LETTERS TO THE EDITOR

Laceration of the eye with a fishing hook

EDITOR,—Perforation of the eye is a challenging emergency in ophthalmology and requires immediate treatment. Visual outcome after penetrating injuries with and without intraocular foreign bodies depends on the visual acuity after injury, age of patient, and the severity of the ocular trauma.1 We report a case of globe laceration following an accident with a fishing hook which was successfully treated without surgery.

CASE REPORT

A 12 year old boy presented with visual disturbance and a small lid wound on the left eye at our emergency department. He had been playing with a fishing rod while sitting on a tree, and was trying to hook fruit on the ground. The line recoiled swiftly and pierced the upper lid of the left eye. He jumped from the tree and the hook jerked itself out of the boy’s eye. A small wound of the left upper lid was found. The visual acuity was 20/20 in the right eye and 20/25 in the left eye. The anterior segment and the intraocular pressure were normal. The fundus of the right eye was normal. The fundus of the left eye showed a mild localised vitreous haemorrhage superiorly near the temporal vessels. From there the scleral rupture went straight up to the posterior vitreous and the arrow shows the area of vitreous incarceration into the scleral wound. The optic nerve is marked with the asterisk.

The ultrasound disclosed a partly detached vitreous with adherence at the site of penetration (Fig 1A). No foreign body was detected by the orbital computed tomograph scan. The patient was admitted to hospital for 8 days and treated with intravenous antibiotics during 1 week followed by an oral antibiotic in the second week. He also received cortisone systemically for 10 days. Three weeks later the visual acuity was 20/20. On funduscopy a sclerochoroidal scar was disclosed. The edges of the retinal tear were attached and vitreous haemorrhage was completely resolved (Fig 2). After 1 year of follow up the situation remained unchanged.

COMMENT

Although penetrating injuries of the posterior segment often require surgical treatment (that is, pars plana vitrectomy), in this case antibiotic therapy was used in order to prevent an endophthalmitis in combination with orally administered cortisone to reduce the inflammatory reaction. Because of an excellent fundus view, postponing surgery seems to be more appropriate, since surgery implies additional risks (for example, cataract formation, retinal detachment, etc) for the eye.1 Significant predictors for a final visual acuity of 20/50 or better are a visual acuity of 20/800 or better and youth (≤ 18 years).1 In a mammalian study it was disclosed that simple penetration of the equator with vitreous loss does not lead to retinal detachment.1 An intact tamponading vitreous at the time of injury seems to prevent fibrous ingrowth due to anti-proliferative effects of the hyalocytes.1 The findings of this case suggest that surgery is not the first treatment strategy for similar penetrations of the posterior segment.

RALF KROTT
K ULRICH BARTZ-SCHMIDT
KLADIUS HEIMANN
Department of Vitreoretinal Surgery, University of Cologne, Germany

Correspondence to: Ralf Krott, MD, Department of Vitreoretinal Surgery, University of Cologne, Joseph-Stelzmann-Strasse 9, D-50924 Cologne, Germany.

Accepted for publication 11 June 1999


Intraocular infestation with the worm, Thelazia callipaeda

EDITOR,—Ocular infections with helminthic parasites are well described. The commonest of these are flarial worms that reside in subcutaneous tissue, and are found as skin infestations or masses in the lids. Some are known to live freely in the conjunctival sac. Worms that are visible to the naked eye are often referred to as “eyeworms”, and are in the larval or adult stage of their life cycle. Thelazia callipaeda, or the oriental eye worm, is a spiruroid nematode which is the causative organism in theliaisis, a well described condition affecting the external eye.1 It is primarily a parasite of the conjunctiva in dogs, and is also found in rabbits and humans.2 Its presence in the conjunctival sac causes laceration and irritation, and its frequent excursions across the cornea may cause marked discomfort and, eventually, corneal scarring. The worm also causes paralytic ectropion through its presence in the lower fornix. At least 40 cases of infection in humans have been described from China, Japan, India, Russia, Thailand, and Korea. All of these report extracocular infection only. We report the first case of Thelazia callipaeda to cause intraocular infection.

CASE REPORT

A 21 year old Chinese woman presented to the ophthalmology department at Shantou Central Hospital, Guangdong, China, complaining of decreased vision in her right eye and a floater in the right visual field. She had no photopsia or field loss, and no pain, redness, or discharge. She had no past ocular or medical history, and was otherwise well.

On examination she had a visual acuity of 6/60 in the right eye, and 6/6 in the left eye. There was no external evidence of trauma; the lids were healthy, the conjunctiva white, the cornea clear, and the anterior chamber quiet. There was no photophobia or field loss, and no pain, redness, or discharge. She had no past ocular or medical history, and was otherwise well.

On examination she had a visual acuity of 6/60 in the right eye, and 6/6 in the left eye. There was no external evidence of trauma; the lids were healthy, the conjunctiva white, the cornea clear, and the anterior chamber quiet. A mild vitreous haze obscured the foveal reflex, and clearly visible within the vitreous cavity was a live, mobile, white worm. There was no retinal abnormality.

One month later she underwent a three port pars plana vitrectomy, and the worm was coaxed into a flute needle and removed intact. The patient made an uncomplicated recovery from surgery. At 6 weeks postoperatively the eye was quiet and she had a visual acuity of 6/24. On detailed examination, the worm was identified as an adult female specimen of Thelazia callipaeda. Treatment with ivermectin is recommended for cases of theliaisis, but in the absence of preoperative and postoperative
Thelazia callipaeda (magnification ×10).

Figure 1 Light micrograph of Thelazia callipaeda (magnification ×10).

