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 left eye was quiet and she had a visual acuity of 6/60 in the right eye, and 6/6 in the left eye. She had no past ocular history, redness, or discharge. She had no past ocular history. She was a 12 year old Chinese woman presenting to Shantou Central Hospital, Guangdong, China, with a complaint 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. A mild vitreous haze obscured the foveal excursion. There was no retinal abnormality.

One month later she underwent a three port pars plana vitrectomy, and the worm was removed. At 6 weeks postoperatively there was no retinal abnormality. 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 thelaziasis, but in the absence of preoperative and postoperative

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 endophthalmitis1 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 20/30 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.

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Intraocular infestation with the worm, Thelazia callipaeda

EDITOR,—Ocular infections with helminthic parasites are well described. The commonest organisms are filarial 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 thelaziasis, 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 lacrimation 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 extraocular 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, with a complaint 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. A mild vitreous haze obscured the foveal excursion. There was no retinal abnormality.

One month later she underwent a three port pars plana vitrectomy, and the worm was removed. At 6 weeks postoperatively there was no retinal abnormality. 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 thelaziasis, but in the absence of preoperative and postoperative


enable it to burrow through or bore its way into any body structure. Little is known about the life cycle of Thelazia. The intermediate host is Amiota variegata, a fly of the Drosophila family. It is known to infect the lacrimal sac, from where it passes into the conjunctival sac. It is probably deposited in the larval form by flies settling on the eyes and this accounts for extracocular infection. The adult worm in our patient’s eye was fully developed and healthy. It had caused little inflammatory response, and appeared to thrive in the conditions provided by the vitreous cavity, suggesting the possibility of immune privilege. We cannot explain how this worm entered the eye. We put forward the suggestion that humans may be the definitive host, with dogs and rabbits being the reservoir. For intraocular colonisation, the infective stage is probably the filariform larva, the portal of entry being the skin. An alternative mode of infection may be as the larva or embryonated egg ingested with raw drinking water.

COMMENT
Thelazia callipaeda (Onchocerca to suck) is a member of the phylum Nematoda, order Spirurida, suborder Spirurida, superfamily Spiruroidea. Adult worms look like creamy white threads; males are 4.5–13 mm long and 0.25–0.75 mm in diameter; females are larger, ranging from 6.2 to 17 mm in length and 0.3 to 0.85 mm in diameter (Fig 1). T. callipaeda is an unsegmented invertebrate with distinct oral and anal parts. The male is identifiable by its curved posterior (Fig 2A). In both the male and the female, the mouth is without lips (Fig 2B) and consists of two concentric rings of flattened papillae around a central opening. It should be noted that there are no hooks or sharp spines in the mouth or elsewhere on the body, and this raises the question of how this worm entered the eye.

Onchocerca spirocerca, a close relative of Thelazia, is quite frequently found within the eye and causes major tissue destruction. Structurally, one of its most distinguishing features is the presence of rows of curved hooks and posteriorly angled spines in a cuff around the mouth used for boring into tissue. T. callipaeda does not possess the organs to

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 1973, ophthalmological examination led to a diagnosis of bilateral hereditary macular dystrophy (malattia leventinese) in a woman born in 1943. Her visual acuity was then 20/30 in each eye. By 1981, subfoveal neovascularisation led to an irreversible decrease in visual acuity in her right eye, down to 20/1000 (Fig 1A).

In 1996, a dense subretinal 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 supertemporal 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 intraretinal 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.

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We have here described the first example of peripheral retinal neovascularisation with ischaemia. The aetiology of this ischaemia probably concerns a retinal vein branch occlusion not linked with macular degeneration. The occlusion had developed very discretely, with the patient aware of no functional sign. In fact, visual acuity was already severely impaired by the neovascular complication of the macula and only the nasal visual field was affected, whence the lack of ocular disturbance. No aetiology could be found for the occlusion. A general check up showed no systemic abnormality.

Thus, this observation suggests the need for great attention to be paid to the retinal periphery in patients suffering from macular dystrophy, whether hereditary or not, so as to prevent the development of potentially serious complications.

