Sebaceous carcinoma of the eyelid: a rarity worth considering

The sebaceous carcinoma is a very rare malignant tumour primarily found in the area of the eyelid. Most of these carcinomas originate in the tarsal meibomian glands although they may in extremely rare cases originate in the glands of Zeis of the eyelashes or the sebaceous glands of the caruncle. The periorbital primaries are reported to behave aggressively with the tendency to metastasise early with a significant mortality in most series.

In many cases correct diagnosis of a sebaceous carcinoma of the eyelid is delayed not only as a result of the rarity of this tumour but also because of its ability to masquerade as a variety of other eye conditions such as chalazion or chronic blepharoconjunctivitis. A high index of suspicion is vital if these tumours are to be adequately treated.

We present a case of sebaceous carcinoma and discuss the clinical behaviour, the diagnosis, and the differential diagnosis difficulties as well as the therapy.

Case report

A 78 year old patient presented with a localised inflammation in the centre of the right upper eyelid. Under local antibiotic therapy for 6 weeks the lesion increased in size and showed purulent secretion on the surface. The patient refused further treatment and presented 22 months later with no healing of the lesion on the upper lid and a visible increase in size. The clinical examination revealed an ulcerating tumour measuring 3 cm in diameter at the right upper lid with a bloody encrusted lid margin (Fig 1A) and an enlarged lymph node on the right preauricular area of the parotid gland. The ocular examination revealed no conjunctival or corneal involvement. A biopsy of the eyelid tumour revealed a carcinoma of a meibomian gland with complete infiltration of the upper lid. Conjunctival map biopsies were negative.

Two weeks later a total resection of the upper lid with rapid section examination of the resection margins was carried out as well as the extirpation of the lymph node and an underlying metastasis beneath the parotid gland. Because the rapid section examination from the lateral mucosal border did not show any infiltration with tumour tissue, we decided to do a three layered primary reconstruction of the upper lid with a pedicled temporal island flap covered with a full thickness skin graft from the contralateral upper lid. The reconstruction of the conjunctiva was achieved by an oral mucosal graft. In order to make the raising of the lid possible, the flap was fixed on the fascial residue of the superior levator palpebrae muscle. The histopathological reappraisal revealed tumour stage T3N1 (Fig 2) and a R0 resection.

Two weeks after the surgery a neck dissection was performed and an adjuvant radiation therapy of the right orbit as well as the cervical lymph draining was begun in 25 cycles with a dose of 50 Gy.

A local relapse or further metastasis 15 months after the surgery did not occur (Fig 1B).

Comment

The low incidence and the non-specific clinical symptoms led us to conclude that the diagnosis of a sebaceous carcinoma of the eyelid often occurs very late. One warning sign is the local loss of the eyelashes resulting from a tumour infiltration of the follicle. Every therapy refractive, one sided, tumour-like eyelid lesion has to be clarified histologically to rule out a sebaceous carcinoma. A full thickness eyelid biopsy combined with conjunctival map biopsies is the appropriate approach to the diagnosis. In our case we found the typical pagetoid infiltration of the overlying epidermis at the margins of the widely exulcerated carcinoma. It presented without sign of multicentric growth, which together with pagetoid spread indicates poor prognosis.

The therapy of choice for the sebaceous carcinoma is primarily a surgical one. If the tumour is very large or recurrent with demonstrated spread to bulbar conjunctiva, to the other eyelid, or to orbital tissue, a subtotal or complete exenteration may be necessary. If evidence of spread to regional lymph nodes is present a lymph node or radical neck dissection should be performed, usually combined with partial parotidectomy. An alternative to the surgical excision is the radiation therapy with a total dose of >50 Gy. Radiation therapy may be considered as an adjunct to local surgery. Recurrence of tumour usually occurs within 3 years following radiotherapy alone.

