vitrectomy, cryotherapy, encircling band, and gas tamponade (5F, 20%). Following discharge he had been troubled by nausea and retching. Past medical history included moderate chronic obstructive pulmonary disease and treated essential hypertension. Medical examination was consistent with this. He was admitted and treated with intravenous Diamox and Maxolon, topical Alphagan and Trusopt. IOP was 33 mm Hg 5 hours later. Early the follow- ing morning he had one episode of blood stained vomitus following which he developed sudden retrosternal chest pain and dys- pnoea. GTN spray and Gaviscon did not alleviate his symptoms. On examination he was very distressed and unable to lie flat. Cardiovascular and respiratory examination demonstrated no new pathology. His blood pressure, ECG, and oxygen saturation were normal. Abdominal examination revealed general tenderness and guarding in all quad- rants. Abdominal and chest x rays were unre- markable as was abdominal ultrasound. Ur- gent surgical and medical opinions were sought. At lunch time he developed further sudden severe dyspnoea and became confused. Blood gases revealed severe acidosis with reduced PaO, and PaCO,. Physicians believed this was a mixed metabolic and respiratory acidosis induced by pulmonary thromboembolism. He was anticoagulated and transferred to the high dependency unit. Repeat chest x ray at 20:00 showed a right sided pneumothorax. A chest drain was inserted. The pleuritic fluid sample contained bile, Gram positive cocci, and Gram negative rods. The patient, who by now was in septic shock, was commenced on broad spectrum intravenous antibiotics. Emergency laparotomy confirmed oesophageal perforation. After discus- sion with his family no further interven- tion was deemed justifiable. Our patient died a few hours later.

Comment

Boerhaave syndrome is transmural perfora- tion of the oesophagus to be distinguished from Mallory-Weiss syndrome, non-transmural oesophageal tear also associated with vomiting.1 It is the result of a sudden rise in intraluminal oesophageal pressure pro- duced during vomiting. Pain is the most consis- tent symptom, present in 70–90% of pa- tients, and is usually related to the site of disruption (Table 1).2 Chest x ray suggests the diagnosis in 90% of patients, but immediately after disruption the chest film may be normal.3 Pneumomediastinum, surgical em- physema, mediastinal widening, or a mediastinal air-fluid level must prompt investigation to rule out oesophageal perforation.4 Survival depends upon early recognition and appro- priate surgical intervention. The overall mor- tality rate is approximately 35%. Surgical repair within 24 hours carries 70–75% chance of survival. This falls to 35–50% if surgery is delayed longer than 24 hours and to approxi- mately 10% if delayed longer than 48 hours.5 Surgical intervention is the optimum man- agement in most cases.6 This case highlights two very important points, firstly, the import- ance of adequate IOP control and associated nausea, secondly, although the above case is rare familiarity of the case and its inclusion in the differential diagnosis can be potentially life saving.
Severe bilateral necrotising retinitis caused by *Toxoplasma gondii* in a patient with systemic lupus erythematosus and diabetes mellitus

Ocular toxoplasmosis may be remarkably atypical in situations of evident immunosuppression such as acquired immunodeficiency syndrome, malignancy, and use of chronic immunosuppressive drug therapy. Aggressive forms in immunocompetent hosts are very rare. We present a case of severe, bilateral necrotising retinitis by *Toxoplasma gondii* initially misdiagnosed as an acute retinal necrosis (ARN) syndrome, in a patient with systemic lupus erythematosus (SLE) and diabetes mellitus type 2, who was taking medium dose prednisone.

**Case report**

A 47 year old woman reported a 3 month history of rapid visual loss in the right eye followed by a decrease in her left eye vision 2 months later. Twenty days before the onset of ocular symptoms the patient had a seizure. Her medical history showed a SLE, ocular symptoms the patient had a seizure, followed by a decrease in her left eye vision. A 47 year old woman reported a 3 month history of rapid visual loss in the right eye.

**References**


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**Figure 1**

(A) Fundus appearance of the left eye. (B) Fundus appearance of the right eye. (C) Light micrograph shows necrotic retina with occasional inflammatory cells and *Toxoplasma gondii* cyst (arrow). (D) Light micrograph shows the necrotic retinal pigment epithelium with *Toxoplasma gondii* cysts (arrow) and the choroid with a dense infiltrate of lymphocyte and plasma cells (haematoxylin and eosin, bar = 10 µm). (E) Immunohistochemistry with alkaline phosphatase. Two *Toxoplasma* cysts (arrows) positively dyed pink (bar = 10 µm). (F) Electron micrograph of a paraffin embedded tissue post processed for electron microscopy of the choroidal biopsy shows two *Toxoplasma* cysts with micronemes (inset, arrow) and white spherules with hazy borders amylopectin granules (inset, asterisk). The inset shows higher magnification of a bradizoite, showing typical apical structures. Magnification is represented in standard bars.
Based on these results, aciclovir was discontinued and oral anti-Toxoplasma treatment was initiated with pyrimethamine, sulfadiazine, and folic acid supplementation. After 1 month treatment retinal lesions started to heal, yet no improvement of visual acuity was observed.

Comment

The present case was atypical because of an extensive bilateral necrotising retinitis and obliterative vasculitis, with no pre-existing choroidal scars. Three factors could have conferred a mild degree of immunosuppression to this patient: SLE, diabetes mellitus, and prednisone at medium dose.6 It is recognised that retinal vasculitis and immunosuppression increase the risk of a herpetic6 and, to a lesser extent, a toxoplasmic7 infection. Ocular toxoplasmosis associated with collagen vascular disorders is a rare event8 and has been described after aggressive immunosuppressive therapy. On the other hand, retinal vasculitis occurs in less than 10% of SLE patients, particularly in those presenting with antiphospholipid syndrome, which makes them susceptible to retinal infection. In the present case, laboratory findings consistent with antiphospholipid syndrome were absent. The pre-existence of widespread SLE retinal vasculopathy could be inferred by the active lupus central nervous system disease.9 Therefore we speculate that the active SLE associated with diabetes mellitus and the use of corticosteroid increased the patient’s susceptibility to infection leading to this atypically severe ocular toxoplasmosis.