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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 pulsatil 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 × 10⁶ 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.

Figure 2. (A) Right eye. Photograph in 1996. Advanced stage of the macular disease with irregular subretinal fibrous metaplasia, hyperplasia of the retinal pigment epithelium, and discrete radial basal laminnar drusen. Note also a wedge-shaped superotemporal area with intraretinal haemorrhages, hard exudates, and sheathed vessels. (B) Right eye. Fluorescein angiogram in 1996. Note the temporal retinal ischaemia with telangectatic vessels, shunt vessels, and microaneurysms, in addition to the neovascularisation.
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and the patient subsequently went on to complications, the treatment failed to show other treatment modalities had already been in the second eye has been described. Several activator in a patient with a non-acute CRVO occlusions.

CRVO. Fibrinolysin 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 in those circumstances where the presentation is acute should the use of intravenous thrombolysis be considered.

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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 perimetreries 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. Ophthalmoscopy 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, perimetreries 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 perimetreries, ERG responses remained undetectable irrespective of visual fluctuations. His healthy parents were first cousins, and his elder brother had night blindness due to classic retinitis pigmentosa. Because of the mild mental retardation, the day to day visual fluctuations or other symptomatic variations could not be confirmed in his brother.

DNA was extracted from the peripheral blood after obtaining informed consent. Eton 11 of the arrestin gene was amplified using polymerase chain reaction. The amplified product was separated on polyacrylamide gel and revealed abnormally rapidly migrated signal suggesting a small deletion. Direct sequencing using an automated nucleotide sequencer (ALF Express, Pharmacia) disclosed a homozygous deletion of adenine at nucleotide 1147 (1147delA). This mutation

Figure 2 Visual field sensitivity, determined by an automated perimeter (Interzeag Octopus 101, low vision program) on different days. Representative visual field charts (right eye) obtained on a “good day” (left chart) and a “bad day” (right chart).

Figure 1 Fundus in the right (A) and left (B) eye of the patient. There are diffuse chorioretinal atrophies accompanied by numerous bone spicule pigments, attenuated retinal vessels, and pale optic discs.
here had advanced retinitis pigmentosa and reported that he had experienced over the years alternating days of good and bad vision and that the day to day visual fluctuations had been modified by physical stress or alcohol drinking. Perimetric evaluations of this case demonstrated daily variability in visual sensitivities which corresponded to the subjective complaints. This form of visual fluctuation appears unusual, and it is remarkable that this patient had such a homozygous mutation in the arrestin gene that gives rise to a premature truncation of translation. The human arrestin, also known as S-antigen, has an inhibitory role in the activated phototransduction cascade. It should be mentioned that the arrestin gene is one of those expressed in the pineal gland that is considered to play a major role in the circadian rhythm. Thus, it is tempting to speculate that the mutation in the arrestin gene could have modified frequent fluctuations to induce daily alternating visual fluctuations in the present retinitis pigmentosa patient. The full coding sequences of the rhodopsin gene were normal in the relevant patient (data not shown). However, we could not deny the association of different polymorphisms in related gene products, or different environmental influences on the daily alternating visual fluctuations.

Recent molecular assessments revealed families with diffuse retinitis pigmentosa with or without features of Oguchi’s disease harboring 1147 delA in the arrestin gene. The patient described here provides evidence that the same arrestin gene mutation is causally related not only to Oguchi’s disease but also to autosomal recessive retinitis pigmentosa. It is emphasised that our patient had classic features of autosomal recessive retinitis pigmentosa with poor visual prognosis and showed unusual visual fluctuations. In view of a rare mutation of arrestin gene, the relevant retinitis pigmentosa might be extremely rare. In a large series of white patients with aetiology of undefined retinitis pigmentosa arrestin gene mutation was not detected.