Thelazia callipaeda (magnification ×1000). (B) Electron micrograph of head end of Thelazia callipaeda (magnification ×1000).

Figure 2 (A) Electron micrograph of tail end of Thelazia callipaeda (magnification ×1000). (B) Electron micrograph of head end of Thelazia callipaeda (magnification ×1000).

Macular dystrophy of malattia leventinese. A 25 year follow up

EDITOR,—Macular degeneration is a clinical term used to describe a variety of diseases characterised by progressive loss of central vision associated with abnormalities of Bruch’s membrane and the retinal epithelium. This dominantly inherited disorder, characterised by a radial pattern of innumerable small elongated basal laminar drusen, was initially reported in a family from the Leventine Valley (Switzerland). The gene responsible for autosomal dominant malattia leventinese has been mapped to the short arm of chromosome 2p16–21. We report the case of a woman who developed unusual complications associated with this disease during a 25 year follow up.

CASE REPORT

In 1981, subfoveal neovascularisation led to an irreversible decrease in visual acuity in her right eye, down to 20/1000 (Fig 1B). In 1996, a dense right vitreous haemorrhage led to a further decrease in acuity. After resorption, fundus examination disclosed an advanced stage of the macular disease with irregular subretinal metaplasia, hyperplasia of the retinal pigment epithelium and discrete radial basal laminar drusen (Fig 2A). The fundus also showed a wedge-shaped superotemporal area with intraretinal haemorrhages, hard exudates, and sheathed vessels. The fluorescein angiogram showed telangiectatic vessels, shunt vessels, and microaneurysms, in addition to neovascularisation (Fig 2B). After laser photocoagulation, no further intraretineal haemorrhage episode occurred. The left eye had a visual acuity of 20/30 and fundus examination revealed a macula identical to that of the right eye, without complication.

COMMENT

In malattia leventinese, the maculopathy is characterised by a radial pattern of innumerable small elongated basal laminar drusen. This maculopathy has been described in a 15 year old patient. The visual acuity of patients suffering from malattia leventinese, however, remains good for a quite a long time. Thus, most patients are asymptomatic until the fourth or fifth decade of life, at which point they have a variety of symptoms, including decreased visual acuity, paraclinical scotomas, photophobia, and metamorphopsia. The main complication is macular subretinal neovascularisation, reported in some patients. In our present case, such macular subretinal neovascularisation caused a severe decrease in visual acuity down to 20/100 in 1981. The contralateral eye was unaffected, fortunately conserving an acuity of 20/30.
Acute central retinal vein occlusion successfully treated with intravenous thrombolysis

Editor,—Central retinal vein occlusion (CRVO) is a condition which often has profound effects on vision. At present there is little to offer patients in the form of treatment to preserve vision. The visual outcome is largely determined by the severity and duration of the vein occlusion. Management is currently aimed at preventing the complications secondary to retinal ischaemia. The incidence of fellow eye involvement with CRVO is believed to be in the order of 1%. We present a patient with “second eye” CRVO, who presented with acute reduction in vision and who responded dramatically to intravenous streptokinase.

CASE REPORT
A 75 year old white man noticed a sudden reduction in vision in his right eye while walking up a hill. He already had poor vision in his left eye from a CRVO 6 years earlier. He went immediately to eye casualty and was seen within 2 hours from the onset of symptoms. His only risk factor for vein occlusion was ocular hypertension treated with timoptol 0.25% twice daily to both eyes.

On examination, visual acuities were 6/36 right eye and counting fingers left eye, there was no relative afferent pupillary defect and intraocular pressures were 21 mm Hg in both eyes. Examination of the right fundus revealed scattered tiny blot haemorrhages and tortuous veins.

A fundus fluorescein angiogram showed pulsatile arterial filling with venous filling delayed until 34.6 seconds (Fig 1). A diagnosis of acute central retinal vein occlusion was made. Because of the previous left CRVO and the short history of symptoms in his right eye, the option of thrombolysis was carefully discussed with the patient, including the small risk of cerebral haemorrhage. Streptokinase, 1.5 x10^6 units, was infused 7 hours after the onset of symptoms. Within 30 minutes, his visual acuity had improved to 6/9. The patient was empirically anticoagulated with heparin and warfarin, keeping the international normalised ratio between 2 and 3. Repeat fluorescein angiogram showed marked improvement in venous filling and loss of pulsatile arterial filling. In addition, several post thrombolysis haemorrhages were evident (Fig 2). The patient took warfarin for 9 months in total and now remains on aspirin alone. His vision in the right eye remains at 6/9.

COMMENT
Central retinal vein occlusion can profoundly affect vision and lead to neovascular complications. Current therapeutic options are limited mainly to the prevention or treatment of secondary complications.

Evidence for thrombus formation in CRVO has been reported by Green et al in a prospective histopathological study of patients with CRVO. They demonstrated recanalised thrombus in 89.7% of eyes and fresh thrombus formation in the remaining 10.3%.

Animal studies have shown encouraging responses to thrombolysis in experimental

Figure 1 Fundus fluorescein angiogram of the right eye demonstrating early venous filling 34.6 seconds after injection.
CRVO. Fibrinolysis given within 2 days of experimental vein occlusion in rabbits was shown to result in resolution of retinal haemorrhages and return of normal retinal circulation within 48 hours. Similar animal studies using recombinant tissue plasminogen activator shortly after experimentally induced vein occlusion resulted in significant retinal vein patency in treated eyes. In one randomised controlled clinical trial, patients with CRVO who received streptokinase followed by full anticoagulation within 7 days of onset of visual symptoms, showed a statistically significant improvement in visual acuity. Late presentation was identified as a possible cause of limited improvement in some cases. Reports of vitreous haemorrhage occurring during treatment (3/20 cases) together with the reported incidence of cerebral haemorrhage with streptokinase (57/13607 cases) account for the absence of a defined role of this drug in the treatment of vein occlusions.

