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.1

Case report

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

Her best corrected visual acuities were 6/18 in the right eye and 6/12 in the left eye. The left peripheral supratemporal cornea showed three areas of 80% corneal thinning, 1 x 1 mm in size, with an overlying epithelial defect (Fig 1). There was adjacent conjunctival injection and moderate anterior chamber cellular activity. The other eye was normal with no signs of inflammation. Examination of her hands showed no obvious rheumatoid abnormalities. Initial investigations performed were full blood count, electrolytes, C reactive protein, erythrocyte sedimentation rate (ESR), and rheumatoid factor. Corneal scraping and cultures were also undertaken to exclude an infective cause. The only significant test result was a raised ESR of 94, with other tests including creatinine being within the normal range. A provisional diagnosis was made of rheumatoid corneal melt and treatment was commenced with topical dexamethasone 0.1% non-preserved hourly and oral prednisolone at 40 mg/day.

After 5 days of steroid treatment, the corneal thinning was resolving and the conjunctival injection settling (Fig 2). Also by the fifth day, the patient’s hearing had dramatically improved. She could now conduct a conversation at normal volume. On further questioning, she said her hearing had deteriorated only over the past few months and had ascribed it to “natural” old age deterioration. This suggested a link between her corneal changes and hearing loss and she was investigated further. A raised ANCA titre was found for her granulomatosis (WG). Her serum antineutrophil cytoplasmatic 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. Best corrected visual acuity in the left eye remained at 6/12. Examination by an otolaryngologist identified no pathologic or inflammatory conditions. Rheumatological review found only mild arthrophy.

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.1 4 Profound sensorineural deafness is a less common feature, occurring in about 6% of patients on presentation.5 6 It occurs either secondary to inflammation of the cochlear vessels or serious otitis media.7 Other important systemic features include pyrexia, weight loss, recurrent epistaxis, sinus discharge, haemoptysis, perineural neuropathy, cerebral vasculitis, and renal failure (major cause of death). Fortunately in this case, the patient had the limited form of WG, with no renal involvement.

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

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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 chemotherapy 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 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. Fundoscopy showed extensive pigmentary clumping in each eye and macular oedema in the left side (Fig 1). She was commenced on cortisone acetate and fluocortisone to attempt to improve her vision.
Comment

Surgical resection is the treatment of choice for adrenocortical carcinoma. Mitotane (o,p’-DDD) is the only drug that causes regression of metastases and improves survival. Its biochemical action is unknown but data suggest that it modifies peripheral steroid metabolism and directly suppresses the adrenal cortex. The incidence of ocular side effects was 4% in a study of 132 patients. These effects include visual blurring, diplopia, lens opacities, optic neuritis, and a toxic retinopathy 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 5 days after discontinuance of mitotane. In our case, the patient’s visual acuity did improve significantly, with drying out of macular oedema after cessation of therapy and initiation of steroid replacement. It is possible, however, that the ocular changes in this patient were not caused by mitotane but were secondary to cancer associated retinopathy (CAR). Its characteristic findings include attenuated retinal arterioles, with limited, if any, clinically apparent retinal pigmented changes and cells in the vitreous humour. CAR was considered to be unlikely in this case because of the normal calibre of the retinal arterioles, the absence of vitreous cells, and the timing of the onset. Patients with CAR experience visual symptoms that often precede or are concurrent with the tumour diagnosis.

Although primary hypoadrenalinism in Addison’s disease has never been reported to cause retinal problems, secondary hypoadrenalinism in adrenoleucodystrophy is associated with pigmentary retinopathy and other oculomotor 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 visual tract 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 17-hydroxy corticosteroids level but an increase in the levels of 6-beta-hydroxyl cortisol, cholesterol, liver enzymes, corticosteroid binding globulin, and sex hormone binding globulin. At the time of writing, there was no published information on whether mitotane treatment affects serum long chain fatty acid levels. The availability of such data will be valuable for evaluating this postulation.

References


Coxsackievirus B4 associated uveoretinitis in an adult

Coxsackievirus is a kind of enterovirus, which may cause respiratory and gastrointestinal symptoms, erythema, 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, scotopic and photopic retinal 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 general malaise. There was no significant medical or family history. At the initial ophthalmic examination, his best corrected visual acuities were 6/24 in the right eye and 6/24 in the left. Fundus examination showed mottled pigmentation at the macula. One week later her visual acuity was 6/18 in the right eye 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.

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Coxsackievirus B4 infection.