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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. 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 ptosis. 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 fundoscopic 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 1%. A magnetic resonance imaging (MRI) scan with
gadolinium contrast was done which showed an extensive superior orbital mass which was not clearly delineated, but involved the upper eyelid (see Fig 1). An orbital biopsy was subsequently performed. Histological sections revealed fibrosis with a marked vascular proliferation and a prominent perivascular inflammation consisting of eosinophils and lymphocytes (see Fig 2). There were also scattered lymphocytes and eosinophils throughout the fibrotic tissue with reactive lymphoid follicles. There was no evidence of necrosis, granulomas, or malignant cells. Immunohistochemical studies revealed no cells of myogenic origin present. Studies for desmin, myoglobin, and vimentin were negative. All of the cells considered lymphoid on the haematoxylin and eosin stain also stained for the leucocyte marker common leucocyte antigen (CLA). A diagnosis of Kimura’s disease (angiolymphoid hyperplasia with eosinophilia) was made.

At biopsy, the tumour was debulked and the patient was thereafter placed on a short 6 week course of oral steroids. The ptosis resolved and at 1 year follow up, there has been no clinical evidence of recurrence.

Figure 2  Section showing vascular proliferation with eosinophilic and lymphocytic perivascular inflammation and fibrosis. Haematoxylin and eosin stain. Original magnification ×200.

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; however, there was only one previously reported orbital case in a child (of Afro-Caribbean descent). To our knowledge, this is the first orbital case reported in a white child. Non-orbital cases of Kimura’s disease have been treated with surgical excision, irradiation and steroid therapy. After active therapy, they tend to recur. Cases have been conservatively managed for up to 12 years successfully although the cosmetic deformity can be quite a handicap. Periorbital cases have so far been treated successfully with surgical excision and biopsy with debulking. Oral steroids or intralessional injection with steroids have not been as successful. There was only one reported case of severe haemorrhage on surgical resection.

Complete surgical excision appears to be the best treatment.
vitreal aminoglycosides we recommend the use of ceftazidime instead of amikacin or gentamicin.

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

The exact mechanism by which neurological complications following vaccination are unknown but various hypotheses exist including immune complex mediated demyelination or neurotoxicity, antigenic mimicry between the vaccine and the nervous system to cause an autoimmune reaction, or several of these mechanisms occurring simultaneously. Neurotoxicity is associated with the MMR vaccine and the monovalent rubella vaccine. Neurological complications also occur 1–3 weeks following vaccination which supports the claim that vaccination was the aetiological factor in this case. To the best of our knowledge, this is the first report of this complication with either of the two individual vaccines. The fact that there are many case reports describing a diverse variety of systemic reactions of an autoimmune nature associated with both vaccines also adds to the claim. Those cases of optic neuritis attributed to other vaccines and to post infectious optic neuritis generally did better with corticosteroids than with other therapies. Our case supports the claim that vaccination was the aetiological factor in this case.

Comment

Other authors also report similar cases. A review of the literature supports the claim that vaccination was the aetiological factor in this case. The B-cell response following vaccination is considered to be the most significant risk factor. The likelihood of a clinical correlation between vaccination and optic neuritis is very low. The B-cell response following vaccination is considered to be the most significant risk factor. The likelihood of a clinical correlation between vaccination and optic neuritis is very low.

REFERENCES


COMMENT

The viral infection of hepatitis B vaccine has been associated with a diverse range of isolated adverse reactions but ocular complications are exceedingly rare. Granell 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 epitheliopathy (APMPE) have been described. Vaccinated vaccine derived antibodies and immune responses to the vaccine have been associated with bilateral optic neuritis.