Selective cannulation of a branch retinal vein and infusion of tissue plasminogen activator in a patient with a non-acute CRVO in the second eye has been described. Several other treatment modalities had already been tried unsuccessfully. While avoiding systemic complications, the treatment failed to show any objective improvement in visual acuity and the patient subsequently went on to develop angle neovascularisation.

Clearly many vein occlusions present late, where irreversible retinal damage has occurred. We propose that only in those circumstances where the presentation is acute should the use of intravenous thrombolysis be considered.

Retinitis pigmentosa with visual fluctuations and arrestin gene mutation

Editor,—We report a case of retinitis pigmentosa with day to day visual fluctuations and a mutation in the arrestin gene.

CASE REPORT
A 45 year old Japanese man first noticed night blindness at junior high school age, followed by a slowly progressive loss of visual acuities and fields. At presentation, the best visual acuity was 10/200 in each eye. Goldmann perimetrices revealed generalised narrowing of the peripheral field and marked loss of central visual sensitivities. Bright flash electroretinogram (ERG) in a fully dark adapted state was not recordable in either eye. Ophthalmoscopics revealed advanced stage of retinitis pigmentosa (Fig 1). There was no Mizuo’s phenomenon in the fundus. This patient reported that he had felt relatively better vision (“good day”) on every other day that alternated with worse vision (“bad day”).

Furthermore, he claimed that bad day was repeated after physical stress or alcohol drinking, followed by a good day to return to the ordinary cycle. In our 10 years’ observation, the profile of day to day variation was reproducible. To quantitatively assess the characteristic visual symptoms, perimetric tests were performed in both the centre and periphery using an automated perimeter on eight different days in a period of 2 months, half on a good day and the other half on a bad day. ERG examinations were also performed. Figures 2 and 3 illustrate the results, demonstrating that the visual sensitivity in the peripheral field varied in a manner consistent with the patient’s reports, although not obvious in the central field. In contrast with...
Figure 4 Direct sequencing results of exon 11 of the arrestin gene in the reported case, demonstrating a homozygous deletion of adenine at nucleotide 1147 (arrowhead).

Orbital Kimura’s disease in a white child

EDITOR,—Kimura’s disease is an uncommon, chronic inflammatory disorder of unknown aetiology which occurs predominantly in orientals and presents with tumour-like swellings mainly in the head and neck region. The condition primarily involves the subcutaneous tissues, parotid glands, and/or lymph nodes. Orbital cases are infrequent and most reported cases are in adults, with only one case in a child of Afro-Caribbean descent.1 Our case report presents orbital Kimura’s disease in an 8 year old white child.

CASE REPORT
An 8 year old white girl presented to us with a 4 week history of a painless swelling of her right upper lid. She initially had an upper respiratory tract infection lasting a month which was treated by her general practitioner with Augmentin. She subsequently developed what appeared to be a right sided stasis. Of note, she had a strong history of allergic eye disease with intermittent conjunctivitis, and also suffers from asthma for which she takes Pulmicort and Bricanyl inhaler regularly. Examination revealed a right partial ptosis, mild right proptosis, and a palpable, painless mass in the superior aspect of her right orbit. There were no pupillary abnormalities, her extraocular movements were full, and funduscopic examination was unremarkable. There was no associated regional lymphadenopathy and the rest of the ocular and general physical examination showed no other abnormalities.

Investigations done included a full blood count, which was within normal limits. WBC 8.57 x10^9/l with a differential of neutrophils 62%, lymphocytes 26%, monocytes 6%, eosinophils 5%, and basophils 0%. A magnetic resonance imaging (MRI) scan with...
COMMENT
Kimura's disease (or angiolymphoid hyperplasia with eosinophilia) is a chronic inflammatory disorder of unknown aetiology which presents with tumour-like swellings mainly in the head and neck region. Most cases have been described in China and Japan, with relatively fewer cases reported in non-orientals. The disease typically affects males in the 20–40 year age group, and presents as single or multiple smooth swellings in the subcutaneous tissues, major salivary gland and/or lymph nodes in the head and neck area.

Histopathologically, the features described in previous cases corresponded with ours; there are eosinophilic infiltrates (mainly in a perivascular pattern), vascular proliferation, fibrosis, and formation of lymphoid follicles. It is still uncertain as to whether it represents a benign lymphoid neoplastic process, a variant of haemangioma or an allergic reaction. There have been no reported cases of malignant change or fatalities. There is usually an insidious onset with a long benign course, but recurrences are common after surgical excision. Systemic associations include asthma and nephrotic syndrome. Regional lymphadenopathy ranges from 50–75% of cases and there is a high occurrence of peripheral blood eosinophilia.

Cases of Kimura’s disease have been reported in the orbits, but the choice of antibiotics remains controversial. Most centres rely on combination therapy and amoxicillin is often used to treat Gram negative organisms. Aminoglycosides have a narrow therapeutic index and therefore a relatively small increase in concentration can cause significant toxicity. Following concerns about the risks of retinal toxicity caused by intravitreal gentamicin many ophthalmologists now use 0.4 mg intravitreal amikacin even though its use has also been associated with retinal toxicity. We report a case of retinal toxicity in a patient treated with intravitreal amikacin for postoperative bacterial endophthalmitis. This case serves to remind ophthalmologists of the risk of amikacin therapy and presents an argument for an alternative strategy in the treatment of bacterial endophthalmitis.

