Wegener’s granulomatosis

Wegener's granulomatosis (WG) is a systemic granulomatous inflammatory disease of unknown origin. It occurs at any age, with the peak incidence in the third and fourth decades. The classic diagnostic triad of WG is necrotising granuloma of the upper or lower respiratory tract, vasculitis, and nephritis. Identifying a raised ANCA titre is virtually diagnostic of the disease, especially with the classic granular cytoplasmic staining pattern of neutrophils (C-ANCA). This disease is usually fatal if untreated; however, early diagnosis and instigation of immunosuppressors such as cyclophosphamide can induce long term remission.1

Case report

A 79 year old woman presented with a 2 week history of a painful, red left eye. She had mild arthritis and was also noted to be profoundly deaf, requiring hand written notes for communication. There was no history of sinus problems.

Her best corrected visual acuities were 6/18 in the right eye and 6/12 in the left eye. The left peripheral supratemporal cornea showed three areas of 80% corneal thinning, 1 x 1 mm in size, with an overlying epithelial defect (Fig 1). There was adjacent conjunctival injection and moderate anterior chamber cellular activity. The other eye was normal with no signs of inflammation. Examination of her hands showed no obvious rheumatoid abnormalities. Initial investigations performed were full blood count, electrolytes, C reactive protein, erythrocyte sedimentation rate (ESR), and rheumatoid factor. Corneal scraping and cultures were also undertaken to exclude an infective cause. The only significant test result was a raised ESR of 94, with other tests including creatinine being within the normal range. A provisional diagnosis was made of rheumatoid corneal melt and treatment was commenced with topical dexamethasone 0.1% non-preserved hourly and oral prednisolone at 40 mg/day. After 5 days of steroid treatment, the corneal thinning was resolving and the conjunctival injection settling (Fig 2). Also by the fifth day, the patient’s hearing had dramatically improved. She could now conduct a conversation at normal volume. On further questioning, she said her hearing had deteriorated only over the past few months and had ascribed it to “natural” old age deterioration. This suggested a link between her corneal changes and hearing loss and she was investigated for Wegener’s granulomatosis (WG). Her serum antineutrophil cytoplasmic antibodies (cANCA) titre was found to be raised (> 160). Rheumatoid factor was negative. Antiretroviral medications was commenced on diagnosis of WG and the oral prednisolone slowly tapered. Her ocular inflammation, hearing, and arthritis continued to improve on treatment and at the 2 month follow up, there was minimal peripheral corneal thinning and no inflammation. Best corrected visual acuity in the left eye remained at 6/12. Examination by an otolaryngologist found mild hearing impairment and no significant sinus pathology. Rheumatological review found only mild arthropathy.

Comment

The majority of the ocular involvement in WG is caused by focal vasculitis, involving the anterior and/or posterior segment of the eye and corneal melting has been reported in 11–16% of patients with WG.2,4 Profound sensorineural deafness is a less common feature, occurring in about 6% of patients on presentation.6,8 It occurs either secondary to inflammation of the cochlear vessels or serous otitis media.9 Other important systemic features include pyrexia, weight loss, recurrent epistaxis, sinus discharge, haemoptysis, peripheral neuropathy, cerebral vasculitis, and renal failure (major cause of death). Fortunately in this case, the patient had the limited form of WG, with no renal involvement.

This patient presented with an uncommon combination of corneal melt and profound deafness. As deafness is a relatively common problem among the elderly patient population, it may be overlooked as an important symptom, especially if ocular pathology is the main feature at presentation. It was not until the patient’s hearing improved on treatment with oral prednisolone that its significance became apparent. WG is a rare condition and early recognition and treatment are essential as the disease carries a significant risk of serious complications. A full systemic history in patients with corneal melt is important in formulating the correct diagnosis and thereby expediting the appropriate treatment.

K S Lim, GA Lee, C E Pavesio, L A Ficker
Moorfields Eye Hospital, London, UK

References


Pigmentary retinopathy, macular oedema, and abnormal ERG with mitotane treatment

Adrenocortical carcinoma is a rare tumour with a poor prognosis. Mitotane (o,p’-DDD), a chemotherapy drug that suppresses the adrenocortical gland and modifies peripheral steroid metabolism has been reported to cause ocular side effects including visual blurring, diplopia, cataract, toxic retinopathy with retinal haemorrhage, oedema, and papilloedema. We present a 32 year old woman with reduced visual acuity, retinal pigmentation, macular oedema, and abnormal ERG after taking mitotane. While primary hypoadrenalism in Addison’s disease has never been reported to cause any retinal problem, secondary hypoadrenalism in adrenoleucodystrophy is associated with pigmentary retinopathy and other ocular findings. We postulate that the retinal problems secondary to mitotane treatment may act via a similar mechanism.

Case report

A 30 year old woman had a left nephrectomy, adrenalec-tomy, and chemotherapy in September 1997 following diagnosis of an adrenal carcinoma. In 1999, she was found to have secondary tumours in her lungs and liver. She was commenced on intra-arterial cisplatin and oral mitotane of up to 4.5 g daily for 6–8 months, ceasing in December 1999 because of weight loss, malaise and, soon after that, marked decrease of visual acuity in both eyes. The patient had no family history of any retinal disease. She had worn glasses for myopia for 9 years with best corrected visual acuity of 6/4 each eye previously.

On 2 March 2000 her visual acuity was 6/12 in the right eye and 6/60 in the left eye. She also had facial pigmentation. Funduscopy showed extensive pigmentary clumping in each eye and macular oedema in the left side (Fig 1). She was commenced on cortisone acetate and fluocortisone to attempt to improve her vision.
One week later her visual acuity was 6/18 in the right and 6/24 in the left. Fundus examination showed mottled pigmentation at the macula and mid-periphery of both eyes but macular oedema in her left eye had resolved. Disc and vessels were normal. Full field electroretinography (ERG) was performed. Her rod response (scotopic, dim blue stimulus) b-wave was 50% of normal, the cone response (photopic, red stimulus) had b-wave 75% of normal amplitude and delayed implicit time (Fig 2). The amplitude of 30 Hz flicker responses was also reduced. There was no significant asymmetry between the two eyes. On 30 March 2000, her visual acuity improved to 6/12 in the right and 6/9 in the left. Her facial pigmentation had also faded.

Comment

Surgical resection is the treatment of choice for adrenocortical carcinoma. Mitotane (o,p’-DDD) is the only drug that causes regression of metastases and improves survival. Its biochemical action is unknown but data suggest that it modifies peripheral steroid metabolism and directly suppresses the adrenal cortex. The incidence of ocular side effects was 4% in a study of 132 patients. These effects include visual blurring, diplopia, lens opacities, optic neuritis, and a toxic retinopathy. Visual loss associated with mitotane has been reported previously. Previous studies also did not mention side effects reversibility except for a case of lenticular opacities that disappeared 5 days after discontinuance of mitotane. In our case, the patient’s visual acuity did improve significantly, with drying out of macular oedema after cessation of therapy and initiation of steroid replacement.

It is possible, however, that the ocular changes in this patient were not caused by mitotane but were secondary to cancer, associated retinopathy (CAR). Its characteristic findings include attenuated retinal arterioles, with limited, if any, clinically apparent retinal pigmented changes and cells in the vitreous humour. CAR was considered to be unlikely in this case because of the normal calibre of the retinal arterioles, the absence of vitreous cells, and the timing of the onset. Patients with CAR experience visual symptoms that often precede or are concurrent with the tumour diagnosis.

Although primary hypoadrenalism in Addison’s disease has never been reported to cause retinal problems, secondary hypoadrenalism in adrenoleukodystrophy is associated with pigmentary retinopathy and other ocular findings. Adrenoleukodystrophy is a group of rare lipid storage disorders with increased serum level of long chain fatty acids (C24–C30). Ocular findings in adrenoleukodystrophy include visual loss secondary to vitreous cells, and the timing of the onset.

Comment

We postulate that the retinal side effects of mitotane could occur via a similar mechanism that affects the metabolism of long chain fatty acids. Mitotane causes a reduction in plasma 17-hydroxy corticosteroids level but an increase in the levels of 6-beta-hydroxyl cortisol, cholesterol, liver enzymes, corticosteroid binding globulin, and sex hormone binding globulin. At the time of writing, there was no published information on whether mitotane treatment affects serum long chain fatty acid levels. The availability of such data will be valuable for evaluating this postulation.

References


Coxsackievirus B4 associated uveoretinitis in an adult

Coxsackievirus is a kind of enterovirus, which may cause respiratory and gastrointestinal symptoms, erythema, meningocerephalitis, myocarditis, pericarditis, and myositis. Although Coxsackievirus B4 is apparently a rare cause of chorioretinitis, Coxsackievirus infection and concurrent posterior segment inflammation have been disclosed in a few cases. However, iridocyclitis, retinal hemorrhagic dots, and occlusive retinal vasculitis have not previously been reported as being features of Coxsackievirus B4 infectious ocular disease.

Case report

A 34 year old man was referred to our hospital with a 2 week history of inferior visual field defect and visual loss in his right eye. His left eye was asymptomatic. A few days before the onset of visual manifestations, the patient noted a prodrome of viral infection, consisting of severe headache and high grade fever of 38.0–39.2°C, joint pain, and general malaise. There was no significant medical or family history. At the initial ophthalmic examination, his best corrected visual acuities were 10/20 in the right eye and 25/20 in the left eye, and intraocular pressure was normal in both eyes. Slit lamp examination showed moderate cellular infiltration in the anterior chamber in both eyes. Humphrey central visual field testing demonstrated relative scotoma inferior fixation in the right eye. Goldman peripheral visual field testing was within normal limits in both eyes. Ophthalmoscopic examination disclosed subretinal exudates with haemorrhage in the parafovea and the mid-periphery of bilateral eyes (Fig 1). Moreover, scattered

Figure 1 Right fundus showing mottled pigmentation at the macula.

Figure 2 Full field ERG.
Coxsackievirus B4 infection.

respectively, we diagnosed the ocular lesions in demonstrating a 16-fold rise between the acute titre to Coxsackievirus B4 had increased, dem-

strated by blocked fluorescence and mild dye leakage from the retinal vessels correspond-

ing to the lesions detected clinically, and irregular choroidal background fluorescence (Fig 2). Laboratory examination findings were normal except a white blood cell count of 10 500 ×10/mm3, GOT of 76 IU/L, GPT of 209 IU/L, gamma-GTP of 88 IU/l, and LDH of 771 IU/l. Serological tests, however, revealed a slight elevation of serum antibody to Coxsackievirus B4, at a titre of 1:8 (neutralisation test).