Reactions of an autoimmune nature associated with retinal complications following vaccination are unexplained. The role of the immune system in the pathogenesis of ocular complications following vaccination is unknown but various hypotheses exist including immune complex mediated demyelination or neurotoxicity, antigenic mimicry between the vaccine and the nervous system, or several of these mechanisms occurring simultaneously. Neurotoxicity is associated with the MMR vaccine and the monovalent rubella vaccine. Neurological complications also occur 1–3 weeks following vaccination which supports the claim that vaccination was the aetiological factor in this case. To the best of our knowledge, this is the first report of this complication with either of the two individual vaccines. The fact that there are many case reports describing a diverse variety of systemic reactions of an autoimmune nature associated with both vaccines also adds to the claim. Those cases of optic neuritis attributed to other vaccines and to post infectious optic neuritis generally did better with corticosteroids than with other therapies. Our case supports the claim that vaccination was the aetiological factor in this case.
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).1–5 Patients may tolerate large cumulative doses (up to 3923 g) of hydroxychloroquine without developing retinopathy.1 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.6–9 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 (6.3 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 pseudocolorimetric 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 pigmentedary 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.10 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.11 All cases of preretinopathy completely resolved after the discontinuation of hydroxychloroquine.5 Five cases of true hydroxychloroquine retinopathy reported in the literature demonstrated that discontinuation of hydroxychloroquine resulted in stabilisation but not resolution of the retinopathy.5 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 therapy, (2) were treated with daily doses >400 mg/day, or (3) exceeded a daily dosage of 6.5 mg/kg of body weight/day.2 Our patient’s daily dose never exceeded 400 mg/day or 6.3 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.

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Figure 1 (A) Colour fundus photograph of the right eye at presentation showing subtle pigmentary changes (arrows) of the retinal pigment epithelium inferior to the fovea. (B) Fluorescein angiogram of the right eye demonstrating transmission defects inferior to the fovea consistent with toxicity.

Figure 2 (Upper) Pattern standard deviation as calculated by Humphrey 10-2 automated static perimetry at presentation showing bilateral paracentral scotoma. (Lower) Follow up visual fields 10 months after cessation of hydroxychloroquine therapy showing partial resolution of the scotoma.
A 12 year old girl was referred to Moorfields Eye Hospital for a second opinion regarding her deteriorating vision following bilateral trabeculectomies with adjunct mitomycin C for raised intraocular pressure.

At the age of 5 years, she was diagnosed as 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 on 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, iris, 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 no further 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 in view of the transient visual obscurations which suggested critically impaired perfusion of the disc. To prevent even further vision loss, a lumbar 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 lumbar 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 CSF opening pressure was recorded as 5.5 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 following 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 refilling with Healon-GV. Her IOPs stabilized 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 needling 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 has 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.

This is a long girl 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 procedure led us to consider that hypotony may have played a role in her deteriorating vision.
surgical procedures made us suspect that she had an inherent problem in wound healing and scleral rigidity, especially as she displayed such dramatic changes in axial lengths (1.6–1.8 mm difference in pre- and postoperative lengths). We therefore wondered if she had an underlying connective tissue disorder.

She was found to have Ehlers-Danlos syndrome type II (EDS II). Examination revealed her to have joint hypermobility (Fig. 2), with a high joint hypermobility score (Table 1) of 7/9. In addition, she showed evidence of skin striae in the lumbar region, thighs and breasts with a papyraceous scar on her right knee, and gave a history of bruising easily with prolonged bleeding when cut. Echocardiography however was normal, with a normal palate, no spinal defects, no arachnodactyly, and a normal arm span to height ratio. However, there was a strong family history of joint hypermobility.

EDS is a heterogeneous group of genetically determined disorders of connective tissue affecting skin, ligaments, joints, blood vessels, and internal organs, of which at least nine subtypes exist, with the following diagnostic triad: extensible skin (extreme examples being described in fairground artists, such as the ‘elephant man’), hyperextensible joints, and connective tissue fragility, and hypermobile joints. Although ocular complications in EDS are mostly associated with EDS type I, a few ocular problems in EDS II have been documented such as prominent epicanthal folds, redundant skin on the upper eyelid, blue sclera, and the absence of periorbital fat. The wound healing process in EDS II is known to be abnormal in the skin because of the anomalous development of scar formation due to aberrant collagen fibrils. An abnormal healing response to the initial filtration surgery in this young girl might have been exaggerated with the use of mitomycin C. Cutaneous wounds in EDS II initially appear with gaping edges—so-called ‘fish mouths’ and heal slowly, and when scar formation eventually occurs, are characteristically shiny and stretched—papyraceous. Our findings on initial exploration of the surgical site in the eye, in this case, revealed a friable, disintegrating scleral flap which would have permitted unguarded passage of aqueous into the subconjunctival space—hence the occurrence of hypotony when combined with a non-healing conjunctiva.