CASE REPORT
A 60 year old highly myopic woman presented with an acute history of increasing ocular pain and redness 5 days after routine phacoemulsification cataract extraction and intraocular lens implant. She had no other ocular or systemic history.

Initial examination revealed a visual acuity of 6/18, significant anterior chamber inflammation with a fibrin deposit over the pupil. A diagnosis of bacterial endophthalmitis was made and the patient was commenced on intravenous ciprofloxacin and hourly gentamicin, vancomycin, and dexamethasone eye drops. A vitreous biopsy was performed and 2 mg of intravitreal vancomycin (0.1 ml of 20 mg/ml) and 0.4 mg amikacin (0.1 ml of 4 mg/ml) were administered. The intravitreal antibiotics were prepared by a senior pharmacist using a typedown protocol. The following day vision had deteriorated to no perception of light and the patient had developed a severe retrobulbar pain and papillitis.

Figure 1  A magnetic resonance imaging (MRI) scan with contrast. There is an extensive superior orbital mass on the right side.

Figure 2  Section showing vascular proliferation with eosinophilic and lymphocytic perivascular inflammation and fibrosis. Haematoxylin and eosin stain. Original magnification x200.

Amikacin retinal toxicity

Editor,—It is well established that early intravitreal antibiotics are the treatment of choice for bacterial endophthalmitis but the choice of antibiotics remains controversial. Most centres rely on combination therapy and amoxicillin is often used to treat Gram negative organisms. Aminoglycosides have a narrow therapeutic index and therefore a relatively small increase in concentration can cause significant toxicity. Following concerns about the risks of retinal toxicity caused by intravitreal gentamicin many ophthalmologists now use 0.4 mg intravitreal amikacin even though its use has also been associated with retinal toxicity. We report a case of retinal toxicity in a patient treated with intravitreal amikacin for postoperative bacterial endophthalmitis. This case serves to remind ophthalmologists of the risk of amikacin therapy and presents an argument for an alternative strategy in the treatment of bacterial endophthalmitis.
vitreal aminoglycosides we recommend the use of ceftazidime instead of amikacin or gentamicin. TIMOTHY L JACKSON
TOM H WILLIAMSON
St Thomas’s Hospital, London
Correspondence to: Tim Jackson, Vitreoretinal Unit, Department of Ophthalmology, St Thomas’s Hospital, Lambeth Palace Road, London SE1 7EH.
Accepted for publication 18 May 1999


Simultaneous administration of hepatitis B and polio vaccines associated with bilateral optic neuritis

EPTON,—Immunisation against hepatitis B is recommended when there is an increased risk of contracting the virus because of lifestyle, occupation, or factors such as close contact with a case. Immunisation against poliomyelitis is routine for infants in the UK with reinforcement during childhood and then again in the teenage years. For those individuals at continued risk of infection, further reinforcing doses are given every 10 years.

Both are commonly used vaccines and serious adverse reactions are extremely rare. We describe a case of severe bilateral, progressive optic neuritis occurring 1 week after vaccination against hepatitis B and poliomyelitis.

CASE REPORT
A 44 year old female health worker presented with gradual reduction of vision in both eyes associated with retrobulbar discomfort exacerbated by ocular movement, 7 days after vaccination against hepatitis B and poliomyelitis. Ophthalmological findings revealed visual acuities of 6/18 right eye, 6/12 left eye associated with bilateral optic nerve swelling. Within the next 48 hours the visual acuities dropped to perception of light in both eyes with absent direct and indirect pupillary light responses. This was despite commencement of therapy with intravenous methylprednisolone. Systemic examination revealed no other abnormalities. All haematological and biochemical investigations were normal and no infective cause was isolated. Computed tomograph imaging was normal and there was no evidence of demyelination on magnetic resonance imaging. Cerebrospinal fluid (CSF) examination revealed no abnormality either biochemically, after culture, or on electrophoresis. In addition, CSF pressure was within normal limits. Visually evoked potentials revealed absent responses. Despite 5 days of intravenous methylprednisolone (1 g per day) followed by a slow tapering of oral prednisolone (1 mg/kg/day) her vision remained poor (counting fingers at 1 metre in both eyes) after 3 months.

COMMENT
The recombinant hepatitis B vaccine has been associated with a diverse range of isolated adverse reactions but ocular complications are exceedingly rare. Granel et al attributed four cases of central retinal vein occlusion in patients under 50 years of age to the vaccine, and associations with multiple evanescent white dot syndrome (MEWDS) and acute posterior multifocal placoid pigment epideimyopathy (APMPPE) have been described. Stimulating vaccine derived anti-eggs and neutralizing antibodies of an autoimmune nature have been reported including CNS demyelination. Bilateral optic neuritis occurs occasionally in acute hepatitis B infection. Vaccines derived from live attenuated viruses such as the trivalent oral polio vaccine can cause direct viral infections of the central nervous system. The incidence of vaccine derived paralytic poliomyelitis is reported as one in three million in recipients of the vaccine or their close contacts. The vaccine has also been linked to some cases of Guillain-Barré syndrome but ocular complications have not been reported.

Others such as the trivalent measles, mumps, and rubella (MMR) vaccine and the monovalent rubella vaccine, have also been associated with bilateral optic neuritis.