The presumptive agent was initially diagnosed as *T gondii* based on PCR analysis of a vitreous sample and confirmed by histopathological findings of the chorioretinal biopsy. In the present case, the biopsy was also useful to discharge any tumoral infiltration.10 Although the local production of anti-Toxoplasma antibodies is also considered a useful parameter, the high intraocular total IgG titre found in this case, probably caused by a breakdown of the blood-aqueous barrier, was responsible for the false negative Desmont’s coefficient. The simple observation of *Toxoplasma* cysts is not indicative of active infection. However, the presence of multiple cysts, aside from a positive immunohistochemistry test in a symptomatic patient, allowed an accurate diagnosis.

Taking into account that the fundus characteristics strongly suggested a herpetic acute retinal necrosis, one could interpret the PCR as being a false negative result because of both, either a previous aciclovir treatment11 or a lack of sensitivity. Nevertheless, the disease progression despite antiviral therapy, the absence of any viral particles on electron microscopy, in conjunction with all the evidence pointing to a toxoplastic retinitis makes the possibility of a combined infection still possible, but remote.

Based on the present case that showed an atypically severe form of ocular toxoplasmosis in a patient with a vasculopathy because of collagen and metabolic vascular disorders, we suggest that cases of severe necrotising retinitis be evaluated expeditiously to exclude conditions that mimic ARN. A less invasive vitreal PCR and, when indicated, an early retinocorialdial biopsy may yield a definite diagnosis avoiding any delay in introducing effective treatment.

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References


Idiopathic dilated episcleral veins and increased intraocular pressure

Intraocular pressure (IOP) is dependent on the rate of aqueous production, facility of outflow, and episcleral venous pressure. Increased IOP and visual field loss may result from an elevation of episcleral venous pressure, the causes of which include large vessel venous obstruction (venous sinus thrombosis and superior vena cava syndrome), arteriovenous shunts or fistulas, Sturge-Weber syndrome, scleritis, thyroid related orbitopathy, and orbital tumours.1 Increased episcleral venous pressure may result in elevated IOP and optic neuropathy, even in the absence of these entities. We report a 34 year old man with idiopathic dilated episcleral veins (IDEV) and increased IOP with visual field loss.

Case report

A 34 year old man presented for neuro-ophthalmic evaluation because of decreased vision in the right eye and injection of both eyes. His intraocular pressure (IOP) had been elevated in the right eye more than the left for the past 13 years, reaching 32 mm Hg in the right eye. His eyes appeared red for as long as he could remember, but details of his previous ophthalmic examinations were unavailable. Because of the bilateral injection he saw two other neuro-ophthalmologists, who suspected an arteriomegic fistula. One year before presentation he underwent magnetic resonance imaging (MRI) of the head and orbits and catheter arteriography on two occasions, which were both normal. He had no other previous medical problems, was taking no medications except diazepam for anxiety, and had no history of hypertension, diabetes, or smoking.

On examination, visual acuity was 20/25 OD and 20/40 OS. The right eye was significantly more injected than the left. Intraocular pressure was 26 mm Hg OD and 32 mm Hg OS. The anterior segment was normal and the diagnostic gonioscopy revealed open angles. The fundus examination was normal. A 1 month treatment retinal lesions started to heal, yet no improvement of visual acuity was observed.
drugs, and had no recent history of trauma to the head or neck.

External examination revealed no evidence of cutaneous angiomatosis. Best corrected visual acuity was 20/25 in the right eye and 20/20 in the left eye. Although he correctly identified eight of eight Ishihara pseudoisochromatic colour plates with each eye, he had a 35% subjective red desaturation on the right, along with a rightrelative afferent pupillary defect (RAPD). There was no exophthalmos and no increased resistance to retropropulsion of the globes into the orbits. Dilated episcleral veins were apparent in each eye (Fig 1), while the conjunctival vessels were normal. Intraocular pressure was 22 mm Hg on the right and 16 mm Hg on the left, and the mires of the Goldmann applanation tonometer were normally pulsatile. Gonioscopy revealed bilaterally open iridocorneal angles to the ciliary body band for 360 degrees with blood in the canals of Schlemm in both eyes, greatest within the inferior 90 degrees, but without evidence of neovascularisation or angle recession. Automated perimetry revealed a nasal step on the right and decreased sensitivity inferiorly on the left. Asymmetric cupping of the optic disc, with vertical elongation on the right, was noted on funduscopy (Fig 2).

Orbital colour Doppler imaging revealed normal flow parameters in the ophthalmic and central retinal arteries of each eye. The superior ophthalmic vein was of normal calibre and there was no evidence of reversal of flow in either orbit (Fig 3).

**Comment**

Arteriovenous fistula is the most frequent cause of ocular injection with increased IOP due to elevated episcleral venous pressure.*** The ocular injection results from engorged, arterialized conjunctival vessels, which is different from the injection seen in IDEV. Traumatic high flow fistulas may present with proptosis, eyelid oedema, and conjunctival chemosis, and are often easily identified clinically. Spontaneous, frequently low flow fistulas may present with progressive unilateral or bilateral injection, and may be more difficult to diagnose.

There is, however, an infrequent syndrome of ocular injection and increased IOP which is idiopathic. Although originally described by Minas and Podos, the same entity is termed the Radius–Maumenee syndrome in the German literature.12,13 In one report 10 of 64 patients with glaucoma due to elevated episcleral venous pressure had this syndrome.12 In another report of six patients with unilateral dilated episcleral veins and one patient with bilateral involvement, the IOP in the affected eyes ranged between 21 and 44 mm Hg.14 In all six patients with apparently unilateral involvement the IOP was higher in the eye with the dilated vessels. Episcleral venous pressure was measured to be twice the normal value in the affected eyes of these patients.

