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The use of magnetic resonance imaging in the diagnosis of suspected giant cell arteritis

Giant cell arteritis (GCA) is a vasculitis of unknown origin that has a predisposition for the cranial arteries in the elderly. It has potentially devastating visually complications and produces a broad range of symptoms and signs that mimic many other medical and surgical conditions. Blood tests reflect the underlying inflammatory process, yet the erythrocyte sedimentation rate (ESR) may be normal in 8% of patients with biopsy proved GCA.¹ Nevertheless, making a definitive diagnosis has importance therapeutically

as patients are committed to a lengthy oral corticosteroid regimen. Non-invasive techniques, such as colour Doppler or duplex ultrasonography, have been studied in an attempt to improve patient preselection for temporal artery biopsy (TAB).²⁻³ Magnetic resonance imaging (MRI) has been shown to improve the diagnosis of early Takayasu arteritis.⁴ More recently several case reports have described the diagnostic potential of MR angiography and gadolinium contrast MRI in demonstrating the vessel changes of GCA.⁵⁻⁶ We compared the ability of MRI to detect changes in the temporal arteries with TAB in patients clinically suspected of having GCA.

Methods and results

A prospective, pilot, single masked study of seven female patients (age range 60-88 years, mean 76 years) with suspected giant cell arteritis, and two age matched healthy controls was undertaken. Local research ethical approval and informed written consent were obtained. All patients underwent a standard clinical examination including a detailed history and clinical examination. Investigations included ESR and C reactive protein (CRP). Each patient was given a GCA criteria "score" based on the 1990 ACR (American College of Rheumatology) classification⁷ (Table 1). Within 48 hours of presentation patients underwent a unilateral temporal artery MRI scan on a 1.5T scanner using a surface coil and small field of view. T₁ and T₂ weighted images perpendicular to the temporal artery and a time of flight sequence were obtained. The MRI visualised the location of the temporal artery that was

subsequently biopsied in a standard manner within 24 hours of the scan. Two healthy age matched controls also underwent a medical assessment, ESR and CRP, and an MRI as detailed above, but a TAB was not performed. The MRI scans were reported by an independent, masked neuroradiologist.

Each patient's ACR criteria "score" and the results of the MRI scan and TAB are shown in Table 2. The finding of three out of five ACR criteria is associated with a 94% sensitivity and 91% specificity for the diagnosis of GCA.⁷ There were two positive and one equivocal TAB result from the seven patients, but no positive MRI findings were identified. However, when using the ACR criteria as "gold standard," there were two true negative MRI scan results compared with three false negative scan results. The two remaining MRI scans were described as equivocal, in comparison with the ACR criteria—one patient was positive for GCA and the other patient's ACR criteria "score" was negative for GCA. From the data the negative predictive values of MRI scanning and TAB for GCA were 40% and 50%, respectively. Of the five patients who showed a prompt response to oral corticosteroid, the MRI scan was negative in four and equivocal in the other.

Comment

Although our study sample was small our findings suggest that MRI scanning was unable to distinguish between a normal and an affected artery. We conclude that there is no potential for the use of MRI scanning without contrast enhancement in the evaluation of patients with suspected GCA.

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Table 1 1990 American College of Rheumatology criteria for the classification of giant cell (temporal) arteritis (traditional format)

Criterion	Definition
1 Age at disease onset >50 years	Development of symptoms or findings beginning at age 50 or older
2 New headache	New onset of or new type of localised pain in the head
3 Temporal artery abnormality	Temporal artery tenderness to palpation or decreased pulsation, unrelated to arteriosclerosis of cervical arteries
4 Elevated ESR	>50 mm in the first hour by the Westergren method
5 Abnormal artery biopsy	Biopsy specimen with artery showing vasculitis characterised by a predominance of mononuclear cell infiltration or granulomatous inflammation, usually with multinucleated giant cells

Table 2 Summary of results

Patient	Age	ACR criteria score	Prompt response to steroids	TAB	MRI scan
1	78	5	Yes	Positive	Negative
2	67	4	Yes	Negative	Negative
3	83	2	Yes	Equivocal	Negative
4	88	3	Yes	Negative	Negative
5	60	2	No	Negative	Negative
6	67	2	No	Negative	Equivocal
7	87	4	Yes	Positive	Equivocal
8	67	NA	NA	NA	Negative
9	69	NA	NA	NA	Equivocal

TAB=temporal artery biopsy; ACR=American College of Rheumatology, MRI=magnetic resonance imaging; NA=not applicable, *=control.

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Bilateral ischaemic optic neuropathy and stroke after multiple bee stings

Despite the common occurrence of insect stings and local and systemic allergic reactions,¹ there are few reports of optic neuropathy or stroke following bee or wasp stings and, to our knowledge, there has been no report of both cerebral infarction and optic neuropathy occurring in the same patient after such an event. We report on a middle aged woman who sustained both a stroke and ischaemic optic neuropathy after multiple bee stings.

Case report

A 57 year old white woman reported being stung by 30–40 bees, identified as Africanised honey (killer) bees, in the back of her neck, head, right eye, face, and right arm. She was treated with intravenous antihistamines and antiemetics at a local emergency room and released.

Two days later, the patient experienced a severe headache with nausea and vomiting and noticed a left homonymous visual field loss. She went to see her primary doctor and while there became unresponsive, leading to hospitalisation. Head computed tomography (CT) showed a right occipital ischaemic infarct.

Shortly thereafter, the patient experienced acute nausea and vomiting with neck rigidity and was readmitted. A head CT scan and brain magnetic resonance image (MRI)/magnetic resonance angiography (MRA) were performed showing a large right temporo-occipital haemorrhagic infarct (fig 1A, B). An ocular examination revealed best corrected visual acuity (BCVA) of 20/20–1 right eye and 20/30–2 left eye at distance and 20/20 right eye and 20/200 left eye at near, with left homonymous hemianopia, a left inferior altitudinal defect, and bilateral arcuate defects (fig 1C) with bilateral haemorrhagic disc oedema.

Past medical and surgical history are significant only for controlled arterial hypertension and pseudophakia.

Neuro-ophthalmic examination 5 weeks after her sting episode showed BCVA of 20/15 right eye and 20/25 left eye at distance and 20/20 right eye and 20/30+1 at near. Amsler grid and automated perimetry showed a left homonymous hemianopic defect with a right inferior arcuate defect and a left inferior altitudinal defect.

Pupil examination showed isocoria with a 0.3–0.6 log unit relative afferent pupillary defect in the left eye. Motility was unremarkable, as was anterior segment both eyes. Intraocular pressures were 20 mm Hg right eye and 18 mm Hg left eye. Funduscopic examination showed bilateral disc oedema with pallid swelling superiorly and temporally in both eyes and peripapillary

Figure 1 MRI (A), MRA (B), and Humphrey 30-2 visual fields (C) of our patient.

haemorrhage and cotton wool spots in both eyes consistent with anterior ischaemic optic neuropathy (AION). Both maculas were unremarkable without exudative changes. Both retinas were flat with normal vasculature out to the periphery.

Three months after the sting event, the patient reported some improvement of peripheral vision, and repeat visual fields improved slightly inferiorly but were otherwise unchanged. Both optic discs were now flat and showed superior temporal pallor with corresponding nerve fibre layer dropout.

Comment

In their literature review of five cases and report of two additional cases of optic neuropathy occurring after bee and wasp sting, Maltzman, *et al*² describe common characteristics, such as acute to subacute onset of symptoms, moderate to severe visual loss followed by significant recovery (except in one case of a sting directly to the eye); oedematous and haemorrhagic optic discs, and central or caecocentral scotomas. Although our patient had subacute vision loss associated with haemorrhagic disc oedema, her case differs because of minimal recovery of vision and altitudinal visual loss consistent with an ischaemic neuropathy, rather than a transient optic neuritis.

Seven cases of wasp and bee sting associated cerebral infarction were found in the literature.^{3–9} Reported neurological

complications included seizure, hemiparesis, aphasia, apraxia, dysarthria, ataxia, and coma, none of which were experienced by our patient. None of these patients had a full eye examination, although in one patient⁸ a right homonymous superior quadrantanopia was demonstrated (table 1).

The pathophysiology explaining the associated stroke is unknown. Hypotension caused by anaphylaxis may certainly induce cerebral and optic nerve ischaemia; however, this was not documented in our case. Similar to acute myocardial infarction after hymenoptera stings, it has been suggested that vasoconstriction secondary to mediators released after the sting, aggravated by exogenous adrenaline, and platelet aggregation also contribute to cerebral ischaemia.⁸ Bee venom itself contains histamine, thromboxane, leucotrienes, and other vasoactive and inflammatory mediators. In our patient, we postulate that the systemic immune mediated reaction to the bee sting caused vasoconstriction and a prothrombotic state with subsequent ischaemia leading to both the stroke and AION. In addition, a neuropharmacological (sympathetic) mechanism of endothelial permeability involving the cerebral vasculature with a concurrent systemic thrombogenic or immune response has also been postulated.^{5–6}

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Table 1 Reports of cerebral hypoxia and infarction following bee/wasp sting

Author/ref	Age/sex	Type of stings: location	Onset of neurological deficit	Examination findings and symptoms	Eye examination	MRI/CT findings	Treatment	Recovery
Day ³	36/M	Wasp: multiple on neck, face, and arms	<1 hour	Headache, seizure, right hemiplegia, coma	Equal and reactive pupils	NR: necropsy showed left haemorrhagic cortical infarct	Cortisone, antihistamines, phenobarbital	Deceased
Starr and Brasher ⁴	37/M	Wasp: 3 stings on arms	<1 hour	Seizure, right hemiplegia	NR	Left cerebral infarction (CT done 14 months later)	Barbiturates, corticosteroids, adrenaline	Partial right hemiplegia, one seizure
Riggs <i>et al</i> ⁵	38/M	Wasp: multiple on left face and neck	2 days	Right hemiplegia, dense global aphasia	NR	Ischaemic infarction in the distribution of the left MCA; angiogram: left ICA occlusion	NR	NR
Riggs <i>et al</i> ⁶	52/M	Wasp: single, location NR (previous history of wasp sting allergy)	A few hours, with worsening 24 days later	Anaphylactic shock with respiratory arrest, slurred speech and left hemiparesis initially, then 24 days later, acute obtundation and quadriparesis	NR	Initially, three small focal ischaemic infarcts, two in the right centrum semiovale and one in the right temporal lobe. After worsening, diffuse bilateral ischaemic white matter lesions and left parietal and insular cortical infarctions. MRA and angiogram: complete and near complete occlusions of the right and left ICA, respectively	IV adrenaline, methylprednisolone, diphenhydramine	NR
Speech <i>et al</i> ⁷	30/M	Bee: single, location NR	<1 hour	Decerebrate posturing, extensor plantar reflexes, left hemiparesis, hyporeflexia; after coma, patient had motor apraxia and left sensory neglect	NR	Normal MRI and CT	IV diphenhydramine, steroids and nebulised β_2 agonist and anticholinergic medications	Residual ideomotor apraxia
Crawley <i>et al</i> ⁸	30/F	Wasp: left arm	<1 hour	Facial and arm swelling, widespread urticaria, acute pulmonary oedema, visual loss	Right homonymous superior quadrantanopia	SPECT: hyperperfusion of the left dorsolateral frontal cortex, but no areas of hypoperfusion or other abnormalities Left occipital ischaemic infarct	SQ adrenaline, IV gelofusine, IV hydrocortisone, IM chlorpheniramine, IV furosemide	Full recovery from quadrantanopia
Bhat <i>et al</i> ⁹	35/M	Bee: multiple "all over the body"	<1 day	Multiple swellings all over the body, vomiting, dysarthria, tinnitus, vertigo and swaying gait, hypertension, bilateral cerebellar signs, rhabdomyolysis with acute renal (respiratory?) failure	No papilloedema	Bilateral cerebellar haemorrhagic infarct	Dexamethasone, antihistamines, mannitol, insulin, haemodialysis	Deceased
Present report	57/F	Bee: multiple on neck, head, R eye, R side of her neck, face and R arm	2 days	Nausea, vomiting, vision loss	BCVA of 20/15 right eye, 20/25 left eye; left homonymous hemianopia, left inferior arcuate and right altitudinal defect; Bilateral oedema (right eye < left eye) w/ pallid haemorrhagic swelling	Haemorrhagic infarct 2 days post-ischaemic stroke	IV antihistamines and antiemetics	Left homonymous hemianopia with inferior arcuate defects; central vision unaffected right eye and only mildly affected left eye

NR = none reported.

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Cause of V pattern strabismus in craniosynostosis: a case report

Strabismus is a common association in patients with craniosynostosis or craniofacial dysostosis (60–70%).^{1–3} V pattern exotropia is the most common ocular motility problem.

Various theories have been proposed to explain the cause of the V pattern and surgical attempts to correct it with weakening procedures of the inferior oblique have been disappointing.^{2,3}

This is a case report of one child with this disorder who underwent orbital computed tomography (CT) scans and had a marked improvement of the V pattern following strabismus surgery based on the CT findings.

Figure 1 Preoperative V pattern exotropia, over-elevation in adduction, under-depression in adduction in both eyes.

Figure 2 Preoperative coronal views on CT scan of both orbits showing evidence of rectus muscle heterotopy. A vertical line joining the centre of the belly of the vertical rectus muscles shows the relative temporal displacement of the superior rectus muscle compared to the nasal displacement of the inferior rectus muscle. A horizontal line joining the centre of the belly of the horizontal rectus muscles shows the relative inferior displacement of the lateral rectus muscle compared to superior displacement of the medial rectus muscle.

Figure 3 Intraoperative photograph. The left eye is adducted with a muscle hook placed under the lateral rectus muscle (LR). The lower line highlights the downward slanting of the left lateral rectus muscle. A curved ruler is used to show the normal horizontal path of the lateral rectus muscle.

Figure 4 Postoperative clinical pictures showing improvement in V pattern exotropia with improvement in versions in adduction.

Case report

This child with craniosynostosis had undergone six previous cranial surgeries. She had three strabismus surgical procedures including anterior transpositions of the inferior

obliques in an attempt to correct a large V pattern. She presented to us with a chin up position, V pattern exotropia (60 prism dioptres), over-elevation in adduction, limitation of depression in adduction, and concomitant hypertropias in side gazes (fig 1). Objective fundus excyclotorsion was noted.

