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 immunosuppressants such as cyclophosphamide can induce long term remission.

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 showing no obvious rheumatoid abnormalities. The patient was commenced with topical dexamethasone 0.1% non-preserved hourly and oral prednisolone at 40 mg/day.

On the fifth day, the patient’s hearing had deteriorated only over the past few months and was investigated. A provisional diagnosis was made of Wegener’s granulomatosis (WG). Her serum antineutrophil cytoplasmic antibodies (cANCA) titre was found to be raised (>160). Rheumatoid factor was negative.

Oral cyclophosphamide 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. 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. Profound sensory deafness is a less common feature, occurring in about 6% of patients on presentation. It occurs either secondary to inflammation of the cochlear vessels or serious otitis media. Other important systemic features include pyrexia, weight loss, recurrent epistaxis, sinus discharge, haemoptysis, peripheral neuropathy, cerebral vasculitis, and renal failure (major cause of death). Unfortunately 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.

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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 chemotherapeutic drug that suppresses the adrenal cortex 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, adrenalectomy, 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 in 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 fludrocortisone to attempt to improve her vision.
Figure 1 Right fundus showing mottled pigmentation at the macula.

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 adenocortical 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 with features of papilloedema and retinal haemorrhage. In another study involving 19 patients, three patients had toxic retinopathy that included papilloedema, small retinal haemorrhages, and oedema and another patient had a subcapsular cataract. To our knowledge neither pigmentary retinopathy nor abnormal ERG findings have been reported previously. Previous studies also did not mention side effects reversibility except for a case of lenticular opacities that disappeared 3 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 adrenoleucodystrophy is associated with pigmentary retinopathy and other ocular findings. Adrenoleucodystrophy is a group of rare lipid storage disorders with increased serum level of long chain fatty acids (C24–C30). Ocular findings in adrenoleucodystrophy include visual loss secondary to vitreous demyelination and primary retinal ganglion cell degeneration, squint, cataracts, loss of corneal sensation, abnormal visual evoked potentials, and macular pigmentary changes. The retinal pigmentary changes observed histologically were different from those in retinitis pigmentosa.

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 6-beta-hydroxyl cortisol, cholesterol, liver enzymes, corticosteroid binding globulin, and sex hormone binding corticosteroids level but an increase in the levels of 17-hydroxy corticosteroids. Mitotane could occur via a similar mechanism that affects the metabolism of long chain fatty acids. It is possible, however, that the ocular 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 6-beta-hydroxyl cortisol, cholesterol, liver enzymes, corticosteroid binding globulin, and sex hormone binding corticosteroids level but an increase in the levels of 17-hydroxy corticosteroids.

References

Coxsackievirus B4 associated uveoretinitis in an adult

Coxsackievirus is a kind of enterovirus, which may cause respiratory and gastrointestinal symptoms, erythema, meningoencephalitis, 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, scattered hemorrhagic dots, and occlusive retinal vasculitis have not previously 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 generalized malaise. There was no significant medical or family history. At the initial ophthalmic examination, his best corrected visual acuities were 20/20 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 field testing was within normal limits in both eyes. Ophthalmoscopical examination disclosed subretinal exudates with haemorrhage in the paracentral and mid-periphery of bilateral eyes (Fig 1). Moreover, scattered
haemorrhagic dots and blots were observed along retinal vessels. The optic discs appeared normal. Fluorescein angiography demonstrated occlusive retinal vasculitis characterised by blocked fluorescence and mild dye leakage from the retinal vessels corresponding 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 \( \times 10^3/\)l, 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).

Eight months after the initial examination, bilateral exudative decrease. The patient's general condition improved and antibody scars, and most of the retinal haemorrhagic lesions had improved and were replaced by irregular choroidal background fluorescence.

**Figure 1** Photographs of right eye (A) and left eye (B) at the initial ophthalmic examination. Subretinal exudates with haemorrhage around the macula and near the disc, 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 and choroid along the macula, and irregular choroidal background fluorescence.

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. The other was an adult case, with choriorretinitis similar to the multiple evanescent white dot syndrome. 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 clinical setting, since it is possible that overt clinical manifestations and complications may occur depending on the severity and the location of the uveoretinal lesions.

**Comment**

Viruses are one of the most common causes of infections involving the posterior segment of the eye. Such infections may be congenital or 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, subsequent to an acute viral systemic illness. Coxsackievirus associated choriorretinitis is thought to elicit the same clinical course as in other viral choriorretinitis. Coxsackievirus infection and concurrent posterior segment inflammation have been reported in a few cases, in which clinical and antibody titres suggested that Coxsackievirus was the responsible agent. 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. The other was an adult case, with choriorretinitis similar to the multiple evanescent white dot syndrome. 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 clinical setting, since it is possible that overt clinical manifestations and complications may occur depending on the severity and the location of the uveoretinal lesions.

**References**


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

Ophthalmic surgery is well suited to the use of telemedicine in diagnosis and management, but diagnostic accuracy and reliability are critical if it is to be widely introduced. Moorfields Eye Hospital is located in central London and runs outreach clinics. One clinic and operating list in Ealing is 17 miles from the central hospital. The surgical teams operate in Ealing and return to the central hospital. We therefore investigated the use of a telemedicine link for review of postoperative cataract surgical patients aiming to assess what can be seen clearly and reliably using telemedicine and to identify what the observer using this link may potentially miss.