This patient as uveoretinitis induced by respectively, we diagnosed the ocular lesions in demonstrating a 16-fold rise between the acute titre to Coxsackievirus B4 had increased, dem-

scotoma persisted in his right eye. Antibody restored to 25/20 in the right eye and 30/20 in

ophthalmic examination, bilateral exudative decrease. Eight months after the initial

title:1 the lesions detected clinically, and leakage from the retinal vessels correspond-

white lesions in mid-periphery of the retina. The other was an adult case, with chori-

oretinitis similar to the multiple evanescent white dot syndrome. However, the clinical features observed in our patient consisted of iridocyclitis, scattered haemorrhagic dots and haemorrhagic blots are evident.

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 correspond-

ing to the lesions detected clinically, and irregular choroidal background fluorescence.

(Fig 2). Laboratory examination findings were normal except a white blood cell count of 10 500 ×10⁹/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).

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

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

ing a 16-fold rise between the acute (1:8) and convalescent sera (1:128). Retro-

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

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 chorioiditis caused by viruses such as measles, influenza, Epstein-Barr virus, and Rift Valley fever virus typically occur on an acquired basis, subse-

quent to an acute viral systemic illness. Coxsackievirus associated chorioretinitis is thought to elicit the same clinical course as in other viral chorioretinitis. Coxsackievirus infection and concurrent posterior segment inflammation have been reported in a few cases, in which clinical and antibody titres suggested that Coxsackievirus was the responsible agent. Among them, two cases of chorioretinitis associated with Coxsackie-

virus 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 chori-

oretinitis 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 clini-

cal setting, since it is possible that overt clini-

ical manifestations and complications may occur depending on the severity and the loca-

tion of the uveoretinal lesions.

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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 community outreach clinics. One clinic and operating list in Ealing is 17 miles from the central hospital. The surgical teams operate in Ealing and then return to the central hospital. We therefore investigated the use of a telemedicine link for review of postoperative cataract surgical pa-

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 the study and full informed written consent was obtained. A tel-

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

ty. 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.

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

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

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

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

The video display also affects the infor-

mation observed. Broadcast video monitors are accurately colour calibrated, whereas digital compressed video to 384 K bandwidth has
Reduced colour depth. Recognition of corneal oedema is less about well-defined structure as subtle display of colour and "haze."

It is felt by the authors that telemedicine is a wholly different experience from a simple telephone conversation and for many situations is capable of facilitating postoperative assessment with trained operators. H-261 video at 384 K (triple ISDN) is sufficient for anterior segment overall examination but is not useful for a detailed corneal segment assessment. Other practitioners have used data rates up to four times faster for more detailed examination.1

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Table 1 Consistency of scoring by the two observers

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

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Inherited retinal dystrophy and asymmetric axial length

The prenatal and postnatal development of the eye is determined by complex interactions between a number of genes, their products, and certain environmental factors.1-3 Since each eye is influenced by precisely the same processes as its fellow, mutations in regulatory genes usually lead to symmetric phenotypes. In this report, we describe two siblings of Asian ethnicity, born to unrelated parents with no family history of ocular disease, who have an unusual bilateral retinal dystrophy associated with very asymmetrical ocular growth.

Case report

Sib 1 was an 8 year old boy with epilepsy who had been born at term after an uneventful pregnancy. Pendular nystagmus had been noted soon after birth and his visual acuity in each eye was 3/60; refraction –17.00 DS right, plano left. Anterior segment examination was unremarkable with normal intraocular pressures. Examination of the right eye (Fig 1A and B) revealed generalised retinal pigment epithelial and choroidal atrophy, a macula "coloboma," sheathed and occluded retinal vessels inferotemporally, and a shallow 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.

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

Electrophysiology was performed on both children based on the ISCEV standards for adults, but initially using surface electrodes on the lower eyelids. No definite ERG response could be recorded from the emmetropic left eye of either child (even with high intensity stimulation or photopic flicker stimulation with averaging), a finding consistent with severe generalised dysfunction involving rod and cone photoreceptors. The electrical responses of both children’s myopic fellow eye showed a milder degree of dysfunction: there were reduced b:a ratios in both scotopic and photopic responses which, together with the abnormal 30 Hz flicker ERGs, suggest a mid-retinal locus affecting postphototransductional cone and rod systems.1

The skin recordings obtained with surface electrodes were confirmed under anaesthesia using gold foil corneal electrodes. It is reasonable to conclude that these two siblings represent original probands with a novel inherited, probably autosomal recessive, retinal dystrophy. The particularly interesting feature of this disorder is the asymmetry of the axial lengths and ocular volumes of the eyes, rare in genetically determined ocular diseases. Indeed, we are aware of only one other report that identifies ocular asymmetry in association with retinal degeneration. Lafat et al4 reported a single patient who had bilateral Stargardt’s disease and unilateral myopia.