The other was an adult case, with chori-

oretinitis similar to the multiple evanescent white dot syndrome.1 However, the clinical features observed in our patient consisted of iridocyclitis, scattered haemorrhagic dots, and occlusive retinal vasculitis were apparently different from the two earlier reports.

Coxsackievirus infection is apparently a rare cause of uveoretinitis but, nevertheless, should be considered in the appropriate clini-

Comment

Figure 1 Photographs of right eye (A) and left eye (B) at the initial ophthalmic examination. Subretinal exudates with haemorrhage around the macula, and scattered haemorrhagic blots are evident.

Figure 2 Fluorescein angiograms of the right eye. Note blocked fluorescence and slight dye leakage from the retinal vessels around the macula, and irregular choroidal background fluorescence.

acquired, and may affect primarily the retina or the choroid. Retinitis and choroiditis caused by viruses such as measles, influenza, Epstein-Barr virus, and Rift Valley fever virus typically occur on an acquired basis, subse-

quent to an acute viral systemic illness. Coxsackievirus-associated chorioretinitis is thought to elicit the same clinical course as in other viral chorioretinitis. Coxsackievirus infection and concurrent posterior segment inflammation have been reported in a few cases, in which clinical signs and antibody titres suggested that Coxsackievirus was the responsible agent.1 3 Among them, two cases of choriorretinitis associated with Coxsackievirus B4 infection were reported. One was a paediatric case, and the ophthalmoscopic manifestation was described as scattered white lesions in mid-periphery of the retina.1 The other was an adult case, with chori-

The patient's general condition improved gradually without specific medication. Cellu-

lar infiltration in the anterior chamber gradu-

ally disappeared. The retinal exudative lesions reduced in size, and haemorrhage tended to decrease. Eight months after the initial ophthalmic examination, bilateral exudative lesions had improved and were replaced by scars, and most of the retinal haemorrhagic lesions had disappeared. Visual acuity was restored to 25/20 in the right eye and 30/20 in the left eye. However, relative parafoveal scotoma persisted in his right eye. Antibody titre to Coxsackievirus B4 had increased, demon-

strating a 16-fold rise between the acute

(1:8) and convalescent sera (1:128). Retro-

spectively, we diagnosed the ocular lesions in this patient as uveoretinitis induced by Coxsackievirus B4 infection.

Ophthalmic surgery is well suited to the use of telemedicine in diagnosis and management,1 2 but diagnostic accuracy and reliability are critical if it is to be widely introduced.3 Moorfields Eye Hospital is lo-

Table 1

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Evaluation of telemedicine for slit lamp examination of the eye following cataract surgery

Ophthalmic surgery was recruited to the study and full informed written consent was obtained. A telelink of a pair of Global Telemed mobile workstations connected by three ISDN lines, with a video transmission rate of 384 bits/s, was used for examinations with high resolution and the examination being achieved by static images. Two surgeons conducted examinations, one using the slit lamp and the other using the telemedicine link. Slit lamp signs graded by the two observers comprised the presence and degree of the following—central corneal oedema, corneal oedema at the limbal section, Siedel's sign, folds in Descemet's membrane, anterior chamber depth, flare and cells, intraocular lens decentration, and lens stabili-

Both observers in person independently examined a group of postoperative cataract patients in order to determine agreement between practitioners.

Twelve consecutive postoperative cataract patients were recruited to the study with 10 others as a control group to assess interobserver agreement by in-person examination. The results of the study are given in Table 1.

Comment

Although interobserver variation means that telelink reliability cannot be precisely calcu-

lated in terms of sensitivity and specificity, these findings give an indication of the accu-

racy of slit lamp examination after cataract surgery using 384 K bandwidth. Examination by video telelink was relatively reliable in detecting oedema at the central cornea but did not consistently detect oedema at the cor-

neal section, or anterior chamber flare and failed entirely to detect DM folds or anterior chamber cells. Patients enjoyed the telen-

cine experience finding it reassuring to see as well as interact with their surgeon via the tele-

link.

Video compression algorithms used in streaming video are “lossy” and higher band-

width increases video quality. The process begins with a good video source with low noise, since noise does not compres.

s well. A poor quality video image is very difficult to stream successfully because of this difficulty in compression. The videoconferencing standard H.261 was implemented to provide for video compression to a given ISDN band-

width. With increasing bandwidth there is correspondingly better quality video. A tele-

video link for corneal assessment in Canada using six ISDN lines (768 K) was found to be acceptable, whereas three lines (384 K) were unacceptable (personal communication, Dr M Pop, Montreal, Canada).

The video display also affects the infor-

mation observed. Broadcast video monitors are accurately colour calibrated, whereas dig-

ital compressed video to 384 K bandwidth has

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reduced colour depth. Recognition of corneal oedema is less about well defined structure as subtle display of colour and “haze.”

It is felt by the authors that telemedicine is a wholly different experience from a simple telephone conversation and for many situations is capable of far more postoperative assessment with trained operators. H-261 video at 384 K (triple ISDN) is sufficient for anterior segment overall examination but is not suitable for detailed corneal assessment. Other practitioners have used data rates up to four times faster for more detailed examination.

L F F Smith, J Bainbridge, J Burns, J Stevens, P Taylor, I Murdoch
Moorfields Eye Hospital, City Road, London EC1V 2PD, UK

Correspondence to: Lindsey Smith; 101506.12711@compuserve.com
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Table 1 Consistency of scoring by the two observers

<table>
<thead>
<tr>
<th></th>
<th>Control group (n=10)</th>
<th>Study group (n=12)</th>
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</thead>
<tbody>
<tr>
<td>Oedema of the incision</td>
<td>7/10 (70%)</td>
<td>4/12 (33%)</td>
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<tr>
<td>Oedema of the central cornea</td>
<td>7/10 (70%)</td>
<td>10/12 (83%)</td>
</tr>
<tr>
<td>Descemet’s folds</td>
<td>10/10 (100%)</td>
<td>7/12 (58%)</td>
</tr>
<tr>
<td>Anterior chamber cells</td>
<td>9/10 (90%)</td>
<td>0/12 (0%)</td>
</tr>
<tr>
<td>Anterior chamber flare</td>
<td>9/10 (90%)</td>
<td>6/12 (50%)</td>
</tr>
<tr>
<td>Lens implant stability</td>
<td>9/10 (90%)</td>
<td>12/12 (100%)</td>
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<tr>
<td>Management decisions</td>
<td>9/10 (90%)</td>
<td>11/12 (93%)</td>
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Inherited retinal dystrophy and asymmetric axial length

The prenatal and postnatal development of the eye is determined by complex interactions between a number of genes, their products, and certain environmental factors. Recognition of corneal oedema is less about well defined structure as subtle display of colour and “haze.” Since each eye is influenced by precisely the same processes as its fellow, mutations in regulatory genes usually lead to symmetric phenotypes. In this report, we describe two siblings of Asian ethnicity, born to unrelated parents with no family history of ocular disease, who have an unusual bilateral retinal dystrophy associated with very asymmetrical ocular growth.

Case report

Sib 1 was an 8 year old boy with epilepsy who had been born at term after an uneventful pregnancy. Pendular nystagmus had been noted soon after birth and his visual acuity in each eye was 3/60; refraction =17.00 DS right, plano left. Anterior segment examination was unremarkable with normal intraocular pressures. Examination of the right eye (Fig 1A and B) revealed generalised retinal pigment epithelial and choroidal atrophy, a macula “coloboma,” sheathed and occluded retinal vessels inferotemporally, and a shallow inferotemporal retinal detachment with subretinal fibrosis. The right eye appeared similar; however there was no macula abnormality. By ultrasound, the axial length and ocular volume were 26.70 mm and 9.9 ml right, 20.70 mm and 4.6 ml left respectively.

Sib 2 was a 6 year old girl born at term with no significant past medical history. Her corrected visual acuity was 3/60 in each eye; refraction =17.00 DS right, plano left. Anterior segment examination was normal. Funduscopy of the right eye revealed a “macula coloboma,” extensive retinal pigment epithelial and choroidal atrophy, intraretinal pigment migration and preretinal fibrosis. The appearances of the left eye were similar. No retinal detachment was noted. Axial lengths and ocular volumes were 26.1 mm and 9.4 ml right, 20.7 mm and 4.6 ml left respectively.

Electrophysiology was performed on both children based on the ISCEV standards for adults, but initially using surface electrodes on the lower eyelids. No definite ERG response could be recorded from the emmetropic left eye of either child (even with high intensity stimulation or photopic flicker stimulation with averaging), a finding consistent with severe generalised dysfunction involving rod and cone photoreceptors. The electrical responses of both children’s myopic fellow eye showed a milder degree of dysfunction: there were reduced b/a ratios in both scotopic and photopic responses which, together with the abnormal 30 Hz flicker ERGs, suggest a mid-retinal locus affecting phototransductional cone and rod systems.

The skin recordings obtained with surface electrodes were confirmed under anaesthesia using gold foil corneal electrodes.

It is reasonable to conclude that these two siblings represent original probands with a novel inherited, possibly autosomal recessive, retinal dystrophy. The particularly interesting feature of this disorder is the asymmetry of the axial lengths and ocular volumes of the eyes, rare in genetically determined ocular diseases. Indeed, we are aware of only one other report that identifies ocular asymmetry in association with retinal degeneration. Lafaet et al. reported a single patient who had bilateral Stargardt’s disease and unilateral myopia.

Normally, axial elongation and ocular enlargement are carefully coordinated to equalise growth of fellow eyes in the quest for emmetropia.1 The differences observed in the two siblings described here reflect a decoupling of this developmental synchrony. It has been demonstrated that form deprivation in various species produces progressive axial growth and myopia of the postnatal eye. However, in both our sibs the ERG responses were much better preserved in the highly myopic right eye where, in addition, there was a low b/a ratio. Similar findings do not occur in myopia without retinal pathology.2

The bilateral symmetrical retinal dystrophy noted in both our patients may have a genetic basis for the disease but a single genetic defect cannot explain the additional asymmetry of eye size and electrophysiological measurements. Most probably, the phenotype results from at least two separate events, a germline mutation in a retinal or retinal pigment epithelial specific gene leading to degeneration, thereby creating a susceptible background on which a second event could occur. This event, leading to the asymmetry of ocular volume, may be an environmental factor or a mutation in a second gene important in the regulation of eye growth. If this were a somatic rather than germline mutation it would explain the asymmetry of the disease.
Adenoma of ciliary pigment epithelium: a case series

Adenoma of ciliary pigment epithelium is a rare tumour. Many are diagnosed retrospectively either after excision or enucleation, as malignant melanoma is suspected. We report a series of four patients found to have adenoma of ciliary pigment epithelium and discuss the clinical features and unusual behaviour of these neoplasms.