Catheter arteriography in these patients is normal, without abnormal arteriovenous communications.15,16 Other adjunctive testing, including neuroimaging, orbital venography, and fluorescein angiography are also normal. Although abnormalities in ocular blood flow have been suggested, the pathogenesis of this condition remains unclear.

Orbital colour Doppler imaging provides a non-invasive means of imaging orbital and ocular blood flow and, in patients with fistulas, characteristically shows reversal of blood flow in a dilated superior ophthalmic vein. The superior ophthalmic veins were of normal calibre in our patient, and both orbits showed flow reversal in the episcleral vessels. Similar findings were reported in the only other patient with this syndrome who underwent orbital colour Doppler imaging, and increased arterial and venous flow in the episcleral vessels was noted in the eye with dilated episcleral vessels; however, the images of the ultrasonography were not published.17 We did not measure the episcleral blood flow in our patient.

Although the cause of this disorder remains elusive it should be distinguished from other disorders, most commonly the arteriovenous fistula (Table 1), which can result in dilated episcleral vessels and increased IOP. In patients with IDEV a congenital abnormality in

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**Table 1** Differentiating features of arteriovenous fistula and IDEV and increased IOP

<table>
<thead>
<tr>
<th>Feature</th>
<th>Arteriovenous fistula</th>
<th>Idiopathic dilated episcleral veins (IDEV)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Onset</td>
<td>Most often acquired</td>
<td>May be congenital</td>
</tr>
<tr>
<td>Predisposing factors</td>
<td>Trauma, hypertension</td>
<td>Possible familial predisposition</td>
</tr>
<tr>
<td>Symptoms</td>
<td>Pulsatile bruit or tinnitus, pain, diplopia</td>
<td>None</td>
</tr>
<tr>
<td>Neurological signs</td>
<td>Ocular misalignment, unilateral or bilateral sixth nerve paresis, oculomotor paresis</td>
<td>None</td>
</tr>
<tr>
<td>External examination</td>
<td>Dilated, corkscrew conjunctival and episcleral vessels, conjunctival chemosis, eyelid oedema, proptosis</td>
<td>Dilated episcleral veins, conjunctival vessels of normal calibre</td>
</tr>
<tr>
<td>Anterior segment</td>
<td>Cataract, iris, and angle neovascularisation with orbital ischaemic syndrome</td>
<td>Normal</td>
</tr>
<tr>
<td>Orbital signs</td>
<td>Bruit, proptosis</td>
<td>None</td>
</tr>
<tr>
<td>Goldmann tonometry</td>
<td>Abnormally pulsatile mires</td>
<td>Normally pulsatile mires</td>
</tr>
<tr>
<td>Funduscopy</td>
<td>Dilated, engorged retinal vessels. Occasionally ocular ischaemic syndrome, with optic disc oedema and macular oedema. Rarely central retinal vein occlusion.</td>
<td>Optic disc cupping</td>
</tr>
<tr>
<td>Imaging</td>
<td>Dilated superior ophthalmic vein or cavernous sinus</td>
<td>Normal</td>
</tr>
<tr>
<td>OCDD</td>
<td>Dilated superior ophthalmic veins with reversal of orbital venous flow</td>
<td>Normal</td>
</tr>
<tr>
<td>Catheter angiography</td>
<td>Arteriovenous fistula</td>
<td>Normal</td>
</tr>
</tbody>
</table>

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**Figure 2** The left optic disc (A) is normal and the right optic disc (B) shows a vertically elongated cup. The retinal vessels are of normal calibre.

**Figure 3** Orbital colour Doppler imaging shows a normal calibre of the superior ophthalmic veins in the right (A) and left (B) orbits. The blue colour represents normal retrograde flow of the orbital veins.
vasculature and familial predisposition have been suggested as the cause; however, some reports suggest that the ocular injection is acquired.¹⁰

Treatments have focused on reducing IOP with aqueous suppression. Filtration surgery has been favoured by some, and in one patient who underwent trabeculectomy the IOP was reduced but the dilated episcleral veins remained unchanged.¹¹ Presumably an alteration in aqueous outflow would preclude the use of IOP lowering agents which increase uveoscleral outflow; however, the earlier reports of IDEV preceded the availability of these agents.

Although no clear aetiology has been identified for IDEV, the findings in this syndrome have been characterised well enough to obviate the need for catheter angiography in patients with the clinical features of this syndrome: venous stauation and familial predisposition have been identified for IDEV, the findings in this syndrome.

Treatments have focused on reducing IOP to avoid delaying the treatment of elevated IOP.

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References


Trypan blue assisted descemetorhexis for inadvertently retained Descemet’s membranes after penetrating keratoplasty

The presence of a retrocorneal membrane following an uneventful keratoplasty is a known entity.¹² While the inflammatory membranes usually disappear with the use of topical steroids,¹³ a YAG laser treatment has been used to make an opening in the opacified retained membrane. We describe a new technique of removal of retained Descemet’s membrane using trypan blue dye.

Case 1

A 27 year old man underwent penetrating keratoplasty in his left eye for congenital hereditary endothelial dystrophy (CHED). The host cornea was initially trephined to 90% of the corneal depth with a 7.5 mm trephine (Storz Ophthalmics, St Louis, MO, USA). The anterior chamber was entered with a blade and the host cut was completed with corneal scissors. An 8 mm donor button was anchored with 16 interrupted sutures (10–0 monofilament nylon). On the first postoperative day, slit lamp biomicroscopy revealed a retained Descemet’s membrane and a supernumerary anterior chamber. The graft was clear with a best corrected visual acuity (BCVA) of 20/40. At 1 week, an additional inflammatory membrane was detected between the retained Descemet’s membrane and iris at 6 o’clock position extending from the inferior margin up to the lower third of the anterior chamber (Fig 1). The inflammatory membrane did not resolve with increased frequency of topical steroid and at 4 weeks we decided to excise these membranes.