**Case report**

This study had Moorfields Eye Hospital research and ethics committee approval. Patients consecutively admitted for cataract surgery were recruited to this study and fully 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 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 stability. 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 calculated in terms of sensitivity and specificity, these findings give an indication of the accuracy 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 corneal section, or anterior chamber flare and failed entirely to detect DM folds or anterior chamber cells. Patients enjoyed the telemedicine experience finding it reassuring to see as well as interact with their surgeon via the telelink.

Video compression algorithms used in streaming video are “lossy” and higher bandwidth increases video quality. The process begins with a good video source with low noise, since noise does not compress 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 bandwidth. With increasing bandwidth there is correspondingly better quality video. A televideo 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 information observed. Broadcast video monitors are accurately colour calibrated, whereas digital compressed video to 384 K bandwidth has
The prenatal and postnatal development of the eye is determined by complex interactions between a number of genes, their products, and certain environmental factors. Since each eye is influenced by precisely the same genetic processes as its fellow, mutations in regulatory genes usually lead to symmetrical 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 inferior longstanding retinal detachment with subretinal fibrosis. The left 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.

<table>
<thead>
<tr>
<th>Oedema of the incision</th>
<th>m:2.933, p=0.087</th>
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<tr>
<td>Control group (n=10)</td>
<td>Study group (n=12)</td>
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<tr>
<td>Oedema of the central cornea</td>
<td>7/10 (70%)</td>
</tr>
<tr>
<td>Descemet's folds</td>
<td>7/10 (70%)</td>
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<tr>
<td>Anterior chamber cells</td>
<td>10/10 (100%)</td>
</tr>
<tr>
<td>Anterior chamber flare</td>
<td>9/10 (90%)</td>
</tr>
<tr>
<td>Lens implant stability</td>
<td>9/5 (90%)</td>
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<tr>
<td>Management decisions</td>
<td>9/10 (90%)</td>
</tr>
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</table>

### References


### 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. Since each eye is influenced by precisely the same genetic processes as its fellow, mutations in regulatory genes usually lead to symmetrical 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 inferior longstanding retinal detachment with subretinal fibrosis. The left 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.

![Figure 1](color graphics of right fundus of sib 1 (A) showing macular “coloboma,” generalised retinal pigment epithelial and choroidal atrophy, and evidence of sheathed and occluded retinal vessels inferotemporally; (B) same eye, shallow inferior retinal detachment with subretinal fibrosis.)

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 offering a much more comprehensive postoperative assessment with trained operators. H-261 video at 384 K (triple ISDN) is sufficient for assessment with trained operators. H-261 video at 384 K (triple ISDN) is sufficient for a wholly different experience from a simple telephone conversation and for many situations is capable of offering a much more comprehensive postoperative assessment with trained operators.
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 dating 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 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 allergic conjunctivitis. The tumour was a relatively small and dark brown lesion arising from the ciliary body. The vitreous showed presence of pigment clumps and the degree of anterior chamber invasion appeared disproportionate to the size of the tumour.

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 intraocular 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 1 Clinical details of the patients with adenoma of the pigment epithelium of ciliary body

<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>
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<tr>
<td>1</td>
<td>40/M/W</td>
<td>6/5</td>
<td>8/7×1</td>
<td>Angle invasion</td>
<td>Local resection</td>
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<tr>
<td>2</td>
<td>62/F/A</td>
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<td>5×3×1</td>
<td>Angle invasion, cataract</td>
<td>Local resection</td>
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<tr>
<td>3</td>
<td>65/F/W</td>
<td>6/18</td>
<td>8×4×4</td>
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<td>Local resection</td>
<td>No</td>
<td>1998</td>
<td>RD repair</td>
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<tr>
<td>4</td>
<td>54/M/W</td>
<td>1/60</td>
<td>5×4×4</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, tractional 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. Ultrasound 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 tractional retinal detachment.

Histopathologically, these tumours showed haemorrhage, mitotic activity which 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 the presentation of features 1 and 2. Blurred vision due to pigment dispersion was noted in patients 3 and 4. This was mainly in the vitreous resulting in the presentation of patient 3. Angle closure glaucoma and pigment dispersion were the main features of patient 4. Shields et al reported 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 patients 3 and 4 was unique and has not been reported before. Chang et al in 1979 reported the presence of pigment in the retrostellar 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 to our knowledge not 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 1999, 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 thought to be associated with malignancy. However, this is not always the case. Presence of a sentinel vessel indicates ciliary body involvement. Fine needle aspiration biopsy may be considered for aiding diagnosis of malignancy. However, its role in the diagnosis of these lesions may be limited. Absence of malignant cells does not always rule out the presence of malignancy.

Our study highlights the paradoxical behaviour of adenoma of pigment epithelium of ciliary body that has not been emphasised before. Adenoma of the pigment epithelium of the ciliary body should be kept in mind if there is extensive pigment dispersion by larger tumours and invasion of the anterior chamber angle by relatively small tumours.