Orbital imaging demonstrated that all extraocular muscles in each eye were present, normal in size and shape but anatomically displaced. The extraocular muscles in the left eye were rotated clockwise and in the right eye were rotated counterclockwise (fig 2). Ineffectiveness of inferior oblique weakening procedures and the presence of muscle heterotopy led us to consider that the over-elevation in adduction was most likely related to the anatomical displacement of the rectus muscles.

Surgical exploration confirmed muscle heterotopy. The lateral recti were found slanting inferiorly (fig 3). Repositioning of the lateral recti superiorly to a more horizontal position and suturing the superior border of the muscle belly to the adjacent sclera about 18 mm from the limbus using a

non-absorbable suture was the first surgical procedure performed by us on this patient. This led to some improvement of the V pattern. This was followed by recession and nasal repositioning of the superior recti suturing the nasal border of the muscle belly to the adjacent sclera about 18 mm from the limbus using a non-absorbable suture. This achieved good alignment in the primary position and eliminated the anomalous chin up position, markedly reduced the V pattern, eliminated the over-elevation in adduction, and improved depression in adduction (fig 4).

Comment

V pattern strabismus in craniosynostosis may be related to anatomical malposition of the rectus muscles. This may be documented by orbital imaging, which could also aid in planning the surgical approach. In these cases the over-elevation in adduction and under depression in adduction may be due to the anatomical displacement of the rectus muscles.²

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West Nile virus chorioretinitis

West Nile virus has been described in Africa, Europe, the Middle East, west and central Asia, Oceania, and has emerged in recent years in temperate regions of Europe and North America.¹ West Nile virus was first isolated from a febrile adult woman in the West Nile District of Uganda in 1937 and became recognised as a cause of severe human meningoencephalitis in elderly patients during an outbreak in Israel in 1957.² In 1999, the plight of city birds and a collection of human encephalitis cases in New York heralded the arrival of West Nile virus on this side of the Atlantic. From 1999 through 2001, there were 149 cases of human West Nile virus infection in the United States, including 18 deaths, but in 2002 alone more than 3500 cases and 200 deaths were reported.³ In 2003, over 9000 cases were reported with more than 300 cases of neuroinvasive disease.³

The Centers for Disease Control notes that neuroinvasive disease includes those cases resulting in meningitis, encephalitis, or meningoencephalitis.³ Cases with ocular involvement should probably be included in this category as well. As our clinical experience in such cases evolves so does our understanding of the ophthalmic manifestations of the disease. Here, we present a case

of ocular involvement with West Nile virus, highlighting the typical ocular findings.

Case report

An 80 year old man convalescing in a nursing home from neurological complications of recently acquired West Nile virus meningoencephalitis presented with bilateral visual loss of unspecified duration. The patient had been hospitalised 4 months previously for serologically confirmed West Nile virus encephalitis. His infectious course was complicated by residual right sided paresis, dysarthria, and generalised mental status changes with dementia. Over the following months as he regained his mental faculties he complained to family members of decreased vision and central scotomas, worse in his left eye than right. His best corrected visual acuity at this time was 20/40 in the right eye and 20/60 in the left eye. The patient's ophthalmic and medical histories were otherwise non-contributory. Biomicroscopic examination revealed normal anterior segments without inflammation and moderate nuclear sclerotic and cortical changes involving both crystalline lenses. Funduscopy examination revealed mild vitreous debris with moderately large areas of retinal pigment epithelial and choroidal atrophy in the posterior segment (fig 1A and B, right and left eyes, respectively) in addition to partially atrophic and pigmented chorioretinal foci throughout the retinal periphery (fig 2A and B, right and left eyes, respectively).

Over the next 3 months the patient developed problems with his activities of daily living at night and glare with lighting. Subsequent examination revealed progression of the lenticular changes and the patient was referred for cataract extraction. He returned 3 months later after uneventfully cataract surgery. He was not on any medications at this time. Best corrected visual acuity measured 20/30 in the right eye and 20/40 in

Figure 1 Moderately large areas of retinal pigment epithelial and choroidal atrophy with mild pigmentary disturbance of both foveae are present bilaterally, to a greater degree in the left eye (B) compared to the right (A).

Figure 2 Partially atrophic and pigmented chorioretinal foci analogous to those noted in previous reports are distributed throughout the retinal periphery in the right (A) and left (B) eyes respectively.

the left eye. Normal anterior segments without inflammation and well placed posterior chamber intraocular lenses were noted. The vitreous debris persisted and his funduscopy examination was without change bilaterally. Examination 6 months later and approximately 16 months after initial West Nile virus infection demonstrated stable ophthalmic findings and visual acuity.

Comment

Although ocular symptoms associated with West Nile virus were first reported in 1956 ocular findings in West Nile virus infection were first described in the medical literature soon after the West Nile virus epidemic in North America in 2002.⁴⁻⁸ Initial reports described analogous clinical findings consisting of mild anterior segment inflammation, vitritis, and discrete nummular outer retinal/choroidal lesions which were often linear in distribution and varied in appearance from "creamy whitish-yellow" to atrophic with various degrees of pigmentation.^{3-7,9} Mild retinal haemorrhage was also occasionally present. Fluorescein angiography revealed these "target" lesions to be hypofluorescent centrally and hyperfluorescent peripherally. Leakage from the optic nerve is sometimes present as optic neuritis and papilloedema may be associated with contiguous central nervous system involvement.⁸⁻¹⁰ Later reports confirmed these findings and suggested that active lesions associated with vitritis may appear "creamy" in nature eventually progressing to foci of well circumscribed chorioretinal atrophy as the disease becomes inactive and subsequently becoming more prominent with time.^{9,10} Occlusive vasculitis without chorioretinal findings has also been noted in an isolated case.¹¹

Various ocular inflammatory and infectious processes such as toxoplasmosis and juvenile rheumatoid arthritis have been associated with periods of recurrence and

exacerbation after intraocular surgery.¹² This highlights an important issue with regard to West Nile virus infection as the risk for neuroinvasive disease is higher for people 50 years of age and older, many of whom are currently or soon will be candidates for cataract extraction. Our patient did well with routine postoperative care and surveillance after uncomplicated cataract extraction in an eye previously affected by West Nile virus chorioretinitis. The eye remained quiescent without evidence of uveitis or reactivation of previously affected fundus lesions. Although surveillance would be recommended for these patients, our findings suggest that chorioretinitis associated with West Nile virus appears to be an acute self limited process without residual sequelae after subsequent intraocular surgery.

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Swimming goggles suck

We present a complication arising from the use of swimming goggles in a patient with glaucoma drainage blebs.

Figure 1 (A) (B) Right eye at 4 months postoperatively showing corneal dellen and nasal bleb extension (A) and the adjoining isthmus (B) with arrows at each end. (C) Regression of the right accessory bleb after needling, 5-fluorouracil, and topical steroids. (D) Left eye at 7 months postoperatively with smaller and slightly inflamed nasal accessory bleb. (E) Pressure transducer setup measuring "intragoggle" pressure using AD Instruments Powerlab (www.adinstruments.com) and IOP transducer (gold disc). (F) Transducer recording showing several goggle applications (positive pressure, "a" labels) and the transient negative pressure spikes produced on removing them ("r" labels); In area 1 of the trace, the goggles were overtight and in area 2 they were comfortable.

Case report

A 73 year old white man with poorly controlled primary open angle glaucoma underwent routine trabeculectomy with adjunctive 5-fluorouracil to the right eye, followed by the same procedure to the left eye 6 weeks later. Preoperatively the intraocular pressures were 28 mm Hg bilaterally and cup:disc ratios were 0.95 right, 0.8 left. Early post-operative intraocular pressure (IOP) in the right eye was low (5 mm Hg at weeks 2 and 6), but uncomplicated. The recovery of the left eye was uneventful, and at 3 months the IOPs were 10 mm Hg right eye, 12 mm Hg left.

However, at 4 months the patient presented with discomfort and redness in the right eye. A large extension of the bleb had formed at the nasal limbus, with an associated corneal dellen (fig 1A and B). The IOP had increased in the right eye, which was treated with a needling procedure and 5-FU injection, repeated 3 weeks later. Subsequently the bleb extension receded and the previously elevated right nasal conjunctiva was found to be firmly adherent to the underlying sclera (fig 1C).

He re-presented 7 months after the initial surgery with redness and swelling, this time in the left nasal conjunctiva (fig 1D). At this point the patient mentioned that he was a keen swimmer and inquired whether his problem could have been caused by the use of swimming goggles. He had resumed regular swimming 2 weeks before developing the right eye complication, then stopped. He had resumed again 3 months before developing the left.

With this in mind, we set out to investigate the pressure changes inside swimming goggles. With a pressure transducer fixed to one eyepiece (fig 1E), we recorded a comfortable range of -1 to -5 mm Hg, discomfort over -10 mm Hg and a maximum suction of -44 mm Hg. Upon removing the goggle, a transient negative pressure spike was also produced (fig 1F). Given these observations and the timing of the clinical events, we surmise that the patient's bleb extensions

were plausibly consequent upon his aquatic activities.

Comment

Previous reports of barotrauma sustained while wearing overtight goggles include suction petechiae¹ and changes in the eyelid skin,² but we are not aware of any information concerning the effects of swimming goggles on glaucoma drainage blebs. When goggles are applied, firm pressure displaces a small volume of air and creates a negative "intragoggle" pressure, the basis by which a seal is maintained. In a person who has undergone trabeculectomy, an increase in the transconjunctival pressure gradient could open up a weakness in the perimeter of the bleb and cause it to extend in the direction of least resistance.

Other experimental work has examined the pressure changes occurring in the mask space during scuba diving.³ This is a rather different system as the nose is included in the mask, allowing the pressure to be equalised by exhaling through the nose. The eye and periorbital structures can be subjected to significant negative pressures if this is not done, but the duration is usually limited by this pressure gradient acting across the tympanic membrane, causing pain and prompting the diver to ascend or equalise. Ocular barotrauma can result in subconjunctival haemorrhage and chemosis, and it has been recommended that patients wait a minimum of 2 months after glaucoma filtering surgery before resuming scuba diving.⁴

We do not believe patients who have undergone trabeculectomy need to cease swimming, but they should be aware that goggles may be able to produce excessive negative pressure if they form a very tight seal.

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Immune recovery disease: a case of interstitial keratitis and tonic pupil following bone marrow transplantation

Immune recovery disease results from an immunological response to circulating viral antigens in the host after bone marrow transplant (BMT) mediated immune reconstitution. It may also occur after successful antiretroviral therapy in patients with HIV and AIDS. We report a case of a child with severe combined immune deficiency (SCID) and disseminated varicella zoster virus (VZV) infection who developed interstitial keratitis and a tonic pupil after BMT.

Case report

An 8 month old male infant was referred to the ophthalmology clinic at Great Ormond Street because of suspected congenital glaucoma. The past ophthalmic and family history were unremarkable. The child was born with multiple congenital anomalies of the lower limbs which included bilateral tibial deficiencies, and an extreme talipes equinovarus of the right foot.

The child had a known history of disseminated varicella infection caused by SCID (fig 1). On examination it was noted that he had a generalised vesicular rash throughout his body extending to his eyelid margins. The eyes were white with clear corneas and he was alert, fixing and following well with full extraocular eye movements. Both pupils were reactive to light with no afferent pupillary defect. The anterior chambers were unremarkable and the intraocular pressure with the Perkins tonometer was 16 mm Hg bilaterally. Examination of the fundus, including cup to disc ratio was normal. He was reviewed periodically over the next 6 months while an inpatient undergoing treatment for SCID. During this period, he was persistently positive for VZV DNA in his blood determined by polymerase chain reaction (PCR) analysis but was negative for Epstein-Barr virus (EBV), herpes simplex virus (HSV), cytomegalovirus (CMV), and adenovirus DNA. He suffered a number of exacerbations of the varicella infection and was treated with systemic aciclovir, foscarnet, and cidofovir.

At 12 months of age he underwent allogeneic BMT from a one antigen mismatched unrelated donor following reduced intensity conditioning with fludarabine, melphalan and alemtuzumab. Engraftment was very rapid with neutrophils appearing by day 10. Before BMT the CD3+ CD4+ count was

0.04×10⁹/l but 7 weeks after BMT the CD3+ CD4+ count was 0.47×10⁹/l.

Four weeks after BMT his mother noted bilateral corneal haze which was more marked on the left eye. He was reviewed in his isolation cubicle with a hand held slit lamp and Perkins tonometer and was found to have bilateral corneal stromal haze and corneal vascularisation (fig 2). There was no conjunctival injection, and his intraocular pressure was 13 mm Hg in each eye. Both pupils were reactive to light. The lymphocyte count had recovered at this point to more than 1.0×10⁹/l.

An examination under anaesthesia was arranged and a diagnosis of interstitial keratitis without epithelial involvement was made. He was treated with intensive topical prednisolone acetate 1% (one drop every 2 hours) and cyclopentolate 1% twice daily to both eyes. Betaxolol 0.5% twice daily was prescribed prophylactically to prevent raised intraocular pressure which could exacerbate the corneal haze. The child was reviewed regularly and the stromal vascularisation was seen to regress. He was thus gradually weaned off the steroid drops to one drop daily and the cycloplegics were stopped. Three months after BMT his mother reported a change in pupillary size in the right eye. On examination the right pupil was mid-dilated and oval in shape, not reactive to light and there was no evidence of posterior synechiae (fig 3). A diagnosis of a right tonic pupil was made.

At the most recent review, 6 months following BMT he had clear corneas centrally in both eyes with some persistent peripheral stromal vessels, and a right tonic pupil. Unaided visual acuity was 0.60 logMAR with both eyes using the Cardiff acuity test (Keeler Ltd, Windsor, UK). There was a left fixation preference and amblyopia therapy was commenced with occlusive patches.

Currently, the child has an ongoing mild chronic graft versus host disease affecting the skin and intestine which is controlled with low dose systemic steroids. His systemic

Figure 1 Photograph of the child with severe systemic varicella a few days after admission.

Figure 2 Photograph of the left eye showing deep and superficial corneal stromal vessels.

Figure 3 Photograph of the right eye showing the tonic pupil and corneal stromal vascularisation.

medications also include aciclovir 120 mg four times daily.

