Itive diagnosis has importance therapeutically proved GCA.

be normal in 8% of patients with biopsy underlying inflammatory process, yet the signs that mimic many other medical and the cranial arteries in the elderly. It has unknown origin that has a predisposition for Giant cell arteritis (GCA) is a vasculitis of suspected giant cell arteritis imaging in the diagnosis of

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. 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). Magnetic resonance imaging (MRI) has been shown to improve the diagnosis of early Takayasu arteritis. More recently several case reports have described the diagnostic potential of MR angiography and gadolinium contrast MRI in demonstrating the vessel changes of GCA. 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) classification (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. 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.

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

<table>
<thead>
<tr>
<th>Criterion</th>
<th>Definition</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>Age at disease onset &gt;50 years</td>
</tr>
<tr>
<td>2</td>
<td>New headache</td>
</tr>
<tr>
<td>3</td>
<td>Temporal artery abnormality</td>
</tr>
<tr>
<td>4</td>
<td>Elevated ESR</td>
</tr>
<tr>
<td>5</td>
<td>Abnormal artery biopsy</td>
</tr>
</tbody>
</table>

<table>
<thead>
<tr>
<th>Criterion</th>
<th>Definition</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>Development of symptoms or findings beginning at age 50 or older</td>
</tr>
<tr>
<td>2</td>
<td>New onset of or new type of localised pain in the head</td>
</tr>
<tr>
<td>3</td>
<td>Temporal artery tenderness to palpation or decreased pulsation, unrelated to arteriosclerosis of cervical arteries &gt;50 mm in the first hour by the Westergren method</td>
</tr>
<tr>
<td>4</td>
<td>Biopsy specimen with artery showing vasculitis characterised by a predominance of mononuclear cell infiltration or granulomatous inflammation, usually with multinucleated giant cells</td>
</tr>
</tbody>
</table>

Table 2 Summary of results

<table>
<thead>
<tr>
<th>Patient</th>
<th>Age</th>
<th>ACR criteria score</th>
<th>Prompt response to steroids</th>
<th>TAB</th>
<th>MRI scan</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>78</td>
<td>5</td>
<td>Yes</td>
<td>Positive</td>
<td>Negative</td>
</tr>
<tr>
<td>2</td>
<td>67</td>
<td>4</td>
<td>Yes</td>
<td>Negative</td>
<td>Negative</td>
</tr>
<tr>
<td>3</td>
<td>83</td>
<td>2</td>
<td>Equivocal</td>
<td>Negative</td>
<td>Equivocal</td>
</tr>
<tr>
<td>4</td>
<td>88</td>
<td>3</td>
<td>Negative</td>
<td>Negative</td>
<td>Negative</td>
</tr>
<tr>
<td>5</td>
<td>60</td>
<td>2</td>
<td>No</td>
<td>Negative</td>
<td>Equivocal</td>
</tr>
<tr>
<td>6</td>
<td>67</td>
<td>2</td>
<td>No</td>
<td>Negative</td>
<td>Equivocal</td>
</tr>
<tr>
<td>7</td>
<td>87</td>
<td>4</td>
<td>Yes</td>
<td>Positive</td>
<td>Equivocal</td>
</tr>
<tr>
<td>8</td>
<td>67</td>
<td>NA</td>
<td>NA</td>
<td>NA</td>
<td>NA</td>
</tr>
<tr>
<td>9</td>
<td>69</td>
<td>NA</td>
<td>NA</td>
<td>NA</td>
<td>NA</td>
</tr>
</tbody>
</table>

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

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4 Tanigawa K, Eguchi K, Kitamura Y, et al. Magnetic resonance imaging in detection of arterial and pulmonary artery wall thickening in the acute stage of Takayasu arteritis: improvement of
Bilateral ischaemic optic neuritis and stroke after multiple bee stings

Despite the common occurrence of insect stings and local and systemic allergic reactions, 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 antiepileptics 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 imaging (MRI)/magnetic resonance angiography (MRA) were performed showing a large right temporoparietal haemorrhagic infarct (Fig 1A, B). An ophthalmological 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 pseudohypoparathyroidism. 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.