Case reports

We reviewed the histopathological reports in the ophthalmic pathology archive dateing from 1980 to date and identified four patients who had the histopathological diagnosis of adenoma of ciliary pigment epithelium. We crosschecked the details with the clinical oncology database. We reviewed their notes for features that would help us to identify this ciliary body tumour clinically. The salient features of these patients are given in Table 1.

Patient 1 was reported elsewhere in 1994. He had a dark brown multinodular mass in the inferotemporal anterior chamber angle of the left eye. His tumour was a relatively small but invasive lesion. Patient 2 was the only non-white patient with this condition in our series. Her tumour was an incidental finding when she presented to an ophthalmologist with extensive pigment dispersion made the media hazy (Fig 1D). The tumour was a solid dark brown lesion arising from the ciliary body between the 12 and 1 o’clock meridians. The clinical differential diagnosis was between malignant melanoma and adenocarcinoma of the pigment epithelium of ciliary body.

Patient 4 was initially treated for acute angle closure glaucoma in another hospital. Trabeculectomy was performed to achieve control of intraocular pressure. Postoperatively, he was found to have a lesion behind the crystalline lens. He underwent phacoemulsification with intracocular lens implantation to improve visualisation of the lesion. A black ciliary body mass was seen (Fig 1E). This prompted his referral to the oncology service in June 1999. Control of the intraocular pressure proved refractory even with additional medical treatment.

<table>
<thead>
<tr>
<th>No</th>
<th>Age/sex/race</th>
<th>VA</th>
<th>Size (mm)</th>
<th>Clinical features</th>
<th>Surgery</th>
<th>Recurrence</th>
<th>Year of diagnosis</th>
<th>Complication</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>40/M/W</td>
<td>6/5</td>
<td>8x2x1</td>
<td>Angle invasion</td>
<td>Local resection</td>
<td>No</td>
<td>1992</td>
<td>RD repair</td>
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<td>2</td>
<td>62/F/A</td>
<td>6/6</td>
<td>5x3x1</td>
<td>Angle invasion, cataract</td>
<td>Local resection</td>
<td>Yes</td>
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<tr>
<td>3</td>
<td>65/F/Arabic</td>
<td>7/18</td>
<td>5x4x4</td>
<td>Sentinel vessel, pigment dispersion</td>
<td>Local resection</td>
<td>No</td>
<td>1998</td>
<td>RD repair</td>
</tr>
<tr>
<td>4</td>
<td>54/M/W</td>
<td>1/60</td>
<td>5x4x4</td>
<td>Secondary glaucoma, pigment dispersion</td>
<td>Enucleation</td>
<td>No</td>
<td>1999</td>
<td></td>
</tr>
</tbody>
</table>

M = male, F = female, W = white, A = Asian, RD = retinal detachment, VA = visual acuity.
Pigment dispersion was seen in patients 3 and 4. This was mainly in the vitreous of patient 3 but in both the vitreous and the anterior segment of patient 4. There was heterochromia of the iris in patient 4. No angle invasion was seen in these two patients.

Other associated features were a localised cataract in patient 2, trahcetal retinal detachment and secondary glaucoma in patient 4. Patient 3 had an episcleral sentinel vessel over the tumour. None of these patients had any history of ocular trauma or intraocular inflammation. Ultrason biomicroscopy (Fig 1C) helped us to evaluate these tumours in more detail.

The first three patients underwent local resection of the tumour in the form of iridocyclectomy under hypotensive anaesthesia. The last patient had enucleation as he opted to have the eye removed because of the poor visual prognosis for that eye as a result of secondary glaucoma, extensive pigment dispersion, and trachetal retinal detachment.

Histopathologically, these tumours showed hormonal activity. Mitotic activity was absent or low. Invasion of ciliary muscle and the iris root was seen in patients 1 and 2. Patient 3 had a cystic adenoma with cells forming gland-like structures around central cysts.

Comment

Our series highlights the paradoxical behaviour of adenoma of ciliary pigment epithelium. Smaller lesions invaded the angle and larger lesions caused extensive pigment dispersion although non-invasively. Angle invasion resulted in these tumours being seen and resulted in the presentation of patients 1 and 2. Blurred vision due to pigment dispersion in the vitreous resulted in the presentation of patient 3. Angle closure glaucoma and pigment dispersion were the main features of patient 4. Shields et al. presented a series of eight patients with adenoma of ciliary pigment epithelium and described their clinical features. In their series they found an association with cataract, vitreous haemorrhage, and neovascular glaucoma.

Pigment dispersion was seen in two of our patients. The presence of pigment clumps and extensive pigment dispersion in the vitreous of patient 1 (Fig 1D) is unique and has not been reported before. Chang et al. in 1979 reported the presence of pigment in the retrotemporal space adjacent to the tumour in a case of adenoma of ciliary pigment epithelium. Extensive pigment dispersion in vitreous had been reported in malignant melanoma of the choroid but not to our knowledge in adenoma of ciliary pigment epithelium.

Secondary glaucoma from intraocular tumours is well known. In their survey of intraocular tumours causing secondary glaucoma Shields et al. reported on 2704 eyes. Of the five adenomas of the ciliary body one was from ciliary pigment epithelium. None of these had secondary glaucoma. Of the ciliary body melanomas, 17% had secondary glaucoma. Angle closure was responsible for secondary glaucoma in 12% of the eyes with ciliary body melanoma. In their series in 1997, Shields et al. had one patient who had neovascular glaucoma secondary to adenoma of ciliary pigment epithelium. Patient 4 in our series presented with secondary angle closure glaucoma.

Malignant melanoma of ciliary body is known to invade the anterior chamber angle. Chang et al. reported angle invasion in adenoma of the pigment epithelium of ciliary body. Shields et al. reported a patient in whom invasion of the iris stroma by an adenoma of ciliary pigment epithelium was documented with progressive growth. They initially suspected this to be a tumour of the iris but on later evaluation showed the origin from the ciliary body. The presenting feature in patients 1 and 2 in our series was similar, although the ciliary body origin was recognised initially. In patient 2 the diagnosis of adenoma of ciliary body was strongly suspected preoperatively, as the tumour that had invaded the angle was very small. Invasion of the angle by ciliary body melanomas usually does not occur until they have attained a larger size. Invasion of the angle has also been described in melanocytoma of the ciliary body. They too tend to be relatively larger when they invade the angle, unlike the adenomas that we described. Iris melanocytomas undergo central necrosis and cause pigment dispersion and glaucoma.

However, the necrotic centre is absent in adenomas.

One of our patients (patient 1) had a sentinel vessel. Sentinel vessels are typically larger lesions. Mitotic activity was absent or low. Invasion of ciliary muscle and the iris root was seen in patients 1 and 2. Patient 3 had a cystic adenoma with cells forming gland-like structures around central cysts.

References

9. Pheneylephrine 2.5% and 10% in phacoemulsification under local anaesthesia: is there an effect on systemic blood pressure?

Pheneylephrine 10% leads to a faster and more pronounced mydriasis but cardiovascular effects like hypertension and arrhythmias have been reported. In a young healthy adult the upper limit of safety for intravenous administration of pheneylephrine is 1.5 mg and Kumar et al. had found pheneylephrine plasma levels after administration of topical 10% viscos solution to their patients to be 1.842–11.526 ng/ml after 20 minutes while topical 2.5% aqueous solution produced plasma levels of 0.1–2.720 ng/ml after 20 minutes. They concluded that the mean pressure tended to be higher with the 10% viscos solution.

Chin et al. in their study on 89 patients concluded that significant hypertensive effects can arise after topical pheneylephrine; however, no significant difference between the two groups (2.5% v 10%) was shown. Symons et al. reported no significant change in the mean systolic and diastolic blood pressure in 126 patients receiving 10% pheneylephrine. Malhotra et al. in their study on 54 cases showed no difference in systemic cardiovascular effects of either the 2.5% or the 10% concentration.

Methods

We carried out a prospective randomised and double masked study on 53 patients undergoing phacoemulsification under topical anaesthesia (Amethocaine 0.1% and Marcaine 0.75%) and no exclusion of cases with hypertension, heart failure, or diabetes was made. All patients received one drop of each of tropicamide 1%, cyclopentolate 1%, and Voltarol 0.1% Pheneylephrine drops. 2.5% and 10%, were administered in three doses starting half hour before surgery at 5 minute intervals. All patients had their blood pressure measured before dilatation and in the anaesthetic room preoperatively then postoperatively.

Results

The study included 49 cases aged 52–91, 24 cases were known to have cardiovascular problems and seven were also diabetic. Comparison of the blood pressure changes before

Figure 1 Blood pressure (mm Hg) with pheneylephrine 2.5%. Numbers 1 and 2 = systolic and diastolic reading preanaesthetic; 3 and 4 = systolic and diastolic reading in the anaesthetic room; 5 and 6 = systolic and diastolic reading in recovery.
dilatation and in the anaesthetic room preoperatively in the 2.5% group (Fig 1 showed a mean rise in systolic BP of 22 mm Hg (p=0.003) in the normotensive group. In the hypertensive group all patients recorded statistically significant rises in systolic and diastolic BP (respectively p=0.010, p=0.009). In the 10% group (Fig 2, 10 patients in the normotensive group showed a mean rise in systolic BP of 34.4 mm Hg (p=0.001) and 10.5 mm Hg in diastolic BP (p=0.077). In the hypertensive group six patients recorded a rise in systolic BP with a mean of 22.8 mm Hg (p=0.015) and a diastolic rise with a mean of 16.8 mm Hg (p=0.033) (Table 1). (Figs 3 and 4).

Comment

Our study showed that patients in both groups of phenylephrine doses experienced a rise in systolic blood pressure which was statistically significant; the mean of which is higher in the 10% group. The maximal systolic blood pressure rise is around 10–20 minutes after administration, which corresponds to the time of maximal plasma levels of phenylephrine as demonstrated by other authors, but the paired t test failed to show any significance between the results of the groups of either concentration. Postoperative results showed some elevations in systolic and diastolic BP but weren't statistically significant. Using Altman's normogram the study was limited in its power. Although the aim of this study was not designed to investigate the effect of anxiety on systemic blood pressure, this element should be taken into consideration since it is known to elevate the systolic blood pressure. We have also used topical anaesthesia so that adrenaline 1:200 000, which is always included in local anaesthesia, is excluded which might cause elevation of the blood pressure although the concentration is too low for any significant systemic effect.