References

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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 hyper tension, heart failure, or diabetes was made. All patients received one drop of each of tropicamide 1%, cyclopentolate 1%, and Voltarol 0.1% Phenylephrine 10% and 0.1%, 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 and 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 phenylephrine 2.5% and 10% in phacoemulsification under topical anaesthesia: is there an effect on systemic blood pressure?

Phenylephrine 2.5% tends 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 phenylephrine is 1.5 mg and Kumar et al had found phenylephrine plasma levels after administration of topical 10% viscous 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% viscous solution.

Chin et al, in their study on 89 patients, concluded that significant hypertensive effects can arise after topical phenylephrine; however, no significant difference between the two groups (2.5% vs 10%) was shown. Symons et al reported no significant change in the mean systolic and diastolic blood pressure in 126 patients receiving 10% phenylephrine. 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.

Phenylephrine 2.5% and 10% in phacoemulsification under topical anaesthesia: is there an effect on systemic blood pressure?
effect. The blood pressure although the concentration is excluded which might cause elevation of which is always included in local anaesthesia, this element should be taken into consideration since it is known to elevate the systolic BP but weren't statistically significant; the mean of which is 22.8 mm Hg.

Using Altman's normogram the study diastolic BP but weren't statistically significant; the mean of which is 22.8 mm Hg. In the hypertensive group six patients recorded a 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 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.

We therefore recommend the routine use of the 2.5% phenylephrine in ophthalmic surgery and only use 10% solution for cases where the lower concentration may not be as effective.

References


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 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 (82%, 9/11) was detected preoperatively than in those in whom displacement was not detected (0%, 0/4) (p = 0.01, χ² test with Yates’s correction). The mean preoperative VA tended to be better and the mean VA measured 3 months postoperatively was significantly better in patients in whom photoreceptor displacement was detected preoperatively (0.15 and 0.79, respectively) than in those in whom it was not detected (0.08 and 0.25, respectively) (p = 0.06 and p = 0.001, respectively, unpaired t test). Visual acuities were converted to the logarithm of the minimum angle of resolution (logMAR) for statistical analysis.

Comment

Jensen and Larsen reported that in two macular hole patients with the longest duration of symptoms, photoreceptor displacement was not detected. The reason why patients with a macular hole with longer duration of symptoms did not have photoreceptor displacement might be that there was more damage to the displaced photoreceptors. A longer disease duration probably induced functional damage to the displaced photoreceptors, and consequently no photoreceptors were detected. We also reported that the detection of photoreceptor displacement preoperatively should affect postsurgical visual function, and photoreceptor damage might occur in eyes in which photoreceptor displacement was not detected preoperatively, resulting in worse postoperative visual function.

Our results demonstrated that in patients with a macular hole in whom photoreceptor displacement was detected preoperatively, the postoperative foveal findings and visual acuities were better when compared to patients in whom displacement was not detected preoperatively. It was also reported that better anatomical foveal recovery in eyes after macular hole surgery resulted in better improvement of postoperative visual function. Although limitations of this study are based in its sample size, we think that a degree of preoperative photoreceptor damage influences not only postoperative visual function but also anatomical recovery.

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

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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. 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 53 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 left eye was unremarkable. The ultrasound images of the macular hole were normal. The postoperative visual function was better than in those in whom displacement was not detected preoperatively, which might occur in eyes in which photoreceptor damage influences not only postoperative visual function but also anatomical recovery.

Our results demonstrated that in patients with a macular hole in whom photoreceptor displacement was detected preoperatively, the postoperative foveal findings and visual acuities were better when compared to patients in whom displacement was not detected preoperatively. It was also reported that better anatomical foveal recovery in eyes after macular hole surgery resulted in better improvement of postoperative visual function. Although limitations of this study are based in its sample size, we think that a degree of preoperative photoreceptor damage influences not only postoperative visual function but also anatomical recovery.

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

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Figure 1  Scleral nodule of the left eye with tenderness in a patient with sarcoidosis.

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 51.5 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 lymphocyte cell infiltration, with non-casecting 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-Neelson and Golorni’s silver method showed no evidence of acid fast bacilli. Scanning electron microscopy revealed a progressive interstitial lung disease typical of sarcoidosis.

Comment

Scleral involvement has been previously reported in a few patients with sarcoidosis. In a series of 537 patients with histologically proved sarcoidosis, 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, 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.

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...
An 18 year old male patient presented to our clinic with contact TSCPC using G-probe delivery, reported in about two thirds of eyes with panophthalmitis following contact diode laser cyclophotocoagulation. The mainstay of treatment for sarcoidosis is corticosteroids, and the scleritis responded in our patient.

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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. Panophthalmitis following contact diode laser cyclophotocoagulation in patients with glaucoma. The spectrum of the ICE syndrome includes the Chandler’s syndrome, the essential iris atrophy, and the Cogan-Reese syndrome. We present a unique case of an ICE syndrome, which we would subclassify as Chandler’s syndrome, with concomitant fingerpfft dystrophy and multiple interepithelial bilaminar bodies within the fingerprint-like striae.