Normally, axial elongation and ocular enlargement are carefully coordinated to equalise growth of fellow eyes in the quest for emmetropia.1 The differences observed in the two siblings described here reflect a decoupling of this developmental synchrony. It has been demonstrated that form deprivation in various species produces progressive axial growth and myopia of the postnatal eye.5 However, in both our sibs the ERG responses were much better preserved in the highly myopic right eye where, in addition, there was a low b:a ratio. Similar findings do not occur in myopia without retinal pathology.6

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

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Figure 1 Colour photographs 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.

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Adenoma of ciliary pigment epithelium: a case series

Adenoma of ciliary pigment epithelium is a rare tumour. Many are diagnosed retrospectively 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 dark brown multinodular mass in the inferotemporal anterior chamber angle of the left eye. His tumour was a relatively small but invasive lesion. Patient 2 was the only non-white patient with this condition in our series. Her tumour was an incidental finding on ophthalmic examination. The salient features of these patients are given in Table 1.

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>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>40/M/W</td>
<td>6/5</td>
<td>8x7x1</td>
<td>Angle invasion</td>
<td>Local resection</td>
<td>No</td>
<td>1992</td>
<td>RD repair</td>
</tr>
<tr>
<td>2</td>
<td>62/F/A</td>
<td>6/6</td>
<td>5x3x1</td>
<td>Angle invasion, cataract</td>
<td>Local resection</td>
<td>No</td>
<td>1996</td>
<td></td>
</tr>
<tr>
<td>3</td>
<td>65/W/Ch</td>
<td>6/18</td>
<td>5x4x4</td>
<td>Sentinel vessel, pigment dispersion</td>
<td>No</td>
<td>1998</td>
<td>Secondary glaucoma, pigment dispersion</td>
<td>RD repair</td>
</tr>
<tr>
<td>4</td>
<td>54/M/W</td>
<td>1/60</td>
<td>5x4x4</td>
<td>Secondary glaucoma, pigment dispersion</td>
<td>Enucleation</td>
<td>1999</td>
<td></td>
<td></td>
</tr>
</tbody>
</table>

M = male, F = female, W = white, A = Asian, RD = retinal detachment, VA = visual acuity.

Figure 1  (A) A small ciliary body adenoma invading the angle in patient 2. (B) Gonioscopic view of the ciliary body adenoma (A) that had invaded the angle and the root of the iris, also showing the localised cataract through the pupil. (C) Ultrasound biomicroscopy of the adenoma of ciliary body shown in (A) and (B). (D) Pigment clumps in the vitreous in patient 3. (E) Shows the adenoma of the ciliary body, the dark iris, and the trabeculectomy site in patient 4.

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.
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, trabecular 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 trabecular retinal detachment.

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

Comment
Our series highlights the paradoxical behaviour of adenoma of ciliary pigment epithelium. Smaller lesions invaded the angle and larger lesions caused extensive pigment dispersion although non-invasively. Angle invasion resulted in these tumours being seen and resulted in the presentation of patients 1 and 2. Blurred vision due to pigment dispersion in the vitreous resulted in the presentation of patient 3. Angle closure glaucoma and pigment dispersion in the vitreous 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 patient 4 (Fig 1D) is unique and has not been reported before. Chang et al. in 1979 reported the presence of pigment in the retrolental 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 secondary 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 the pigment epithelium of ciliary body that has not been emphasised before. Adenoma of the pigment epithelium of 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.

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References


Phenylephrine 2.5% and 10% in phacoanesthesia under phacoemulsification: is there an effect on systemic blood pressure?

Phenylephrine 10% leads to a faster and more pronounced mydriasis but cardiovascular effects like hypertension and arrhythmias have been reported. In a young healthy adult the upper limit of safety for intravenous administration of 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 0–1.720 ng/ml after 20 minutes. They concluded that the mean pressure tended to be higher with the 10% than with the 2.5% concentration.

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.

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

Results
The study included 49 cases aged 52–91, 24 cases were known to have cardiovascular problems and seven were also diabetic. Comparison of the blood pressure changes before
dilatation and in the anaesthetic room preoperatively in the 2.5% group (Fig 1). A mean rise in systolic BP of 22 mm Hg (p=0.003) in the normotensive group. In the hypertensive group all patients recorded statistically significant rises in systolic and diastolic BP (respectively p=0.010, p=0.009). In the 10% group (Fig 2), 10 patients in the normotensive group showed a mean rise in systolic BP of 34.4 mm Hg (p=0.001) and 10.5 mm Hg in diastolic BP (p=0.077). In the hypertensive group six patients recorded a rise in systolic BP with a mean of 22.8 mm Hg (p=0.015) and a diastolic rise with a mean of 16.8 mm Hg (p=0.033) (Table 1) (Figs 3 and 4).