There are some techniques of eyelid reconstruction presently available. An ideal eyelid reconstruction should replace the missing tarsal conjunctival structures and skin, protect the globe, give a natural appearance with only minor donor defect morbidity, and should be done as a one stage procedure. The temporal island flap described in our case report has the advantage of being a one stage procedure, which is relatively easy and quickly done and provides well vascularised tissues. A disadvantage is that the overlying temporal skin of the island flap can be very heavy, which makes...
de-epithelialisation or excision of the relatively thick skin graft necessary.
Postoperative patients must be followed up at short intervals as the tumour has a fast growth potential. The approximate guidelines are 1 monthly interval during the first year, 6 monthly during the second year, and then on a yearly basis for life.7

Alexander Gardetto
Christian Rainer
Department of Plastic and Reconstructive Surgery, University Hospital of Innsbruck and Ludwig Boltzmann-Institute for Quality Control in Plastic Surgery, Austria

Christian Ensinger
Institute of Pathology, University Hospital of Innsbruck, Austria

Ivo Baldissera
Department of Ophthalmology, University Hospital of Innsbruck, Austria

Hildegunde Piza-Katzer
Department of Plastic and Reconstructive Surgery, University Hospital of Innsbruck and Ludwig Boltzmann-Institute for Quality Control in Plastic Surgery, Austria

Correspondence to: Alexander Gardetto, MD, University Hospital of Innsbruck, Department of Plastic and Reconstructive Surgery, Anichstrasse 35, A-6020 Innsbruck, Austria; alexander.gardetto@uibk.ac.at

Accepted for publication 27 April 2001

References

Dramatic regression of conjunctival and corneal acquired melanosis with topical mitomycin C

Conjunctival primary acquired melanosis is the most important precursor of conjunctival malignant melanoma.11 Primary acquired melanosis appears as flat, patchy, non-cystic pigmentation in the conjunctival epithelium and can remain dormant for years or show slow progression.7 Studies have shown that it leads to conjunctival melanoma in approximately 1% to 30% patients.7,14 Treatment of primary acquired melanosis includes observation, excisional biopsy, alcohol epitheliectomy, cryotherapy, and topical chemotherapy.11,14

We illustrate a dramatic case where topical chemotherapy provided complete regression of advanced, aggressive primary acquired melanosis. A 73 year old white man had noted slowly progressive pigmentation on the surface of his right eye for 5 years. A biopsy revealed intraepithelial conjunctival melanosis with atypia. The patient was referred to the Oncology Service at Wills Eye Hospital for evaluation and management. On examination, his visual acuity was 20/20 right eye and 20/40 left eye. Intraocular pressures were 13 mm Hg in both eyes. The interior of both eyes was unremarkable with mild nuclear sclerosis and mild red pigment epithelial mottling. The only abnormality on examination was the presence of patchy, flat, brown conjunctival pigmentation on the right eye for 360 degrees on the bulbar and fornical conjunctiva with involvement of the caruncle, upper tarsal conjunctiva, and entire surface of the cornea (Fig 1). There was no thickening or excessive vascularity to suggest malignant melanoma.

One option for management of this precancerous condition was extensive conjunctival excision and 360 degrees of cryotherapy followed by complete alcohol induced corneal epitheliectomy with anticipation of extensive postoperative scarring, possible symblepharon, and likely conjunctival overgrowth onto the cornea because of the absence of corneal stem cells. The other options included topical chemotherapy with mitomycin C or low dose radiotherapy with a custom designed conformal plaque. Mitomycin C was selected and the patient was treated with a total of 6 weekly cycles of topical mitomycin C 0.04% four times daily interrupted by a week's hiatus of no medication between each cycle.

During the weeks of topical chemotherapy, the patient had occasional symptoms of ocular irritation and periorbital cutaneous erythema and minor oedema. The ocular surface was injected during treatment but, on follow up, there was no chronic ocular surface disease or skin or cilial abnormality.

Topical mitomycin C ophthalmic preparation has been found to be safe and effective for conjunctival squamous neoplasia.10 Its usefulness for conjunctival melanocytic neoplasia is debatable and not yet proved in a large series of patients. A recent study on a small number of patients with conjunctival melanoma and
malignant melanosis indicated a possible role for topical chemotherapy, but the authors advised further investigation. In this regard, we prefer to reserve mitomycin C therapy for patients with biopsy-proven primary acquired melanosis with atypia. An important observation in our case was the effectiveness of topical chemotheraphy for corneal melanosis where other methods of treatment could have led to scarring, conjunctival overgrowth, and ultimate visual loss. We emphasised, however, that patients should be monitored biomicroscopically while taking the medication as overuse could lead to lasting corneal and scleral tissue damage. The recommended duration of treatment has not yet been established in a randomised study. We prefer to limit treatment to two to four cycles with a non-treatment weekly hiatus between cycles to avoid complications. In summary, we report a dramatic case that supports the value of topical mitomycin C for primary acquired melanosis.

Acknowledgments

Support provided by the Eye Tumor Research Foundation, Philadelphia, PA (CS) and the Paul Kayser International Award of Merit in Retina Research, Houston TX (JS).