Case report
Ten years earlier a now 45 year old patient presented with a bilateral ICE syndrome and fingerpfft dystrophy. On slit lamp microscopy the epithelial layer showed fingerprint lines, while the endothelial layer had a “hammered” appearance. Iris atrophy, corectopia, ectropion uveae and anterior synechiae were obvious (Fig 1A) and progressive in both eyes, nevertheless the intraocular pressure and the visual acuity were normal. Eight years later both corneas developed oedema (Fig 1B) and the left eye displayed cataract with reduction of visual acuity from 1.0 to 0.25 and 0.9 to 0.7 in the left and right eye, respectively. Combined perforating keratoplasty and extracapsular cataract extraction were performed on the left eye in June 1998. None of the patient’s family members had a similar condition.

The histological examination of the paraffin embedded corneal specimen revealed fingerprint-like stria containing small basophilic deposits between the cells (Fig 2A). The van Kossa stain for calcium was negative and no hydroxyapatite crystals were seen. At the ultrastructural level the deposits were found to be round and bilaminar showing an electron dense core in the centre (Fig 2B). Some of the deposits coalesced to form oval lesions.

References

Bilaminar interepithelial bodies within fingerprint dystrophy-like changes in bilateral iridocorneal endothelial syndrome
In most cases the iridocorneal endothelial (ICE) syndrome affects women unilaterally and shows endothelial degeneration, thickening of Descemet’s membrane, iris atrophy, and the Cogan-Reese syndrome. We present a unique case of an ICE syndrome, which we would subclassify as Chandler’s syndrome, with concomitant fingerprint dystrophy and multiple interepithelial bilaminar bodies within the fingerprint-like striae.

Case report
Ten years earlier a now 45 year old patient presented with a bilateral ICE syndrome and fingerprint dystrophy. On slit lamp microscopy the epithelial layer showed fingerprint lines, while the endothelial layer had a “hammered” appearance. Iris atrophy, corectopia, ectropion uveae and anterior synechiae were obvious (Fig 1A) and progressive in both eyes, nevertheless the intraocular pressure and the visual acuity were normal. Eight years later both corneas developed oedema (Fig 1B) and the left eye displayed cataract with reduction of visual acuity from 1.0 to 0.25 and 0.9 to 0.7 in the left and right eye, respectively. Combined perforating keratoplasty and extracapsular cataract extraction were performed on the left eye in June 1998. None of the patient’s family members had a similar condition.
the ICE syndrome, owing to obvious morphological changes of the iris despite non-specific endothelial changes. The striking features of Chandler’s syndrome are corneal endothelial dystrophy and subsequent oedema. Corectopia, glaucoma, and iris atrophy are less compared to the two other ICE syndromes—that is, the essential iris atrophy and the iris-naeveus syndrome, possibly owing to differences in endothelial proliferative activity.

Our case is unique in several aspects. Firstly, our patient is a man and is affected bilaterally. Most often women are affected unilaterally by ICE syndrome, nevertheless an exception of essential iris atrophy with bilateral changes have been described. Secondly, an epithelial dystrophy—namely, a fingerprint dystrophy, is present in addition to the ICE associated changes. To our knowledge this association has not been described in the literature before.

Thirdly, the fingerprint striae contain bilaminar deposits with an electron dense core. Several types of deposits have been described in association with epithelial dystrophies. Patches of “unknown substance” were noticed in fingerprint dystrophy along basal plasmalemmas of the basal epithelial cells without any similarity to the bilaminar structures described here. Broderick et al mentioned intercellular dense oblong bodies which were embedded in condensed matrix and were much smaller than the deposits in our case. In a cornea with a Meesmann dystrophy intraepithelial “peculiar electron dense substance” were shown, also with a totally different appearance compared to that seen in the fingerprint striae of the specimen we present here. In a patient with ocular tumoral calcinosis, nodules from the palpebral conjunctiva contained calcified “spherules,” which show some similarity to our—nevertheless, uncalcified—deposits. Our deposits were located within the fingerprint striae, close to the epithelium. It seems to be obvious that the epithelial cells must have produced those bilaminar bodies, since no other cell type was present nearby, but there was no evidence of extrusion from the basal cells.

Our case shows that ICE syndrome can occur bilaterally in men, and can be associated with a fingerprint-like dystrophy. Further studies should be carried out to confirm our electron microscopical observation of bilaminar deposits within fingerprint-like striae and directed towards an analysis of their chemical composition of the extracellular material and origin.

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References

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,1–4 the fundus appearance becomes indistinguishable from other retinal dystrophies with time.5 The advent of confocal scanning laser ophthalmoscopy (SLO) with infrared light has enabled high sensitive examination of the deep layer of the retina.6 We studied the deep retinal abnormalities in a case with a typical Bietti’s crystalline retinopathy and in another case with myopic choriotinal atrophy throughout the posterior pole. Consequently, we detected numerous fine crystalline deposits in both cases, which were not detected with other funduscopic 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 20/20 left eye. Infrared examination of the deep layer of the retina 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 subretinal 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 hypofluorescence lesions surrounded by hyperfluorescent lesions in the early phase. Indocyanine green angiography also showed the atrophy of the choriocapillaris 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 funduscopic examinations (Fig 1). Each crystal deposit appeared larger in size. In addition, numerous fine crystal deposits were evident, which were not observed by conventional funduscopic examinations including ophthalmoscopy and
and no fine crystal deposits were found at
limbus 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 choroid and extending from
the posterior to mid-peripheral retina in
both eyes. Optic disc showed myopic appear-
ance. Retinal vessels and peripheral retina
showed normal appearance. Standard elec-
trotectromegametry was non-recordable. Gold-
mann perimetry showed marked constriction
of all isopters and loss of visibility of I–4–e
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 hyperflu-
orescence in the later phase. Indocyanine green
angiography disclosed the loss of background
veil-like fluorescence throughout the poste-
rior 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 fundu-
scopic examinations, including ophthalmo-
scopy 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 examina-
tions did not show any abnormal value.