**Comment**

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

**References**

displacement (82%, 9/11) was detected preoperatively than in those in whom displacement was not detected (0%, 0/4) \((p = 0.01, \chi^2\) 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 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\(^1\) 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 with a gap were found.

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.\(^1\)

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.\(^1\) 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.\(^1\) 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 right eye was unremarkable. A scleral nodule in the inferior nasal quadrant was found in the left eye (Fig 1), and a moderate tenderness was determined in this area. The intraocular pressures were within normal ranges. On ophthalmoscopy, the posterior segment of both eyes was normal. The ultrasound images did not indicate posterior scleritis.

The medical history was remarkable. An episode of hepatitis A 20 years ago, and the patient suffered from exercise dependent dyspnocia, night sweats, and repeated headaches.

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

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

Comment

Scleral involvement has been previously reported in a few patients with sarcoidosis. In a series of 537 patients with histologically proved sarcoidosis,\(^1\) 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,\(^2\) 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.\(^1\)

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...
has not been found in our patient, a granuloma-like perivascular cell infiltration was seen, consisting primarily of lymphocytes, some histiocytes, and very few giant cells, which are typical for sarcoidosis. To the best of our knowledge, this is the first histopathological description of a sarcoid lesion in the sclera. The mainstay of treatment for sarcoidosis is corticosteroids, and the scleritis responded in our patient.

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References


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

Diode laser trans-scleral cyclophotocoagulation (TSCPC) is a well accepted method for the management of patients with refractory glaucoma. However, it was reported that the procedure is associated with complications, although less frequent than with other cyclodestructive procedures. Conjunctival burns, corneal decompensation, neurotrophic corneal defects, uveitis, IOP spikes, phthisis (0.5%), chronic hypotony (1%), macular pucker (0.5%), intraocular haemorrhage (0.5%), visual loss, and malignant glaucoma have all been reported.1,2 Possible correlation of outcome with perilimbal pigmentation and the hearing of audible “pop,” especially in patients with dark brown irides,3 has been suggested.

Scleral thinning is a recognised risk factor for complications, but scleral perforation following this procedure has been reported earlier in one patient only.4 This patient had scleral thinning following previous cataract surgery. Possibly, the sharp probe edge had cut conjunctival vessels causing bleeding and contamination of the probe tip. Thin adherent debris was then carbonised allowing the laser tip temperature to rise to 300°C, sufficient to cause scleral perforation. This report led to redesigning of the probe tip to protect the vascular structures from its sharp edges.

Our patient was treated as part of a standard protocol using the modified G probe, and bleeding did not occur. The perforation could have resulted from mechanical pressure, or the effect of the laser on the thinned out sclera, which the patient probably had as a result of buphthalmos and previous ocular surgery.4 There are no known “corrective factors” to utilising two thirds of the scleral thinning, and if treatment is absolutely necessary, a 40% lower energy should be used,5 and areas of thinning and possible wound dehiscence, such as the bleb, should be avoided. We emphasise caution in undertaking contact diode laser trans-scleral cyclophotocoagulation in operated eyes with thin sclera.

Our patient was treated as part of a standard protocol using the modified G probe, and bleeding did not occur. The perforation could have resulted from mechanical pressure, or the effect of the laser on the thinned out sclera, which the patient probably had as a result of buphthalmos and previous ocular surgery.4 There are no known “corrective factors” to utilising two thirds of the scleral thinning, and if treatment is absolutely necessary, a 40% lower energy should be used,5 and areas of thinning and possible wound dehiscence, such as the bleb, should be avoided. We emphasise caution in undertaking contact diode laser trans-scleral cyclophotocoagulation in operated eyes with thin sclera.

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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, and a “fingerprint-like” appearance. 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 endothelial layer showed fingerprint lines, while the endothelial layer had a “hammered” appearance. Iris atrophy, corneoctasia, ectropium euvee 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 striae containing small basophilic deposits between the cells (Fig 2A). The von 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

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structures. The cytoplasm of the adjacent epithelial cells was normal without deposits in relation to the cell membranes (uranyl acetate, lead citrate, bar = 250 nm).