References


Table 1 t Test 2.5% and 10% (normotensives and hypertensives)

<table>
<thead>
<tr>
<th></th>
<th>Mean</th>
<th>SD</th>
<th>Significance*</th>
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</thead>
<tbody>
<tr>
<td>Normotensive/sys</td>
<td>−12.44</td>
<td>19.08</td>
<td>0.86</td>
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<tr>
<td>Normotensive/diast</td>
<td>2.88</td>
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<td>0.61</td>
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<tr>
<td>Hypertensive/sys</td>
<td>14.16</td>
<td>28.36</td>
<td>0.27</td>
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<tr>
<td>Hypertensive/diast</td>
<td>−4.16</td>
<td>14.00</td>
<td>0.49</td>
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</table>

Two tailed t test, comparing the 2.5% group and 10% group both hypertensive and normotensive patients.

*p=0.05.

Effect of preoperative detection of photoreceptor displacement on postoperative foveal findings in eyes with idiopathic macular hole

In idiopathic macular hole formation, Gass' hypothesised that macular holes enlarge with displacement of the photoreceptors without tissue loss around the fovea. Jensen and Larsen's developed binocular kinetic perimetry that could evaluate local retinal photoreceptor displacement and also confirm centrifugal photoreceptor displacement away from the fovea in eyes with a macular hole. Using the binocular perimetry technique and scanning laser ophthalmoscope microperimetry, we confirmed that the patients with a unilateral macular hole with preoperative photoreceptor displacement had better postoperative visual function when compared to patients without preoperative photoreceptor displacement. We also previously reported that the postoperative foveal findings were correlated with the postoperative visual function in eyes with a macular hole.

The purpose of the present study was to investigate whether or not detecting photoreceptor displacement preoperatively affects the postoperative foveal findings in eyes with an idiopathic macular hole.

Methods and results

Fifteen patients (12 women, three men; mean age 62 (SD 5) years) with a unilateral idiopathic macular hole that resolved after vitreous surgery were examined. All had undergone surgery at our institution. Eleven eyes were classified as having a stage 3 hole, four stage 2, and one stage 4. The best corrected visual acuity (VA) levels in the unaffected eyes were 0.7 or better, and no patients had ocular diseases except for mild cataract.

To detect photoreceptor displacement in eyes with the disease preoperatively, we performed binocular kinetic perimetry. The anatomical status of the repaired macular holes was assessed 3 months after the operation using optical coherence tomographic equipment (OCT 2000, Humphry Instrument, division of Carl Zeiss, San Leandro, CA, USA). Good postoperative foveal findings were defined as the presence of a foveal depression, as previously reported by Imai and associates (Figs 1 and 2). Photoreceptor displacement was detected in 11 of the 15 (73%) patients preoperatively. Good postoperative foveal findings were observed in nine of the 11 (82%). Photoreceptor displacement was detected preoperatively in all nine patients. The prevalence of good postoperative foveal findings was significantly higher in patients in whom photoreceptor displacement was observed.

Figure 1 Good postoperative foveal findings observed with optical coherence tomography. A foveal depression is observed.
PostScript 507

The authors have no proprietary interest in any instruments used in this study.

Correspondence to: Taichi Hikichi, MD, Department of Ophthalmology, Asahikawa Medical College, 2-1 Midorigaoka-higashi, Asahikawa 078-8510, Japan; hikichi@asahikawa-med.ac.jp

References

Nodular scleritis in a patient with sarcoidosis

Sarcoidosis is an immune mediated disease that may affect any organ. Scleral involvement has rarely been described with sarcoidosis.1,2 We report on a patient with unilateral nodular anterior scleritis who was found to suffer from sarcoidosis. Here, the clinical and histopathological features are described.

Case report

A 33 year old white woman had persistent moderate tenderness in her left eye that lasted for several weeks. Visual acuity was 20/20 in both eyes. The slit lamp appearance of the right eye was unremarkable. A scleral nodule in the inferior nasal quadrant was found in the left eye (Fig 1), and a moderate tenderness was determined in this area. The intraocular pressures were within normal ranges. On ophthalmoscopy, the posterior segment of both eyes was normal. The ultrasound images did not indicate posterior scleritis. The medical history was remarkable for an episode of hepatitis A 20 years ago, and the patient suffered from exercise dependent dyspnoea, night sweats, and repeated headaches.

An extensive examination for associated systemic diseases was performed, including serological tests for syphilis, CRP, ESR, ANCA, RF, CIC, ANA, and PPD tests: all were within normal ranges and consultant examinations by the internal medicine, rheumatology, neurology, and ENT departments were performed. The chest x ray examination revealed a bilateral hilar lymphadenopathy and basal interstitial pulmonary infiltration typical of sarcoidosis stage 2. In the body plethysmography, a mild restriction and a moderately reduced diffusion capacity was determined. The bronchoalveolar lavage revealed an increased proportion of CD3+ T cells and an increased CD4/CD8 ratio, consistent with active pulmonary sarcoidosis. The level of angiotensin converting enzyme was 51.3 U/l (normal, 8–52) in the high normal range. The magnetic resonance images revealed an increased enhancement and thickening of the inferior nasal part of the sclera of the left eye. A biopsy was taken from the scleral nodule under peribulbar anaesthesia. Histopathological studies on the paraffin embedded tissue that was performed by the pathologists revealed a perivascular lymphoid cell infiltration, with non-casing granuloma-like cell accumulation (Fig 2). Some histiocytes and very few giant cells were found. The scleral tissue was oedematous and the vessels were dilated, but necrosis, vasculitis or malignant cell configurations were absent. Additionally, Ziehl–Neelsen and Grocott’s silver methamine stains excluded the possibility of causative infections.

Treatment with oral prednisone with an initial dosage of 1 mg/kg was performed. The corticosteroid dosage was tapered within 6 weeks, and was maintained at 20 mg. Under this regimen, the pain and the scleral nodule resolved immediately, and the ACE level fell to 24 U/l (normal).

Comment

Scleral involvement has been previously reported in a few patients with sarcoidosis. In a series of 537 patients with histologically proved sarcoidosis,3 scleral plaques anterior to the equator have been found in four cases, and this was seen in the acute stage of disease and in association with erythema nodosum and bilateral lymphadenopathy. In another series by Jabs and Johns,4 within a group of 183 patients with chronic sarcoidosis, scleral involvement was detected in one patient. However, the clinical course and histopathological appearances have not been described previously. Posterior scleritis has been seen only rarely with sarcoidosis.5

In our patient, the sparse systemic symptoms made the diagnosis difficult and, therefore, scleral biopsy was done in order to rule out infections or malignancies and to disclose the histopathological evidence for sarcoidosis. Although a classic non-caseating granuloma...
18 year old male patient presented to our Department of Ophthalmology, St Franziskus Hospital, Muenster, Germany

Correspondence to: Arnd Heiligenhaus, MD, Department of Ophthalmology, St Franziskus Hospital, Hohenzollerring 74, 48145 Muenster, Germany; arnd.heiligenhaus@t-online.de

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References

Panophthalmitis following contact diode laser cyclophotocoagulation in a patient with failed trabeculectomy and trabeculotomy for congenital glaucoma

Diode laser trans-scleral cyclophotocoagulation (TSCPC) is a well accepted method for the management of patients with refractory glaucoma. A lasting decrease in IOP has been reported in up to 90% of eyes with severe, medically uncontrolled glaucoma following contact TSCPC using G-probe delivery, with preservation of visual acuity.1 Although the procedure is considered safe,2 complications have been reported.3 Here, we report the first case, to the best of our knowledge, of panophthalmitis following contact diode laser cyclophotocoagulation using the G-probe in a patient with failed trabeculectomy and trabeculotomy for congenital glaucoma.

Case report
An 18 year old male patient presented to our emergency service with a 4 day history of pain, redness, and periorbital swelling in the right eye, accompanied by poor vision. One week earlier he had undergone 360 degree diode laser (trans-scleral) cyclophotocoagulation using contact G-probe in the right eye for uncontrolled glaucoma. He had thrice undergone filtering surgeries in both eyes for congenital glaucoma since 1985. During the last trabeculectomy performed in 1996, intraoperative mitomycin C (0.2 mg/100 ml applied for 3 minutes beneath the scleral flap) had also been used. But it failed in the right eye and the patient had developed a painful blind eye.

His visual acuity was no light perception in the right eye and 20/60 in the left. There was right sided periorbital oedema, restricted motility, and purulent discharge with matting of the eyelashes. Rupture of the globe with prolapse of the iris was evident along the superior limbus of the eye, and the bleb was well formed, and the IOP (Goldmann applanation) was 12 mm Hg. The optic disc showed a cup disc ratio of 0.4 with temporal pallor. A diagnosis of panophthalmitis of the right eye was made. As no response to intravenous antibiotic treatment was observed over the next 48 hours, evisceration was undertaken with the patient’s informed consent. Staphylococcus epidermidis was grown on culture.

Comment
Its efficacy notwithstanding, diode laser cyclophotocoagulation has been associated with complications, although less frequent than with other cyclodestructive procedures. Conjunctival burns, corneal decompensation, neurotrophic corneal defects, uveitis, OIP spikes, phthisis (0.5%), macular pucker (1%), intraocular haemorrhage (0.5%), visual loss, and malignant glaucoma have all been reported.4–6 Possible correlation of outcome with perilimbal pigmentation and the hearing of audible “pop,” especially in patients with dark brown irides,7 has been suggested.

Scleral thinning is a recognised risk factor for complications, but scleral perforation following this procedure has been reported earlier in one patient only.8 This patient had scleral thinning following previous cataract surgery. Possibly, the sharp probe edge had cut conjunctival vessels causing bleeding and contamination of the probe tip. Thin adherent debris was then carbonised allowing the laser tip to rise to 300°C, sufficient to cause scleral perforation. This report led to redesigning of the probe tip to protect the vascular structures from its sharp edges. Our patient was treated as part of a standard protocol using the modified G probe, and bleeding did not occur. The perforation could have been halted from 90% mechanical pressure, or the effect of the laser on the thinned out sclera, which the patient probably had as a result of buphthalmos and previous ocular surgery.9 There are no known “corrective factors” to utilise in such cases, but the probe tip should be avoided. We emphasise caution in undertaking contact diode laser trans-scleral cyclophotocoagulation in operated eyes with thin sclera.