Comment
The diagnosis of Bietti’s crystalline retin-
opathy in case 1 is based on the retinal
crystalline and supported by the characteris-
tic island-like hypofluorescence observed by
fluorescein angiography.1 Fundus examina-
tion by SLO with the infrared light disclosed
numerous tiny crystalline deposits, which were
not observed by conventional fundus examina-
tions. Thus, in Bietti’s crystalline retinopathy,
there are more numerous accum-
ulation of retinal crystalline deposits than
are observed with funduscopic examina-
tions.
In case 2, numerous tiny retinal crystalline
deposits were detected by SLO with infrared
light, although no other examinations dis-
closed 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 depos-
its, 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 angi-
ography, diagnosis of Bietti’s crystalline retin-
opathy in advanced cases without fundu-
scopically apparent crystalline deposits have
been established only when family history
was contributory.1 Because the presence of
crystalline material in the peripheral
lymphocyte5 was not observed by transmis-
sion electron microscopy, whether case 2 can
be categorised as Bietti’s crystalline retin-
opathy or a distinct clinical entity awaits fur-
ther investigation. Additionally, to our knowl-
edge, Bietti’s crystalline retinopathy with
degenerative myopia has not been reported in
the literature. However, the presence of
parental consanguinity of this case might
suggest an autosomal recessive inheritance,
similar with some reported cases with Bietti’s
crystalline retinopathy, and the striking re-
ssemblance of the morphology of the tiny crys-
talline deposits observed by SLO using infra-
red light between the two cases may support
that underlying pathogenesis is similar.

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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 supersen-
sitivity to topical dilute pilocarpine.1 Most
instances do not have an identifiable cause;
however, a variety of conditions including
crystals, herpes zoster, orbital trauma including sur-
ery, and paraneoplastic syndromes may
result in tonic pupils. The mechanisms(s) which
produce tonic pupils are not known, al-
though axonal loss within the ciliary gan-
lon is generally believed to result in super-
sensitivity of the iris sphincter muscle. Giant
cell arteritis (GCA) has been reported as an
uncommon cause of tonic pupils.2 Further-
more, the cause of tonic pupils in GCA is
unknown and direct evidence for an ischae-
ic 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 58 year old woman presented with visual
loss of the left eye for 2 days. She had had

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pupils constricted slowly and tonically to a pupil no longer reacted to direct light. Both eye and neuro-ophthalmic examination was begun after the instillation of 0.08% pilocarpine, with sectoral iridoplegia. Both examination on the right revealed an irregular pupil (Fig 2B) with sectoral iridoplegia. Both pupils were sluggish in the first hour and her jaw claudication was unchanged and her pupils remained non-reactive to light but with denervation supersensitivity, confirmed by constriction to 0.125% pilocarpine, 2 months after the onset of visual loss.

Comment
Five earlier reports have documented tonic pupils in patients with GCA. Davis and coworkers described a 69-year-old woman who developed polymyalgia and anisocoria. The involved pupil constricted to 2.5% methacholine. In another report of tonic pupils from GCA mild supersensitivity to 0.1% pilocarpine was noted; however, the patient had counting fingers vision in each eye from bilateral optic neuropathy, suggesting that the light near dissociation may have been caused by bilateral afferent dysfunction. The authors cited pathological studies which have suggested ischaemia of the extraocular muscles as a cause of ophthalmoplegia in some patients with GCA, and they implied that ischaemia was the cause of tonic pupils in their patient, although the site of damage was not specified. Currie and Lessell reported a 63-year-old woman who had jaw claudication and loss of vision bilateral from sequential anterior ischaemic optic neuropathy owing to biopsy proved GCA. After losing vision in each eye she developed bilateral tonic pupils which constricted markedly to 0.08% pilocarpine. These authors suggested that impaired orbital blood flow, including the blood supply to the ciliary ganglia or its postganglionic ciliary nerves, resulted in loss of axons and denervation supersensitivity, although no orbital blood flow studies were performed. They speculated that tonic pupils in patients with GCA may be overlooked because of the severity of visual loss.

Other authors have suggested that ischaemic damage may not be the sole cause of tonic pupils in patients with GCA. Bilateral tonic pupils with conjunctival injection were the only ocular manifestations in a 60-year-old woman with biopsy proved GCA. Because there was no other clinical evidence of orbital or ocular ischaemia the authors postulated that damage to the ciliary ganglion may be mediated by an immunological mechanism. However, they did not quantitate the orbital or ocular blood flow, and our patient shows that clinical signs of ischaemia, except for tonic pupils, may be absent despite markedly decreased orbital blood flow.