Comment

Severe combined immune deficiencies (SCID) are a rare heterogeneous group of disorders characterised by severe T cell and B cell deficiency with low or absent antibody levels. They usually manifest in the first months of life with severe and recurring infections leading to death often by the age of 2 years.¹

Since 1968 these diseases have been successfully treated with haemopoietic stem cell transplantation.² Varicella infection has been associated with severe immune dysfunction following BMT and it has been shown that severe disseminated varicella infection causes ocular disease that mimics the sequelae of herpes zoster ophthalmicus.³ In the adult population, the commonest cause of interstitial keratitis is HSV infection whereas varicella infection is considered a rare cause.⁴ In children, although varicella infection is extremely common, ocular complications of this disease are rare.⁵

If keratitis develops in association with a childhood viral exanthem it is important to consider a number of possible infectious agents such as HSV, EBV, mumps, syphilis, Lyme disease, or tuberculosis in the differential diagnosis.⁴ In this setting, other documented complications in association with SCID include bilateral viral endophthalmitis,⁶ CMV retinitis, and optic neuritis.⁷ In this case the history and the physical findings were highly suggestive of the diagnosis and were confirmed by PCR testing. As far as we are aware this is the first case of varicella

associated interstitial keratitis and a tonic pupil occurring in a child with SCID following BMT.

The signs of early interstitial keratitis and a right tonic pupil were noted by the child's mother about 4 weeks after the allogeneic bone marrow transplant. We believe that an immunological response to pre-existing varicella was responsible for the development of the eye signs. This signifies a positive response from a nascent immune system in the recipient—an example of immune recovery disease.

There is experimental evidence to support this as it has been shown that whole lymphocyte and splenocyte transfer leads to herpes simplex keratitis in SCID mice.⁸ In other words SCID mice reconstituted with T lymphocytes of the CD4+ phenotype developed subsequent corneal lesions in relation to HSV infection. Conversely Mercadal *et al* have shown that unconstituted SCID mice remained lesion free when infected with HSV.⁸ This suggests that herpes simplex keratitis is a T cell mediated immunopathological reaction to virus in the cornea. In our case the corneal changes occurred following bone marrow reconstitution. Before BMT the CD3+ CD4+ count was $0.04 \times 10^9/l$ but 7 weeks following BMT the CD3+ CD4+ count was $0.47 \times 10^9/l$. The corneal changes become apparent at about 4 weeks after BMT. We believe that our case illustrates a similar mechanism in the human model in relation to varicella infection.

The tonic pupil developed as a consequence of a post-viral ganglionitis affecting the ciliary ganglion and the short posterior ciliary nerves, a rare but previously described complication of varicella infection.⁹ Other reported cases of ophthalmic immune recovery disease include a case of varicella zoster virus associated anterior stromal keratitis in a patient with AIDS¹⁰ and, in another case, in association with CMV retinitis.¹¹

Considering BMT, varicella zoster virus associated disease can be a frequent complication following autologous and allogeneic transplantation.¹² Other complications in relation to BMT include pseudomembranous conjunctivitis,¹³ keratoconjunctivitis sicca,¹⁴ cataracts,¹⁵ and severe graft versus host disease.¹⁶

This child did suffer a graft versus host disease-like rash at the time of the development of the keratitis. While it is possible that the keratitis was purely Graft versus host disease this seems unlikely, given that there was no conjunctival involvement and that the graft versus host disease was extremely mild. Furthermore, in this case the disseminated varicella infection preceded the BMT and formed the basis for identifying a severe immune deficiency in the child. It highlights the importance of frequent ophthalmic follow up in the immediate period following BMT as there is an increased risk of ocular disease.

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Occult macular dystrophy in an 11 year old boy

Occult macular dystrophy (OMD) is an inherited macular dystrophy characterised by a progressive decline of visual acuity without visible fundus abnormalities.^{1–3} In these patients, the fluorescein angiograms and conventional full field electroretinograms (ERGs) are normal, but the amplitudes of the focal macular ERGs and multifocal ERGs are significantly reduced but only in the central retina.^{1–4}

The age at the onset of symptoms in OMD patients is relatively old,^{1–3} and the first visit to the hospital is aged 20 years or more with the youngest being 16 years in most cases. Here, we present an 11 year old boy who was diagnosed as having OMD because of the results of electrophysiological and psychophysical tests.

Case report

An 11 year old boy was referred to our hospital with a complaint of progressive decline of vision in both eyes. His corrected visual acuity was 20/25 in both eyes at 6 years of age, but had decreased to 20/33 at 10 years of age. Family history revealed no other members to have any eye diseases. At the initial examination, his visual acuity was 20/40 right eye and 20/33 left eye with –3.0 dioptres (D) in both eyes. The fundus examination and fluorescein angiograms were normal (fig 1). The peripheral visual fields were intact but a relative central scotoma was detected with the I-2 target within 10 degrees in both eyes. A moderate red-green defect was found on the Ishihara pseudoisochromatic plates, Hardy-Rand-Rittler pseudoisochromatic plates, and Farnsworth-Munsell 100 hue test.

The amplitude of full field ERGs were within the normal range for both rod and cone components (fig 2A). However, focal macular ERGs with 5, 10, and 15 degrees stimulus spots³ were severely reduced and essentially absent (fig 2B). The multifocal ERGs⁴ demonstrated a loss of local responses in the central retina (fig 2C).

Psychophysical rod and cone sensitivity was performed on his right eye with 31 test points across the 60 degree horizontal meridian using a previously described method.⁵ The cone sensitivities were severely affected in the central retina but fell within the normal range in the periphery (fig 2D). The rod sensitivities were at the lower borderline at almost all locations tested (not shown).

At present (August 2003, 13 years old), his acuity has decreased to 20/50 in both eyes, but his fundi still remain normal in both eyes.

Comment

This boy had a progressive decrease of visual acuity in both eyes, and his fundus examinations and fluorescein angiograms were completely normal. The amplitude of the conventional full field ERGs were also within the normal range for both rod and cone components. However, focal macular cone ERGs and multifocal ERGs were severely reduced in the central retina. Results of psychophysical perimetry showed a reduction of cone sensitivity but only in the central retina. These findings are consistent with the clinical characteristics of OMD which we have previously reported.^{1–3,4}

OMD in children is very rare.^{1–3} In our 42 consecutive OMD patients seen at the Nagoya University Hospital from 1988 to 2003, the age at initial visit to the hospital ranged from 16 to 74 years (mean 45.8 years), and 95.2% of patients visited the hospital at 20 year old or more. To the best of our knowledge, this boy is the youngest case with OMD reported anywhere.

We would like to emphasise that OMD can be found even in children. Because the fundus examination and full field ERGs are normal in these patients, these children are apt to be misdiagnosed as optic nerve disease, central nerve disease, or psychological

Figure 1 Fundus photographs (upper) and fluorescein angiograms (lower) of the 11 year old boy.

Figure 2 Results of conventional full field ERG, focal macular cone ERG, multifocal ERG, and cone perimetry in our patient. Full field ERGs, focal macular cone ERGs, multifocal ERG, and cone perimetry are recorded with previously reported methods.³⁻⁵

disorders. Focal or multifocal ERG techniques are the only key to diagnose this rare type of macular dystrophy.

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Pseudomonas aeruginosa microbial keratitis secondary to cosmetic coloured contact lens wear

Cosmetic coloured contact lenses are worn to give the appearance of a different or unusual eye colour and about 60 000 people in the United Kingdom obtain these types of contact lenses through eye care professionals.¹ A subset of these lenses—those with no optical power ("plano" coloured lenses)—falls outside legislation designed to restrict the sale of contact lenses to suitably qualified professionals. We report a severe case of microbial keratitis caused by *Pseudomonas aeruginosa* which has resulted in lasting visual impairment in a patient obtaining cosmetic coloured contact lenses from a fashion shop rather than through an eye care practitioner.

Case report

An 18 year old south Asian male student presented in December 2003 with a 2 day history of a foreign body sensation in his left eye. One day before presentation the eye had become slightly red. He had commenced the use of Brolene eye drops which had been purchased from a large chain supermarket. The eye then became painful with eyelid swelling and he presented to the local district general hospital the following day. He was diagnosed with a corneal ulcer and referred to our institution.

He reported a 12 month history of cosmetic coloured plano contact lens wear, having purchased the lenses from a fashion shop rather than through an eye care professional.

No counselling was provided at the point of purchase regarding a hygiene routine, care of lenses, or possible complications associated with their use. He wore the lenses 12 hours per day, 7 days per week without any overnight use. The lenses were designed to make the eye appear grey or blue (patient's natural eye colour was brown). There was no past medical or ocular history of note including amblyopia.

On examination the unaided vision was 6/6 in the right eye and 6/36 in the left eye. The left eye demonstrated a mid-peripheral corneal infiltrate in the 4 o'clock position with overlying 2.4 mm diameter ulcer, and surrounding stromal swelling (fig 1). There was a 0.5 mm height hypopyon. The intraocular pressure was within the normal range. The right cornea demonstrated a very small peripheral infiltrate with no significant anterior chamber reaction. Both posterior segments were unremarkable. A corneal scrape was performed with the Gram stain demonstrating a small quantity of neutrophils and Gram negative bacilli. Ofloxacin 0.3% drops were commenced every hour to the left eye. The peripheral infiltrate resolved with the corneal epithelial healed by day 10. Topical prednisolone 0.5% was commenced on day 4. A more central mid stromal corneal infiltrate encroaching on the visual axis developed on day 1 after admission and has gradually become less prominent during follow up over 3 months (fig 2) although the visual acuity remains reduced at 6/36. *Pseudomonas aeruginosa* was grown from the corneal scrape, sensitive to ciprofloxacin, ofloxacin, gentamicin, and ceftazidime. The right eye was not scraped, responded well to topical ciprofloxacin drops, and did not develop any scarring. The contact lenses and their cases were also investigated as there was a high degree of suspicion that clinically they would be contaminated. All grew *Pseudomonas aeruginosa* with a sensitivity profile identical to the corneal scrape specimen. Mixed coliform growth was also noted also in one of the contact lens cleaning solutions.

Comment

The use of cosmetic coloured plano contact lenses, sourced via non-professional suppliers is becoming increasingly common and fashionable. Their use over the past 12 months has increased fourfold and stores have reportedly sold more than one million pairs.² Their purchase is currently possible from non-eye care professional retailers without any ocular assessment, customised fitting, or verbal counselling regarding a hygiene routine, care of the lenses, or possible complications associated with their use. In addition, there is often no plan for follow up.

Figure 1 Large corneal infiltrate with overlying area of ulceration on presentation.

Figure 2 Residual central corneal infiltrate at 1 month after presentation.

Potential complications are the same as those for all contact lenses and have been documented in a recent case series in the United States.³ *Pseudomonas aeruginosa* microbial keratitis with vision loss requiring elective penetrating keratoplasty, presumed herpes simplex related corneal scarring causing legal blindness, acute iridocyclitis, corneal hypoxia, microcystic oedema, punctate keratopathy, corneal abrasions, and giant papillary conjunctivitis were all documented.

In the United Kingdom, the Opticians Act 1989 states that a person who is not a registered medical practitioner or registered optician shall not fit contact lenses. Plano (or "afocal") contact lenses are not included in this act because they have no optical power. The General Optical Council has received reports of these lenses being shared and exchanged between wearers and of sales staff demonstrating fitting on themselves before offering the lens to the purchaser.⁴ In November 2000 the General Optical Council submitted recommendations to the Department of Health arguing that primary legislation should be passed stipulating that the fitting and sale of plano contact lenses should also fall within the terms of the act. On 28 October 2003 Mr John Robertson, MP for Anniesland, Glasgow, moved a bill to amend the Opticians Act 1989 to include plano contact lenses in the restrictions already placed on the sale of other contact lenses.⁵

This case report highlights the potential complications of these lenses and supports legislation restricting their sale.

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Severe proliferative retinopathy in a patient with advanced muscular dystrophy

The patient is a 25 year old white man with Duchenne muscular dystrophy (DMD), complicated by respiratory failure requiring ventilatory assistance and impaired cardiac function. His ocular complaints were "floaters" and decreased vision over the preceding 6 weeks. He had no history of ocular disease or trauma. The patient's level of alertness was reported to routinely fluctuate but no new neurological findings were present. The best corrected visual acuity was count fingers in the right eye and 20/70 in the left eye. The intraocular pressures were 14 and 8 mm Hg. The anterior segment examination was unremarkable with no neovascularisation of the iris or angle. Biomicroscopy revealed bilateral vitreous haemorrhage. Indirect ophthalmoscopy showed the retinal periphery to be attached in both eyes. The optic discs and macula were partially obscured by haemorrhage. Fluorescein angiography revealed delayed filling and venous beading in both eyes, without central or branch, vascular occlusion. Hyperfluorescence, consistent with neovascularisation, was present along the temporal vascular arcades and at the optic discs. Fundus photography corroborated the angiographic findings (see figs 1 and 2).

Indirect laser with scleral depression resulted in full treatment of retina outside of the vascular arcades. Treatment appeared to have little effect on neovascular progression. Overwhelming anaesthetic risk prevented intraocular procedures. Both eyes progressed to subtotal traction retinal detachment and counting fingers vision.

Comment

The working diagnosis was retinal ischaemia secondary to hypoperfusion or pan-microvascular occlusive disease. The cardiac ejection fraction was 20% of predicted; the forced vital capacity was 14% of predicted and the forced expiratory volume in 1 second was 15% of predicted. We believe that cardiopulmonary compromise was a primary

Figure 1 Colour fundus photograph of the right eye depicting venous beading (arrow), neovascularisation of the disc (arrowheads), and vitreous haemorrhage.

Figure 2 Fluorescein angiogram of the right eye showing venous beading (arrow), leakage from neovascularisation on the disc (arrowheads) and blocking vitreous haemorrhage; the visible retina is attached.

contributor to the development of retinal neovascularisation. Arterial blood gas analysis was not available. The patient was on Coumadin for cardiac indications. He was not a diabetic and finger stick blood sugars were consistently in the low to normal range. Additional normal evaluation included erythrocyte sedimentation rate, anticardiolipin, C reactive protein, C3, C4, total complement, C1q complex, and a Raji assay. The presentation, appearance, and course were not typical for Terson's syndrome, Valsalva retinopathy, or Takayasu disease.

Duchenne muscular dystrophy is the most common X linked neuromuscular disorder. It has an incidence of one in 3500 male births.¹⁻³ DMD results from a gene mutation that leads to altered or absent dystrophin production.⁴ Dystrophin is normally expressed in the retina and localises to photoreceptor terminals and around retinal vessels. Deficiency of dystrophin produces abnormal transmission between photoreceptors and optic nerve bipolar cells and a diminished electroretinogram (ERG) signal.⁵ Mice lacking the Dp71 isoform of dystrophin suffer greater damage to the ganglion cell layer following transient ischaemia than wild type mice.⁶ Therefore, dystrophin may be involved in the regulation of ischaemic processes in the retina. Cardiopulmonary assist is not routinely associated with proliferative retinopathy in adults. Retinal neovascularisation is not prevalent in the Duchenne population, suggesting that absence of dystrophin is not sufficient to induce neovascularisation alone.