Panophthalmitis following contact diode laser cyclophotocoagulation in a patient with failed trabeculectomy and trabeculotomy for congenital glaucoma

Diode laser trans-scleral cyclophotocoagulation (TSCPC) is a well accepted method for the management of patients with refractory glaucoma. A lasting decrease in IOP has been reported in up to 90% of eyes with severe, medically uncontrolled glaucoma following contact TSCPC using G-probe delivery, with preservation of visual acuity.1 Although the procedure is considered safe,2 complications have been reported.3 Here, we report the first case, to the best of our knowledge, of panophthalmitis following contact diode laser cyclophotocoagulation using the G-probe in a patient with failed trabeculectomy and trabeculotomy for congenital glaucoma.

Case report
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syndrome, which is part of the spectrum of Chandler's syndrome. Most often women are affected unilaterally by ICE syndrome, nevertheless an exception of bilateral cases. Our patient is a man and is affected bilaterally.

Reference


Fine retinal crystalline deposits observed by confocal scanning laser ophthalmoscopic examination using infrared light

Bietti's crystalline retinopathy is a tapetoretinal degeneration characterised by numerous tiny sparkling yellow-white spots mainly located in the deeper layer of the posterior pole retina. Since crystalline deposits tend to become small and decrease in number over time,1,2 the fundus appearance becomes indistinguishable from other retinal dystrophies with time.3 The advent of confocal scanning laser ophthalmoscope (SLO) with infrared light has enabled high sensitive examination of the deep layer of the retina.4 We studied the deep retinal abnormalities in a case with a typical Bietti's crystalline retinopathy and in another case with myopic choriotelial atrophy throughout the posterior pole. Consequently, we detected numerous fine crystalline deposits in both cases, which were not detected with other funduscope examinations.

Case reports

Case 1

A 52 year old man was referred to our hospital by his ophthalmologist who found an abnormal fundus appearance. There was no consanguinity. Best corrected visual acuity was 20/24 right eye and 20/30 left eye. Funduscopic abnormalities were found in the anterior segment and media. Fine crystalline deposits were not detected in the corneal stroma of either eye. Funduscope examination revealed numerous fine reflective crystalline deposits throughout the posterior pole and mid-peripheral retina of both eyes. Most of these deposits were in the deep retina and subretina but some deposits were superficial. Goldmann perimetry showed relative scotomas in a zone within 30–40 degrees of the central field in both eyes. Fluorescein angiography of both eyes showed island-like hypofluorescent lesions surrounded by hyperfluorescent lesions in the early phase. Indocyanine green angiography also showed the atrophy of the choripapillaris in the posterior pole. Crystalline deposits did not show any fluorescence in either fluorescein or indocyanine green angiography.

The retinal crystalline deposits were more clearly visible with the fundus examination by SLO (Rodenstock, Germany) using infrared light compared to the routine funduscope examinations (Fig 1). Each crystal deposit appeared larger in size. In addition, numerous fine crystal deposits were evident, which were not observed by conventional funduscope examinations including ophthalmoscopy and
Figure 1 Ocular fundus of case 1. Fundus photograph (A), grey scale image of the same fundus photograph (B) and SLO image with infrared light (C) are shown. Note that the crystalline deposits are most evident with the infrared imaging.

Figure 2 Ocular fundus of case 2. Fundus photograph (A), grey scale image of the same fundus photograph (B) and SLO image with infrared light (C). Magnified image of the boxed area is shown in (D). Note that numerous crystalline deposits are observed with the SLO imaging.

and no fine crystal deposits were found at limbs of either cornea. Both eyes were aphakic and the posterior lens capsules were intact but relatively opaque. Funduscopic examination revealed the posterior staphyloma and the extensive atrophy of the retinal pigment epithelium and choriocapillaris extending from the posterior to mid-peripheral retina in both eyes. Optic disc showed myopic appearance. Retinal vessels and peripheral retina showed normal appearance. Standard electrrotetinogram was non-recordable. Goldmann perimetry showed marked constriction of all isopters and loss of visibility of 1–4° isopter for both eyes. During a 5 year follow up, visual acuity and visual field examination were stationary. Fluorescein angiography performed at the age of 57 demonstrated hypofluorescence throughout the posterior pole in the early phase and diffuse hyperfluorescence in the later phase. Indocyanine green angiography disclosed the loss of background veil-like fluorescence throughout the posterior pole due to the choriocapillaris atrophy. Fundus examination by the confocal SLO with infrared light disclosed numerous tiny crystalline deposits in the posterior pole (Fig 2), which were not evident with other fundusscopic examinations, including ophtalmoscopy and biomicroscopy with a contact lens, or with fluorescein or indocyanine green angiographic studies. The deposits scattered light when the source of light was moved to cause them sparkle, similar to the fine crystalline deposits observed in the fundus of case 1.

Amino acid analysis and other examinations did not show any abnormal value.

Comment
The diagnosis of Bietti's crystalline retinopathy in case 1 is based on the retinal crystalline and supported by the characteristic island-like hypofluorescence observed by fluorescein angiography.1 Fundus examination by SLO with the infrared light disclosed numerous tiny crystalline deposits, which were not observed by conventional fundus examinations. Thus, in Bietti's crystalline retinopathy, there are more numerous accumulation of retinal crystalline deposits than can be observed by fundusscopic examinations.

In case 2, numerous tiny retinal crystalline deposits were detected by SLO with infrared light, although no other examinations disclosed these crystalline deposits. We have not observed such retinal crystalline deposits in other cases with myopic chorioretinal atrophy, suggesting the accumulation of crystalline deposits is not a generalised feature of myopic chorioretinal degeneration. Crystalline deposits, prerequisite for the diagnosis of Bietti's crystalline retinopathy, are difficult to detect in advanced cases.2,3 Since crystalline deposits do not block or transmit fluorescence in fluorescein4 or indocyanine green angiography, diagnosis of Bietti's crystalline retinopathy in advanced cases without funduscopically apparent crystalline deposits have been established only when family history was contributory. Because the presence of crystalline material in the peripheral lens capsule does not suggest an autosomal recessive inheritance, similar with some reported cases with Bietti's crystalline retinopathy, and the striking resemblance of the morphology of the tiny crystalline deposits observed by SLO using infrared light between the two cases may support that underlying pathogenesis is similar.

Y Yanagi, Y Tamaki, H Fukushima
Department of Ophthalmology, University of Tokyo School of Medicine, 7-3-1 Hongo, Bunkyo-ku, Tokyo 113-8655, Japan
Correspondence to: Yasuo Yanagi, Department of Ophthalmology, University of Tokyo School of Medicine, 7-3-1 Hongo, Bunkyo-ku, Tokyo 113-8655, Japan, yanagi-ty@umin.ac.jp

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References
1 Bietti GB. Su alcune forme atipiche di degenerazione retinica (degenerazione tappetoretiniche e quadri morbosi simili). Boll Oculist 1937;16:1159–244.

Tonic pupils from giant cell arteritis
The tonic pupil is characterised by poor reactivity to light, a slow tonic constriction and redilation to a near target, and supersensitivity to topical dilute pilocarpine.1 Most instances do not have an identifiable cause; however, a variety of conditions including herpes zoster, orbital trauma including surgery, and paraneoplastic syndromes may result in tonic pupils. The mechanism(s) which produce tonic pupils are not known, although axonal loss within the ciliary ganglion is generally believed to result in supersensitivity of the iris sphincter muscle. Giant cell arteritis (GCA) has been reported as an uncommon cause of tonic pupils.2 Furthermore, the cause of tonic pupils in GCA is unknown and direct evidence for an ischaemic cause is not convincing.

We used orbital colour Doppler imaging to study orbital and ocular blood flow in a patient with GCA, unilateral visual loss, and bilateral tonic pupils.

Case report
A 38 year old woman presented with visual loss of the left eye for 2 days. She had had...
near target. Orbital colour Doppler imaging
pupils constricted slowly and tonically to a
pupil no longer reacted to direct light. Both
pupils constricted following the instillation of
pupil (Fig 2B) with sectoral iridoplegia. Both
ination on the right revealed an irregular
followed by slow redilation. Slit lamp exam-
bilateral tonic constriction to a near target
remained amaurotic. In addition, there was
agishly to light (Fig 2A) while the left
However, the right pupil now reacted slug-
sbarof prednisone and with a +2.00 dioptre add over a
nisolone she was discharged on 80 mg of oral
n and tonic pupils from GCA.7 Assessment of
ial blood flow was performed using ultra-
sography in three of these patients. In two
patients no flow was noted in the supratroch-
ar arteries, while blood flow in the third was
ormal. The author noted that the ultrasono-
ographic results supported ischaemia as a
cause for the tonic pupils. In the patient with
ormal orbital blood flow, ischaemia below
the resolution of ultrasonography was pre-
sumed to cause the tonic pupils.

The blood supply to the ciliary ganglion
arises from several sources. Eikickova studied
human cadaveric orbits after injection of
dye into the internal carotid artery.1 The
ciliary ganglion was supplied with blood from
one to four arteries, with the posterior lateral
ciliary artery and the lateral muscular artery
the most common sources, followed by the
ophthalmic and central retinal artery. The
vasculature of the ganglion itself is composed of
a network of capillaries.

Orbital colour Doppler imaging is a non-
vasive way to quantitate blood flow to the
orbit and eye. Decreased blood flow in the
ophthalmic artery, central retinal artery, and
short posterior ciliary arteries may be de-
tected on orbital colour Doppler imaging in
patients with GCA.6 Markedly reduced blood
flow was found with orbital colour Doppler
imaging in both orbits in our patient, despite
her visual loss being unilateral. We were
unable to find a previous report of bilateral
tonic pupils in a patient with unilateral visual
loss from GCA. Although anterior segment
ischaemia can cause loss of iris sphincter tone,
we do not believe that the pupillary findings
in our patient resulted from iris ischaemia.
Iris ischaemia would result in poorly reactive
pupils to both light and accommodation,
without a tonic near reaction or denervation
ehersensitivity. Therefore, the findings in our
patient strongly suggest that the tonic pupils
resulted from ischaemic damage to the ciliary
ganglion or the postganglionic ciliary nerves
which are responsible for pupillary constric-

The authors do not have any proprietary interests in
any of the contents of this manuscript.