Ocular hypotony is associated with various complications including hypotony maculopathy, resulting in a permanent reduction in vision and acuity. This occurrence has previously been attributed to reduced scleral rigidity, as seen in young and myopic eyes where a reduction in axial length has been documented following filtration surgery. Another mechanism put forward for the vision loss in these eyes, has been compression of the submacular sclera due to the resting tone of the inferior oblique muscle and decreased scleral rigidity. The incidence of post filtration ocular hypotony following full thickness procedures is 17–41%. This incidence was reduced significantly with the advent of guarded, partial thickness techniques, but the recent introduction of antiscarring agents such as mitomycin C and 5-fluorouracil has led to its increase once again. We believe that our patient had a complex and extreme response to filtration surgery with adjuvant mitomycin C complicated by her reduced scleral rigidity secondary to EDS II, as evidenced by her large eyes and marked changes in axial length in association with reduction in intraocular pressure.

The development of severe vision loss in our patient was associated with the occurrence of transient obscurations 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 is a recognised complication that is fortunately very rare. Experimental models have suggested that axoplasmic transport is delayed in cases of hypotony induced optic disc swelling giving rise to the same features histologically as seen with raised intracranial pressure papilloedema—for example, axonal swelling, accumulation of mitochondria, and cystoid 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 temporary improvement in the 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 suggests that not only might they have elastic skin but also “elastic globes”.

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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 point each hand (max = 2)</td>
</tr>
<tr>
<td>Passive hyperextension of thumb to flexor aspect of wrist</td>
<td>1 point each hand (max = 2)</td>
</tr>
<tr>
<td>Hyperextension of elbow beyond 10°</td>
<td>1 point each arm (max = 2)</td>
</tr>
<tr>
<td>Hyperextension of knee beyond 10°</td>
<td>1 point each leg (max = 2)</td>
</tr>
<tr>
<td>Forward flexion of trunk, palms flat on floor in front, knees in extension</td>
<td>1 point</td>
</tr>
</tbody>
</table>
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 cal, or oblique, and a week history of right described as inconsistently horizontal, verti-referred to the eye department with a week history of right superior rectus overaction (Fig 1). Secondly, episodes of right upper lid retraction, esotropia, and hypertropia were observed (Fig 1). Although modest variability of symptoms (for example, intermittent diplopia) has previously been described, we can find no reports in the literature of such marked fluctuations in ptosis due to third nerve compression. Furthermore, in this case upper lid retraction and, on eye movement testing, esotropia and hypertropia—that is, excessive activity in third nerve innervated muscles, was observed. A similar phenomenon of co-existing inappropriate neural discharge and block is seen in hemifacial spasm—the third nerve hyperactivity course of resolution to that seen following decompression of the third nerve for hemifacial spasm—the third nerve hyperactivity has been demonstrated. Although this case is important for two reasons. Firstly, the ptosis and ophthalmoplegia were notably variable (Fig 1). Secondly, we report a case in which the facial nerve is the causative factor. This case is important for two reasons. Firstly, 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 emphasises the importance of excluding compression by intracranial aneurysm in a patient with variable signs which could be explained by a partial third nerve palsy. MARGARET R DAYAN

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Letters

COMMENT
The early recognition of oculomotor nerve compression by a posterior communicating artery aneurysm is essential given that the mean interval between the onset of diplopia and bleiding is 4 weeks in patients who develop a subarachnoid hemorrhage. Although the retro-ornital pain was typical, this case is unusual for two reasons. Firstly, the ptosis and ophthalmoplegia were notably variable (Fig 1). Secondly, episodes of right upper lid retraction, esotropia, and hypertropia were observed (Fig 1). Although modest variability of symptoms (for example, intermittent diplopia) has previously been described, we can find no reports in the literature of such marked fluctuations in ptosis due to third nerve compression. Furthermore, in this case upper lid retraction and, on eye movement testing, esotropia and hypertropia—that is, excessive activity in third nerve innervated muscles, was observed. A similar phenomenon of co-existing inappropriate neural discharge and block is seen in hemifacial spasm—the third nerve hyperactivity course of resolution to that seen following decompression of the third nerve for hemifacial spasm—the third nerve hyperactivity has been demonstrated. Although this case is important for two reasons. Firstly, 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 emphasises the importance of excluding compression by intracranial aneurysm in a patient with variable signs which could be explained by a partial third nerve palsy. MARGARET R DAYAN