In summary, rapidly progressive, bilateral proliferative retinopathy may be associated with DMD in the presence of severe cardiopulmonary compromise. Whether an absence of dystrophin contributes directly or indirectly is unknown but consideration of the possibility may lead to novel insights into the development of pathological retinal neovascularisation. The visual prognosis with late presentation in this setting is uncertain despite full panretinal photocoagulation. Patients with advanced DMD may benefit from periodic fundus examination as it is not known whether early treatment has the potential to alter prognosis.

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Bilateral decompression retinopathy after orbital decompression surgery

Decompression retinopathy is defined as retinal haemorrhages that typically occur after glaucoma filtration surgery.^{1,2}

Orbital decompression is a common surgery performed to treat patients with thyroid related orbitopathy for functional or cosmetic indications.^{3,4} Many complications have been described with the surgery, but this surgery has never been associated with retinal haemorrhages.

We describe a case of a 70 year old woman, who developed bilateral retinal haemorrhages after staged bilateral orbital decompression surgeries.

Case report

A 70 year old woman with the diagnosis of euthyroid Graves' disease was referred because of severe proptosis. Past ophthalmic history revealed two previous strabismus surgeries. Past medical history was unremarkable with no history of diabetes or cardiovascular disease, also she was not taking aspirin or any other blood thinning medications.

Ophthalmic examination showed visual acuity of 20/20 in each eye. Both orbits were moderately firm to retropulsion. IOP was within normal limits in primary gaze (14, 19 mm Hg) and slightly elevated in upgaze (17, 26 mm Hg). There were limitations in upgaze and lateral gaze in both eyes as well as upper and lower lids retractions. There was a mild degree of lagophthalmos with exposure keratopathy. Funduscopy was normal and did not show any evidence of microvascular disease or retinal haemorrhages. Hertel measurements were 22 mm on the

Figure 1 Fundus photograph (upper image), left eye, 4 days after orbital decompression surgery on the left side showing scattered retinal haemorrhages both in deep and superficial layers of the retina. VA 20/25. (Lower image) Fluorescein angiography, left eye, late frame showing blocked fluorescence from retinal haemorrhages.

right and 23 mm on the left. Computed tomography scan showed enlargement of the extraocular muscles.

She underwent balanced orbital decompression surgery on the left side, including deep lateral and medial wall decompression with intraconal fat removal. Three days after surgery she noted spots in front of her left eye. Visual acuity in that eye was 20/25. Funduscopy examination disclosed dot and blot haemorrhage with flame shaped haemorrhages in the posterior pole of the left eye (fig 1).

The patient was well informed of the complication in the first eye and the chance of developing retinal haemorrhages in the right eye after orbital decompression. She agreed to undergo surgery and 1 week later she underwent balanced orbital decompression on the right side. Three days later she again noted spots in front of her right eye. Best corrected visual acuity decreased to 20/160, and funduscopy examination revealed posterior pole retinal haemorrhages (fig 2).

Three months postoperatively IOP in primary gaze decreased to 12 mm Hg in both eyes, and 14 and 16 mm Hg in upgaze. Exophthalmos decreased to 18 mm on each side, and the lagophthalmos and exposure keratopathy resolved. Fluorescein angiography showed evidence of blocked fluorescence, suggestive of retinal haemorrhage. There was no evidence of neovascularisation, vasculopathy, or choroidal rupture. Visual acuity gradually improved over the course of 3 months and returned to 20/20 in both eyes.

Figure 2 Fundus photograph (upper image), right eye, 4 days after orbital decompression surgery on the right side showing scattered posterior pole retinal haemorrhages. Note the haemorrhage centred in the fovea. VA 20/160. (Lower image) Fluorescein angiography, right eye, late frame showing blocked fluorescence from retinal haemorrhages.

Comment

Decompression retinopathy is a rare complication that may occur after glaucoma filtration surgery. It is associated with scattered retinal haemorrhages concentrated in the posterior pole. It may be more common in patients with marked elevated preoperative intraocular pressure and after acute decrease of IOP. The haemorrhages may be diffuse, both in deep and superficial layers of the retina, and may even show white centres when first observed.^{1,2}

Retinal haemorrhages associated with ocular decompression appear to be relatively benign and usually resolve within weeks to months with no effect on visual acuity or intraocular pressure. A gradual decrease of IOP preoperatively and intraoperatively is recommended in order to avoid this complication.^{1,2}

Decompression retinopathy has not previously been described as a complication of orbital decompression surgery. Our patient had a relatively tight orbit with restrictive strabismus and marked enlargement of the extraocular muscles. Significant force was required to retract the globe to achieve exposure of the medial and deep lateral orbital walls. Retraction was frequently relaxed to assure perfusion of the retina. We hypothesise that the marked intraocular pressure fluctuation that occurs during these surgical manoeuvres may have contributed to the retinal haemorrhages. It may also be that rapid decrease in retrobulbar pressure has caused ocular hypotony and retinal haemorrhage.⁴

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Retinal nerve fibre layer damage after indocyanine green assisted vitrectomy

Recently, indocyanine green (ICG) has been used to stain and visualise the internal limiting membrane (ILM) during vitrectomy.¹ Some case series showed that visual field defects on the nasal side can occur after the surgery through unknown cause.^{2,3} Here, we report a case in which nasal visual field defects occurred after ICG assisted ILM peeling for epiretinal membrane (ERM). Detailed examination revealed that the superior and inferior retinal nerve fibre is severely damaged in this case.

Case report

A 60 year old woman who received ICG assisted ILM peeling for ERM in her right eye was referred to our hospital. The preoperative best corrected visual acuity (BCVA) was 20/60 in the right eye. According to the referring ophthalmologist, 25 mg of ICG (Diagnogreen; Daiichi Pharmaceuticals) was

dissolved in 10 ml of distilled water, which was further diluted by a viscoelastic material (Healon; Pharmacia) to give 0.16% ICG solution. To stain ILM, ICG was injected into an air filled eye and the dye was washed 2 minutes later. An air infusion cannula was placed at the temporal side. There was no complication during the surgery. Seventeen days after the operation, she noticed nasal visual field loss, which got worse 22 days after the surgery. Sixty days after the surgery, she was referred to our hospital. At the initial visit, the BCVA was 20/25 in the right eye. Goldmann perimetry revealed a nasal visual field defect (fig 1A). In the right eye, a relative afferent pupillary defect was found. Ophthalmoscopic examination and fluorescein angiography showed no abnormalities. The optic disc rim appeared to have lost colour without being associated any cup or rim changes typically seen in glaucoma (fig 1B). Residual ICG was evident at the optic disc and along the nerve fibre (fig 1C). The nerve fibre staining was most evident in the superior and inferior quadrants. ICG angiography revealed ICG staining of the optic disc and superior and inferior nerve fibres, but no other abnormalities. Full field electroretinogram (ERG) and multifocal ERG (VERIS science ver3.8, EDI) revealed no abnormalities. The results of visual evoked potential testing were also non-remarkable.

During our 8 month follow up period, there was no significant change in the visual field defect and the distribution of the residual ICG. Scanning laser polarimetric analysis (GDx VCC, Laser Diagnostic Technologies, Inc, San Diego, CA, USA) performed 8 months after the surgery showed profound nerve fibre loss around the disc, especially evident at superior and inferior quadrants (fig 2).

Comment

In this case, an air infusion cannula was placed at the temporal side and Goldmann perimetry showed nasal visual field defects. Thus, the dehydration injury to the retina during air-fluid exchange, which is observed at the opposite side of the cannula, is unlikely to be the cause of this visual field defects.²⁻⁴ Optic neuropathy unrelated to the use of ICG may be considered as a differential diagnosis. However, this visual field damage is not typically seen in optic neuropathy and may be rather associated with ICG, based on

Figure 1 (A) Goldmann perimetry revealed a nasal visual field defect 4 months after ICG assisted peeling of ILM for ERM. Visual field defect testing was also performed 8 months after the surgery, which demonstrated no remarkable change. (B) Fundus photograph taken 4 months after the surgery shows that the optic disc rim appeared to have lost colour without being associated with any cup or rim changes typically seen in glaucoma. The appearance of the optic disc remained unchanged during our 8 month follow up. (C) Infrared fundus photograph taken 8 months after the surgery with an ICG filter set. Residual ICG was evident at the optic disc and the nerve fibre. The fluorescent intensity and distribution of residual ICG did not change remarkably during our 8 month observations.

Figure 2 The results obtained from the GDx-VCC from the right (top) and left (bottom) eyes taken 8 months after the surgery. The left column shows the retardation image. The centre column shows the results from analysis. The navy, blue, yellow, and red points represent significant loss of nerve fibre layer, $p < 5\%$, 2% , 1% , and 0.5% , respectively. The right column shows that the general double hump pattern is evident in the left eye but is lost in the right eye.

recent clinical findings.^{2,3} Retinal damage was not evident by morphological, angiographic, and functional analysis. However, it was evident that the nerve fibres are damaged in this patient. Although the direct causal relation cannot be proved, it is highly likely that the damage to the nerve fibre was caused by the ICG because of the remarkable correspondence of the distribution pattern of ICG and the nerve fibre defects. This is also supported by our experimental findings that ICG showed neurotoxicity at concentrations lower than clinically employed.³ To our knowledge, this is the first report of ICG induced retinal nerve fibre damage assessed by scanning laser polarimetry.

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NOTICES

Worldwide clinical trials for new technique for early detection of eye disease

A unique new non-invasive technique for high resolution optical imaging of the eye is receiving global acclaim. The technique, pioneered by the University of Kent, is

funded by the Toronto-based company, Ophthalmic Technology Inc (OTI). The University's Applied Optics Group is currently working with university hospitals in New York (USA), Osaka (Japan), Asahikawa (Japan), Amsterdam (Netherlands) and Milan (Italy) to carry out preliminary clinical trials. By combining two high-resolution imaging technologies, the new technique provides doctors with 3-D images of the retina, macula and the optic nerve. Such high resolution images provide clinicians with capabilities for early diagnosis and treatment of common ocular diseases such as glaucoma, diabetes and age-related macula degeneration. OTI is planning in the near future to extend the clinical research to other leading university medical centres in Japan, USA and Europe.

Professor Adrian Podoleanu explained: 'At Kent we created a very cost effective imaging system which simultaneously produces optical coherence tomography (OCT) and scanning laser ophthalmoscope (SLO) images. Its early potential was immediately realised by OTI, who commissioned the assembly of several prototypes to be tested in different clinics worldwide before embarking on commercial exploitation of the invention'.

The clinical investigators together with the Kent team have jointly published in international medical publications and presented at clinical and scientific conferences over 50 publications and presentations related to this research.

For more information, contact the Media Office on 01227 823581/823100 or email MediaOffice@kent.ac.uk News releases can also be found at: <http://www.kent.ac.uk/news>

Glaucoma Society Silver Jubilee Meeting 2004

The Silver Jubilee Meeting and Dinner for the Glaucoma Society will be held on 3 December 2004 at the Royal College of Physicians in Regents Park, London. The meeting will take place between 8.30am and 5pm and the dinner will be held between 6.30pm and 10pm. For further information, please contact: Janet Flowers, Administrator, 29 Quarry Hill, Grays, Essex, RM17 5BT (tel: 01375 383172; e-mail: glausoc@ukeire.freemove.co.uk).

Amsterdam Retina Debate

The Amsterdam Retina Debate will be held on 10 December 2004 at the Academic Medical Centre, Amsterdam, The Netherlands. For further information, please contact: Nicolaes Tulp Institute; tel: +31 20 566 8585; fax: +31 20 696 3228; email: retinadebate@amc.uva.nl

British Oculoplastic Surgery Society

Call for papers for the 5th annual meeting of the BOPSS to be held on 15 and 16 May 2005 at The Belfry, Birmingham. The abstract submission deadline is 4 February 2005, abstracts can be submitted online at www.bopss.org.

PostScript

LETTERS

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Demonstration of identical clonal derivation in a case of "oculocerebral" lymphoma

Primary intraocular lymphoma (PIOL) is a high grade malignant non-Hodgkin's lymphoma (NHL) usually of B cell type, involving the retina and vitreous. PIOL can occur independently or together with primary central nervous system lymphoma (PCNSL; the combination termed "oculocerebral lymphoma"). Because of its slow onset and ability to simulate other conditions, the diagnosis of PIOL remains challenging. A number of techniques, including conventional cytology, immunocytology, flow cytometry, polymerase chain reaction (PCR), and biochemical analysis of vitreous samples, are recommended to aid the diagnostic procedure.¹⁻⁸ We report a case of oculocerebral lymphoma, whereby IgH-PCR and GeneScan analysis confirmed the histological diagnosis by demonstration of the identical clonal B cell populations in both the vitreous and stereotactic biopsy.

Case report

A 51 year old systemically healthy man presented in March 2002 with an epileptic fit. Cranial magnetic resonance imaging

Figure 1 (A) Histological examination of the perivascular orientated neoplastic lymphocytes in the stereotactic brain biopsy (Giemsa, original magnification $\times 40$). (B) Cytology of the vitreous aspirate demonstrating grouped and isolated pleomorphic cells (May-Grunewald-Giemsa, original magnification $\times 40$).

Figure 2 (Top) GeneScan analysis following IgH-PCR (FR2) of the vitreous biopsy demonstrating a monoclonal peak (blue) of 257 base pairs in size. The smaller red peaks represent controls. (Bottom) GeneScan analysis following IgH-PCR of the paraffin embedded cerebral biopsy, with a monoclonal peak of 257 base pairs.

demonstrated a mass with intensive contrast enhancement in the left fronto-parietal area. A stereotactic biopsy was performed, establishing the diagnosis of a high grade malignant B cell NHL (fig 1A). The neoplastic cells consisted of medium to large sized blasts and were orientated perivascularly. They demonstrated immunoreactivity for CD20, a monotypic expression of Ig-kappa, and a large growth fraction (Ki-67 antigen) of 90%. Staging procedures did not reveal any systemic lymphoma. Two cycles of high dose methotrexate chemotherapy (4 g/m^2 intravenously per cycle) were commenced. The patient developed recurrent epileptic attacks, and repeat imaging studies demonstrated tumour size increase. The patient was treated with whole brain irradiation (total dosage, 45 Gy), resulting in complete remission for 14 months. In August 2003, the patient complained of "floaters" and a bilateral decrease in vision. On examination, the

visual acuity (VA) was 20/25 and 20/32 in the right and left eyes, respectively. Funduscopy revealed bilateral dense cellular infiltrates in the vitreous.