The 10 o’clock and the 11 o’clock sutures were removed and the graft host junction was separated. Under air, 0.1 ml of 0.1% trypan blue (Vision blue, DORC, Netherlands) was injected over the host’s retained Descemet’s membrane and was left in contact with it for 10 seconds and then washed out. Both retained Descemet’s membrane and inflammatory membranes were stained with the dye. The inflammatory membrane was first removed using a Utrata forceps. Subsequently, a small flap was created in the retained Descemet’s membrane and the membrane was torn in a circular fashion (Fig 2). The anterior chamber was reformed and sutures were reapplied. On the first postoperative day the graft was clear, the anterior chamber was well formed, and there was no retrocorneal membrane.

Case 2

A 26 year old man underwent penetrating keratoplasty in his right eye for CHED with the same surgical technique and graft size as in case 1. On the first postoperative day, a transparent retained Descemet’s membrane along with formation of a supernumerary chamber was detected on slit lamp biomicroscopy. The graft was clear with no active inflammation in the anterior chamber and BCVA was 20/40.

Based on our experience of case 1 regarding the development of inflammatory membrane, we decided to excise the retained Descemet’s membrane at 1 week using the same technique. Postoperatively, slit lamp biomicroscopy did not reveal any evidence of supernumerary chamber or retrocorneal membrane.

Postoperatively, the patients received 2 hourly 0.1% betamethasone sodium phosphate eye drops, 0.3% ciprofloxacin eye drops four times daily and preservative-free artificial tears eye drops four times daily. At 2 months of follow up, the grafts were clear with a single anterior chamber and the visual acuity of 20/40 was maintained in both the eyes.

Comment

Both these cases with retained host Descemet’s membrane following penetrating keratoplasty were that of CHED. In these cases, longstanding stromal oedema may cause loosening of the attachment of the Descemet’s membrane and predispose its separation from the overlying stroma as has also been reported in a case of interstitial keratitis.¹⁴ Hence, inadvertent, incomplete trephination of such oedematous corneas may cause retention of the host Descemet’s membrane. Although both the patients had good vision and the grafts and retrocorneal membranes were clear, we decided to excise these membranes as an inflammatory membrane started growing in the first case. We thought that the host’s retained Descemet’s membrane is acting as a scaffold and further growth of inflammatory membranes may jeopardise the graft.

Trypan blue is commonly used to stain the anterior lens capsule for capsulorhexis in cataract surgery.¹⁵ As both lens capsule and
Desmet's membrane are basement membranes, we attempted to stain the retained Desmet's layer with trypsin blue to enhance its visibility during the surgery. In both cases an optimal staining of the membranes helped in achieving a controlled circular tearing of the membrane without affecting graft clarity.

We called our technique of dye assisted removal of retained Desmet's membrane “desmemetheresis” as the retained Desmet's membrane were torn in a circular fashion at the graft-host junction.

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References

Orbital haemorrhage complication following postoperative thrombolysis

Thrombolytic agents, such as tissue plasminogen activator (tPA), are commonly employed early in the management of patients with acute myocardial infarction. In this setting the rapid thrombolytic action of tPA is a key advantage over warfarin. However, the risk of intracranial bleeding must be balanced against the potential benefits of the treatment. The complications of thrombolytic therapy include major bleeding, haematoma formation, and cerebral infarction. The use of thrombolytic therapy in the treatment of intracranial haemorrhage is controversial. In this report, we describe the successful use of thrombolytic therapy in the management of a postoperative orbital haemorrhage.

Case report
A 72 year old man underwent uncomplicated lateral injection of local anaesthetic. The patient had no past medical history of a haematological disorder. The patient was admitted to the emergency department with an acute inferolateral myocardial infarct. The attending cardiologist and the patient's ophthalmologist agreed that the benefits of thrombolysis outweighed the risk to the eye and vision. A 10 unit dose of tPA was administered to the patient with a concomitant hepatic infusion; this was followed half an hour later by a second dose. Aspirin was also prescribed.

Two hours after the initial dose of tPA peri-orbital swelling of the right eye was noted. Over the next hour there was increasing periorbital oedema and the patient reported mild ocular discomfort, loss of vision, nausea, and vomiting. During one episode of emesis there was an abrupt onset of frank periorbital haemorrhage. The hepatic infusion was stopped. Examination of the affected eye revealed proptosis, subconjunctival haemorrhage, a retinal patient pupillary defect, a hazy cornea, and severely restricted extraocular movements. Vision was no perception of light. A computed tomograph scan demonstrated a large retrobulbar haematoma in proximity of the lateral rectus muscle (Fig 1). The patient was given intravenous acetazolamide and hydrocortisone. Canthotomy and hydrocelectomy was performed. The following day, after urgent coronary angiography, it was considered unsafe for the patient to proceed to decompression via an extraocular route. The patient's ophthalmologist agreed that the benefits of thrombolysis outweighed the risk to the eye and vision. A 10 unit dose of tPA was administered to the patient with a concomitant hepatic infusion; this was followed half an hour later by a second dose. Aspirin was also prescribed.

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This simple technique is easy to perform even for the first time, and has good repeatability. It is recommended only for soft foldable IOls, and should not be used for rigid one piece poly(methyl methacrylate) (PMMA) lenses. PMMA IOL implantation is usually through an enlarged incision, and it would be simpler to dial out the superior haptic and reimplant the lens. The haptics of PMMA IOLs are also much stiffer than foldable IOls and can damage both the posterior capsule and the endothelium during the flipping process. This manoeuvre is also not recommended for cases where the integrity of the capsular bag is lost or in doubt, as there is a potential for worsening any capsular rent.