Wilhelm reported five patients with bilateral visual loss from anterior ischaemic optic neuropathy or central retinal artery occlusion and tonic pupils from GCA. Assessment of orbital blood flow was performed using ultrasonography in three of these patients. In two patients no flow was noted in the suprachoroidal arteries, while blood flow in the third was normal. The author noted that the ultrasonographic results supported ischaemia as a cause for the tonic pupils. In the patient with normal orbital blood flow, ischaemia below the resolution of ultrasonography was presumed to cause the tonic pupils.

The blood supply to the ciliary ganglion arises from several sources. Eliskova studied 18 human cadaveric orbits after injection of dye into the internal carotid artery. 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 vascularisation of the ganglion itself is composed of a network of capillaries.

Orbital colour Doppler imaging is a non-invasive way to quantitate blood flow to the orbit and eye. Decreased blood flow in the ophthalmic arterial system, central retinal artery, and short posterior ciliary arteries may be detected on orbital colour Doppler imaging in patients with GCA. 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 hypersensitivity. 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 constriction.

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Frozen cucumber as a mount for processing vitreoretinal specimens

Vitreoretinal specimens are extremely difficult to process as a frozen specimen because of their small size and tendency to roll up. 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.

Dua et al. and Scott et al. 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 cm²) 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.

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 cucumber 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 manage 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 studies we found that the specimens attached to the cucumber without the use of albumin.

Comment

Swan and Davis first described the biopsy cucumber unit for processing cervical specimens. Frozen cucumber has been described as a mount for bladder and lung biopsies. Ocular tissues that have been processed in this fashion are conjunctiva and cornea. Whittle et al described a technique using cucumber as a mount for processing cadaveric human retina, which enabled indirect immunofluorescence studies.

Retinal specimens are difficult to process because of their size, tendency to roll up and, hence, difficult orientation. Nevertheless, it is necessary to process specimens in this way to perform certain immunohistochemistry techniques.

We suggest that cucumber is a suitable mount for vitreoretinal membranes that are required as frozen specimens for immunohistochemistry. It should be noted that most modern immunohistochemistry may be performed on fixed tissue.


Each author states that he has no proprietary interest in the development or marketing of any product used in this study.

References


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...
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 occlusive, 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 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 panuveitis. 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 (10 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, 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 anterior uveitis, 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 occlusion 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 the blood vessel wall.

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. This is 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.

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**References**


Angiosarcoma of the eyelid

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 check 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 was 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%-6% (Fig. 1B). Immunohistochemically, the tumour cells showed a positive reaction for vimentin, CD34 (human haemopoietic progenitor cell antigen) and FVIII-RA. They were consistently negative for S100 and HMB45 (results not shown).

Comment

Cutaneous angiosarcoma is a distinct subgroup of angiosarcomas most commonly seen in the skin and superficial soft tissues in patients over 55 years. 1 The majority of these tumours arise in the head and neck area, particularly the scalp. They are aggressive and tend to recur locally and disseminate widely with a 5 year survival of approximately 12%-29%. 2 To our knowledge, there are only seven well documented cases involving the eyelid. 3-5 Clinically, the lesions in our patient resembled those previously reported in view of the purple coloured maculopapular lesions, suggesting a vascular origin. 6 In 50% of cases the lesions are multifocal. 1 Often the lesions are poorly defined and the true extent of disease may be difficult to determine clinically. 5,7-10 The differential diagnosis includes ecchymoses, capillary haemangioma, melanoma, and Kaposi's sarcoma. Advanced lesions may ulcerate and atypical tumours simulating, cellulitis, rosacea and xanthelasma have also been reported. 11-13 Owing to lack of clinical awareness and the varied presentation, diagnosis is often delayed.

Histologically, angiosarcomas display a spectrum from well to poorly differentiated varieties. The present case was consistent with a well differentiated angiosarcoma with characteristic interfacing channels lined by atypical endothelial cells and expression of immunohistochemical markers of vascular differentiation (CD34 and FVIII-RA). 11 Often tumours display both vasoformative and undifferentiated areas. 1 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). 10 Mitotic index has been observed as an independent histological prognostic marker for cutaneous angiosarcoma although other histological markers remain poorly defined. 11 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. 5 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. 5,11 As illustrated by our case, the primary lesions were contained within the anterior lamella of the lid 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. 12 In one series of 28 head and neck angiosarcomas, Mark et al 13 have reported better survival after 32 months (mean) with combined surgery and radiotherapy compared with surgery alone. In cases unsuitable for surgery, radiotherapy alone may be considered, however usually only partial responses are achieved. 12 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 (RMU). The Deutsche Forschungsgemeinschaft, Bonn, Germany (TH), Alergan Australia in conjunction with The Royal Australian and New Zealand College of Ophthalmologists and Sydney Eye Hospital (RMC).

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References

Permanent visual loss in a child with a rash

Endophthalmitis is an unusual but well described complication of bacteremia. 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. Spots appeared on her limbs later the same day but they blanched beneath a glass bottle and several developed into small blisters. A classmate was known to have chickenpox. Next day some new spots appeared on her buttocks, and some of the older lesions appeared to be crusting over. She was noticed rubbing her right eye and later said she felt warm, and vomited. Next morning she complained of pain in the right eye and could not open it. She said she felt warm, and vomited. Next morning she complained of pain in the right eye and could not open it.