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Foundation, Cleveland, OH, USA.
Frozen cucumber as a mount for vitreoretinal specimens

Vitreoretinal specimens are extremely difficult to process as a frozen specimen because of their small size and tendency to roll up.1 However, in order to perform immunohistochemical tests it is sometimes necessary to have frozen specimens as antigens may be destroyed if a fixative agent is used.2 Dua et al3 and Scott et al4 suggested the use of frozen cucumber as a mount for conjunctival and corneal tissue; we modified this method for vitreoretinal specimens. We describe our technique and provide examples of our results.

Case report

Fresh cucumber (obtained from a greengrocer) was cut into small (1 cm3) blocks; the part devoid of seeds was used. We found that with cucumbers older than 2 days the membranes did not adhere sufficiently well. These blocks were then stored at 4°C until required. Pig eyes were obtained and stored at 4°C until required. Subsequently, the eye was placed under the dissecting microscope and basic salt solution injected in through the vitreous cavity to enable easier dissection. The cornea was removed and a vitrectomy performed. The retinal specimens were stained with Indian ink in order to facilitate subsequent localisation.

Following vitrectomy, membranes were removed from humans (these included diabetic membranes, subretinal neovascular membranes, and epiretinal membranes); they were initially placed in Hartmann’s solution. Subsequently, they were placed on the cucumber under a dissecting microscope; it was possible to place the membrane flat without it rolling up because of the texture of the cucumber. These membranes were also stained with Indian ink before placing on the cucumber. The membrane with the membrane on its “side” surface was placed in an aluminium foil cup and covered with a cryomatrix of Tissue-Tek OCT compound (Fig 1). The foil cup was then put in a plastic container and the contents flash frozen in liquid nitrogen. The membranes were cut with a cryostat in 4-5 μm sections. We stained one slide from each specimen with haematoxylin and eosin and performed immunohistochemistry on the others (Figs 2 and 3).

No specimens were lost while performing this technique of processing specimens. We were able to maintain the orientation of the specimens and managed to obtain sufficiently satisfactory specimens to perform our immunohistochemical studies. The use of Indian ink allowed us to locate the specimen easily when cutting sections. Unlike previous immunohistochemical techniques, most modern immunohistochemistry may be performed on fixed tissue.

Inflammatory optic neuropathy as the presenting feature of herpes simplex acute retinal necrosis

Acute retinal necrosis (ARN) is a rare but serious ophthalmic manifestation of infection by the herpesvirus family. In the immunocompetent, the viral agent most frequently identified is varicella zoster (VZV) followed by herpes simplex (HSV-1 and HSV-2) and rarely

**References**


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**Figure 1** The biopsy cucumber unit.

**Figure 2** Diabetic membrane on cucumber mount stained with haematoxylin and eosin.

**Figure 3** Diabetic membrane on cucumber mount stained with CD31.
cytomegalovirus and Epstein-Barr virus. The condition, which may present with synchronous or metachronous systemic or cerebral herpetic infection (encephalitis) is now recognised to occur at all ages, though VZV tends to affect the more elderly and herpes simplex the young.

The majority of patients present with a short history of blurred vision, floaters, and mild ocular discomfort. In the immunocompetent, clinical examination reveals one or more well demarcated foci of retinal necrosis outside the major arterial arcades typically with a dense vitritis that may preclude a detailed view of the posterior segment. Disease progression is rapid with coalescence and posterior extension of areas of retinitis. Often there is an exclusively, sometimes haemorrhagic, vasculitis either adjacent to areas of retinitis or at the optic nerve. The visual prognosis for untreated cases is very poor. We report three immunocompetent patients with no evidence of concurrent herpesvirus infection in whom ipsilateral optic neuritis associated with a panuveitis was the presenting feature of their acute retinal necrosis syndrome.

Case reports

Case 1
A 20 year old healthy white male, with no history of herpesvirus infection, presented with a 10 day history of progressive blurred right vision with floaters, significant pericentral disc discomfort, and pain on ocular motility. Acuity was 20/200 right eye, 20/20 left eye, with a right relative afferent pupillary defect (RAPD). There was swelling of the right optic nerve (Fig 1A) together with increasing non-granulomatous pan uveitis. The left eye was unaffected. Fluorescein angiography (Fig 1B) showed marked optic disc hyperfluorescence and peripheral retinal ischaemia. Seven days later (Fig 1C and D), several enlarging foci of retinitis, typical of ARN, developed in the retinal mid-periphery. PCr of a vitreous biopsy amplified HSV-1. There was no further reduction in visual acuity following intravitreal foscarnet and encephalitic doses (30 mg/kg three times daily) of intravenous aciclovir for 10 days.

Case 2
A 47 year old healthy white female, with no history of prior herpesvirus infection, presented with a 3 day history of progressive blurred left vision. She also complained of significant pericentral discomfort exacerbated by eye movement. Acuity was 20/40 left eye, 20/15 right eye, with a left RAPD. There was swelling of the left optic nerve head together with a moderate non-granulomatous panuveitis. The right eye was unaffected. Fluorescein angiography showed marked optic disc hyperfluorescence and peripheral retinal ischaemia. Six days later, several enlarging foci of retinitis, typical of ARN, developed in the inferior retinal periphery. Polymerase chain reaction (PCR) of a vitreous biopsy amplified HSV-1. There was no further reduction in visual acuity following encephalitic doses (10 mg/kg three times daily) of intravenous aciclovir for 10 days.

Case 3
An 81 year old white male, who had suffered herpes simplex meningoencephalitis and synchronous right acute retinal necrosis (acuity reduced to 20/200) 17 years earlier, presented with a 2 day history of profound reduction in left acuity to perception of light only. Clinical findings included a left RAPD, haemorrhagic optic disc swelling, and increasing hypertensive panuveitis. Five days later, three demarcated areas of retinal necrosis appeared superiorly in the same eye. PCR of a vitreous biopsy amplified HSV-1. There was no further reduction in visual acuity following intravitreal foscarnet and 10 days of encephalitic doses of intravenous aciclovir.

Comment
There have been two previous reports in immunocompromised patients of optic neuritis preceding the development of ARN. In these, preceding or concurrent cutaneous herpes zoster infection suggested altered viral behaviour in the context of deficient immunity. Even in the healthy individual, there is evidence of an immunogenetic predisposition to the development of the disease. The novel mode of presentation of our three immunocompetent patients suggests they might share a similar background of immunity that modifies viral behaviour, thereby predisposing to involvement of the optic nerve before the development of retinitis. In this regard, it is interesting that none of our cases developed clinically evident encephalitis. This is a common accompaniment to human HSV-1 ARN and observed in the Szily animal model in which inoculation of the anterior chamber or vitreous with HSV virus produces in ipsilateral optic neuritis, relative sparing of the ipsilateral retina, and necrotising contralateral chorioretinitis with encephalitis. It has been shown that viral spread to the brain and the fellow eye results from viral invasion of the optic nerve. The ability of the host to resist this appears determined by the animal’s immune background.

The recent report in a single case of ARN subsequent to central retinal vascular obstruction in the fellow eye is most likely a manifestation of a different VZV mediated disease process but emphasises the ability of members of the herpesvirus family to directly invade blood vessel walls.

The combination of optic neuritis, peripheral retinal ischaemia, and panuveitis is very unusual. In white patients, the differential diagnosis would include conditions such as sarcoidosis and demyelinating disease for which steroid therapy is often routine and may prove beneficial. This report highlights that herpesvirus infections may also present in this fashion. Since progression to profound and irreversible visual loss is rapid, close daily retinal examination, and early diagnostic vitreous biopsy must be recommended for these patients before commencement of immunosuppressives.

P J Francis, H Jackson, M R Stanford, E M Graham
St Thomas’s Hospital, London, UK
Correspondence to: Peter Francis, St Thomas’s Hospital, London, UK, peterjamesfrancis@btopenworld.com

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References
Cutaneous angiosarcoma of the eyelids

Angiosarcoma of the eyelid is a rare potentially life threatening tumour arising from the vascular endothelium. Through the presentation of a case, the clinicopathological features and management of this condition are described.

Case report

A 69-year-old white man presented with multifocal red-blue maculopapular lesions located in his right cheek and eyelids (Fig.1A). The patient had first noticed the lesions 9 months previously. An incisonal biopsy from the upper eyelid was performed. In subsequent staging investigations no metastases were found. The patient was otherwise in good health and had HIV negative. He underwent surgical resection of all clinically detectable lesions followed by adjuvant radiotherapy (total dose 45 Gy). After 34 months of follow up there was no evidence of local recurrence or distant metastases.

Histopathologically, the specimens consisted of skin and subcutaneous tissue. The lesions proved to be well differentiated angiosarcoma characterised by interlacing blood vessels lined by plump endothelial cells with hyperchromatic nuclei. Using Ki-67, the proliferation rate was assessed as 5%–7% (Fig. 1B). Immunohistochemically, the tumour showed a positive reaction for vimentin, CD34 (Fig.1B) and CD31, and expression of immunohistochemical markers of vascular differentiation (CD34 and FVIII-RA). Often tumours display both vasosclerotic and undifferentiated areas. The present case was noteworthy, in that the entire tumour appeared to be well differentiated. The proliferation rate was low to moderate compared to the reported rate for angiosarcoma (>10% in 72% of cases assessed by Ki-67). Miotic index has been observed as an independent histological prognostic marker for cutaneous angiosarcoma although other histological markers remain poorly defined. In the present case low proliferation index and well differentiated histological features may have contributed to a better outcome.

The most important clinical prognostic factor is lesion size, with tumours <5 cm in diameter having a better prognosis. This underlies the importance of early diagnosis and appropriate management. The mainstay of treatment for cutaneous angiosarcoma includes surgery and/or radiotherapy although, due to the rarity of this condition, there are no standard guidelines. As illustrated by our case, the primary lesions were contained within the anterior lamella of the lids and it was possible to surgically encompass the primary lesions avoiding exenteration and without compromising prognosis. In such cases, the surgical aim is to resect all clinically identifiable disease. In areas of doubt, microscopic control of surgical margins may have a role in guiding the extent of resection. Angiosarcomas usually respond to radiotherapy to some degree and most authors suggest that combined surgery and radiotherapy offers the best chance for long term control. In one series of 28 head and neck angiosarcomas, Mark et all9 have reported better survival after 32 months (median) with combined surgery and radiotherapy compared with surgery alone. In cases unsuited to surgery, radiotherapy alone may be considered, however usually only partial responses are achieved. The role of chemotherapy remains undefined.