The idea of the technique is to cause a rotational movement about the diameter of the IOL optic, similar to flipping a coin, while at the same time protecting the corneal endothelium and posterior capsule. The technique proceeds as follows:

1. The anterior chamber is deepened with viscoelastic to create space between the optic/haptics and the endothelium. The capsule bag behind the optic is distended with viscoelastic to facilitate safe flipping of the IOL.
2. This step flips the IOL. The rotational force on the optic is generated by the careful placement of two intraocular instruments. Non-sharp instruments are preferable, such as a Sinskey hook and mushroom. Their long horizontal segments ensure a controlled rotation of the optic about the central axis. For the right handed surgeon, a Sinskey hook held in the right hand is placed adjacent to the optic, to the right of the optic centre (Fig 1A). To initiate the flip, the mushroom is then raised towards the cornea while simultaneously the Sinskey hook is pushed posteriorly. These movements take place about an axis passing through the diameter of the optic (Fig 1B), forcing the IOL to flip. The haptics then do a 180 degree slow, controlled sweep into the eye. In the AMO Unfolder system, the use of injectable lens haptics and optic. Recognition of the “S” must signal to the surgeon to “stop” and check. Occasionally the trailing haptic may be trapped within the incision by the jaws of the inserting forceps, inducing spontaneous flipping within the eye. Recognised early, this can be aborted by placing the forceps tips over the optic to prevent further rotation, while releasing the trapped haptic with Kelman-McPherson forceps.

Although inverted lens implantation is eminently preventable with proper attention at each step of lens implantation, it still is a problem that occurs with enough frequency to be of concern. Should the surgeon decide that correction of the reversal is appropriate, the IOL flip offers a simple, quick and safe way out of a tight spot.

Acknowledgements

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Goldmann perimetry findings 5 months after commencing antioxidant treatment. The average central response shows a marked improvement to 95 nV and the average peripheral has improved to 83 nV. The delay is reduced by 12–54 ms. (D) Shows the mfERG of the patient's right eye performed 1 year later, 6 months after starting the antioxidant preparation. The average central response is reduced to 21 nV and the average peripheral response is 30 nV. The implicit time is normal.

Birdshot chorioretinopathy (BSCR) is a rare inflammatory disease, which generally follows a chronic course of progressive loss of vision to 6/60 or worse. However, the vision may stabilise or, rarely, improve slightly. Commonly prescribed treatment regimens, including oral steroids, and one or two immunosuppressive agents, may stabilise but generally do not cure the condition.

We describe a case of BSCR in which there was a marked improvement in Snellen acuity and retinal function, measured with wide field multifocal electroretinography (WF-mERG), in a patient intolerant of conventional treatment, who self medicated with an antioxidant preparation.

Case report
A 53 year old white woman was referred with a 2 year history of decreased vision and floaters affecting both eyes. Snellen acuity was 6/24, N8 in the right eye and 6/12, N8 in the left eye. Moderate vitritis, cystoid macular oedema, swelling of the optic discs, and the presence of scattered, deep, pale creamy white lesions, were compatible with a diagnosis of BSCR, supported by positive HLA-A29 serology.

A reducing dose of oral prednisolone in conjunction with cyclosporin was commenced. There was no improvement in visual symptoms and, owing to the development of cellulitis around the umbilicus, cyclosporin was stopped.

She was referred to the regional uveitis service. WF-mERG was performed at presentation, using a custom built system comprising a 61 hexagonal array. This test facilitates localised assessment of retinal function, within a 90° retinal field. WF-mERG showed a global reduction of P1 amplitude responses in both eyes (Fig 1B). A lower dose of cyclosporin was tried but subsequently discontinued because of significant gastrointestinal symptoms.

Over the next 18 months, tacrolimus, azathioprine, and mycophenolate were all prescribed separately in conjunction with a maintenance dose of 5 mg of prednisolone. Tacrolimus 1 mg twice daily was discontinued after 2 weeks because of intolerable side effects. Treatment with azathioprine 50 mg three times daily continued for 7 months with no improvement in vision. Mycophenolate 1 mg twice daily was stopped at the patient's request after a year, owing to progressive deterioration of vision and a general feeling of malaise. In addition, prednisolone was tailed.

**Figure 1** Multifocal electroretinogram (mfERG) of the patient's right eye compared to a normal response. (A) Shows a normal mfERG; the central and peripheral responses are grouped and averaged. The normal range of the central response is 74–122 nV and the normal range of the peripheral response is 61–108 nV. The normal range for mfERG implicit times is 32–42 ms. The ranges are the 5% and 95% confidence limits derived from 50 age-matched controls. (B) Shows the mfERG of the patient's right eye performed in 1999 at presentation to the regional uveitis clinic. The average central response is 30 nV and the average peripheral response is 31 nV. (C) Shows the mfERG of the right eye performed 1 year later. The average central response is reduced to 21 nV and the average peripheral response is 30 nV. The implicit time is delayed by 12–54 ms. (D) Shows the mfERG of the patient's right eye performed 1 year later, 6 months after starting the antioxidant preparation. The average central response shows a marked improvement to 95 nV and the average peripheral has improved to 83 nV. The implicit time delay has also improved by 6 ms to 48 ms. (E) Goldmann perimetry findings before commencing antioxidant treatment. (F) Goldmann perimetry findings 5 months after commencing antioxidant treatment.

**References**

off and stopped after 20 months of steroid treatment. The visual acuity at this stage was 6/36, N14 and 6/12, N14, and the patient was taking no conventional medication. The amplitude and implicit time of the mfERG responses recorded a year after presentation had deteriorated further (Fig 1C).

Three months after discontinuing all conventional treatment, the patient began to self medicate with an antioxidant preparation, the contents of which included white and marigold petals, grape seed extract, and vitamins C and E (Table 1). After 6 months, Snellen acuity had improved to 6/15, N10 in the right eye and 6/9, N8 in the left. In addition, the amplitude of the central WF-mfERG responses had improved from 21 nV to 95 nV in the right eye and from 35 nV to 167 nV in the left eye (normal range 74–122 nV). The amplitude and implicit time of the average peripheral WF-mfERG responses had also improved in both eyes (Fig 1D).