Conventional and immunocytological examination of a diagnostic vitrectomy of the left eye disclosed an intraocular manifestation of B cell NHL. The infiltrating atypical lymphocytes (fig 1B) expressed CD20, and displayed a monotypic expression of Ig-kappa. The remaining vitreous aspirate and the paraffin embedded cerebral biopsy material were submitted for clonality analysis using IgH-PCR and GeneScan techniques. For the detection of IgH rearrangements, three single step PCRs were performed employing family specific framework (FR) 1, FR2, and FR3 Bio-Med 2 primers together with a common JH consensus primer (JH22).⁹ The cycling conditions (50 rounds of amplification) for all PCRs are described in detail elsewhere.⁹ Both samples revealed dominant PCR products of the same size (FR1 327 base pairs, FR2 257 base pairs, FR3 125 base pairs), demonstrating the identical neoplastic B cell population in both lymphomatous manifestations (fig 2). Further, DNA sequencing of the amplicates revealed a functional VH3/JH4 rearrangement of the tumour cells.

Thorough imaging studies revealed neither a cerebral recurrence nor evidence of systemic lymphoma. The patient was commenced on high dose ifosfamide (1500 mg/m^2 intravenously daily over 3 days/cycle). In January 2004, follow up examinations demonstrated a complete resolution of lymphomatous infiltrates in both eyes, and the VA was 20/20 bilaterally.

Comment

Cytological studies of vitreous biopsies remain the first step in the histomorphological

diagnosis of PIOL. Previous reports have described the use of PCR examining for monoclonal rearrangements of immunoglobulin heavy (IgH) or light (IgL) chains in B cell lymphoma or T cell receptor genes in T cell lymphoma as an adjunctive diagnostic tool in the evaluation of vitreous specimens for PIOL.³⁻⁸ The success of these analyses is dependent on the quantity of material provided and the extent of DNA degradation. The quality of DNA extracted from paraffin embedded biopsy material can be compromised by fixation solutions, and the duration of fixation. Improved primers for IgH-PCR and TCR-PCR have recently been developed, thereby increasing the chances of detection of clonal B and T cell populations in tissues and fluids.⁹ In oculocerebral lymphoma, it is assumed on the basis of clinical, morphological, as well as immunohistochemical findings that the cerebral and ocular infiltrations represent the same tumour. To our knowledge, this association between PIOL and PCNSL has not yet been proved genetically. This case, therefore, represents the first in the literature, whereby molecular biological evidence is provided showing that the lymphomatous manifestations in oculocerebral lymphoma consist of the identical neoplastic B cell population and that they derive from the same tumour precursor cell. Furthermore, DNA sequencing of both specimens demonstrated a similar VH gene usage to that previously reported by PCNSL.¹⁰

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The prevalence of pseudoexfoliation syndrome in Chinese people: the Tanjong Pagar Survey

Young and colleagues report that pseudoexfoliation syndrome (PXS) was uncommon in 500 Chinese people aged 60 years and older attending general ophthalmic clinics in Hong Kong with a presumed diagnosis of cataract.¹ We have previously carried out a population based assessment of the prevalence of prevalence and risk factors for glaucoma in a district of Singapore, which allowed us to assess the prevalence of PXS in a representative Chinese adult population.²

Case report

This study was approved by the ethics review board of Singapore National Eye Centre. All subjects gave written, informed consent. A total of 2000 Chinese Singaporeans aged 40 years and older were identified from the electoral register of Tanjong Pagar district. A total of 283 were considered ineligible (they had died, moved, or were medically unfit to be examined), leaving 1717 potential subjects. From this number 1232 underwent a slit lamp examination before and after pharmacological dilatation of the pupil with phenylephrine (2.5%) and tropicamide (1%). Gonioscopy was carried out before dilatation on all subjects. Pseudoexfoliation material (PXM) on the anterior lens surface and/or pupil margin was specifically sought. Glaucoma was diagnosed on the basis of structural abnormalities of the optic nerve

(dimensions of the cup:disc ratio lying outside the 97.5th percentile) combined with a reproducible visual field defect, or advanced structural damage consistent with glaucoma (dimensions of the cup:disc ratio lying outside the 99.5th percentile) if the subject could not complete formal field testing.³

We identified PXM in six eyes of four people (table 1). Two people had definite glaucomatous optic neuropathy, and three had undergone glaucoma surgery. None was using topical or oral medication for glaucoma when seen. The age and sex standardised rate of PXS in Chinese Singaporean adults aged 40 years and older is 0.2% (95% confidence interval; 0.0 to 0.4). In the over 60s, this rose to 0.7% (95% CI: 0.5 to 0.9).

Comment

In contrast with the report from Hong Kong, we identified two people in Singapore with PXS and angle closure sufficient to require incisional surgery. Only one person had PXS with glaucoma and open drainage angles.

In previous studies in east Asia, PXM was identified in three of 22 (13.6%) people with glaucomatous optic neuropathy in a Mongolian population. Two of these were classified as open angle glaucoma. The third was blind in both eyes from primary angle closure glaucoma.⁴ The relative scarcity of PXS in east Asian people contrasts with rates of 6-7.7% in adult black South Africans.⁵ Similarly, PXS appears relatively common (3.8-6.0%) in south India.^{6,7} The slightly lower rate among European settlers in Australia aged 49 years and older (2.3%, 95% CI:1.8 to 2.8) is probably genuinely higher than that seen among east Asians.⁸

In summary, we agree with Young and colleagues that PXS is uncommon in the Chinese cohorts studied to date. However, the tendency for the condition to cluster geographically and in racial subgroups suggests that it may occur with greater frequency in areas not yet studied.

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Table 1 Characteristics of patients with pseudoexfoliation syndrome

	Age/sex	Intraocular pressure		Cup:disc ratio		Surgery		Glaucoma
		Right	Left	Right	Left	Right	Left	
1	66/M	16*	13*	0.6*	0.7*	Nil	Nil	Nil
2	74/F	13*	10	1.0*	0.9	Trab	Trab	PACG
3	77/F	14*	16*	0.3*	0.7*	Nil	Trab	OAG
4	79/F	13*	12	0.5*	0.6	SPI	Trab	PAC

*Eye with pseudoexfoliation material.

Trab, trabeculectomy; SPI, surgical iridectomy; PAC, primary angle closure; PACG, primary angle closure with glaucomatous optic neuropathy; OAG, open angle glaucoma.

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Hoarse voice and visual loss

Giant cell arteritis (GCA) often presents atypically. An under-recognised presentation is with speech or respiratory involvement. We report a case of GCA with dysphonia and visual loss, a previously unreported combination.

Case report

A 67 year old woman presented with sudden, non-progressive, painless blurring of the vision in her right eye for 4 days. Her left eye, which had been poor since childhood, was unchanged. She had no other ophthalmic symptoms or history. She smoked and was on treatment for hypertension. On questioning, she reported pain in her throat for 3 weeks, worse on swallowing, which had not responded to oral antibiotics. Her voice had been markedly hoarse for the same period. She denied other respiratory symptoms. She did not have anorexia, weight loss, myalgia, muscle stiffness, scalp tenderness, or jaw claudication, but had been experiencing a dull ache all over her head for 6 weeks.

On examination, she had deep tissue tenderness on the anterolateral aspect of her neck, bilaterally. She had no scalp tenderness, and her temporal arteries were pulsatile. Her corrected visual acuity was 6/24 in

both eyes. Her right optic disc was swollen, consistent with an anterior, ischaemic, optic neuropathy. The left optic disc was normal. The remaining ophthalmic and systemic examinations were unremarkable. Blood tests revealed an erythrocyte sedimentation rate of 130 mm in the first hour, a C reactive protein of 125 mg/L, and a mild, normochromic, normocytic anaemia. A diagnosis of right anterior, ischaemic optic neuropathy secondary to giant cell arteritis (GCA) was made, and she was admitted for pulse, intravenous methylprednisolone and high dose oral prednisolone. Temporal artery biopsy confirmed the diagnosis of GCA (fig 1).

The following day, her headache and throat pain were much better, her right optic disc was less swollen, and her inflammatory markers began to fall. She was referred to an otorhinolaryngologist regarding her prolonged hoarse voice. Examination and flexible laryngoscopy showed no abnormalities. Chest radiograph was normal. On day 2, her voice began to improve, and by day 3 it was back to normal. She was discharged on oral prednisolone.

Comment

Giant cell (temporal) arteritis is the most common of the vasculitides, and presents with varied and often non-specific symptoms. Diagnosis may be further hindered by the possibility of non-elevated inflammatory markers¹ and negative temporal artery biopsy.² Ophthalmic artery involvement may cause irreversible, bilateral blindness, and may occur even in the absence of systemic symptoms and signs, a scenario termed occult GCA.³

Speech and respiratory features in giant cell arteritis have been described only infrequently.^{4–5} They include cough, sore throat, pain on swallowing, anterior neck tenderness, and dysphonia (hoarse voice). Nevertheless, it has been estimated that 4% of patients have respiratory symptoms as the initial presentation of GCA, and that as many as 9% will display them at some time during the course of the disease.⁴

Voice changes have been reported in eight patients with GCA, seven with hoarseness,^{4–7} and one with a broken, falsetto voice.⁸ None of these were reported to have had visual involvement. It had been suggested that the vasculitis in cases of GCA with speech or respiratory features might show a preference for branches of the external carotid artery (which supplies the larynx) over the internal carotid (which supplies the eye).⁵ Our case, however, demonstrates the possibility of dual involvement.

In considering the diagnosis of GCA, non-classic features, such as speech and respiratory symptoms, can be easily overlooked. In the absence of classic symptoms they should be specifically asked about. The label of occult GCA, therefore, should not be applied without first excluding the whole spectrum of recognised GCA features. In addition, GCA should be considered in cases of prolonged dysphonia with concomitant visual symptoms.

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Subconjunctival migration of silicone oil through a Baerveldt pars plana glaucoma implant

Extravasation of intraocular silicone oil through a sclerotomy into the subconjunctival space has been described.¹ Oil migration through Molteno and Ahmed implants has also been reported in the literature.^{2,3} However, literature search revealed no cases of oil migration through a Baerveldt pars plana implant after a vitrectomy. We report such a case.

Case report

In May 2001, a 58 year old white man presented with dense vitreous haemorrhage, hyphaema, and neovascular glaucoma (intraocular pressure (IOP) was 55 mm Hg by applanation tonometry) associated with proliferative diabetic retinopathy in his phakic right eye (RE), and background diabetic retinopathy in his left eye (LE). Visual acuity (VA) was RE: counting fingers, LE: 20/20. There were mild cortical and nuclear sclerotic cataracts in both eyes. The high IOP along with severe headache and nausea was refractory to maximal medical therapy. A pars plana vitrectomy, endophotocoagulation, and placement of a Baerveldt pars plana glaucoma implant (Model BG-102–350, surface area: 350 mm² Pfizer Inc, New York, NY, USA) in the superior temporal quadrant allowed an immediate relief of severe ocular and systemic discomfort, and normalisation of the IOP to 14 mm Hg (applanation), RE, after surgery. Owing to the increased cataract, his general ophthalmologist performed cataract extraction by phacoemulsification and inserted a posterior chamber implant, RE, in October 2001. Subsequently, intraocular silicone oil tamponade for multiple retinal breaks including a large inferior relaxing retinotomy was required during a repeat vitrectomy for proliferative vitreoretinopathy, RE, in January 2002. Examination in May 2002 revealed disappearance of 50% of intravitreal

Figure 1 Section of temporal artery biopsy stained for elastin.

Figure 1 (A) External photograph of inflamed and elevated superior conjunctival bleb infiltrated with extravasated silicone oil from the vitreous cavity of RE. (B) Fundus photograph shows a high intraocular oil-fluid level as a result of extravasation of more than 50% of oil. Note retina is still completely attached despite loss of a large volume of oil.

oil because of its migration into the superior subconjunctival space via the Baerveldt shunt (fig 1). Patient complained of increasing ocular discomfort as a result of conjunctival inflammation and IOP rise to 30 mm Hg by applanation tonometry, associated with an enlarging superior conjunctival bleb with underlying infiltration of emulsified oil in the subsequent weeks. Application of dorzolamide hydrochloride-timolol maleate and brimonidine tartrate 0.2% ophthalmic solutions lowered the IOP to 18 mm Hg, RE. Removal of intraocular and subconjunctival silicone oil was performed on 28 May 2002. Surgical exploration showed widespread oil infiltration involving the posterior plate of the implant and the subconjunctival soft tissues. Extensive resection of swollen subconjunctival tissues infiltrated with oil droplets was performed (fig 2). The surgical dissection involved primarily the anterior subconjunctival tissues associated with most of the oil infiltration, and stayed away from the posterior orbital space where fibrous encapsulation around the implant plate was noted. The Baerveldt implant was not removed. The ocular inflammation subsided and the IOP was brought down to 16 mm Hg (applanation) without ocular hypotensive medications, RE, within 1 week after surgery. Ocular hypotensive medical therapy was no longer required afterwards. The VA was 20/200 and the IOP was 15 mm Hg (applanation) with complete retinal attachment, RE, 6 months later.

Figure 2 High power photomicrograph of excised conjunctival tissue infiltrated with silicone oil shows a ring of epithelioid cells (large arrow), foreign body material (small arrows), and giant cell (arrowhead).