In summary, cutaneous angiosarcoma should be included in the differential diagnosis of vascular or atypical eyelid lesions as early recognition is critical to optimise outcome. Classification of the mitotic index and degree of histopathological differentiation may be useful criteria in predicting the biological behaviour. Treatment needs to be individualised to the patient and in selected cases complete excision of clinically evident disease may be appropriate. Adjunctive radiotherapy is recommended due to the aggressive natural history of angiosarcoma.

Acknowledgements

The excellent technical assistance of Carmen Hofmann-Rummelt is acknowledged. The study was supported in part by the Alexander von Humboldt Foundation, Bonn, Germany (RMC). The Deutsche Forschungsgemeinschaft, Bonn, Germany (TH), Allergan Australia in conjunction with The Royal Australian and New Zealand College of Ophthalmologists and Sydney Eye Hospital (RMC).

R M Conway, T Hammer, A Viestenz, L M Holbach
Department of Ophthalmology and Eye Hospital, University of Erlangen-Nuremberg, Erlangen, Germany

R M Conway
Alexander von Humboldt Fellow and Lecturer Save Sight Institute, University of Sydney, Australia
Correspondence to: R Max. Conway, MD, PhD, Department of Ophthalmology and Eye Hospital, University Erlangen-Nuremberg, Schwabachanlage 6, D91054 Erlangen, Germany, rmconway@hotmail.com

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References

Permanent visual loss in a child with a rash

Endophthalmitis is an unusual but well described complication of bacteraemia. Here we report endophthalmitis arising in the course of a mild illness resembling chickenpox.

Case report

A previously healthy 6 year old white girl awoke in the early hours feeling hot and complaining of headache. She was given ibuprofen and she went back to sleep. She appeared on her limbs later the same day but they blanched beneath a glass bottle and some of the older lesions appeared to be infiltrated and thinned.

The right eye was chemotic, with a pain in the right eye and could not open it. She vomited. Next morning she complained of crusting over. She was noticed rubbing her eye and some of the older lesions appeared to be apparent septicaemia and meningitis, occurred in the setting of a mild illness thought to be chickenpox. Although the usual hallmarks of meningococcal sepsis were absent, evidence for seeding of the eye in the course of a transient meningococcalemia was provided by Gram negative intracellular diplococci in the aqueous humour and detection of meningococcal DNA in both the aqueous humour and cutaneous material. Reports of meningococcal endophthalmitis in clinically well, non-bacteraemic patients are very rare, with just four published cases in English. The patients were 13 months, 15 years, 17 years, and 23 years old. Both eldest and youngest had no skin lesions, but C-reactive protein 22 mg/l. Material was collected from the base of the sole remaining cataract, forming in two, and pthisis in the 23 year old who underwent vitrectomy. The 13 month old underwent vitrectomy and lenectomy, and the plan at publication was to provide the patient with a new lens.

Vesicles are not generally regarded as lying within the spectrum of rashes caused by meningococcal infection. The acute ocular complications of varicella infection include spread of vesicles to the eyelid, papillary conjunctivitis and, uncommonly, dendritic keratitis, acute retinal necrosis, and optic neuritis. Hypopyon, observed in this case, is not a feature of primary VZV infection. This report serves to emphasise the broad clinical spectrum of meningococcal disease, and highlights the potential for severe sequelae despite an otherwise effective host response to bacteraemia. Seeding of group C meningococci resulted in visual loss in this young patient for whom the UK conjugate vaccine programme came just a year too late.

References


MAILBOX

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Providing it isn’t libellous or obscene, it will be posted within seven days. You can retrieve it by clicking on “read eletters” on our homepage.

The editors will decide as before whether or not to also publish it in a future paper issue.

Those excrescences on Descemet’s membrane

We read with interest the recent report, “Screening human donor corneas during organ culture for the presence of guttae.” Even the title made us question whether we were the only ones still using the original name for the condition cornea guttata. A quick perusal of Medline reveals a trend away from guttata, and towards guttata (Fig 1). This trend may be even more pronounced, as the literature tends to be conservative. The next generation of cornea specialists, or at least the small sample that goes through our training programme, tends to fall into three categories: those who say guttata, those who correct others for saying guttata, and those who try their best not to mention the term for the fear of saying the wrong thing. Many of our faculty have now entered this last group.

When asked what is so bad about guttata, some insist that it is not the plural for gutta, the Latin word for drop. We’re not sure that anyone ever said it was. More erudite dissenters recognise that not only is guttata not a plural, it is not even a noun—it’s an adjective. The closest English counterpart to guttata is probably guttate, drop-like, or speckled, so it’s incorrect to use guttata alone as a noun, or so the thinking goes. Some have chosen to use cornea guttata when referring to the condition, and guttata when referring to the excrescences themselves. The patient was apyrexial and remained clinically well during her hospital admission. At discharge after 5 further days of parenteral antibiotics except vancomycin administered. Vitrectomy could not be performed because of the acute ocular infection. Electron microscopy, immunofluorescence staining and polymerase chain reaction (PCR) testing of the vesicular material was negative. Material was collected from the base of the sole remaining cataract, forming in two, and pthisis in the 23 year old who underwent vitrectomy. The 13 month old underwent vitrectomy and lenectomy, and the plan at publication was to provide the patient with a new lens.

Vesicles are not generally regarded as lying within the spectrum of rashes caused by meningococcal infection. The acute ocular complications of varicella infection include spread of vesicles to the eyelid, papillary conjunctivitis and, uncommonly, dendritic keratitis, acute retinal necrosis, and optic neuritis. Hypopyon, observed in this case, is not a feature of primary VZV infection.

This report serves to emphasise the broad clinical spectrum of meningococcal disease, and highlights the potential for severe sequelae despite an otherwise effective host response to bacteraemia. Seeding of group C meningococci resulted in visual loss in this young patient for whom the UK conjugate vaccine programme came just a year too late.

P A Berry, J K Dort, E Graham, M Matheson, E M E MacMahon
St Thomas’s Hospital, London UK
We believe that the suggested correlation by the authors of acanthamoeba keratitis with water hardness is spurious and demonstrates the danger of projecting “correlation” to “causation” within their study. The authors do not have definitive evidence to support their case because of sampling bias in part from evaluating associations between risk factors and a disease well after the risk exposure first occurs. During that time interval, cases of short duration and cases of severity or asymptomatic, and cases in which the presence of disease alters or entirely removes the exposure, are missed.

Water “hardness” remains unproved as a risk factor for acanthamoeba keratitis being no more than an observational quirk. While we have good molecular based evidence for the association of acanthamoeba keratitis with home tap water, recent work using 18S rDNA typing and tracking has shown that there can be different subtypes of keratitis causing strains of Acanthamoeba spp (T4) between the tap water in the home and the cornea, thus making the epidemiology more involved. 1

In conclusion, the incidence of acanthamoeba keratitis reported for England and Wales by the authors is an underestimate of the true figure by at least 33%. The finding that the reporting of cases was “static” is of concern as it means that there is still need to reinforce ways of preventing this infection. 1

While considerably less frequent since the introduction of multipurpose solutions 7 years ago, the infection remains a problem to be avoided by CL wearers as predicted by one of us 8 years ago. 1 If good hygiene is practised, however, with monthly changing of storage cases, avoidance of tap water, and use only of sterile multipurpose solutions or overnight storage in hydrogen peroxide, then the risk of infection becomes negligible. The all too disposable lens wear has also been virtually free of acanthamoeba infection.

D V Seal
Applied Vision Research Centre, City University, Northampton Square, London EC1V 0HB, UK

T K Beatie, A Tomlinson
Department of Vision Sciences, Glasgow Caledonian University, Cowcaddens Road, Glasgow G4 0BA, UK

D Fan, E Wong
Departments of Ophthalmology and Centre for Clinical Trials and Epidemiological Research (School of Public Health), Chinese University of Hong Kong, Prince of Wales Hospital, Shatin, N\i, Hong Kong

Correspondence to: David Seal; 1007/56, 3112/sealseal@compuserve.com

References
Surgery for glaucoma

We would like to thank Drs Mermod and Rabinet for their online comments on the editorial and also for their comments on O’Brart’s article (letter posted on www.bjophthalmol.com 5 Aug 2002).

We are delighted that there is agreement that subconjunctival drainage is an important component of non-penetrating filtration surgery. Blebs, albeit diffuse, are clearly visible both in cases of deep sclerectomy, and in many cases of viscocanostomy. However, although Rabinet and Mermod and ourselves are certain that this is an important component for drainage in non-penetrating filtration surgery, it is important to clarify this point. Although we are certain of the importance, there is still controversy over the relative importance of subconjunctival drainage versus other routes such as trans-scleral or via Schlemm’s canal in the case of viscocanostomy.

On the second point, although Mermod and Rabinet use antimetabolites before cutting into the sclera, they do not apply antimetabolites including mitomycin into the deep scleral bed. Again, although they do not do this, we know that deep application has been used in other centres. Clearly, the risk of intraocular entry is higher in these situations, and that was the reason for making this comment in the editorial, to warn people of the possible dangers of applying mitomycin after a deep scleral dissection.

Regarding the question of additional hypotony after macroperforation, clearly the team are very experienced in doing this surgery. However, from canvassing personal opinion from individuals who are doing this surgery, who are less experienced than them (like the majority of surgeons doing non-penetrating surgery), and also from reports in the literature including randomised prospective trials, macroperforation is associated with early hypotony. This may well be exacerbated if intraocular application of antimetabolites, particularly mitomycin, have been used.

With regard to the comments on the paper by O’Brart et al, we have forwarded the contents of the letter on to the authors and they have replied separately to the comments.

In conclusion, we are pleased that the article and editorial have led to further healthy discussion. Mermod and his colleagues are to be commended for the work they continue to do in non-penetrating filtration surgery. However, as we said in our editorial, current studies do reveal that none of the current operations for glaucoma are totally ideal yet, and further research, particularly on surgical methods and wound healing control, is needed so that optimal long term pressure control can be achieved for all our patients with a minimum of complications.

P T Khaw, A P Wells, K S Lim

Surgery and the prevention of amblyopia

In conclusion, we are pleased that the article and editorial have led to further healthy discussion. Mermod and his colleagues are to be commended for the work they continue to do in non-penetrating filtration surgery. However, as we said in our editorial, current studies do reveal that none of the current operations for glaucoma are totally ideal yet, and further research, particularly on surgical methods and wound healing control, is needed so that optimal long term pressure control can be achieved for all our patients with a minimum of complications.

A randomised controlled trial of written information

Newsham’s effort to inform parents of children with amblyopia about occlusion therapy is laudable but incomplete. Ethical considerations informed consent require full disclosure of all aspects of the proposed treatment. The following points might be considered for inclusion.