Comment
We consider our case as one of Chandler’s syndrome, which is part of the spectrum of 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. Corneal epithelium, iris atrophy and anomalous iris are less compared to the two other ICE syndromes: that is, the essential iris atrophy and the iris-naevus 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—notably, 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 with no 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 an 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 these 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 microscopic 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 sparking yellow-white spots mainly located in the deeper layer of the posterior pole retina. Since these crystalline deposits tend to become small and decrease in number over time,1 the fundus appearance becomes indistinguishable from other retinal dystrophies with time. The advent of confocal scanning laser ophthalmoscope (SLO) with infrared light has enabled high sensitive examination of the deep layer of the retina.2 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 20/200 left eye. No abnormalities were found in the anterior segment and media. Fine crystalline deposits were not detected in the corneal stroma of either eye. Funduscopic examination revealed numerous fine reflective crystalline deposits throughout the posterior pole and mid-peripheral retina of both eyes. Most of these deposits were in the deep retina and subretina but some deposits were superficial. Goldmann perimetry showed relative scotomas in a zone within 30–40 degrees of the central field in both eyes. Fluorescein angiography was normal in both eyes. 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 limbs of either cornea. Both eyes were aphakic and the posterior lens capsules were intact but relatively opaque. Fundusscopic examination revealed the posterior staphyloma and the extensive atrophy of the retinal pigment epithelium and choroid at the posterior pole, extending from the posterior to mid-peripheral retina in both eyes. Optic disc showed myopic appearance. Retinal vessels and peripheral retina showed normal appearance. Standard electroretinogram was non-recordable. Goldmann perimetry showed marked constriction of all isopters and loss of visibility of 1-4-e isopters for both eyes. During a 5 year follow-up, visual acuity remained stationary. Fluorescein angiography performed at the age of 57 demonstrated hypofluorescence throughout the posterior pole in the early phase and diffuse hyperfluorescence in the later phase. Indocyanine green angiography disclosed the loss of background veil-like fluorescence throughout the posterior pole due to the choriocapillaris atrophy.

Fundus examination by the confocal SLO with infrared light disclosed numerous tiny crystalline deposits in the posterior pole (Fig 2), which were not evident with other fundusoscopic examinations, including ophthalmoscopy and biomicroscopy with a contact lens, or with fluorescein or indocyanine green angiographic studies. The deposits scattered light when the source of light was moved to cause them sparkle, similar to the fine crystalline deposits observed in the fundus of case 1.

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

Comment

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

In case 2, numerous tiny retinal crystalline deposits were detected by SLO with infrared light, although no other examinations disclosed these crystalline deposits. We have not observed such retinal crystalline deposits in other cases with extensive chorioretinal atrophy, suggesting the accumulation of crystalline deposits is not a generalised feature of myopic chorioretinal degeneration. Crystalline deposits, prerequisite for the diagnosis of Bietti’s crystalline retinopathy, are difficult to detect in advanced cases. Since crystalline deposits do not block or transmit fluorescence in fluorescein or indocyanine green angiography, diagnosis of Bietti’s crystalline retinopathy in advanced cases without funduscopically apparent crystalline deposits have been established only when family history was contributory. Because the presence of crystalline material in the peripheral lymphocyte was not observed by transmission electron microscopy, whether case 2 can be categorised as Bietti’s crystalline retinopathy or a distinct clinical entity awaits further investigation. Additionally, to our knowledge, 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 resemblance of the morphology of the tiny crystalline deposits observed by SLO using infrared light between the two cases may support that underlying pathogenesis is similar.
neck discomfort and intermittent jaw claudication over the previous 2 weeks. She had no past medical or ocular problems.

Visual acuity was 20/20 in the right eye and no light perception in the left eye. The right pupil reacted briskly to light, with a normal consensual response, the left was amaurotic. She identified all of the Ishihara pseudoisochromatic colour plates with the right eye, and automated perimetry of the right eye was normal. Extraocular motility was normal. Funduscopy of the right eye (Fig 1A) was normal, and the left optic disc was pale and swollen, with retinal cotton wool infarcts within the retina, chiefly within the left macula (Fig 1B). The superficial temporal arteries were supple, pulsatile, and non-tender. The Westergren erythrocyte sedimentation rate (ESR) was 114 mm in the first hour.

There was no flow detected in the short posterior ciliary arteries of the left eye, and attenuated flow in the short posterior ciliary arteries on the right. Although her ESR fell to 4 mm in the first hour and her jaw claudication resolved, the prednisone dose was maintained because of the attenuated blood flow within the right orbit. Her vision remained 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 bilaterally 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 quantify 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 vein 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 supratrochlear arteries, while blood flow in the third was normal. The authors 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. Elisova 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 vasculature 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 artery, 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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Figure 1  [A] The right optic disc and macula are normal. [B] The left optic disc is pale and swollen. Scattered cotton wool infarcts are seen within the macula. [C] The left optic disc is pale and swollen. Scattered cotton wool infarcts are seen within the retina, chiefly within the left macula. (B) The left optic disc is pale and swollen. Scattered cotton wool infarcts are seen within the macula. (B) The left optic disc is pale and swollen. Scattered cotton wool infarcts are seen within the macula.
Frozen cucumber as a mount for processing vitreoretinal specimens

Vitreoretinal specimens are extremely difficult to process as a frozen specimen 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.