Examination

Investigations included a white cell count of 8.0 x 10^9/l (neutrophils 4.0 x 10^9/l) and C reactive protein 22 mg/l. Material was collected from the base of the sol remaining vesicle, on her skin. Ciprofloxacin was commenced parenterally at a dose of 100 mg twice daily (10 mg/kg/day). Examination under general anaesthesia revealed a panophthalmitis with total corneal abscess and hypopyon. There was no evidence of a puncture wound to suggest exogenous infection. Aqueous humour was aspirated, and intravitreous ampicillin and vancomycin administered. Vitrectomy could not be performed because of the apparent septicaemia and meningitis.

A tentative diagnosis of meningococcal endophthalmitis in a clinically well, non-bacteraemic patient was suggested. The diagnosis was confirmed by PCR testing of the vesicular material was negative for varicella zoster virus (VZV) and herpes simplex virus. Neither IgM nor IgG against VZV was detected in the blood. Cultures of blood and aqueous humour were negative but urgent Gram stain of the aqueous humour revealed scanty intracellular Gram negative diplococci. Meningococcus C DNA was detected in the aqueous and in the skin scraping, but not in blood, by PCR testing.

Comment

In this case meningococcal endophthalmitis, usually seen in association with clinically apparent septicaemia and meningitis, occurred in the setting of a mild illness thought to be chickenpox. Although the usual hallmark of meningococcal sepsis were absent, evidence for seeding of the eye in the course of a transient meningococcaemia 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 the 17 year-old had a transient meningococcaemia was provided as scarlet fever. The 17 year old appeared completely “non-septic” with a 4d a yhistory of the condition. The 15 year old had a low grade fever and a transient fever. A vesicular rash on the lower extremities appeared vasculitic on histological examination.

In all of these cases only one eye was involved. Meningococcus type C was isolated from the eye in two cases, type B in a third, and in the fourth case the type was not stated. The outcome was bad in each case. In one of the three older patients vision was reduced to the perception of light only, with dense cataracts forming in two, and phthisis in the 23 year old who underwent vitrectomy. The 13 month old underwent vitrectomy and lensectomy, 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. Hyponychia, 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 bacteremia. 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.

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MAILBOX

If you have a burning desire to respond to a paper published in the BJO, why not make use of our “rapid response” option? Log on to our website (www.bjophthalmol.com), find the paper that interests you, and send your response via email by clicking on the “eletters” option in the box at the top right hand corner. 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 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 guttata.” We note the close similarities between the condition guttata and 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 fear they’re 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.
“Guttata were present” may well have
granted the ears of an ancient Roman, since
gutta is feminine singular, and Romans
liked their subjects to agree with their verbs in
number as much as the rest of us. However, it
is not so clear that “Flavia’s guttata is impres-
sive” would be at all offensive, particularly
since Flavia herself was named after an
adjectival, as were Augustus and, some say,
even Caesar himself. In fact, early Roman
grammarians barely distinguished between
adjectives and nouns, using them inter-
changeably any chance they could.1 The
learned still do, at least when they refer to
themselves as the learns. Those particularly
offended by using an adjectival substantively
(as a noun) should be careful not to slip up
with words like cornea. Both cornea and con-
junctiva started out their ophthalmological
career as adjectives, modifying tunic (coat,
feminine singular), which is why cornea and,
in turn, guttata are feminine singular in the
first place.

What then should we call these excres-
cences on Descemet’s membrane? Well, if you
think they are drops, then by all means call
them drops, although you probably don’t need
to invent yet another term from a dead
language. Vogt called them “tröpflic” (drop-
like) Prominenzen der Hornhautrückfläche
because of the way the cornea guttata (his quotes
not ours).1 If, like Dr Vogt, you want to convey
that the cornea looks speckled with drop-like
excrescences, then you could use the substan-
tive adjectival guttata as a singular noun, or
the English guttate as an adjective. You could
even use guttata as a plural noun—if criticised
before he coined cornea guttata (his quotes,
like) Prominenzen der Hornhautrückfläche
for the whole population.2 Furthermore, the
authors estimated the “incidence” rate using the
“complete” cases, which is a biased sample. Even
then, if the non-response rates were below 30% in this study, the authors did
not provide the precision of the “incident”
rate and attempt to apply a statistical weight-
ing approach to remove or reduce non-
response bias.

Of 179 positive reports collected, only 106
(59%) met their case definition criteria. This
further reduced their number of cases but will
have contributed a percentage inaccuracy.
A further five cases were excluded as non-UK
residents but where did they acquire their
infection—in the United Kingdom or over-
seas?

The incidence figures calculated for Eng-
land and Wales for 1997-8 and 1998-9 of 0.21
and 0.18 per 10 000 contact lens wearers
(CDW) are thus an underestimate of the
expected true incidence rate. The expected
true figure needs to be increased by a factor of
1.493 (100/67) to account for the missed cases
with the low reporting rate used. 67%. The
English figures then become 0.31 (0.21 x 1.493) and 0.27 (0.18 x 1.493) per 10 000 CDW
respectively, of whom the majority (88%)
were soft hydrogel lenses.

These latter higher figures for England
and Wales are remarkably similar to the true
figure estimated at the same time in a 17
month prospective cohort study in Hong Kong3
of 0.33 per 10 000 “soft” hydrogel CLW.
Reporting “questionnaire” studies of infec-
tion 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
containers, avoidance of tap water, and use only of
terile multipurpose solutions or overnight
storage in hydrogen peroxide, then the risk of
infection becomes negligible. The all too
common exposure of daily disposable lens wear has also been virtually
free of acanthamoeba infection.