Carol L Shields, Hakan Demirci, Jerry A Shields, Christopher Spanich

Ocular Oncology Service, Wills Eye Hospital Service, Thomas Jefferson University, Philadelphia, PA, USA

Correspondence to: Carol L Shields, MD, Ocular Oncology Service, Wills Eye Hospital, 900 Walnut Street, Philadelphia, PA 19107, USA

Accepted for publication 15 June 2001

References


Recurrent corneal ulceration as late complication of toxic keratitis

Abuse of topical anaesthetic agents is a rare but potentially dangerous cause of toxic keratitis. Persistent use of topical anaesthetics may result in severe pain, hyperaemia, mucopurulent discharge, chronic epithelial defects, stromal infiltrates with oedema and neovascularisation, and even hypopyon or hyphaema. Permanent corneal scarring and visual loss may lead to penetrating keratoplasty and enucleation. Patients have obtained anaesthetic drops from eye care practitioners and pharmacists, from theft of the practitioners’ offices, and over the counter in some developing countries. \(^*\) We present a case of toxic keratitis secondary to topical anaesthetic abuse that is unique in two aspects: Firstly, the patient received the drops directly from an emergency department physician and, secondly, the patient developed an ulcerative keratitis months after the cornea had re-epithelialised.

Case report

A 24 year old woman presented with complaints of decreased vision and severe eye pain bilaterally. Two weeks before presentation, she had a history of intensely pruritic allergic conjunctivitis. She was examined by a community hospital emergency department physician, who reportedly treated her for a corneal abrasion in the right eye. Two ophthalmologists subsequently treated her for presumed bilateral bacterial keratitis that worsened despite aggressive topical antibiotic therapy, and the patient was referred to the UMDNJ–New Jersey Medical School ophthalmology department. The patient was not a contact lens wearer, and she denied tap or pool water exposure. Past medical history was unremarkable.

On examination, visual acuity was hand movements in both eyes. The eyelids were oedematous and the palpebral and bulbar conjunctiva were severely hyperaemic. The corneas had relatively symmetrical 5 by 7 mm epithelial defects with underlying dense stromal ring infiltrates (Fig 1). The stromas were mildly thinned, and endothelial folds and fine keratitic precipitates were noted. Anterior chamber examination revealed a mild cell and flare reaction, but no hypopyon. Iris, pupils, and lens examinations were unremarkable, and retinal examination revealed normal macula, disc, and blood vessels in both eyes.

Initial differential diagnosis included bacterial, fungal, viral, and Acanthamoeba infection. However, after repeated review of the history, the patient admitted that the initial emergency department physician had given her a bottle of topical anaesthetic and told her “not to tell anyone.” The patient confessed to using the oxybuprocaine (proparacaine) drops every hour in both eyes over the previous 2 weeks. The anaesthetic drops were discarded, and treatment with erythromycin ointment, diclofenac drops, and homatropine 5% was instituted. A bandage soft contact lens reduced the patient’s pain. Corneal cultures, performed to rule out concurrent or subsequent infection, were negative. Over the next week, visual acuity improved to 20/80 in the right eye and 20/40 in the left, though residual bilateral 4 mm epithelial defects with stromal rings remained. The patient was then lost to follow up for 2 months.

Follow up examination revealed a residual epithelial defect in the right eye, and healed epithelium in the left eye. Over the next month, the epithelial defect resolved in the right eye, but bilateral ring-like stromal haze persisted. Four months later the patient presented with a new 2 by 3 mm epithelial defect with underlying stromal infiltrate and neovascularisation in the left eye (Fig 2). The patient vehemently denied any further use of topical anaesthetic drops. Cultures were performed, and treatment with ofloxacin and erythromycin was instituted. The cultures were negative, and the epithelial defect slowly resolved. The underlying corneal stroma remained opacified and neovascularisation progressed. At last examination, visual acuities were 20/40 in the right eye, and counting fingers at 1 foot in the left eye.