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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 oclusive, 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 periocular discomfort, and pain on ocular motility. Acuity was 20/200 right eye, 20/20 left eye, with a right relative afferent pupillary defect (RAPD). There was swelling of the right optic nerve (Fig 1A) together with increasing non-granulomatous pan uveitis. The left eye was unaffected. Fluorescein angiography (Fig 1B) showed marked optic disc hyperfluorescence and peripheral retinal ischaemia. Seven days later (Fig 1C and D), several enlarging foci of retinitis, typical of ARN, developed in the retinal mid-periphery. PCR of a vitreous biopsy amplified HSV-1. There was no further reduction in visual acuity following intravitreal foscarnet and encephalitic doses (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 and syncytial retinitis preceding the development of ARN (Fig 1E), presented with a 3 day history of progressive right vision with floaters, significant periocular discomfort exacerbated by eye movement. 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 together with increasing non-granulomatous pan uveitis. The left eye was unaffected. Fluorescein angiography showed marked optic disc hyperfluorescence and peripheral retinal ischaemia. Seven days later, several enlarging foci of retinitis, typical of ARN, developed in the retinal mid-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.

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 obstruction in the fellow eye is most likely a manifestation of a different VZV mediated disease process but emphasises the ability of members of the herpesvirus family to directly invade blood vessel walls.

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

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References

Cutaneous angiosarcoma of the eyelids

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

Case report

A 69-year-old white man presented with multifocal red-blue maculopapular lesions located in his right cheek and eyelids (Fig 1A). The patient had first noticed the lesions 9 months previously. An incisional 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 radiation therapy (total dose 45 Gy). After 34 months of follow up there was no evidence of local recurrence or distant metastases.

Histopathologically, the specimens consisted of skin and subcutaneous tissue. The lesions proved to be well differentiated angiosarcoma characterised by interlacing blood vessels lined by plump endothelial cells with hyperchromatic nuclei. Using Ki-67, the proliferation rate was assessed as 5%–7% (Fig 1B). Immunohistochemically, the tumour 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. The majority of these tumours arise in the head and neck, particularly the scalp. They are aggressive and tend to recur locally and disseminate widely with a 5-year survival of approximately 12%–29%. To our knowledge, there are only seven well documented cases involving the eyelid. Clinically, the lesions in our patient resembled ecchymoses, capillary haemangioma, melanoma, and Kaposi's sarcoma. Advanced lesions may ulcerate and atypical cases simulating, cellulitis, rosacea and xanthelasma have also been reported. 4–6 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 interlacing channels lined by atypical endothelial cells and expression of immunohistochemical markers of vascular differentiation (CD34 and FVIII-RA). 7,8 Often tumours display both vasoformative and undifferentiated areas. 9 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). 9 Mitotic index has been observed as an independent histological prognostic marker for cutaneous angiosarcoma although other histological markers remain poorly defined. 10 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. 10 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. 11 As illustrated by our case, the primary lesions were contained within the anterior lamella of the lids and it was possible to surgically encompass the primary lesions avoiding exenteration and without compromising prognosis. In such cases, the surgical aim is to resect all clinically identifiable disease. In areas of doubt, microscopic control of surgical margins may have a role in guiding the extent of resection. Angiosarcomas usually respond to radiotherapy to some degree and most authors suggest that combined surgery and radiotherapy offers the best chance for long term control. 12 In one series of 28 head and neck angiosarcomas, Mark et al. 12 have reported better survival after 32 months (median) with combined surgery and radiotherapy compared with surgery alone. In cases unsuited to surgery, radiotherapy alone may be considered, however usually only partial responses are achieved. 10 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 nature of cutaneous angiosarcoma.

Acknowledgements

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

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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 pain in the right eye. 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 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. Following assessment by her general practitioner she was referred urgently to an eye clinic.

On examination she appeared well and was afebrile. The right eye was chemotic, with an absent red reflex and a large hypopyon (Fig 1). Erythematous maculopapular lesions were present on her arms, legs, and face.

Investigations included a white cell count of 8.0 × 10⁹/l (neutrophils 4.0 × 10⁹/l) and C reactive protein 22 mg/l. Material was collected from the base of the sole 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 intravitreal amikacin and vancomycin administered. Vitrectomy could not be performed because of the severe anterior segment inflammation.

Electron microscopy, immunofluorescence staining and polymerase chain reaction (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.