The exact mechanisms by which neurological complications following vaccination are unknown but various hypotheses exist including immune complex mediated demyelination or neurotoxicity, antigenic mimicry between the viruses such as the trivalent oral polio vaccine and to post infectious optic neuritis generally did better with corticosteroid therapy than in our case but high dose intravitreal corticosteroids are generally considered to be the treatment of choice in these rare but potentially devastating complications.

OWEN STEWART
BERNARD CHANG
JOHN BRADBURY
Department of Ophthalmology, Bradford Royal Infirmary, Bradford, West Yorkshire.
Correspondence to: Mr Owen Stewart, Department of Ophthalmology, St James’s University Hospital, Leeds, West Yorkshire, LS9 7TF.
Accepted for publication 18 May 1999


Retinopathy after long term, standard doses of hydroxychloroquine

EDITOR,—While the antimalarial drug chloroquine has frequently been reported to cause retinopathy, there have been very few documented cases occurring with hydroxychloroquine (Plaquenil, Winthrop Pharmaceuticals, New York, USA).1–3 Patients may tolerate large cumulative doses of hydroxychloroquine without developing retinopathy.3–5 Doses of ≤400 mg/day and ≤6·5 mg/kg of body weight/day of hydroxychloroquine have been used safely and some authors have suggested that ophthalmic screening is not necessary for patients on these doses.3–5 We present a case of hydroxychloroquine toxicity that developed in a patient after long term use of 400 mg (6·3 mg/kg) of hydroxychloroquine daily.

CASE REPORT
A 61 year old white woman presented with a 1 year history of increased glare in both eyes without change in visual acuity. She had a history of rheumatoid arthritis for which she took hydroxychloroquine 400 mg daily (6·5 mg/kg/day) for 10 years. Her total hydroxychloroquine dose was 1460 g. The patient had never taken chloroquine and had no history of macular disease. Her weight had been stable over the 10 year period. Family history was negative for macular dystrophy or retinal degeneration. Visual acuity at presentation was 20/20 in both eyes. Colour vision testing with pseudoisochromatic plates was normal in both eyes. Fundus examination showed subtle retinal pigment epithelium (RPE) pigmentary changes in a crescent pattern around the inferior fovea of both eyes (Fig 1A). Fluorescein angiogram confirmed the fundus findings (Fig 1B) which were felt to be early bull’s eye maculopathy. No drusen or signs of macular degeneration were appreciated in either eye. A central visual field performed with Humphrey automated static threshold perimeter using a white test object revealed bilateral paracentral ring scotoma corresponding to the macular pigmentary changes (Fig 2, upper). A diagnosis of hydroxychloroquine retinopathy was established and the drug was discontinued.

Ten months later the patient’s vision remained 20/20 with normal colour vision in both eyes. Her symptom of glare in both eyes had resolved. The RPE changes in the retina were unchanged and visual field testing showed some improvement of the paracentral scotoma (Fig 2, lower).

COMMENT
Hydroxychloroquine retinopathy is a rare condition characterised by bull’s eye maculopathy or pigmentary changes in the macula, corresponding scotomas on visual field testing, and irreversible signs and symptoms once hydroxychloroquine is stopped.6,7 Cases of preretinopathy have been described in which visual field defects were elicited using a red test object but not a white test object on Humphrey perimeter.7,8 All cases of preretinopathy completely resolved after the discon- tinuation of hydroxychloroquine.7 Five cases of true hydroxychloroquine retinopathy reported in the literature demonstrated that discontinuation of hydroxychloroquine resulted in stabilisation but not resolution of the retinopathy.7,8 Unlike the other reported cases of true hydroxychloroquine retinopathy, our patient had some resolution of visual field loss.

Much controversy exists as to whether the daily or cumulative dose of hydroxychloroquine contributes the greater risk for retinopathy. While there have been reported cases of hydroxychloroquine toxicity at cumulative doses similar to our patient’s (1460 g over a 10 year period), these patients had (1) received chloroquine therapy before hydroxychloroquine, (2) were treated with daily doses >400 mg/day, or (3) exceeded a daily dosage of 6·5 mg/kg of body weight/day.9 Our patient’s daily dose never exceeded 400 mg/day or 6·5 mg/kg of body weight/day. Her renal function was normal, thereby making the possibility of inadequate clearance of hydroxychloroquine unlikely.

Hydroxychloroquine may cause retinopathy when used in recommended doses over a long period of time. Since a threshold dose for retinal toxicity has not been established, careful screening examinations should be performed especially as the cumulative dose increases. Prompt cessation of hydroxychloroquine may result in stabilisation of maculopathy at a clinically benign stage.
CASE REPORT
A 12 year old girl was referred to Moorfields Eye Hospital for a second opinion regarding her deteriorating vision following bilateral trabeculectomies with adjuvant mitomycin C for raised intraocular pressure.

At the age of 3 years, she was diagnosed with having megalocornea with ocular hypertension. Two years later, because of the persistent degree of IOP elevation (between 30–40 mm Hg), she was commenced on bilateral medical treatment. However, at the age of 11, she developed such severe problems with her topical treatment that surgery was considered necessary. In May 1996 right and subsequently left trabeculectomies were performed and during the procedures subconjunctival mitomycin C (0.3 mg/ml) was applied for 5 minutes. Her postoperative recovery was complicated by the development of ocular hypotony.