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Acanthamoeba keratitis
We read with interest the report by Radford et
al on the incidence of acanthamoeba keratitis and the suggested relation
using water hardness of water.3 However, we disagree with their findings and believe their study is flawed for
two reasons.

Firstly, the incidence of acanthamoeba keratitis has been underestimated by approx-
imately 33% since their monthly questionnaire
reporting system yielded an average return of
only 67%. A high non-response rate over 30%
is considered unreliable for making inferences
for the whole population.4 Furthermore, the
authors estimated the “incidence” rate using the “complete” cases, which is a biased sample. Even
then, if the non-response rates were below 30% in this study, the authors did
not provide the precision of the “incident”
rate and attempt to apply a statistical weight-
ing approach to remove or reduce non-
response bias.

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 inter-
val, cases of short duration can cause an under-
estimation 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 had 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 making the epidemiology more
involved.5

In conclusion, the incidence of acan-
thamoeba 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.6

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
containers, 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.

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characterisation of a new corneal pathogen.

Figure 1 References to guttata and guttatae over the past 40 years in Medline. Some articles use both terms.1
Surgery for glaucoma

We would like to thank Drs Mermod and Ravinet for their online comments on the editorial and also for their comments on our article (letter posted on www.bjophthalmol.com 5 Aug 2002). 1

We are delighted that there is agreement that subconjunctival drainage is an important component of non-penetrating filtering surgery. Blebs, albeit diffuse, are clearly visible both in cases of deep sclerectomy, and in many cases of viscocanalostomy. However, although Ravinet 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 viscocanalostomy.

On the second point, although Mermod and Ravinet 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 doing non-penetrating surgery), and also from reports in the literature including randomised prospective trials, macroperforation is associated with early hypotony. 2 This may well be exacerbated if intraoperative antimetabolites, particularly mitomycin, have been used.

With regard to the comments on the paper by O’Brart et al. 3 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.

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References


Ocular and financial health

We read with interest the article by Khandekar et al detailing the results of the 1996–7 Oman Eye Study. 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.1

“Vision 2020—the right to sight” was launched by the WHO and JAPB 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.2 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 42 million blind person years avoided over the next 20 years, and a total saving in excess of $100 billion, by avoidance of lost productivity.3 This would make the effective implementation Vision 2020 not only ethically important but also a cost effective strategy for poverty alleviation.

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2 www.census.gov/ipc/prod/wpg98/wpg98.pdf

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


For those who specialise in the treatment of ocular motility disorders, it seems at times as though time has stood still. Compared with the technical and surgical innovations that strabismology developed and evolved not just in Europe and the United States, but in countries and regions such as Mexico, Japan, South America, and Australia. In thoroughly researched and referenced chapters, the authors describe, time and again, how an ophthalmologist visiting Europe learns a technique, returns to his or her country, refines the technique and applies it to the patient population unique to his or her country. We learn of scoundrels and rogues who foist themselves on the public as miracle workers, only to be publicly exposed and discredited by ethical ophthalmologists of the day (lessons we could stand to learn again in the 21st century). We learn that aesthetic ideals are relative to time and place; such as the fact that in pre-Columbian times a slight degree of esotropia was found to be attractive and convergence was stimulated in infants by attaching a ball of beeswax to the child’s hair to be left dangling between the eyes (vision therapy at its birth).

An often neglected but historically important discipline within the field of strabismology is the practice of orthoptics. In a carefully researched and beautifully illustrated chapter, section author Roger-Ball outlines the origins of the specialty and introduces us to the pioneers of orthoptics—von Graefe, Muller, Donders, and Helmholtz—placed the study and treatment of strabismus at an equal level of importance as the treatment of diseases of the lens, cornea, and retina serves to contemporary vision scientists and ophthalmologists not to neglect this most challenging discipline of ophthalmology—strabismus.

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NOTICES
Role of optometry in Vision 2000

The latest issue of Community Eye Health (No 43) discusses the mobility issues that need 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 Steventons, International Centre for Eye Health, 11–43 Bath Street, London ECI V9EL, 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 Eye ConditionS (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, Zerzabelhofstr 29, 90478 Nürnberg, Germany (tel: +49 911 3913612; fax: +49 911 3913620; email: doc@mcnag.info; web site: www.doc-nuernberg.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: MartinIdi@hotmail.com; web site: www.btp.org.uk). 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 Terezie Pelikanova (Czech Republic) on Pathophysiology of diabetic microvascular complications; Dr Tomas Sosna (Czech Republic) on Risk and protective factors of diabetic retinopathy.

**IOIS—VIIIth International Symposium on Ocular Inflammation**

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

Programme and Organising Committee: Prof. Antonio Secchi, Chairman of IOIS and Convener of Congress (email: ag.secchi@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 Mashehkh, Dr Siamaq Moradian, Dept of Ophthalmology, Labbanfinejad Medical Center, Pasdaran Ave, Boostan 9, Tehran, 16666, Iran (fax: +98 21 254 9039; email: labball@hotmail.com).