Pupillar 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 haemorrhage and cotton wool spots in both eyes consistent with anterior ischaemic optic neuropathy (AION). Both maculas were remarkable 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 bilateral 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. 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. 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 patient 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. Bee venom itself contains histamine, thromboxane, leukotrienes, 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.
<table>
<thead>
<tr>
<th>Author/Ref</th>
<th>Day</th>
<th>Age/sex</th>
<th>Type of stings: location</th>
<th>Onset of neurological deficit</th>
<th>Examination findings and symptoms</th>
<th>MRI/CT findings</th>
<th>Treatment</th>
<th>Recovery</th>
</tr>
</thead>
<tbody>
<tr>
<td>Starr and Brasher</td>
<td>36/M</td>
<td>36/M</td>
<td>Wasp: multiple on neck, face, and arms</td>
<td>1 hour</td>
<td>Headache, seizure, right hemiplegia, coma</td>
<td>NR; necropsy showed left haemorrhagic cortical infarct</td>
<td>Cortisone, antihistamines, phenobarbital</td>
<td>Deceased</td>
</tr>
<tr>
<td>Riggs et al</td>
<td>38/M</td>
<td>37/M</td>
<td>Wasp: 3 stings on arms</td>
<td>1 hour</td>
<td>Seizure, right hemiplegia</td>
<td>NR</td>
<td>Left cerebral infarction (CT done 14 months later)</td>
<td>Partial right hemiplegia, one seizure</td>
</tr>
<tr>
<td>Riggs et al</td>
<td>52/M</td>
<td>38/M</td>
<td>Wasp: multiple on left face and neck</td>
<td>2 days</td>
<td>Right hemiplegia, dense global aphasia</td>
<td>NR</td>
<td>Ischaemic infarction in the distribution of the left MCA; angiogram: left ICA occlusion</td>
<td>NR</td>
</tr>
<tr>
<td>Speach et al</td>
<td>30/M</td>
<td>30/M</td>
<td>Bee: single</td>
<td>&lt;1 hour</td>
<td>Decerebrate posturing, extensor plantar reflexes, left hemiparesis, hyporeflexia; after coma, patient had motor apraxia and left sensory neglect</td>
<td>NR</td>
<td>Normal MRI and CT</td>
<td>NR</td>
</tr>
<tr>
<td>Bhat et al</td>
<td>35/M</td>
<td>35/M</td>
<td>Bee: multiple</td>
<td>1 day</td>
<td>Swellings all over the body, vomiting, dysarthria, tinnitus, vertigo and swaying gait, hypertension, bilateral cerebellar signs, rhabdomyolysis with acute renal failure</td>
<td>No papilloedema</td>
<td>Bilateral cerebellar haemorrhagic infarct; SPECT: hyperperfusion of the left dorsolateral frontal cortex, but no areas of hypoperfusion or other abnormalities</td>
<td>Deceased</td>
</tr>
<tr>
<td>Present report</td>
<td>57/F</td>
<td>57/F</td>
<td>Bee: multiple on neck, head, R eye, R side of her neck, face and R arm</td>
<td>2 days</td>
<td>Nausea, vomiting, vision loss</td>
<td>BCVA of 20/15 right eye, 20/25 left eye; left homonymous hemianopia, left inferior arcuate and right altitudinal defects; Bilateral oedema (right eye, left eye) with pallid haemorrhagic swelling</td>
<td>Haemorrhagic infarct 2 days post-ischaemic stroke</td>
<td>Normal VF</td>
</tr>
</tbody>
</table>

NR = none reported.
Strabismus is a common association in patients with craniosynostosis or craniofacial dysostosis (60–70%). 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.

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.

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 incomitant 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 rectus 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 overelevation in adduction and under depression in adduction may be due to the anatomical displacement of the rectus muscles.7

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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.3 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 1957.4 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 demonstrated stable ophthalmic findings and visual acuity.

Comment

Although ocular symptoms associated with West Nile virus were first reported in 1936, 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 2002.8 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.8–12 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.14 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 choroidal atrophy as the disease becomes inactive and subsequently becoming more prominent with time.15–17 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. 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.

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 postoperative 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 rim, 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 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 eye piece (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 petechiae and changes in the eyelid skin, 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. 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.

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 are not tightly sealed.

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

References

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 (“b” labels). In area 1 of the trace, the goggles were overtight and in area 2 they were comfortable.

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www.bjophthalmol.com
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 transplantation (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 severe 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 through out his body extending to his eyelid margins. He had a generalised vesicular rash throughout his body, a generalised vesicular rash, and a tonic pupil after BMT.

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. Daily 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.

Since 1968 these diseases have been successfully treated with haemopoietic stem cell transplantation. Varicella infection has been associated with severe immune dysfunctions following BMT and it has been shown that severe disseminated varicella infection causes ocular disease that mimics the sequelae of herpes zoster ophthalmicus.

In the adult population, the commonest cause of interstitial keratitis is HSV infection whereas varicella infection is considered a rare cause. In children, although varicella infection is extremely common, ocular complications of this disease are rare.