The patient was treated and remained clinically well during her hospital admission. At discharge after 5 further days of parenteral antibiotics (ceftriaxone) the rash had completely resolved. In the absence of a positive culture sensitivities could not be confirmed, although all the appropriate antibiotics except vancomycin are generally effective against meningococcus. At follow up the eye had become phthisical, with visual acuity reduced to perception of light only.

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-bacteremic patients are very rare, with just four published cases in England. The patients were 13 months, 15 years, 17 years, and 23 years old. Both eldest and youngest had no skin lesions, but C reactive protein was elevated at presentation. The 15 year old had a low grade fever and a generalised macular rash misdiagnosed as scarlet fever. The 17 year old appeared completely “non-septic” with a 4 day history of myalgia, arthralgia, sore throat, and 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 the three older patients vision was reduced to light perception. 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. Hypopyon, observed in this case, is not a feature of primary VZV infection.

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

References

“Guttata were present” may well have grazed them, before being sieved by Romans, since guttata 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 impressive” would be at all offensive, particularly since Flavia herself was named after an adjective, as were Augustus and, some say, even Caesar himself. In fact, early Roman grammarians barely distinguished between adjectives and nouns, using them interchangeably any chance they could. The learned still do, at least when they refer to themselves as the learned. Those particularly offended by an adjective substantively (as a noun) should be careful not to slip up with words like cornea. Both cornea and conjunctiva started out their ophthalmological careers as adjectives, modifying tunica (coat, feminine singular), which is why cornea and, in turn, guttata are feminine singular in the first place.

What then should we call those excrences on Descemet’s membrane? Well, if you think they are foams, then by all means, call them drops, although you probably don’t need to invent yet another term from a dead language. Vögtl called them “tropfige” (drop-like) Prominenzen der Hornhautrückfläche because they are cornea guttata (this question not ours). If, like Dr Vöglt, you want to convey that the cornea looks speckled with drop-like excrences, then you could use the substantive adjective guttata as a singular noun, or the English guttate as an adjective. You could even use guttata as a plural noun—if criticised the English guttate as an adjective “would be at all offensive, particularly number as much as the rest of us. However, it |

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. However, we disagree with their findings and believe their study is flawed for two reasons.

Firstly, the incidence of acanthamoeba keratitis has been underestimeted by approximately 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. 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 weighting 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 to diagnostic accuracy. A further five cases were excluded as non-UK residents but where did they acquire their infection —in the United Kingdom or overseas? The incidence figures calculated for England and Wales for 1997-8 and 1998-9 of 0.21 and 0.18 per 10 000 contact lens wearers (CLW) are thus an underestimate of the expected true infection 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 of 67%. The English figures then become 0.31 (0.21 × 1.493) and 0.27 (0.18 × 1.493) per 10 000 CLW respectively, of whom the majority (88%) wore soft hydrogel lenses.

These latter results indicate that the incidence figures for England and Wales are remarkably similar to the true figure estimated at a 17 month prospective cohort study in Hong Kong1 of 0.33 per 10 000 “soft” hydrogel CLW. Reporting “questionnaire” studies of infectious disease, albeit prospective and of new cases only, usually underestimate the true incidence figure by at least twofold and that used by the authors (BOSU) is no exception. Incidence figures for rare cases are best estimated by performing a prospective cohort study, in which all cases occurring within a community of two million to three million people over a defined period (often a year or more) are examined and tested in several referral hospitals according to defined criteria in advance.

Secondly, the authors suggest that there is a relation between water hardness and acquisition of acanthamoeba keratitis, on the basis of correlation of their questionnaire reports with water quality. They suggest a higher risk in areas of hard water, but such findings are not supported by our results. In the west of Scotland, where there is very soft water (mean value for calcium carbonate 15.22 mg/l), we recorded the highest incidence ever of acanthamoeba keratitis with a 1.49 per 10 000 “soft” hydrogel CLW in 1994-5 in a prospective cohort study.1 In Hong Kong, with an incidence of 0.35 per 10 000 CLW, the water is also soft (mean value for calcium carbonate 40 mg/l).

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 biases in part from evaluating associations between risk factors and a disease well after the risk exposure first occurs. During that time interval, cases of short duration on cases to severity or asymptomatic, and cases in which the presence of disease alters or entirely removes the exposure, are missed.

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

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

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

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References
Surgery for glaucoma

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

We are delighted that there is agreement that subconjunctival drainage is an important component of non-penetrating filtering surgery. Blebs, albeit diffuse, are clearly visible both in cases of deep sclerectomy, and in many cases of viscocanulostomy. However, although Ravinet and Mermoud 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 Mermoud and Ravinet use antimetabolites before cautery to the sclera, they do not apply antime- 

tobiotics and antimetabolites before cautery. They do not do viscocanalostomy.