She continued on the antioxidant regimen and 6 months later, the visual acuity and retinal function in both eyes, recorded with the WF-mfERG, had remained stable.

Comment
WF-mfERG investigations showed a marked improvement in visual function of both eyes after commencing the antioxidant preparation. This observation mirrored the recovery of Snellen acuity but did not correlate with Goldmann perimetry findings (Fig 1E and F).

Antioxidants are thought to scavenge free radicals produced in the retina following light absorption, and thus prevent cellular damage that they may produce. The pathophysiology of BSCR is unclear; therefore the role of antioxidants in reversing the damage caused by BSCR is purely speculative.

In this case it is unclear whether the improvement in visual function, recorded with WF-mfERG was spontaneous, whether it was a late result of previous conventional treatment, or whether it was secondary to the commencement of a complex non-prescribed antioxidant preparation. The role of antioxidant treatment in BSCR needs further appraisal, perhaps using the HLA-A29 transgenic mouse, which has been shown to spontaneously develop a retinopathy that is histologically very similar to that found in BSCR.

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References

Central retinal venous occlusion with co-existent thrombotic thrombocytopenic purpura and antiphospholipid syndrome

Central retinal venous occlusion usually occurs in elderly patients with known risk factors such as raised intraocular pressure, arterial hypertension, atherosclerotic disease, or diabetes mellitus. In addition, central retinal venous occlusion infrequently occurs in individuals with hyperviscosity from leukaemia, polycthyma, Waldenstrom’s macroglobulinaemia, sickle cell disease, and periphlebitis. However, it has also been described in patients with antiphospholipid antibodies and rarely in patients with thrombotic thrombocytopenic purpura (TTP) (pentad of fever, microangiopathic haemolytic anaemia, thrombocytopenia, neurological abnormalities, and renal impairment). We describe central retinal venous occlusion in a 26 year old female patient with systemic lupus erythematosus (SLE), in whom both TTP and antiphospholipid syndrome (APS) appear to have contributed to the pathogenesis. To our knowledge, co-existence of TTP and APS has not been previously reported in a case of central retinal venous occlusion.

Case report
The patient presented in 1994, aged 19, with arthralgia and fatigue. Haemoglobin was 3.5 g/dl and platelet count 15 × 10^9. Investigations revealed SLE (antinuclear antibodies were positive at a titre of 1:640 and anti-double stranded DNA antibodies were 1:320)
with Evans’ syndrome (combined auto-immune haemolytic anaemia and immune thrombocytopenia). Tests for APS were not performed. The patient responded to corticosteroids and azathioprine, which were reduced and finally stopped in April 1997.

She returned 1 week until hospitalised in February 2001 with fever of 38°C, gross haematuria, malaise, poor concentration, and headache. Haemoglobin was 7.5 g/dl (Direct Coomb’s test negative) and platelet count was 1 x 10^9/l. Serum creatinine was 244 µmol/l and activated partial thromboplastin time was prolonged at 93 seconds. IgG antcardiolipin antibodies were elevated (18 GPL units/ml) and thrombocytopenia was detected by a positive kaolin clotting time index and a dilute Russell viper venom test. C3 and C4 complement components were reduced to 6.6 g/l (normal, 7.5–17 g/l) and 0.4 g/l (normal, 1.4–5.4 g/l) respectively, indicating activation of the classic complement pathway by immune complexes.

She was treated with intravenous antibiotic therapy and, for the presumptive diagnosis of immune thrombocytopenia, high dose corticosteroids. Early next morning, she complained of sudden onset loss of vision in the right eye. On ophthalmic examination, haemorrhagic central retinal vein occlusion was noted in the right eye and 6/6 in the left eye. She had a dense right relative afferent pupillary defect and right fundal examination revealed completely frosted retinal veins and a severe haemorrhagic central retinal vein occlusion with diffuse superficial and deep retinal haemorrhages (Figs 1 and 2). As the platelet count remained low at 2 x 10^9/l, a 3 day course of intravenous immunoglobulins (400 mg/kg/day) was commenced. On the third day of hospitalisation, vision in the right eye had not improved and the platelet count remained <5 x 10^9/l. Review of daily blood films revealed increased numbers of fragmented red cells (schizocytes) while serum LDH was 2049 units/l (normal <450 units/l). A diagnosis of TTP secondary to SLE was made and treatment with 1 litre of cryoprecipitate poor fresh frozen plasma (FFP) twice daily was started. Within 36 hours, the platelet count had risen to 39 x 10^9/l and asparin and azathioprine were added. After a week’s therapy with cryoprecipitate poor FFP platelet count, serum creatinine and serum LDH were all within normal range with normal blood film.

Three months later, the right eye had developed retubed oedema, raised intraocular pressure of 34 mm Hg, disc new vessels, and early vitreous haemorrhage. Following treatment with topical β blockers and multiple sessions of panretinal argon laser photocoagulation, the disc new vessels and retubed oedema resolved but visual acuity remained poor at hand movements. Ten months later, the patient is taking prednisolone, azathioprine, and aspirin without evidence of TTP but without recovery of vision in the right eye.

**Comment**

In a recent review, antcardiolipin antibodies were detected in eight of 17 patients with SLE and antphospholipid antibodies therefore, may contribute to the pathogenesis of some cases of TTP in association with SLE, possibly by causing endothelial damage in the microvasculature. TTP is a rare but recognised association with central retinal vein occlusion’ while a high prevalence of anticardiolipin antibodies has been reported in patients with vaso-occlusive retinopathy exempt from conventional risk factors. However, to our knowledge, this present case of central retinal venous occlusion is unique because of co-existent APS and TTP secondary to SLE. Although aggressive immunosuppressive therapy, aspirin and cryoprecipitate poor FFP led to control of APS and TTP, the severe visual impairment due to central retinal venous occlusion failed to recover.