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. In this setting, other documented complications in association with SCID include bilateral viral endophthalmitis, CMV retinitis, and optic neuritis. 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 infection caused by SCID.
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 putative 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.2 In other words SCID mice reconstituted with T lymphocytes of the CD4+ phenotype developed subsequent corneal lesions in relation to HSV infection. Conversely Mercedal et al have shown that unconstituted SCID mice remained lesion free when infected with HSV.2 This suggests that herpes simplex keratitis is an HLA mediated immunopathological reaction to virus in the cornea. In our case the corneal changes occurred following bone marrow reconstitution. Before BMT the CD4+ CD44+ count was 0.04 × 10^9/l but 7 weeks following BMT the CD44+ count was 0.47 × 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 ganglinitis affecting the ciliary ganglion and the short posterior ciliary nerves, a rare but previously described complication of varicella infection.5 Other reported cases of ophthalmic immune recovery disease include a case of varicella zoster virus associated anterior stromal keratitis in a patient with AIDS6 and, in another case, in association with CMV retinitis.7

Considering BMT, varicella zoster virus associated disease can be a frequent complication following autologous and allogeneic transplantation.8 Other complications in relation to BMT include pseudomembranous conjunctivitis,9 keratoconjunctivitis sicca,10 cataracts,11 and severe graft versus host disease.12

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 this is an increased risk of ocular disease.

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Ocular mucular dystrophy in an 11 year old boy

Ocular mucular dystrophy (OMD) is an inherited ophthalmopathy characterised by a progressive decline of visual acuity without visible fundus abnormalities.4,5 In these patients, the fluorescein angiograms and conventional full field electroretinograms (ERGs) are normal, but the amplitudes of the focal mucular ERGs and multifocal ERGs are significantly reduced but only in the central retina.4,6

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 old. Here, we present an 11 year old boy who was diagnosed as having OMD because of the results of electrophysiological and psychological 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 in 6 years of age, but had decreased to 20/33 at 10 years of age. Family history revealed that 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 1-2 target within 10 degrees in both eyes. A moderate red-green defect was found on the Ishihara pseudosochromatic plates, Hardy-Rand-Rittler pseudosochromatic 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 spots’ were severely reduced and essentially absent (fig 2B). The multifocal ERGs demonstrated a loss of focal 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.2 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.1,4

OMD in children is very rare.5,7 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
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. 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 1 2 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 epithelium 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 ceftazidine. The right eye was not scraped, responded well to topical ciprofloxacin, and ceftazadine. The right eye was not sensitive to ciprofloxacin, ofloxacin, gentamicin, and ceftazadine.

Potential complications are the same as those for all contact lenses and have been documented in a recent case series in the United States.1 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, punctuate 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. “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.2 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.3

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 cardio-pulmonary compromise was a primary vendor's Eye Contact Lenses 2003;29:196–200.

1 Cosmetic contact lenses draft explanatory notes.
2 The United Kingdom Parliament Bill moved by Mr John Robertson (Glasgow, Anniesland): Non-Prescription Contact Lenses. 28 October 2003: Column 176, 1.21 pm.

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

Figure 2 Residual central corneal infiltrate at 1 month after presentation.
Mice. The Dp71 isoform of dystrophin suffices to induce neovascularisation alone. This is not prevalent in the Duchenne population, pathology in adults. Retinal neovascularisation is routinely associated with proliferative retinopathy, or choroidal rupture. Visual acuity in that eye was 20/25. Funduscopic examination disclosed dot and blot haemorrhages 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.


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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.1–3

Orbital decompression is a common surgery performed to treat patients with thyroid related orbitopathy for functional or cosmetic indications.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 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 intracanal fat removal. Three days after surgery she noted spots in front of her left eye. Visual acuity in that eye was 20/25. Funduscopic examination disclosed dot and blot haemorrhage with flame shaped haemorrhages in the posterior pole of the left eye (fig 1).

In summary, rapidly progressive, bilateral proliferative retinopathy may be associated with DMD in the presence of severe cardipulmonary 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.
Retinal haemorrhages associated with ocular decompression surgery are rare complications that may occur after glaucoma filtration surgery. It is associated with scattered retinal haemorrhages concentrated in the posterior pole. The haemorrhages may be diffuse, both in deep and superficial layers of the retina, and may even show white centres when first observed.

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.

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.

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Recent clinical findings.\(^5\) Retinal damage was not evident by morphological, angio-
graphic, 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.\(^5\) To our knowledge, this is the first report of ICG induced retinal nerve fibre damage assessed by scanning laser polarimetry.

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Competing interest: none.

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with and without indocyanine green-assisted peeling of the internal limiting membrane.
epiretinal membrane with indocyanine green-assisted internal limiting membrane peeling.

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), 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

Glaucma Society Silver Jubilee Meeting 2004
The Silver Jubilee Meeting and Dinner for the Glaucma 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 8BT (tel:01375 3835172; e-mail: glaucos@uceirc freeserve.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 20 666 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.
Cause of V pattern strabismus in craniosynostosis: a case report

F G Velez, N Thacker, M T Britt and A L Rosenbaum

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