She was seen at Moorfields for the first time with progressive bilateral vision loss and recent onset of transient obscurations, 5 months after her filtration surgery and the onset of hypotony. Her visual acuities had fallen from right eye 20/20 (~0.5–0.25 × 180°) to hand movements (HM), and left eye 20/30 (~1.5 DS) to counting fingers (CF). Her visual fields confrontation with a red target showed an enlarged blind spot. Both eyes had large, draining, diffuse and avascular filtration blebs with signs of hypotony as evidenced by bilateral IOPs of 0 mm Hg, superficial corneal epithelial staining, striae, and macular and choroidal folds. Both optic discs were swollen although no haemorrhages or cotton wool spots were observed. The patient was sensitive to light and good quality photographs could not be obtained. An ultrasound showed bilateral anterotemporal choroidal detachments involving the ciliary body (right larger than left) and axial lengths recorded as 23.0 mm in both eyes.

She underwent several surgical procedures. The day after she first presented, she underwent a right subconjunctival autologous blood injection to the right bleb under general anaesthesia. The following day, however, her vision was worse with visual acuities being recorded as patchy areas of perception of light on the right and hand movements temporally on the left. Her IOPs were 4 and 0 mm Hg in the right and left respectively and the higher degree of disc swelling (as judged by further elevation of the disc) was noted bilaterally. It was considered by a neurologist (GTP) that the disc swelling may itself be contributing to the visual loss, particularly the persistent visual obscurations which suggested critically impaired perfusion of the disc. To prevent even further vision loss, a limbus puncture to reduce the CSF pressure was suggested, to provide short term improvement in disc perfusion, which it was hoped in the longer term would be helped by raising her intraocular pressure. She underwent further surgery to both her eyes for hypotony, later that evening, which comprised revision of both blebs with scleral patch grafts. This was preceded by a limbus puncture under general anaesthesia at which there was an opening pressure of 22.5 cm H2O. A volume of 30 ml of CSF was removed and the intraocular pressure was recorded as 5.3 cm H2O with microbiological and biochemical analysis of CSF revealing no abnormality.

The day after surgery, the fourth day after initial presentation, examination revealed improved visual acuities of HM in both eyes with IOPs of 19 and 23 mm Hg in the right and left eye respectively. The disc swelling was felt to be reduced, and over the next few days improved gradually. Seven days after presentation, her IOPs had fallen to 0 and 8 mm Hg, right and left respectively, and she was taken back to theatre for anterior chamber re-creating with Healon-GV. Her IOPs were around 20 mm Hg again 10 days later, with decreasing disc swelling, and a repeat ultrasound showed reduced choroidal detachments and axial lengths of 24.8 mm in the right and 24.6 mm in the left.

She underwent one other further surgical procedure, 7 weeks after presentation, when her IOPs became elevated again at 35 and 28 mm Hg right and left respectively, to a degree where it was felt her optic discs would be compromised. Treatment consisted of needle procedures to both blebs with subconjunctival injections of 5-fluorouracil to reduce scar formation. Her IOPs responded very quickly to this final procedure and she remains on no antiglaucoma medication with normal intraocular pressures (less than 20 mm Hg) now 7 months after presentation. Both drainage blebs are Seidel negative with good evidence of aqueous drainage into the filtration blebs (Fig 1, top right). Her visual acuities and visual function have now recovered quite substantially. However she does have a relative afferent pupillary defect in the left eye, and her current refraction and visual acuities are right eye ~1.0–4.0 × 05° (RVA 20/30) and left eye ~1.75–6.5 × 180° (LVA 20/120). Her optic discs show no evidence of swelling, and she has neither choroidal detachments nor macular folds (Fig 1, bottom left and right) as confirmed on ultrasound.

COMMENT
This unusual case presented to us with severe ocular hypotony following bilateral primary glaucoma filtration surgery with mitomycin C. She developed vision loss, with episodes of transient obscurations attributable to hypotony and disc swelling. Her response to the initial surgery and subsequently to the corrective

Figure 1 Although neonatal examination had been normal, our patient had been noted to have "large" eyes at a young child. This photograph taken at age 2 years (top left). Reversal of ocular hypotony occurred following filtration bleb revision with scleral patch grafts, with drainage into blebs apparent at 2 months (top right) postoperatively. Hypotony induced papilloedema resolved after surgery with no disc swelling apparent in right (bottom left) or left (bottom right) fundus, shown 7 months postoperatively.

Correspondence to: Jennifer E Thorne, MD, Scheie Eye Institute, 51 North 39th Street, Philadelphia, PA 19104, USA.

Accepted for publication 10 May 1999

REFERENCES

Ipsilateral papilloedema. To our knowledge this is the first report of post filtration hypotony and reduced scleral rigidity. We present a case of ocular hypotony following mitomycin C as suggested that hypotony related complications may be more common in myopes as they have associated with disc swelling. We feel it there are inherent problems in wound healing patients with connective tissue disorders where Scar formation. Her IOPs responded very quickly to this final procedure and she remains on no antiglaucoma medication with normal intraocular pressures (less than 20 mm Hg) now 7 months after presentation. Both drainage blebs are Seidel negative with good evidence of aqueous drainage into the filtration blebs (Fig 1, top right). Her visual acuities and visual function have now recovered quite substantially. However she does have a relative afferent pupillary defect in the left eye, and her current refraction and visual acuities are right eye ~1.0–4.0 × 05° (RVA 20/30) and left eye ~1.75–6.5 × 180° (LVA 20/120). Her optic discs show no evidence of swelling, and she has neither choroidal detachments nor macular folds (Fig 1, bottom left and right) as confirmed on ultrasound.