Comment

In recent years, Baerveldt pars plana glaucoma implants have become increasingly popular for control of refractory glaucoma in eyes with vitreoretinal complications that also require a pars plana vitrectomy.⁴ Frequently, silicone oil tamponade may also be indicated for such eyes. Emulsification of intraocular silicone oil usually takes many months after surgery to develop, the exact timing of which varies and depends on multiple factors, including the purity and viscosity of the oil.⁵ It is interesting that extraocular migration of silicone oil did not occur until 4 months after its placement, coincidental with the start of oil emulsification in this case. Despite the loss of intraocular oil, previous long term retinal tamponade with oil proved sufficient for maintaining retinal attachment after oil removal. The drainage tube was not removed or ligated during the second vitrectomy when silicone oil was inserted to avoid recurrent excessive rise of IOP after surgery in the absence of a patent drainage channel, potentially aggravated by reduced volume of the vitreous cavity for posterior aqueous flow due to the intravitreal silicone oil. Measures that may delay or prevent extraocular oil migration through a drainage tube include placement of the pars plana drainage tube in an inferior quadrant, replacement of the pars plana shunt with another tube shunt inserted into an inferior quadrant of the anterior chamber, and use of highly purified and super-viscous oil with lower tendency for emulsification. In addition, the patient is encouraged to sleep on the side of the drainage tube, since oil may rise from the dependent side and away from the tube. Eventually, emulsified oil droplets may find their way into the drainage tube for extraocular migration. However, this case shows that Baerveldt pars plana implant and silicone oil may coexist for a prolonged period for select cases. Silicone oil extravasation through a glaucoma shunt is not unique for a Baerveldt pars plana implant, but a phenomenon associated with other types of shunt implants as well, as shown by previous case reports.²⁻³ To our knowledge, however, this is the first written report of silicone oil migration through the drainage tube of a Baerveldt pars plana implant.

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Rosai-Dorfman disease: isolated epibulbar masses in two adult patients

Rosai and Dorfman first characterised sinus histiocytosis with massive lymphadenopathy in 1969.¹ This condition most commonly presents as a massive painless cervical adenopathy in children or young adults of African ancestry. The lymphadenopathy typically has a protracted course, lasting for several years before spontaneously resolving. Complications can include compression of vital organs or associated anaemia or leucopenia. The results of chemotherapy or radiation treatments have generally been disappointing; however, surgical debulking, when necessary, has been effective.

Microscopic examination of the lymph nodes reveals a polymorphous infiltrate composed of plasma cells, other lymphocytes, and histiocytes. The histiocytes often contain phagocytised lymphocytes, a histological finding termed emperipolesis. Since these histiocytes fill and expand lymph node sinuses, the disease was first named morphologically as sinus histiocytosis with massive lymphadenopathy. Extranodal involvement, most commonly in the upper respiratory tract and stomach, displays a histology similar to lymph node infiltrates. Because extranodal infiltrates are often found in the absence of lymphadenopathy, the eponym Rosai-Dorfman disease is now preferred.²

The orbit is a common extranodal site of RDD.³ Four cases of RDD manifesting as an epibulbar conjunctival mass have also been reported.⁴⁻⁷ In two of these cases, both in children, the epibulbar mass was an isolated finding.⁵⁻⁷ We present RDD occurring as an isolated epibulbar mass in two adult patients.

Figure 1 Clinical photograph of patient 1 (A), demonstrating the vascularised epibulbar mass. Haematoxylin and eosin stained section through the excised mass patient 1 (B). Chronic inflammatory infiltrate with lymphocytes, plasma cells, and histiocytes is present. Inset shows higher magnification of histiocyte containing lymphocytes and plasma cells within its cytoplasm, demonstrating the characteristic histological finding termed emperipolesis. Haematoxylin and eosin stained section for patient 1 (C), demonstrating small focus of necrosis. Clinical photograph of patient 2 (D), demonstrating the vascularised epibulbar mass.

Case reports

A 71 year old African-American man with a history of hypertension, benign prostatic hyperplasia, asthma, gout, and degenerative joint disease was evaluated for a painless 1.5 cm episcleral mass on the medial aspect of the right eye, adjacent to the limbus (fig 1A). The mass had been growing for

4 months. The patient was examined by an internist, who found no lymphadenopathy, anaemia, or leucopenia. The mass was excised for histopathological diagnosis. Haematoxylin and eosin stained sections of the episcleral nodule revealed a mixed cellular infiltrate, predominantly composed of histiocytes mixed with lymphocytes,

including plasma cells and polymorphonuclear leucocytes (fig 1B). The histiocytes exhibited a large, round, vesicular nucleus with abundant pale staining and finely vacuolated cytoplasm. Several of these cells displayed the presence of polymorphonucleocytes, lymphocytes, and plasma cells within the cytoplasm (fig 1B, inset). Several foci of necrosis were noted, without the formation of granulomas (fig 1C). Stains for bacteria, acid fast bacilli, and fungi were negative. On immunohistochemistry, histiocytes stained positive for S-100, CD-68, lysozyme, and α -1-antitrypsin and negative for CD-1a (fig 2). The lymphoid infiltration showed the presence of kappa and lambda immunoglobulin chains.

A 51 year old African-Brazilian man with no medical problems presented with a 5 mm erythematous, subconjunctival mass. The mass was adjacent to the limbus and appeared to be adherent to the underlying tissues (fig 1D). Systemic evaluation was negative and there was no lymphadenopathy. The patient underwent a superficial sclerectomy with excision of the mass, and the lesion was submitted for histopathology. Follow up examination at 4 months showed no signs of recurrence. Haematoxylin and eosin stained sections of the nodule revealed a mixed cellular infiltrate, predominantly composed of histiocytes mixed with lymphocytes, including plasma cells and polymorphonuclear leucocytes. Several of these histiocytes showed emperipolesis, displaying phagocytosed polymorphonucleocytes, lymphocytes, and plasma cells. Stains for bacteria, acid fast bacilli, and fungi were negative.

Comment

The cases show that RDD can present as an isolated epibulbar mass in the elderly, as late as the eighth decade. Two previous cases of RDD manifesting as an isolated epibulbar mass in children have been described.^{5,7} One case of epibulbar and cutaneous RDD in a 40 year old has also been described.⁶ Although most cases of RDD occur in children or young adults, the disease is known to manifest in the elderly as well. A review of 423 cases of RDD showed a median age at presentation of 20 years (SD 20 years).⁸ The oldest patient in that series was 74 at the time of presentation. The mean age in cases with ocular involvement was 6 years. Patients with soft tissue lesions are known to be older than patients with nodal or solid organ involvement, with a mean age of 46 years in one series.⁹

Although clinical features of RDD may vary from benign soft tissue masses or lymphadenopathy to life threatening compression of vital organs, anaemia, or leucopenia, the characteristic histological features are histiocytic infiltration admixed with lymphocytes and other inflammatory cells. One typical feature of this entity has been emperipolesis, with histiocytes displaying phagocytosed lymphocytes and plasma cells. Histiocytes in RDD, Langerhans cell histiocytosis, and other histiocytoses express S-100, a neural tissue specific protein; however, the pathophysiology of this S-100 expression remains obscure. Although positive staining for S-100 strongly suggests RDD, it is not absolutely required to make the diagnosis in the presence of typical histology for RDD.⁹ CD68 is a monocyte/macrophage marker frequently expressed by histiocytes in all histiocytic disorders and believed to be associated with lysosomal

Figure 2 Immunohistochemistry of histiocytes in patient 1 (A), consistent with RDD (400 $\times$). Stain for S-100 was positive. Stain for α -1-antitrypsin (B) was positive. Stain for lysozyme (C) was positive. Stain for CD-1a (D) was negative.

granules. In all histiocytoses other than Langerhans cell histiocytosis, the histiocytes frequently stain positive for lysozyme, indicating that these cells are activated and have strong phagocytic potential. α -1-Antitrypsin is a proteinase inhibitor expressed by monocytes, inhibiting overexpressed proteinases during inflammation. It has been variably expressed in histiocytes in RDD.⁹ CD-1a is a marker for the Langerhans histiocyte and may be involved in antigen presentation. Staining for CD-1a is negative in RDD. Early reviews of RDD stress the lack of necrosis on histopathology. Despite the lack of documentation in the early literature, small foci of necrosis are sometimes present in RDD. Consistent with our case 1, small foci of necrosis resembling microabscesses were observed in a series of soft tissue RDD.⁹

Rosai-Dorfman disease should be considered by the clinician and pathologist when evaluating epibulbar masses in children, adults, and the elderly. The diagnosis is made by the pathologist based on the typical histology, including emperipolesis and confirmatory immunohistochemistry. Surgical excision is the treatment of choice. Patients should be managed in conjunction with the internist to evaluate for lymphadenopathy, multiple extranodal lesions, anaemia, and leucopenia.

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Systemic FK506 improved tear secretion in dry eye associated with chronic graft versus host disease

Dry eye is one of the major symptoms of chronic graft versus host disease (CGVHD).^{1,2} Although effective therapy for dry eye associated with CGVHD has not been well established, successful treatment with systemic FK506,^{3,4} topical retinoic acid,⁵ topical cyclosporin A,⁶ and topical autologous serum have been reported.^{7,8} However, improved tear secretion was reported only in one patient with systemic FK506 by Masaoka *et al*,³ with limited description of ocular findings. We present a patient with dry eye associated with CGVHD, where systemic administration of FK506 resulted in improved ocular surface findings along with the Schirmer test value.

Case report

A 33 year old man had HLA matched sibling allogeneic peripheral blood stem cell transplantation in December 2000 for acute myeloid leukaemia in remission. He received cyclosporine A as GVHD prophylaxis, but it was discontinued on day 85. On day 98, he was referred to an ophthalmologist for screening of CGVHD. On examination, slight tarsal and bulbar conjunctival injection, and slight superficial punctate keratitis (SPK) were noted bilaterally. The Schirmer test value was 10 mm in the right eye and 9 mm in the left eye.

On day 105, he developed bilateral pseudo-membranous conjunctivitis, lichenoid oral lesions, skin eruptions in the upper half of the body, and liver dysfunction. The diagnosis of CGVHD was made. On day 123, oral cyclosporin A (5 mg/kg /day) and oral prednisolone (60 mg/day) were started, and then skin eruptions and liver dysfunction gradually subsided. On day 140, although pseudo-membranous conjunctivitis was settled down, SPK worsened (fig 1) in both eyes. Prednisolone was discontinued on day 186. On day 196, the Schirmer test values decreased to 1 mm in the right and 3 mm in the left eye, and tear break up time (BUT) was 1 second bilaterally. Because lichenoid oral lesions and SPK were unchanged and skin eruptions gradually worsened, oral cyclosporine A was discontinued and oral FK506 was started on day 221.

On day 287, the trough level of FK506 was 8.5 ng/ml and the Schirmer test values improved to 4 mm in the right and 5 mm in the left eye. SPK had gradually improved in both eyes. On day 322, the Schirmer test

Figure 1 Fluorescein staining of the corneal epithelium on day 140, showing diffuse superficial punctate keratitis.

Figure 2 Fluorescein staining of the corneal epithelium on day 553. There was no recurrence of superficial punctate keratitis.

value improved to 16 mm and SPK disappeared in both eyes. On day 421, the trough level of FK506 was 7.9 ng/ml. The Schirmer test value was maintained over 10 mm up to day 553 without recurrence of SPK (fig 2) in both eyes, although BUT remained 1 second bilaterally.

Comment

Ogawa *et al*⁴ reported that in two patients with CGVHD, the symptoms of dry eye and the findings of the ocular surface markedly improved after the administration of systemic FK506 with corticosteroids. However, in their cases the results of Schirmer tests were not normalised in contrast with the result of Masaoka *et al*.³ Ogawa *et al*⁴ speculated that this difference is probably the result of the degree of lacrimal gland destruction. They demonstrated the result of biopsy of the lacrimal gland with prominent interstitial fibrosis and T cell infiltration in one of their patients.

The degree of lacrimal gland destruction may vary with the duration and/or severity of CGVHD. In two patients reported by Ogawa *et al*,⁴ FK506 had been administered 246 days after the onset of CGVHD in one patient, and the other had mild dry eye before haematopoietic stem cell transplantation. The lacrimal gland in these patients might have been irreversibly damaged before the administration of FK506. We speculate that in our patient, because FK506 substituted for cyclosporin A 101 days after the onset of CGVHD before irreversible damage of the lacrimal gland occurred, thereby may effective in improving tear secretion. The lack in the improvement of BUT in our case may be the result of severe damage to goblet cells with preceding pseudomembranous conjunctivitis.

This case indicates that systemic administration of FK506 is effective for dry eye associated with CGVHD, although the degree of improvement in tear secretion may vary between cases with the duration and/or severity of CGVHD.

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3D ultrasound coronal C-scan imaging for optic nerve sheath meningioma

The use of three dimensional (3D) ultrasonography (3DUS) for optic nerve measurements has been described in normal eyes utilising coronal 'C-scans'.^{1,2} This study demonstrates the use of 3DUS generated C-scans for optic nerve measurements in orbits with optic nerve sheath meningioma and compares those with measurements obtained from computed tomography (CT) scans.

Case reports

A 69 year old woman with a left optic nerve sheath meningioma was treated with external beam radiation therapy 6 years earlier.

On 3DUS coronal C-scans, an optic nerve sheath diameter (ONSD) 3 mm behind the globe was measured to be 7.4 mm in the left eye (fig 1, top left) and 6.4 mm in the right (fig 1, top right).

CT of the orbits was obtained. The centre of each optic nerve could clearly be identified on the axial imaging. At a distance of 3 mm posterior to the junction of the optic nerve with the sclera, the diameter of the optic nerve was measured. The ONSD was found to be 7.6 mm in the left eye (fig 1, bottom left) and 6.4 mm in the right (fig 1, bottom right).

A 74 year old woman with left optic nerve sheath meningioma was treated by external beam radiation 12 years before our evaluation. On 3DUS coronal C-scan imaging, the ONSD 3 mm behind the globe was measured to be 7.2 mm in the left eye (fig 2, top left) and 5.4 mm in the right (fig 2, top right).

CT of the orbits was obtained. The ONSD 3 mm behind the junction of the optic nerve with the sclera was measured to be 7.2 mm in the left eye (fig 2, bottom left) and 5.4 mm in right (fig 2, bottom right).

Comment

C-scan ultrasound imaging provided ONSDs similar to those obtained by CT of the orbits. Each was consistent with tumour related thickening of the left optic nerve. At 3 mm posterior to the globe, an ONSD discrepancy of at least 1 mm between the left and the right eyes was independently observed by both 3DUS C-scans and by CT axial scans.

Values obtained by C-scan correlated well with CT scan measurements. The diameters of the left optic nerves were thicker than the normative CT scan range of 4-6 mm.³

Figure 1 Case 1. The patient's left (top left) and right (top right) optic nerve sheath diameters are shown by 3DUS coronal C-scans. The patient's left (bottom left) and right (bottom right) optic nerve sheath diameters are shown by axial CT scans.

Figure 2 Case 2. The patient's left (top left) and right (top right) optic nerve sheath diameters are shown by 3DUS coronal C-scans. The patient's left (bottom left) and right (bottom right) optic nerve sheath diameters are shown by axial CT scans.