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CASE REPORT

A 59 year-old white man was admitted to our outpatient department in July 1997 with acute onset of vision loss in his left eye within the previous 2 weeks. The patient had been diagnosed with HIV infection in 1991. No opportunistic infections defining AIDS had been present so far. Antiretroviral therapy consisted of zidovudine since June 1991 as well as additional indinavir and nevirapine since January 1997. Best corrected visual acuity was right eye 20/20 and left eye 6/20. Further ophthalmic examination revealed a left afferent pupillary defect and an absolute central scotoma in the left eye. Indocyanine green angiography and fluorescein angiography disclosed a slight cedema of the optic disc without leakage. Visual evoked potentials (VEP) of the left eye showed reduced central amplitudes and prolonged latencies. Ophthalmic examination of the right eye at the time of presentation was normal.

(LHON) Therefore the patient was specifically asked for past or present additional medical or environmental factors postulated to trigger LHON. The patient denied any alcohol or tobacco consumption. Exposure to chemicals, solvents, toxins, or dangerous factory processes was not reported. Significantly larger amounts of foods with high cyanide content were not described. Additional metabolic or neurological diseases were not present.

COMMENT

LHON is a bilateral acute or subacute optic neuropathy caused by mutations in the mtDNA. Point mutations of the mtDNA at nucleotide position 11778, 3460, and 14484 involving NADH dehydrogenase (ND) subunits 1, 4, and 6, respectively, of respiratory chain complex I, are responsible for the mitochondrial changes that depend more on the duration of zidovudine application than on the applied dose. The onset of visual loss in LHON typically occurs between the ages of 15 and 35 years in most pedigrees. We hypothesise that the exceptionally long period of zidovudine treatment since diagnosis of HIV infection in 1991 and/or the infection itself may account for the unusual late expression of LHON in our patient. This is the first report on a patient with LHON suffering from additional HIV infection.

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Acute onset comitant esotropia as presenting sign of demyelinating disease

EDITOR—Comitant esotropia is most often acquired at an early age, and is generally benign. However, acute comitant esotropia has also been reported in patients with intracranial diseases including hydrocephalus, Arnold-Chiari malformations, and central nervous system tumours. I describe a case of acute onset comitant esotropia associated with acute demyelinating disease.

CASE REPORT

An 11 year old boy was referred for evaluation of diplopia. The patient stated he woke up the previous day with double vision. He denied decreased visual acuity, photophobia, pain with eye movements, headache, or nausea and vomiting. One week before the onset of esotropia, he had an upper respiratory infection with a fever that resolved without antibiotics. His mother also noted that his diplopia had gradually resolved. Lumbar puncture was not performed because neurological examination was entirely unremarkable. Follow up ocular examination revealed normal ocular alignment and motility.

COMMENT

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 neurological disease. It has been reported in association with hydrocephalus.1 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.
esotropias. Patients may not necessarily present with papilloedema even when hydrocephalus is present.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.1 1 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.1 2 3 4 5 6 7 Tumours in this group include cerebellar astrocytomas, and medulloblastomas, as well as pontine gliomas.1 An A-pattern esotropia is unusual in these patients. 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. 1–7 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.

Acute comitant esotropia has been associated rarely with a hypertensive haemorrhage in the thalamus.1 However, these patients also present with other neurological signs leading rapidly to the diagnosis. In addition, acute comitant esotropia has been reported rarely with myasthenia gravis and seizures.1 8 9 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.7 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 disease.