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**REFERENCES**


**MAILBOX**

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Further considerations of retinopathy with renal failure

We were interested to read the letter by Patel et al reporting the unusual and striking fundus appearance of retinal arterial calcification in association with chronic renal failure. However, we feel that two important points have been omitted from the comment, which merit further discussion.

Firstly, Monckeberg’s sclerosis, which in its classic form is characterised by “pipe stem” calciﬁcation deposition in the medial coat of muscular arteries in middle aged and elderly individuals, is described with widespread systemic distribution. Interestingly though, a juvenile form is seen, particularly in association with chronic renal failure and diabetes. The pathological mechanism of Monckeberg’s scleroderma is uncertain, but is thought to encompass elements both of dystrophic and metastatic calcification. In the reported case, it would be of interest to know if there was any evidence of vascular calcification elsewhere in this individual, or whether it was localised to the retinal arteries only.

Secondly, Senior-Loken syndrome (renal-retinal syndrome, nephropathosis associated with retinitis pigmentosa, or retinal aplasia) is also germane to this discussion. Nephropathosis is a major cause of progressive medullary cystic renal disease leading to chronic renal failure in adolescents. It is associated with a variable retinal phenotype, a reflection of both clinical and genetic heterogeneity. When associated with retinitis pigmentosa, there is an absence of the classic bone spicule pigmentation, but optic nerve head pallor and attenuation of the blood vessels is seen. Also, electroretinographic alterations have been reported in nephropathosis despite normal or a normal fundus examination. In view of the phenotypic overlap seen with the reported case, results of electoretinography would be pertinent to the discussion. We would also advocate a molecular genetic analysis at the known loci associated with Senior-Loken syndrome, which could potentially reveal an interesting new allele in this case. Finally, while the fundus photograph elegantly demonstrates the stark changes seen in the retinal vasculature; this evidence could be further augmented by the addition of an ultrasonic B-scan image demonstrating the pathophenocognomic echogenic features of intraretinal calcification.

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**References**

Comments on myopia in secondary school students in Tanzania

I refer to the interesting article by Wedner et al published recently in the BJO. The authors have concluded, without enough justification, that “an eye screening programme for significant refractive errors is indicated in this population.” I believe more data are required for such a conclusion.

Weft et al could have reported the true positive predictive rate for the detection of myopia using the stated referral criterion of “worse than 6/12” on the Snellen visual acuity chart: 141/174 or 81% (95% CI: 74 to 86). However, they have referred to the prevalence of “significant” myopia being 141/2511 or 5.6%. In epidemiological convention it is incorrect to apply this denominator and report this prevalence rate because the reference standard for the determination of refractive errors—that is, refraction, has not been performed in the majority (2337) of the 2511 students. The positive predictive value of a screening test is influenced by the prevalence of the condition to be screened (that is, myopia). Nevertheless, without experimental data on the false negative and false positive predictive rate for the detection of ‘significant’ myopia being 141/2511 or 5.6%. Out of 29 eyes, nucleus hardness was grade 3 or more in 17 eyes in our series. It would be worthwhile to know the nuclear hardness in the series reported by Kageyama et al as the manoeuvres in hard cataract is more difficult to perform with the non-dominant hand.

The mean ultrasound power used in our cases was 25.7% (9.5%) and the mean effective phacoemulsification time was 18 (11) seconds. No posterior capsular tear or vitreous loss was present in our cases and this could be attributed to the fact that an experienced surgeon performed the surgery. However, were there any anterior capsular tears noted by Kageyama et al, which occurred in two eyes in our series during the chopping procedure? Nevertheless, none of these cases had a posterior extension of the tear with vitreous loss. Twenty seven eyes (93.1%) achieved a best corrected visual acuity of 20/20 at 4 weeks after the surgery, while two eyes had a best corrected visual acuity of 20/40 because of the presence of age related macular degeneration. It would be worthwhile to know the amount of astigmatism preoperatively and postoperatively reported by Kageyama et al, as one of the advantages of a temporal incision phaco is less surgically induced astigmatism. The mean preoperative keratometric ATR astigmatism was 0.54 (0.44) D (range 0.25 to 2 D) and the mean postoperative ATR astigmatism was 0.25 (0.64) D (range −0.75 to 1.75D) (Wilcoxon signed rank test, p = 0.0267) in our series. We had a similar mean endothelial loss of 6.02% at the end of 12 weeks.

Clear corneal non-dominant hand phacoemulsification is indeed a safe and efficacious method. This obviates the need for a surgeon to change position between cases. In addition, the dominant hand is utilised for chopping, which makes the surgeon the most active instrument, while the non-dominant hand holds the ultrasound hand piece as a passive instrument in the centre of the capsular bag. This helps to reduce the use of ultrasound energy for nuclear management, as the chopper is the instrument used for mechanical disintegration of the nucleus.

References


Left handed non-dominant hand phacoemulsification

We appreciate the commendable work by Kageyama et al, which was published in the BJO.1 We have also studied the results of left handed non-dominant hand phacoemulsification prospectively, performed in left eyes of 29 patients with an immature senile cataract and against the rule astigmatism. Phacoemulsification was performed by an experienced surgeon (RBV) under peribulbar anaesthesia. The surgeon sat at the head end of the patient while performing the operations (on left eyes). In all the cases, a 3.2 mm clear corneal temporal incision was made with a diamond knife held in the left hand of the surgeon. Following a continuous curvilinear capsulorhexis and hydroprocedures a phaco chop nucleotomy technique was used using Storz Pro-tégé phaco machine. The chopper was held in the right hand of the surgeon and inserted from the 12 o’clock side port and the phaco probe was held in the left hand of the surgeon and introduced from the main incision on the temporal side. This was followed by automated irrigation aspiration and implantation of an Acrysof foldable intraocular lens (Alcon MA 60 BA) in the capsular bag.