Although the right optic nerve of case 1 was slightly beyond the normative range by 3DUS and by CT, the right optic nerve of case 2 was within normal limits by both tests.

We have found that 3DUS could image the optic nerve up to 15 mm behind the globe. However, the full coronal outline of the optic nerve was no longer apparent starting 7 mm posterior to the globe. Proceeding from this point towards the posterior orbit, parts of the optic nerve sheath outline became indistinct, blending with the blackness of the optic nerve shadow. This is complicated by the twisting manner by which the optic nerve traverses the orbit and sound attenuation that occurs at these distances from the transducer. In contrast, CT allows for a better overall view of the optic nerve (and tumour) as they traverse the orbit.^{4,5}

Three dimensional ultrasound C-scan imaging is a non-invasive, quantitative, and inexpensive method to screen for optic nerve asymmetry and optic nerve tumours.

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An infected hydrogel buckle with *Corynebacterium pseudotuberculosis*

Scleral buckling is still the most common procedure to repair a rheumatogenous retinal

detachment. Acute or chronic infection of scleral explant is rare but well recognised serious postoperative complication threatening the eye and jeopardising the retinal attachment and visual outcome. They may present acutely as painful red eye with purulent discharge or chronically with extrusion of the explants. The reported incidence varies between 0.5% and 5.6%.¹ Surgical technique, different synthetic materials of scleral explants, duration of surgery, size, and position of buckle affect the rate of infection.

In the largest retrospective review of 757 patients with episcleral buckle for rheumatogenous retinal detachment, Roldan-Pallares and associates had reported 1.3% patients requiring removal of the implant with the commonest seen in silicone sponge (9%) and the least common encountered in hydrogel implant (1.3%).²

Smiddy *et al* have studied 45 cases of scleral buckling infection and identified coagulase negative staphylococci being the most common isolates (17 of 33 positive cultures), and the others include *Staphylococcus aureus*, *Bacillus*, and *Mycobacterium*.¹ *Corynebacterium pseudotuberculosis* is a rare zoonosis and, apart from its rare description in human lymphadenitis, it has not been reported in the ophthalmology literature. The isolates from the scleral buckle infection of our case was susceptible to penicillin and vancomycin. The treatment regimen and possible sources of the infection have been explored.

Case report

A 63 year old white man presented with 8 week history of dull ache over his left eye coupled with mucopurulent discharge. He had received an uneventful scleral buckling surgery with encircling silicone rubber band, 5 mm radial hydrogel episcleral sponge, and cryopexy for his left eye retinal detachment 8 years earlier. On examination, the visual acuity was 20/20 in his right eye and 20/50 in his left. Examination revealed exposed hydrogel scleral buckle with surrounding conjunctival oedema and hyperaemia (fig 1). Fundus examination showed a clear view and an attached retina with good buckle support. There was no feature of erosion and the chorioretinal adhesion from previous retinal cryopexy looked adequate. An infected buckle was diagnosed and the removal of buckle was arranged. Intraoperatively, the hydrogel buckle was noticed to be decomposed into a mess and it had to be removed in pieces. The scleral bed was irrigated with copious gentamicin solution. Gram smear of the specimen showed Gram positive bacilli and culture

Figure 1 Exposed hydrogel scleral buckle with conjunctival chemosis, hyperaemia, and purulent discharge.

grew *Corynebacterium pseudotuberculosis*, which was sensitive to penicillin and vancomycin but resistant to erythromycin. One week of systemic ampicillin and cloxacillin together with 4 weeks of topical penicillin and vancomycin were prescribed. The conjunctival wound healed without sequelae and the retina remained attached. Upon inquiry, he did not have any history of trauma or gross contamination over the ocular surface. There was no recent travel history within 6 months from buckle exposure. He had constant contact with animals, as a dog was kept as a pet at home.

Comment

A hydrogel episcleral implant is the most resistant material to be infected in buckling surgery.² This peculiar clinical and bacteriological pattern may be related to the intricate physiochemical and biocompatibility characteristics of hydrogel. The low infective incidence of hydrogel implant was believed to be because of the lack of dead spaces and probable antibiotic absorption and depot effect.³ However, observations related to the fragility, swelling, and fragmentation of the hydrogel material with time were made since the first report in 1997 and the dead spaces created could possibly contribute to delayed episcleral implant infection years after surgery.^{2-4,6}

Corynebacterium pseudotuberculosis is a veteran infection and throughout the literature only 25 cases have been reported in humans and 22 of them have been reviewed.⁷ Exposure is usually occupational especially with a history of contact with sheep. The sheep farming industries within New Zealand and Australia are particularly involved. Infected humans generally presented with lymphadenitis, abscess, and constitutional symptoms.

Animal acquired infection was deemed as the most probable source of infection in our patient since he had a contact history with domestic animals, but otherwise no gross ocular soiling or contaminations was noted. Just like other human infection, the presentation of scleral buckling infection is closely related to the virulence and infective dose of the offending organisms. Our case may represent the first human ocular *Corynebacterium pseudotuberculosis* infection involving a scleral buckle after retinal reattachment operation.

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A deficit in visits to the optometrist by preschool age children: implications for vision screening

Vision screening in children is aimed primarily at detecting non-strabismic amblyopia (other forms of vision defect are generally evident to parents). Such non-strabismic amblyopia occurs mostly as a result of uncorrected refractive errors.^{1,2} In the December 2003 report by the Child Health Sub-group³ it was recommended that all 4-5 year olds should receive vision screening. The Health For All Children 4 (HFAC4, 2003) "Hall Report"⁴ and the Children's Eye Health Working Party guidelines⁵ similarly suggest vision screening should be undertaken in all 4-5 year olds. This advice is in accord with the results of the first randomised controlled trial of treatment for amblyopia,² which found that treatment of moderate amblyopia (acuity 6/36-6/18) in

preschool aged children was effective. However, currently the coverage of vision screening is patchy, and numbers of specialist screening personnel may be insufficient to meet demand if the recommendation to screen all 4-5 year olds were to be implemented.⁶ In districts where vision screening is not carried out, optometrists might act as an important safety net by providing an additional route for referral of non-strabismic amblyopes.

Methods

As part of an investigation into the genetics of myopia,⁷ we investigated the age distribution of individuals attending for a sight test at 19 optometry practices in northern England during the period January 2000-December 2001. For subjects attending more than once, only the most recent visit was recorded. Of the 90 884 attendees, age was known for 90 750. None of the optometry practices operated in a manner that would be expected to discourage the attendance of children. The age distribution of this optometric cohort was compared with data from the census of England and Wales, conducted in 2000.

Results

Figure 1 shows the age distribution of the optometric cohort compared with that of the year 2000 census. Although the optometry practices were not selected according to defined epidemiological sampling criteria, the high similarity in the age distribution of the two datasets after the age of 10 suggests the optometry attendees are generally representative of the UK population. However, there was a clear deficit in visits to optometrists in the preschool age group, which was highly significant ($\chi^2 = 4186.4$, $df = 1$;

Figure 1 Age distribution of subjects visiting optometric practices ($n = 90\,750$) and in the 2000 population census for England and Wales ($n = 52\,041\,916$). Note the deficit in numbers of children under the age of 10 years (see inset figure for detail), and the increased attendance of patients >45 years old coinciding with the onset of presbyopia.

$p < 0.0001$). Attendance to optometrists appeared to increase linearly until about age 11 when it reached adult levels (fig 1, inset). Our analysis suggests that only ~7% of children aged 0–5 years visit an optometrist (1.48% of visits in the optometric cohort were for infants aged 0–5 years, and there were 16.6 million sight tests carried out in Great Britain in total,⁸ in the year 2000, suggesting 246 000 tests on the 3.7 million infants in this age group). Because infants in whom a refractive error has been detected are likely to visit their optometrist each subsequent year, this figure must be an overestimate of the proportion attending for the first time—that is, in a screening context.

Comment

The fact that a visit to the optometrist is such an exception to the rule at this age underlines the importance of vision screening programmes, and suggests that every effort should be made to implement a comprehensive system of screening at age 4–5 in order to detect children likely to benefit from early treatment for amblyopia. However, where such programmes are not in place, we suggest that encouraging children to visit an optometrist should help in the early referral of non-strabismic amblyopes.

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"Only rarely seen in dreams"—visual experiences during cataract surgery

Cataract surgery is the most commonly performed elective surgery in many countries including the United Kingdom.¹ With the majority of procedures performed under local anaesthesia, it is important for surgeons to recognise if patients are indeed visually aware of their environment. Understanding their experience would be a step forward in providing the safest and the most effective ophthalmic care to cataract patients.

Clinical significance of patients' visual experience lies in the fact that a large number of patients are frightened by their experience, which potentially leads to a number of problems.^{2–3} This could range from poor cooperation during surgery to a sympathetic surge with undesirable adverse effects of hypertension, tachycardia, hyperventilation, and acute panic attack.

Since the visual disturbances during cataract surgery can cause fear and anxiety and adversely affect patient satisfaction, any measure that could reduce its negative impact would contribute to making the operation safer and more bearable.

Visual experiences during cataract surgery have not been discussed in any major ophthalmic textbooks and have not been well studied until recently.^{2–6}

It is commonly expected by the majority of ophthalmologists that patients are not able to perceive much with the eye being operated on during surgery. Even the patient information leaflet published by the Royal College of Ophthalmologists, London, states, "you will not be able to see what is happening, but will be aware of a bright light."¹ This advice, unfortunately, may not be accurate in a sizeable proportion of patients undergoing cataract surgery.⁷

A number of artists have expressed their experience during cataract surgery previously.^{3–8} Two of our patients also wrote back describing their visual experiences. Both underwent uneventful cataract surgery by phacoemulsification and intraocular lens implantation in our unit. One was a professional artist and the other a local poet. The artist sent us an elaborate drawing resembling a "colourful monkey" which portrayed his visual experience (fig 1). The poet sent us a poem, inspired by his visual perception (fig 2). His words clearly reflect the drawing. Taken together the drawing and the poem can in fact provide a tangible insight into how patients may visually experience cataract surgery under local aesthetic.

Figure 1 Artist's impression of his visual experiences during cataract surgery.

Wondrous light from laser beams
To show such strong dramatic scenes
Only rarely seen dreams
This helps the eye to see

Bright and beautiful coils of light
Crystal clear to heal the sight
Soft and warm and glowing bright
Fascinating mystery

Subtle shades of pink and blue
Smoky white and yellow too
Will these show the same for you
As they did for me?

Our thanks to those who show the light
Their skills and loving care delight
And much improve our failing sight
A wondrous place to be

Figure 2 Poem inspired by visual experiences during cataract surgery.

This documentation of visual experiences during cataract surgery could prove helpful to counsel patients on what to expect during the procedure. An explanation of possible visual experiences during local anaesthesia may relieve patient anxiety and should be included in patient information leaflets regarding cataract surgery. This could provide a useful tool to offer some reassurance to the anxious patients about to undergo the procedure. Patient counselling in this way may increase patient comfort and cooperation during the entire procedure.

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Spontaneous closure of microaneurysms in diabetic retinopathy with treatment of co-existing anaemia

Pathogenesis of diabetic retinopathy is multifactorial. Various potential risk factors include hypertension, proteinuria, and duration of diabetes, use of insulin, chronic renal disease, and anaemia. Management of diabetic macular oedema has mainly focused on laser photocoagulation of leaking microaneurysms. While anaemia has been found as an independent risk factor for the development of high risk proliferative diabetic retinopathy,¹ its correction has not received due attention in the management of diabetic retinopathy. We report a patient with insulin dependent diabetes mellitus (IDDM) with coexisting nutritional anaemia, who showed spontaneous closure of the microaneurysms on correction of anaemia and metabolic control.

Case report

A 39 year man with IDDM for 12 years, presented with bilateral decrease in vision for 3 months. Examination revealed a visual acuity of 20/40 in both eyes and normal anterior segment. Fundus examination showed multiple microaneurysms, cotton wool spots, and superficial retinal haemorrhages scattered throughout the posterior pole in both eyes. Fundus fluorescein angiography showed multiple microaneurysms with focal leakage in both eyes (fig 1). Review of his systems was essentially normal. Laboratory results showed low haemoglobin (4.7 g%), raised erythrocyte sedimentation

Figure 2 Same patient 3 months later shows resolution of retinopathy (A and B). Multiple microaneurysms in posterior pole of both eyes show spontaneous closure (C and D).

rate (ESR) (65 mm in the first hour) and hyperglycaemia (fasting blood sugars -242 mg/dl). Peripheral blood film showed moderate anisocytosis and microcytosis of red cells. Total leucocyte count, differential leucocyte count, platelet count, serum electrolytes, urea, creatinine, 24 hours urinary proteins, and bilirubin were within normal limits. Chest x ray, ultrasound abdomen, stool for occult blood, duodenal biopsy, and serum electrophoresis for Waldenstrom's microglobulinaemia were normal. He was labelled as a case of nutritional (iron deficiency) anaemia. He received blood transfusion (two units) and started on iron, folic acid, vitamin B1, B6, and B12 supplements.

His insulin regimen was modified. After 3 months of therapy, his haemoglobin improved to 14 g/dl and blood sugars were normal (fasting blood sugars 110 mg/dl). His visual acuity improved to 20/20 in both eyes. Fundus examination showed spontaneous closure of majority of microaneurysms and resolution of superficial haemorrhages and cotton wool spots in both eyes (fig 2).

Comment

In our patient the retinopathy was characterised by multiple microaneurysms, cotton wool spots, and haemorrhages, which were highly suggestive of moderately severe non-proliferative diabetic retinopathy. Anaemia is known to produce a retinopathy that is characterised by haemorrhages and cotton wool spots, and occasionally hard exudates. To our knowledge development of microaneurysms has not been reported in nutritional anaemia. The Diabetes Control and Complications Trial (DCCT)² has shown that intensive management of diabetes reduces the development and progression of retinopathy in the long run but spontaneous closure of microaneurysms was not noted in this study. A large cross sectional study³ found a twofold increase in risk of retinopathy in patients with haemoglobin less than 12 gm/dl, when controlled for other known risk factors. Shorb *et al*⁴ reported three diabetic patients with severe iron deficiency anaemia, who rapidly progressed to severe proliferative retinopathy. Friedman and associates⁵ reported resolution of macular hard exudates in five patients who were treated with erythropoietin for coexisting anaemia. The authors did not speculate on the mechanism of resolution of hard exudates. It is unlikely that a better metabolic control alone led to spontaneous closure of microaneurysms in our patient. It is more likely that anaemia induced retinal hypoxia played a major part in the development of microaneurysms and other retinopathy changes. We postulate that correction of

Figure 1 Fundus photographs of right eye (A) and left eye (B) showing superficial haemorrhages and cotton wool spots. Corresponding fluorescein angiograms (C and D) show multiple microaneurysms in posterior pole of both eyes.

hypoxia may be the possible mechanism in improvement of the retinopathy.