COMMENT
This unusual case presented to us with severe ocular hypotony following bilateral primary glaucoma filtration surgery with mitomycin C. She developed vision loss, with episodes of transient obscurations attributable to hypotony and disc swelling. Her response to the initial surgery and subsequently to the corrective

Figure 1 Although neonatal examination had been normal, our patient had been noted to have “large” eyes at a young child. This photograph taken at age 2 years (top left). Reversal of ocular hypotony occurred following filtration bleb revision with scleral patch grafts, with drainage into blebs apparent at 2 months (top right) postoperatively. Hypotony induced papilloedema resolved after surgery with no disc swelling apparent in right (bottom left) or left (bottom right) fundus, shown 7 months postoperatively.
syndrome type II (EDS II). Examination
We therefore wondered if she had an underly-
ing skin man” of the 19th century),
described in fairground artistes, such as the
diad: extensible skin (extreme examples being
ecting skin, ligaments, joints, blood vessels,
a normal palate, no spinal defects, no arach-
boilage in pre- and postoperative lengths).
However, there was a strong family
history of joint hypermobility.
Table 1 Screen for joint hypermobility

<table>
<thead>
<tr>
<th>Action</th>
<th>Score (maximum = 9)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Passive hyperflexion of 5th metacarpophalangeal joint beyond 90°</td>
<td>1 (max = 2)</td>
</tr>
<tr>
<td>Passive hyperextension of thumb to flexor aspect of wrist</td>
<td>1 (max = 2)</td>
</tr>
<tr>
<td>Hyperextension of elbow beyond 10°</td>
<td>1 (max = 2)</td>
</tr>
<tr>
<td>Hyperextension of knee beyond 10°</td>
<td>1 (max = 2)</td>
</tr>
<tr>
<td>Forward flexion of trunk, palms flat on floor in front, knees in extension</td>
<td>1</td>
</tr>
</tbody>
</table>

The development of severe vision loss in our patient was associated with the occurrence of transient obstructions and hypotonous disc swelling. Resolution of the disc swelling was 4–6 weeks after correction of the hypotony, and improvement in visual acuity was noted thereafter. Its natural history—that is, the pattern of resolution and visual recovery, is compatible with disc swelling. Visual loss associated with hypotonous maculopathy, however, may never recover despite reversal of hypotony. Prolonged disc swelling in ocular hypotony may be related to that occurring in raised intracranial pressure papilloedema—for example, axonal swelling, accumulation of mitochondria, and cytoid body (cotton wool spots) formation.

The pathogenesis of disc swelling in ocular hypotony may be related to that occurring in raised intracranial pressure if it is postulated that in both situations the CSF pressure in the subarachnoid space around the optic nerve exceeds the perfusion pressure of the optic disc. Therefore in our patient, it was considered that lowering the CSF pressure to well below normal levels (<20 mm H₂O) might provide a temporary improvement in disc perfusion—hence our target for a very low closing pressure of 5.5 mm H₂O in the lumbar puncture. Two previous reports of unilateral disc swelling following trabeculectomy also suggest that optic disc oedema could result from a disturbed equilibrium between ICP and IOP—though none of the patients described in these papers had hypotony or profound vision loss.

The development of profound vision loss in association with hypotony induced optic disc swelling in our patient, was as a complication of mitomycin C assisted glaucoma filtration surgery. However, the degree of hypotony was most probably related to reduced scleral rigidity. We believe that her abnormal wound healing response and defective connective tissue contributed to the severity of the hypotony that followed initial surgery with mitomycin C. We suggest that antiproliferatives be used with extreme caution in patients who you suspect may have a connective tissue disorder. A quick screening procedure might include a joint hypermobility score as shown in Table 1. Patients with Ehlers-Danlos syndrome, such as our patient, may be at high risk of post filtration hypotony and developing “soft eyes” with sight threatening sequelae, as our case suggested that not only might they have elastic skin but also “elastic globes”.

Supported in part by the Wellcome Trust (grant no 048474) (to MFC), Guide Dogs for the Blind (MFC), and the Medical Research Council (to PTK).

M FRANCESCA CORDEIRO
Wound Healing Research and Glaucoma Units, Institute of Ophthalmology and Moorfields Eye Hospital, London

GORDON T PLANT
Moorfields Eye Hospital and National Hospital for Neurology and Neurosurgery, London

ANNE CHILD
St George’s Hospital Medical School, London

BARRY JONES
Moorfields Eye Hospital, London

PENG T KHAW
Wound Healing Research and Glaucoma Units, Institute of Ophthalmology and Moorfields Eye Hospital, London
Fluctuating oculomotor hyperfunction and hypofunction caused by aneurysmal compression of the third cranial nerve

Editor,—Aneurysms of the posterior communicating artery classically present with a painful progressive palsy of the third cranial nerve producing ptosis, ophthalmoplegia, and mydriasis, and mydriasis. We present a case in which painful progressive palsy of the third cranial nerve, or oblique, and a 2-week history of right eye tension and ischaemic heart disease was referred to the eye department with a 3-week variable limita-
ties were right 6/6 and left 6/9. There was a right upper lid retraction (Fig 1). The results of intermittent diplopia—third nerve hyperactivity resolved immediately following clipping of the aneurysm while the ophthalmoplegia improved rapidly but took several weeks to resolve fully. The degree of third nerve hypofunction, however, much greater than the mild facial weakness seen in hemi-
facial spasm and may have been due to the greater degree of compression of the third nerve in this case. This case is important for two reasons. Firstly, this is the first report of compression of the third cranial nerve producing alternating oculomotor hypo- and hyperfunction as a result of a compressive lesion. Secondly, it emphasizes the importance of excluding com-
pression by intracranial aneurysm in a patient with variable signs which could be explained by a partial third nerve palsy.