In our study of 29 patients the mean age of the patients was slightly younger, 63.50 (SD 9.61) years (range 43–80 years), than patients of Kageyama et al. Out of 29 eyes, nucleus hardness was grade 3 or more in 17 eyes in our series. It would be worthwhile to know the nuclear hardness in the series reported by Kageyama et al as the manoeuvres in hard cataract is more difficult to perform with the non-dominant hand.

The mean ultrasound power used in our cases was 25.7% (9.5%) and the mean effective phacoemulsification time was 18 (11) seconds. No posterior capsular tear or vitreous loss was present in our cases and this could be attributed to the fact that an experienced surgeon performed the surgery. However, were there any anterior capsular tears noted by Kageyama et al, which occurred in two eyes in our series during the chopping procedure? Nevertheless, none of these cases had a posterior extension of the tear with vitreous loss. Twenty seven eyes (93.1%) achieved a best corrected visual acuity of 20/20 at 4 weeks after the surgery, while two eyes had a best corrected visual acuity of 20/40 because of the presence of age related macular degeneration. It would be worthwhile to know the amount of astigmatism preoperatively and postoperatively reported by Kageyama et al, as one of the advantages of a temporal incision phaco is less surgically induced astigmatism. The mean preoperative keratometric ATR astigmatism was 0.54 (0.44) D (range 0.25 to 2 D) and the mean postoperative ATR astigmatism was 0.25 (0.64) D (range −0.75 to 1.75D) (Wilcoxon signed rank test, p = 0.0267) in our series. We had a similar mean endothelial loss of 6.02% at the end of 12 weeks.
Second Sight
Second Sight, a UK based charity whose aims are to eliminate the backlog of cataract blind in India by the year 2020 and to establish strong links between Indian and British ophthalmologists, is regularly sending volunteer surgeons to India. Details can be found at the charity website (www.secondsight.org.uk) or by contacting Dr Lucy Mathen (lucymathen@yahoo.com).

Specific Eye Conditions (SPECS)
Specific Eye Conditions (SPECS) is a not for profit organisation which acts as an umbrella organisation for support groups of any conditions or syndrome with an integral eye disorder. SPECS represents over fifty different organisations related to eye disorders ranging from conditions that are relatively common to very rare syndromes. We also include groups who offer support of a more general nature to visually impaired and blind people. Support groups meet regularly in the Boardroom at Moorfields Eye Hospital to offer support to each other, share experiences, and explore new ways of working together. The website www.eyeconditions.org.uk acts as a portal giving direct access to support groups’ own sites. The SPECS web page is a valuable resource for professionals and may also be of interest to people with a visual impairment or who are blind. For further details about SPECS contact: Kay Parkinson, SPECS Development Officer (tel: +44 (0)1803 524238; email: k@eyeconditions.org.uk; web site: www.eyeconditions.org.uk).

The British Retinitis Pigmentosa Society
The British Retinitis Pigmentosa Society (BRPS) was formed in 1975 to bring together people with retinitis pigmentosa and their families. The principle aims of BRPS are to raise funds to support the programme of medical research into an eventual cure for this hereditary disease, and through the BRPS welfare service, help members and their families cope with the everyday concerns caused by retinitis pigmentosa. Part of the welfare service is the telephone help line (+44 (0)1280 860 363), which is a useful resource for any queries or worries relating to the problems retinitis pigmentosa can bring. This service is especially valuable for those recently diagnosed with retinitis pigmentosa, and all calls are taken in the strictest confidence. Many people with retinitis pigmentosa have found the Society helpful, providing encouragement and support through the help line, the welfare network, and the BRPS branches throughout the UK. (tel: +44 (0)1280 821 334; email: lynda@brps.demon.co.uk; web site: www.brps.demon.co.uk)

Surgical Eye Expeditions International
Volunteer ophthalmologists in active surgical practice are needed to participate in short term, sight restoring eye surgery clinics around the world. Contact: Harry S Brown, Surgical Eye Expeditions International, 27 East De La Guerra, C-2, Santa Barbara, CA 93101-9858, USA (tel: +805 963 3303; fax: +805 965 3564; email: hsbrown.md@cox.net or seeintl@seeintl.org; web site: www.seeintl.org).

XXXII Retinal Detachment Course
The XXXII Retinal Detachment Course with International Faculty and Case Presentations will be held 5–6 June 2003, in Beijing, China. The Congress language is English with simultaneous translation in Chinese. Further details: Scientific programme: Prof Ingrid Kreissig, University of Tuebingen, Schleichstr. 12, Breuningerbau, 72076 Tuebingen, Germany (tel: +49 7071 295209; email: ingrid.kreissig@med.uni-tuebingen.de). Local organisation: Ningli Wang, Liang Xu, Wu Liu. Congress office: Wu Liu, Beijing Tong Ren Eye Center, Beijing 100730, China (email: wulujb@yahoo.com).

Detachment Course with international faculty on: Retinal and Vitreous Surgery with Case Presentations preceding the Annual Meeting of Iranian Society of Ophthalmology
The detachment course with international faculty on: Retinal and Vitreous Surgery with Case Presentations preceding Annual Meeting of Iranian Society of Ophthalmology will be held on 29–30 November 2003 and 1–4 December 2003 respectively, at the Razi Conference Center, Hemmat Hyw, Tehran, Iran. Further details: Scientific programme: Prof Ingrid Kreissig, University of Tuebingen, Schleichstr. 12, Breuningerbau, 72076 Tuebingen, Germany (tel: +49 7071 295209; email: ingrid.kreissig@med.uni-tuebingen.de.). Local organisation: Dr Arman Masheyekhi, Dr Siamak Moradian, Dept of Ophthalmology, Labbafinejad Medical Center, Pasdaran Ave, Boostan 9, Tehran, 16666, Iran (fax: +98 21 254 9039; email: labbai@hotmail.com).
LASIK induced choroidal infarcts

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