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References

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References
Ocular and financial health

We read with interest the article by Khadekar 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 along with the under 5 years of age population, which is estimated to see a 6% growth in the same period.

“Vision 2020—the right to sight” was launched by the WHO and IAPB in 1999, aiming for the elimination of avoidable blindness by the year 2020. In 1995 the estimate of global blindness (<3/60 better eye) was 44 million, and this is projected to rise to 76 million by 2020 if there is no change in current trends. 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 save $292 million blind person years avoided over the next 20 years, and a total saving in excess of $100 billion, by avoidance of lost productivity. 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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References
2 www.census.gov/ipc/prod/w98/w98p99.pdf

Ocular Pathology, 5th ed.


The fifth edition of this well established ocular pathology text is excellent. The presentation, the photographs, the histopathology, and micrograph sections provide the reader with an array of outstanding examples of ocular pathology. The text is easy to read, agreeably not always in depth 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 in the history of the treatment of cornelia and retinal diseases, strabismologists have continued to use techniques and instruments devised in the mid-19th century. However, after reading von Noorden’s The History of Strabismology, it becomes clear that the history of our specialty has been characterised by physicians and scientists possessing remarkable creativity and intellect striving to understand how the eyes move and the brain sees.

The History of Strabismology is the ninth volume of The History of Ophthalmology, a planned 21 volume series of monographs intending to comprehensively review the history of ophthalmology from ancient times to the current day. The author has chosen to review the history of strabismology not with a traditional “art through the ages” chronologcal approach, but rather with a geocentric approach. By enlisting prominent strabismologists from around the world as contributing authors, von Noorden allows the reader to discover how the discipline and practice of 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, in detail, to the international audience, how an ophthalmologist visiting Europe learns a technical innovation concerning the aetiology of strabismus. The reader cannot help but see how current concepts and techniques in strabismology flesh out the names we associate with instruments and techniques. Each section author provides numerous references of seminal papers on strabismus published from the corners of the globe. It is refreshing to see a book on history recognise the contributions from those outside the traditional medical centres of Europe/United Kingdom and the United States.

This book will be a valuable reference for all those who specialise in the area of strabismus and those interested in the history of ophthalmology. The illustrations and historical references will greatly enhance the quality of lectures on the topic of strabismus. Knowledge of the origins of critical thought and technical innovation concerning the aetiology and treatment of strabismus will stimulate further interest in today’s students of ophthalmology. Finally, the knowledge that the pioneers of ophthalmology—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 notice to contemporary vision scientists and strabismologists not to neglect this most challenging discipline of ophthalmology—strabismus.

D R Fredrick
San Francisco

Role of optometry in Vision 2000

The latest issue of Community Eye Health (No 43) discusses the mobilisation 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 Medicine, Equipment, Instruments and Optical Supplies (2001) for eye care services in developing countries. It is compiled by the Task Force of the International Agency for the Prevention of Blindness. Further details: Sue Stevens, International Centre for Eye Health, 11–43 Bath Street, London EC1V 9EL, UK (tel: +44 (0)20 7608 6910; email: eyeresource@ucl.ac.uk).

Second Sight

Second Sight, a UK based charity whose aims are to eliminate the backlog of cataract blind in India by the year 2020 and to establish...
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@bhrs.demon.co.uk; web site: www.bhrs.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 seuintl@seuintl.org; web site: www.seuintl.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 3931621; fax:+49 911 3931620; 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: mariti@iheartsol.wmids.nhs.uk; web site: www.bopss.org).

13th Meeting of the EASD Eye Complication Study Group
The 13th Meeting of the EASD Eye Complication Study Group will be held on the 23–25 May 2003, in Prague, Czech Republic. The scientific programme includes keynote lectures from Professor John H Fuller (UK) on The epidemiology of diabetic retinopathy; Dr P Martin van Hagen (The Netherlands) on Growth factors and diabetic retinopathy; Professor Terezia Pelikanova (Czech Republic) on Pathophysiology of diabetic microvascular complications; Dr Tomas Sosna (Czech Republic) on Risk and protective factors of diabetic retinopathy.

Further details: Ortopedické Centrum, s.r.o., Steckovské nabrezi 51, 400 03 Usti nad Labem, Czech Republic (tel: +420 47 521 6588; fax: +420 47 533 40 77; email: orcentrum-ul@volni.cz; web site: www.ortopedicke-centrum.cz).

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

Programme and Organising Committee: Prof. Antonio 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).

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, 72076 Tuebingen, Germany (tel: +49 7071 293209; 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: wulijb@yahoo.com).

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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).

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, 72076 Tuebingen, Germany (tel: +49 7071 293209; 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: wulijb@yahoo.com).
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