MARGARET R DAYAN
Oxford Eye Hospital, Radcliffe Infirmary, Woodstock Road, Oxford OX2 6HE

Correspondence to: Miss Dayan

Accepted for publication 31 May 1999

2 Barrion JD, Trautmann JC, Sundt TM. Min-
5 Schultz-Bonhage A, A. Electrophysio-
6 Ryu H, Yamamoto S, Sugimori K, et al. Hemifac-

Late onset of Leber’s hereditary optic neuropathy in HIV infection

Editor,—We report a case of late onset of Leber’s hereditary optic neuropathy (LHON) in a 59 year old patient with HIV infection being treated long term since 1991 with zido-
vudine. The onset and course of the patient’s eye disease as well as the diagnostic process were analysed. Molecular genetic testing revealed the mitochondriald DNA (mtDNA) mutation in nucleotide position 11778 confir-
moving the diagnosis of LHON. In our patient late disease expression may be interpreted as a result of the rare combination of the 11778 mutation with HIV infection and long term zidovudine treatment. This is the first report on a patient with LHON suffering from additional HIV infection.

Correspondence to: Peng T Khaw, Wound Healing Group and Glaucoma Unit, Pathology Department, Institute of Ophthalmology, Bath Street, London EC1V 9EL.

Accepted for publication 18 May 1999

8 Minckler D, Tso M, Zimmerman L. A light microscopic autoradiographic study of axono-
9 Greenfield DS, Waniwchcharungruang B, Lieb-
mann JM, et al. Pseudotumour cerebri appear-
ing from addi-
10 Kawasaka A, Purvin V. Unilateral optic disc edema following trabeculectomy J Neuro-
In previous studies HIV was only exposed to two epigenetic factors at the time of presentation. The CD4+ T lymphocyte count was 360 × 10^6/l and HIV RNA in plasma was 8500 VECT. On examination, visual acuity without correction was 20/20 bilaterally. Pupils were equally reactive without an afferent pupilary defect. Extraocular motility was full, and saccades were grossly normal and symmetrical. The patient had been diagnosed with HIV infection in 1991 and/or the diagnosis of HIV infection in 1991 and/or the expression of LHON in our patient. This is the first report on a patient with LHON suffering from additional HIV infection.

Christoph Lüke Klinik und Poliklinik für Augenheilkunde
Oliver A Cornely Klinik für Innere Medizin, Universität Köln, Germany
Julia Frickel Elfi Lehner Karl Ulrich Bartz-Schmidt Klinik und Poliklinik für Augenheilkunde, Universität Köln, Germany
Bernd Wissinger Molekulargenetisches Labor, Universitätsgesundheitsklinik Tübingen, Germany
Richard Brunner Klinik und Poliklinik für Augenheilkunde, Universität Köln, Germany

Correspondence to: Christoph Lüke, Klinik und Poliklinik für Augenheilkunde, Universität Köln, Joseph-Stelzmann-Strasse 9, D-50924 Köln, Germany

Accepted for publication 17 June 1999

COMMENT


Acute onset comitant esotropia as presenting sign of demyelinating disease

Acute onset comitant esotropia is most commonly benign when occurring in infancy or early childhood. Examination to rule out a parietic deviation is essential. However, acute onset of comitant esotropia has also been reported in association with serious neurologistic disease. It has been reported in association with hydrocephalus. They may have a meningomyelocele or encephalocele, and an A-pattern esotropia.1,2 In cases of comitant esotropia associated with shunt failure, all the esotropias resolve when the shunts are revised. These patients do not have A-pattern.
esotropias. Patients may not necessarily present with papilloedema even when hydrocephalus is present.\textsuperscript{1} In the present case, no hydrocephalus was noted on neuroradiographic study.

Arnold-Chiari malformation has also been reported in association with acute comitant esotropia.\textsuperscript{1,2} These cases may present with an A-pattern, co-existing nystagmus and hydrocephalus. Arnold-Chiari malformations sometimes do not manifest until late childhood or adulthood, and can be mild. Neuroradiographic study did not reveal Arnold-Chiari malformation in this patient.

Central nervous system tumours have also been reported in association with acute onset comitant esotropias.\textsuperscript{3,4} Tumours in this group include cerebellar astrocytomas, and medulloblastomas, as well as pontine gliomas.\textsuperscript{1} An A-pattern esotropia is unusual in these patients.\textsuperscript{2} In a report by Williams and Hoyt, three of their six patients had some form of nystagmus associated with the comitant esotropia. They suggested neurological evaluation in any patient with both nystagmus and acute comitant esotropia. In addition, acute comitant esotropia is often reported rarely with myasthenia gravis and seizures.\textsuperscript{5} This patient had a brainstem lesion that was consistent with an acute central nervous system demyelination. Its location in the midbrain may have interfered with neurons associated with horizontal gaze centres or vergence mechanisms. This is the first reported case of acute demyelinating disease presenting with acute comitant esotropia. Acute disseminated encephalomyelitis is generally post viral and is characterised by abrupt headache, fever, drowsiness, and focal neurological dysfunction. Cerebrospinal fluid analysis may exhibit pleocytosis. It may resolve completely or may result in permanent impairment and seizures.\textsuperscript{6} Whether this lesion represents a localised variant of acute disseminated encephalomyelitis or multiple sclerosis is not clear. However, cases of acute onset comitant esotropia with suspicious presentations warrant neuroimaging to rule out intracranial pathology including central nervous system demyelinating lesions.

BARRY N WASSERMAN
Chester County Eye Care Associates, 606 East Marshall Street, Suite 104, West Chester, PA 19380, USA

Accepted for publication 8 June 1999