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MAILBOX

In answer to "Who is Ivan Schwab?"

Generally, a man must commit a heinous crime, donate prodigious sums of money, or have mortality intervene to have an editorial directly questioning who he is. With this letter, I certify that, at least, as of this writing, none of the above has occurred.

None the less, some concern has been expressed regarding my credentials to write the essays that accompany the cover photographs for the *BJO* of the past few years.¹

As was hinted in the editorial "Eyeshots to eyeshine"² early in this series, my education in this regard is not appreciably different from that of most of the readers, but that education has been a powerful tool. Presumably, Papalkar and Francis³ are both ophthalmologists and have been trained with a science background, medical school, and the appropriate residency requirements to qualify for their chosen profession. This education allows us to understand optics, neurology, and biology at both a clinical and a basic level. I am also certain that these authors have a highly curious intellect. For proof of that proposition, I offer the fact that they read their journals, ask critical questions of the authors, and question credentials. This is key to the question at hand.

As ophthalmologists our training, curiosity, and the pursuit of truth and honesty will provide the dividends of self education. We are, after all, entirely self educated. As a teacher, I can only hope to recruit, stimulate

and, with luck, inspire my students to become better ophthalmologists than I—a teacher's ultimate goal. I can help to open the door to knowledge; the student must walk through it.

With these essays, I hope to teach a bit of comparative ophthalmology and optics and to stimulate your interest and thinking. All essays are written with the assistance of scientific evidence previously published on the topic and often vetted directly by those who did the original work or by others in the field. In the interest of space, I reference only a few of these publications. If the reader discerns mistakes, notifying me will enable me to correct them.

The editorial asking the question "Who is Ivan Schwab?"³ can be answered simply by "one of you." I am flattered by the interest in my qualifications, because that tells me that you are reading your journals; in particular, you are reading my essays, and above all, you are asking questions. Stay tuned.

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Ciprofloxacin in endophthalmitis: an alternative to ceftazidime and amikacin!

I read with great interest the letter by Doft *et al*¹ suggesting amikacin to be a better alternative to ceftazidime, in response to the article by Galloway *et al*,² that suggested the converse. I would like to suggest that ciprofloxacin is a better alternative to both these drugs. There are certain points that I would like to mention to support my statement.

(1) It has been shown that vancomycin and ceftazidime are incompatible upon mixing, with precipitate formation.³ In addition, Kwok *et al* have suggested ceftazidime to be relatively ineffective owing to its higher rate of precipitation in the vitreous at body temperature resulting in a free antibiotic concentration much less than the MIC₉₀ of the organisms.⁴ Interestingly, in the study, ceftazidime was precipitated to a significant extent, especially when prepared in balanced salt solution plus (BSS Plus) rather than in normal saline (NS), with up to 88% loss in concentrations of the measurable free antibiotics. Such a low antibiotic concentration would be inadequate for the treatment of a potentially blinding disease like infective endophthalmitis. Hui *et al*, in an elegant study, measured the concentrations of vancomycin and ciprofloxacin in an equilibrium dialysis chamber by high performance liquid chromatography and fluorescence polarisation immunoassay.⁵ They did note that ciprofloxacin precipitates in vitreous, but to a much lesser extent than ceftazidime and, significantly, the remaining ciprofloxacin concentration was many times above the MIC₉₀ of the drug against the common Gram

negative bacteria encountered. This suggests that the problem of precipitation might not be so important in the use of intravitreal ciprofloxacin. The precipitation of ciprofloxacin was also found to be independent of the medium, which means that there is no need to avoid the use of BSS Plus during preparation of the ciprofloxacin for intravitreal injection or during intraocular surgery.

(2) Various studies have shown the efficacy of ciprofloxacin. Benz *et al* have shown that 92% of Gram negative organisms in culture proved endophthalmitis were susceptible to ciprofloxacin.⁶ In the Indian scenario too ciprofloxacin is considered to be a very dependable drug. In fact, 88.4% of even the Gram positive organisms in the series of Anand *et al*,⁷ were sensitive to ciprofloxacin. This is a significantly lower rate of resistance of Gram positive organisms to ciprofloxacin than that found in the ex vivo study. The series of post-traumatic endophthalmitis over a period of 2 years from our institute also shows 26 of the 39 isolates to be sensitive to ciprofloxacin and a hitherto unreported poor rate of susceptibility to ceftazidime (four out of the 39 isolates) (unpublished data).

(3) The intravitreal combination of choice for the initial empirical treatment of endophthalmitis could be vancomycin and ciprofloxacin. A certain amount of synergy could be expected with this combination, with vancomycin inhibiting the cell wall synthesis of the bacteria allowing ciprofloxacin to penetrate into the cell and inhibiting the DNA supercoiling. This synergy and the resultant greater bactericidal activity would be all the more important considering that there is no assistance from the body's immune system in combating the intraocular infection. Although it has been proved to be non-toxic in animal models,⁸ this substitution of ceftazidime with ciprofloxacin of course, would necessitate further studies on the safety profile of intravitreal ciprofloxacin.

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No evidence for severe retinopathy of prematurity following sildenafil

Marsh and colleagues¹ raise the spectre of a possible association between the use of sildenafil and the development of retinopathy of prematurity (ROP) in a baby of 26 weeks gestation with pulmonary hypertension. We are concerned that this report offers no real evidence for its claims and that a potentially lifesaving agent is being unfairly maligned.

The report describes the use of intravenous sildenafil of unspecified dose for 16 days in a 525 g preterm infant with a very difficult intensive care course. The management included a litany of recognised causes of ROP, including extreme prematurity, >6 weeks of mechanical ventilation with 80–100% oxygen, and bacterial and fungal infections.

Despite this, Marsh *et al* chose to incriminate sildenafil as the causal agent. The suggestion is even more perplexing as the baby had already received inhaled nitric oxide at high levels (40 ppm for 2–3 weeks) before the sildenafil; both are vasodilators and have the same mechanism of action.

The authors make the further statement that they observed a recent increase in treatable ROP in their unit, coinciding with the use of sildenafil. Where is their evidence?

As far as we are aware there is no evidence in the literature that sildenafil has any significant effect on either retinal or choroidal blood vessels. Pache *et al* reported² that in adults, sildenafil induced a 5.8% dilatation of retinal vessels but this was not confirmed by Grunwald *et al* on either retinal or choroidal circulations.^{3,4} To date there are no data on the effect of sildenafil on the developing ocular circulations.

We entirely agree that vigilant monitoring and responsible reporting of side effects are mandatory for any new drug application. To our knowledge the only available intravenous sildenafil is being released on a named patient basis in a prospective study in neonates. How did the authors obtain and administer the drug in neonates? Sildenafil and inhaled nitric oxide are experimental therapies within the preterm population and as clinicians we have a responsibility to ensure that they are used as part of prospective randomised controlled trials with the appropriate short and long term follow up. Although being well intentioned, such unconvincing reports may impede the use of agents that might have an important future role in the management of primary pulmonary hypertension of the newborn.

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Identification of silicone oil in ocular tissues

I read with interest the article by Miyamoto *et al*.¹ Numerous studies have examined the inter-relation between silicone oil and various ocular structures, such as the retina, iris, or anterior chamber. However, the silicone oil itself was never evident in these studies, but rather small vacuoles suspected to be ghosts of the incorporated silicone oil were apparent. Nevertheless, the vacuoles also could have been artefacts² because silicone, which is solubilised in the organic solvents during the preparation,³ is never detected. We previously demonstrated silicone oil emulsion in the rabbit retina using phthalocyanine blue as a marker.⁴ This compound contains a copper molecule that imparts a blue colour and remains in the tissue after the silicone oil is washed out and enables visualisation of the silicone emulsion in the trabecular meshwork at the light and electron microscopic levels. When we injected a suspension of the dye into the anterior chamber, the dye filled the small vacuoles within the cells. In contrast, when silicone droplets containing the dye were injected into the chamber, the blue dye formed clusters in small cellular vacuoles (light microscopy) and touched the limiting membranes of the vacuoles (electron microscopy). The silicone droplets were washed out by the organic solvents used to prepare the specimens, and since the dye was insoluble in the organic solvent, it probably precipitated around the vacuoles.⁵ However, figure 5 of this article did not show any limiting membranes or the low magnification of this figure precluded their identification. In addition, energy dispersive x ray analysis is also a useful method to detect silicone oil in tissues.⁶

The authors injected silicone oil that was not emulsified into the vitreous cavity after vitrectomy. However, silicone oil that is not emulsified has a large surface area and high interfacial surface tension and is not incorporated into the tissues. The authors did not show by gross examination whether silicone oil became emulsified during the experiment. They should discuss why they could see residual silicone oil in the rest of the anterior capsule. The readers were not able to obtain information about silicone oil structure.

Although the authors described emulsification of silicone oil related to protein, many factors are involved in this process. Contamination of low molecular weight siloxanes may enhance silicone oil emulsification.

In the discussion, the authors state: "It is likely that lens epithelial cells attaching to oil droplets might be stimulated to express many wound healing related molecules including extracellular matrix components." This is speculation. The central area of the posterior surface of the rest of the anterior capsule is covered with accumulated fibrous extracellular matrix in figures 2B and 4B. However, there were no differences in the expression of collagen types I, III, V, and cellular fibronectin by immunohistochemistry. The authors did not provide these data in the text. If this information is related to their hypothesis, they should demonstrate differences by providing immunohistochemical data.

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Severe ocular trauma caused by an ostrich

We read with great interest the case report of severe vision loss caused by ostrich pecking trauma and would like to bring readers' attention to a case we recently reported about an adult farm worker who lost his vision as a result of an ostrich attack.¹

In our case, a 34 year old man was attacked by the giant bird with consequent severe pain and immediate loss of vision to no light perception. On examination, patient's right eye had significant proptosis with severe limitations of the globe in all directions and irregular full thickness lacerations of the skin. Exploration of the wound revealed two fragments of bony-like tissue but no fractures. Ultrasound examination and computed tomography scan of the orbits revealed a disorganised right globe with multiple scleral ruptures without any bony fractures. Microscopic examination of bony fragments was consistent with avian rostrum.

Human eye injuries caused by pecking of birds are uncommon and are usually labelled as humorous or incidental, and,

consequently, most go unreported. Serious injuries to humans caused by birds have been sparsely reported in the English literature. In the non-English literature, Kuhl reviewed a series of 14 patients with severe eye injuries from 1875 through 1970 caused by birds.² All were penetrating ocular injuries, and some caused permanent visual injuries and/or blindness.

In general, birds are viewed as presenting less of a danger because of the assumption that the bird will take flight if frightened. On the contrary, some birds show aggressive behaviours related to territoriality or breeding. The male ostrich (a flightless bird) is known to establish territory, display aggressive territorial behaviour, and may attack potential predators.³ These two reports of an ostrich attack causing permanent visual loss in adult humans are the first in the ophthalmic literature and emphasise the potential for serious ocular injuries from birds. People living in rural areas and those who work or plan to visit farms should be aware that territorial behaviour of many domestic animals and birds may be a potential risk factor.

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Cost accounting in cost utility analysis of screening and treatment

I would like to make some comments regarding the cost utility analysis in the paper by Hopley *et al*¹ because it is important to understand that the costs should be accounted at the same time and with the same degree of accuracy as outcome data. The economic definition of costs should be used in cost valuation, not the financial definition.²⁻³ The concept implies that all resources consumed by an intervention should be valued, not just those constituting a budgetary line item.

All methods (that is, cost effectiveness, cost minimisation, cost utility, and cost benefit) of economic evaluation in health care have one principle in common: they examine one or more possible interventions and compare the inputs or resources necessary to carry out such interventions with their consequences or effects.⁴ Cost utility analysis aimed to compare different interventions in terms of both quantity and quality of life; we express them as utilities. In this case, competing interventions are compared in terms of cost per utility (for example, cost per QALY).⁴⁻⁷ Values of resources in the cost utility analysis are assigned by defining costs. In accounting costs both tangible items (for example, equipment, drugs, materials, money etc) and intangible items (for example, time and treatment mode) must be taken into account, regardless of whether they are used by and accrue to health services, society, or the single individual.⁶ Costs for some resources may vary because of market forces—for example, rent, exploitation, so it is important to present results not only in monetary terms but also in quantity of resources used.

To allow comparability across different interventions, a 3% discount rate must be used as recommended by most guidelines if economic evaluations are made at different times.⁷

While this is increasingly becoming the practice, most studies have either attempted to estimate costs for alternative therapies retrospectively or, using literature reviews, budgetary line items and healthcare insurance costs sheets. This should be avoided from economic evaluations because it mainly reflects on budgetary formulations and has very little in common with the real cost of intervention.

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NOTICES

Worldwide clinical trials for new technique for early detection of eye disease

A unique new non-invasive technique for high resolution optical imaging of the eye is receiving global acclaim. By combining two high-resolution imaging technologies, the new technique provides doctors with 3-D images of the retina, macula and the optic nerve.

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Vision 2020 Priority Diseases

The latest (redesigned) issue of *Community Eye Health* (No 51) deals with the gaps between aims of Vision 2020 and how far we are still from them, especially in Africa. For further information please contact: Journal of Community Eye Health, International Resource Centre, International Centre for Eye Health, Department of Infectious and Tropical Diseases, London School of Hygiene and Tropical Medicine, Keppel Street, London WC1E 7HT, UK (tel: +44 (0)20 7612 7964; email: Anita.Shah@lshtm.ac.uk; online edition: www.jceh.co.uk). Annual subscription (4 issues) UK£28/US\$45. Free to developing country applicants.

British Oculoplastic Surgery Society

Call for papers for the 5th annual meeting of the BOPSS to be held on 15 and 16 May 2005 at The Belfry, Birmingham. The abstract submission deadline is 4 February 2005, and abstracts can be submitted online at www.bopss.org.

CORRECTION

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In the letter titled Bilateral ischaemic optic neuropathy and stroke after multiple bee stings (*Br J Ophthalmol* 2004;88:1596-8) the authors were listed incorrectly. The correct listing is as follows; J S Schiffman, R A Tang, E U Dorotheo, S S Singh, H M Bahrani. The journal apologises for this error.