(1) Occlusion therapy has never been scientifically validated with a randomised, controlled study.

(2) The dose/response relationship has never been defined. Flynn et al stated that “Success was not related to the duration of occlusion therapy, type of occlusion used … ” The variety of treatment protocols accentuates another dilemma “owing to our paucity of knowledge on the dose-effect relation—a situation one finds hard to imagine for any comparably established therapy outside ophthalmology. In other words we have no understanding of the dose-effect relation of occlusion in amblyopia therapy.”

(3) The application of “greater levels of occlusion being prescribed for more severe amblyopia” is compromised by the observation “that success was related to … the depth of visual loss before treatment…”

(4) The benefits of treatment are likely to deteriorate following cessation of patching.

(5) Visual acuity improves as children become more mature, literate, and familiar with vision testing protocols. This is also true for amblyopic eyes. In amblyopic children between 3 and 7 years old without treatment visual acuity was shown to consistently improve in each older age group.

(6) Both the occluded and the amblyopic eyes improve at the same rate during treatment.

(7) Success in amblyopia treatment is usually defined as improvement by a minimum of three lines. Many of the successfully treated patients, by that criterion, will still not have normal vision at the end of presumably successful treatment. One quarter of treated patients with initial acuity of better than 20/20 do not even achieve these limited goals.

Therefore, the comments about achieving normal vision may raise expectations that will not be achieved.

(8) Occlusion therapy does have potential adverse effects beyond disruption of family and social life and interference with education.

Despite decades of occlusion therapy the prevalence of amblyopia in the adult population is similar to that of the school age population. Moreover, “the prevalence of unilateral amblyopia was not found to be statistically different by age group.” This suggests that long term benefits of conventional therapy are not demonstrated in demographic studies.

Patients and their families should be provided with comprehensive information concerning proposed treatments. Physicians are obliged to make this information accurate and inclusive.

P Lempert

Cornell University School of Veterinary Medicine, Ithaca, NY 14850, USA; eyechartplus@aol.com

References


www.bjophthalmol.com
Ocular and financial health

We read with interest the article by Khademkar et al detailing the results of the 1996–7 Oman Eye Study.1 They found a prevalence of blindness of 16.8% in those 60+ years of age, and comment that the population of this age group in Oman is predicted to double by the year 2020. This has serious implications for planning the provision of health care and specifically for eye care services.

This observation is true for most countries; the global population for the over 65 age group is projected to increase from approximately 400 million to 800 million people by the year 2020, contrasting with the under 5 years of age population, which is estimated to see a 6% growth in the same period.2 “Vision 2020—the right to sight” was launched by the WHO and IAPB in 1999, aiming for the elimination of avoidable blindness by the year 2020. In 1995 the estimate of global blindness (<3/60 better eye) was 44 million, and this is projected to rise to 76 million by 2020 if there is no change in current trends.3 Vision 2020 prioritises five diseases for global attention—cataract, refractive errors, trachoma, onchocerciasis, and vitamin A deficiency. Action against diabetic retinopathy and glaucoma is also deemed important in countries where ocular infections have been controlled. Vision 2020 contends that the current increase in blindness, estimated at 1–2 million people per year, can be reversed if human and financial resources are targeted at these priority diseases in the countries with the highest prevalence and number of blind people. It is estimated that the result of a successful programme, achievable at a cost of $2 billion, will be $29 billion blind person years avoided over the next 20 years, and a total saving in excess of $100 billion, by avoidance of lost productivity.4 This would make the effective implementation Vision 2020 not only ethically important but also a cost effective strategy for poverty alleviation.

J C Buchan, C Gilbert
London School of Hygiene and Tropical Medicine, Keppel Street, London WC1E 7HT, UK
Correspondence to: John Buchan; john.buchan@lshtm.ac.uk

References
2 www.census.gov/ipc/prod/wpg98/wpg98.pdf
3 Frick KD, Foster A. The magnitude and cost of global blindness: an increasing problem that can be alleviated. Am J Ophthalmol [in press]

Ocular Pathology, 5th ed.

The fifth edition of this well established ocular pathology text is excellent. The presentation, the photographs, the histopathology, and micrograph sections provide the reader with an array of outstanding examples of ocular pathology. The text is easy to read, agreeably not always in depths, but gives enough information or direction in references for further reading. This is being hypercritical of an enjoyable text and there is no more to say except that every unit and library should update their edition.

A Dick

NOTICES

Role of optometry in Vision 2000

The latest issue of Community Eye Health (No 43) discusses the mobility of optometry to deal with uncorrected refractive error, which is now a major cause of functional blindness. For further information please contact: Journal of Community Eye Health, International Centre for Eye Health, Institute of Ophthalmology, 11–43 Bath Street, London EC1V 9EL, UK (tel: +44 (0)20 7608 6910; fax: +44 (0)20 7250 3207; email: eyeresource@ucl.ac.uk; web site: www.jceh.co.uk). Annual subscription (4 issues) UK£25/US$40. Free to workers in developing countries.

International Centre for Eye Health

The International Centre for Eye Health has published a new edition of the Standard List of Medicines, Equipment, Instruments and Optical Supplies (2001) for eye care services in developing countries. It is compiled by the Task Force of the International Agency for the Prevention of Blindness. Further details: Sue Stevens, International Centre for Eye Health, 11–43 Bath Street, London EC1V 9EL, UK (tel: +44 (0)20 7608 6910; email: eyeresource@ucl.ac.uk).

Second Sight

Second Sight, a UK based charity whose aims are to eliminate the backlog of cataract blind in India by the year 2020 and to establish
strong links between Indian and British ophthalmologists, is regularly sending volunteer surgeons to India. Details can be found at the charity web site (www.secondsight.org.uk) or by contacting Dr Lucy Mathen (lucymathen@yahoo.com).

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

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

**Surgical Eye Expeditions International**
Volunteer ophthalmologists in active surgical practice are needed to participate in short term, sight restoring eye surgery clinics around the world. Contact: Harry S Brown, Surgical Eye Expeditions International, 27 East De La Guerra, C-2, Santa Barbara, CA 93101-9858, USA (tel: +805 963 3303; fax: +805 965 3364; email: hsbrown.md@cox.net or seeintl@seeintl.org; web site: www.seeintl.org).

**16th Annual Meeting of German Ophthalmic Surgeons**
The 16th Annual Meeting of German Ophthalmic Surgeons will be held 8–11 May 2003 in Nürnberg, Germany, Messezentrum. Organised by the Professional Association of German Ophthalmologists Ophthalmic Surgery Group the conference will cover cataract surgery, refractive surgery, glaucoma surgery, vitreoretinal surgery, corneal surgery, eye surgery in developing countries, and orbita, lacrimal and lid surgery. Further details: MCN Medizinische Congress organisation Nürnberg AG, Zerrabzehofsitz 29, 90478 Nürnberg, Germany (tel: +49 911 3931621; fax: +49 911 3931620; email: doc@mcnag.info; web site: www.doc-nurnberg.de).

**3rd British Oculoplastic Surgery Society Meeting**
The 3rd British Oculoplastic Surgery Society Meeting will be held 18–19 May 2003 in Birmingham, UK. For further details please contact the Secretary of the British Oculoplastic Surgery Society Jane Olver (tel: +44 (0)121 424 5464; fax: +44 (0)121 424 4464; email: MaritiDI@heartsof.wmlids.nhs.uk; web site: www.bopps.org).

**13th Meeting of the EASD Eye Complication Study Group**
The 13th Meeting of the EASD Eye Complication Study Group will be held on the 23–25 May 2003, in Prague, Czech Republic. The scientific programme includes keynote lectures from Professor John H Fuller (UK) on The epidemiology of diabetic retinopathy; Dr P Martin van Hagen (The Netherlands) on Growth factors and diabetic retinopathy; Professor Terezije Peliakanova (Czech Republic) on Pathophysiology of diabetic microvascular complications; Dr Tomas Sosna (Czech Republic) on Risk and protective factors of diabetic retinopathy. Further details: Ortopedie Centrum, s.r.o., Steckkovské nabrezi 51, 400 03 Usti nad Labem, Czech Republic (tel: +420 47 521 6588; fax: +420 47 533 40 77; email: ortcentrum-ul@volni.cz; web site: www.oropedie-centrum.cz).

**IOIS—VIIth International Symposium on Ocular Inflammation**
The IOIS—VIIth International Symposium on Ocular Inflammation will be held 25–30 May 2003, in Padova, Italy.

Programme and Organising Committee: Prof. Antonio Secchli, Chairman of IOIS and Convener of Congress (email: ag.secchli@unipd.it). Further details: Congress Secretariat, Meet and Work, Mario Sbalchiero (tel: +39 049 860 1818; fax: +39 049 860 2389; email: info@iois-2003.com; web site: www.meetandwork.com).

**XXXII Retinal Detachment Course**
The XXXII Retinal Detachment Course with International Faculty and Case Presentations will be held 5–6 June 2003, in Beijing, China. The Congress language is English with simultaneous translation in Chinese. Further details: Scientific programme: Prof Ingrid Kreissig, University of Tuebingen, Schleichstr. 12, Breuningerbau, 72076 Tuebingen, Germany (tel: +49 7071 295209; email: ingrid.kreissig@med.uni-tuebingen.de).

Local organisation: Ningli Wang, Liang Xu, Wu Liu. Congress office: Wu Liu, Beijing Tong Ren Eye Center, Beijing 100730 China (email: wuliu@yahoo.com).

**Detachment Course with international faculty on: Retinal and Vitreous Surgery with Case Presentations preceding the Annual Meeting of Iranian Society of Ophthalmology**
The detachment course with international faculty on: Retinal and Vitreous Surgery with Case Presentations preceding Annual Meeting of Iranian Society of Ophthalmology will be held on 29–30 November 2003 and 1–4 December 2003 respectively, at the Razi Conference Center, Hemmat Hyw, Tehran, Iran. Further details: Scientific programme: Prof Ingrid Kreissig, University of Tuebingen, Schleichstr. 12, Breuningerbau, 72076 Tuebingen, Germany (tel: +49 7071 295209; email: ingrid.kreissig@med.uni-tuebingen.de).

Local organisation: Dr Arman Masheyekhi, Dr Sihamak Moradian, Dept of Ophthalmology, Labbanfinejad Medical Center, Pasdaran Ave, Boosan 9, Tehran, 16666, Iran (fax: +98 21 254 9039; email: labbali@hotmail.com).