Comment

Topical anaesthetic agents are routinely used in ophthalmic examinations and procedures. The most commonly used preparations include oxybuprocaine and amethocaine (tetraacaine). They are topical ester anaesthetics with fast onset of action (10–20 seconds), and relatively short durations of action (10–15 minutes).\(^*\) The agents act by interfering with sodium transmission through neuronal membranes, thereby stabilising the transmembrane potential, inhibiting the generation of action potentials, and blocking the sensation of pain.\(^*\)

The toxic effects of topical anaesthetics are both direct and indirect. Epithelial cell organelles, desmosomes, and cytoskeletal elements can be directly damaged by these agents, altering cellular metabolism and function.\(^*\) Indirectly, loss of epithelial microvilli can cause tear film instability, promoting corneal desiccation and inhibiting re-epithelialisation.\(^*\) Secondary neurotrophic changes may then occur.

Electron microscopy of corneal endothelial cells exposed to topical anaesthetics shows loss of the normal mosaic of hexagonal cells and abnormalities of the intercellular junctions and apical attachments.\(^*\) Immune complexes released from the cornea may be deposited on the epithelium, thereby promoting increased corneal permeability. We present a case of toxic keratitis secondary to topical anaesthetic abuse that is unique in two respects.
Ataxia and vision loss: flow cytometric diagnosis of primary central nervous system lymphoma

Approximately 20% of patients with primary central nervous system lymphoma will have eye involvement, which often precedes diagnosis by a number of months.1 2 The diagnosis of intraocular and/or CNS lymphoma depends on histological tissue obtained from a CNS biopsy or cytological demonstration of malignant cells in the vitreous or cerebrospinal fluid (CSF).3

Cytological differentiation of reactive lymphoid cells from differentiated lymphoma using morphological characteristics depends on observer skill and the preservation of adequate numbers of cells.2 Immunochemistry staining of cell surface antigens assists in this differentiation and can detect cell population monoclonality, a common feature of large B cell lymphomas.4 Cytofluorography or flow cytometry is a semi-automated method of immunocytochemistry which has some advantages over slide based immunocytochemistry including objective and quantitative data on cell surface markers.5

We present a patient with ataxia and vision loss who was diagnosed with primary CNS lymphoma using cytofluorographic analysis of a vitreous biopsy specimen.

Case report

A 53 year old woman was diagnosed with bilateral panuveitis. Best corrected visual acuity (BCVA) was 20/30 right eye and 24/40 left eye. Bilateral mild anterior uveitis and vitritis were present. Diagnostic tests for uveitis were negative. The patient was treated with topical (1%) and oral prednisolone (35 mg) over 16 weeks resulting in a BCVA of 20/30 right eye and 20/25 left eye, resolution of the anterior uveitis, and marked reduction in the vitreous cells bilaterally.

Three months later, the patient presented to the Beth Israel Deaconess Medical Center emergency room with ataxia and vision loss. She had recurrence of her bilateral panuveitis, but no chorioretinal lesions were noted on ophthalmoscopy. A magnetic resonance image (MRI) with contrast demonstrated multiple well defined, enhancing lesions of varying size located at the grey-white matter junction consistent with either metastatic neoplasia or lymphoma (Fig 1). Pars plana vitrectomy was performed and an undiluted and diluted vitreous specimen was sent for cytology. The patient recovered a BCVA of 20/16 right eye and 20/40 left eye.

The diluted vitreous was analysed by flow cytometry using three colour gating (light scatter versus CD45) to optimise lymphoid yield. Approximately 58% of the gated events were composed of abnormal B cells as evidenced by expression of surface antigen CD 19, a pan-B cell marker (Fig 2). The cells showed monoclonal lambda light chain restriction and did not express CD20, CD5, CD10, or CD23 surface antigens (Fig 2). These cytofluorographic findings combined with cytological features of large cell size, moderately abundant amphophilic cytoplasm, and large irregular nuclei with coarse, clumped chromatin and conspicuous nucleoli seen in the undiluted vitreous were consistent with a high grade B cell non-Hodgkin’s lymphoma.

The patient declined chemotherapy and underwent whole brain external beam radiotherapy consisting of a total of 30 Gy administered over 4 weeks and adjunctive intraocular dexamethasone, 1.5 mg twice daily tapered off over 3 weeks. Treatment resulted in a reduction in the size of the cerebral lesions on follow up MRI.

Comment

This case highlights typical features of intraocular lymphoma including uveitis of unknown aetiology presenting before evidence of CNS-NHL, bilateral involvement, and an initial response to steroids.6 7 Decisions regarding the method of diagnosis weigh the lower morbidity and mortality of vitrectomy and lumbar puncture compared to CNS biopsy, against a lower diagnostic yield using cytology.8 Flow cytometry is a semiautomated method of immunocytochemistry and can rapidly identify abnormal B and T lymphocyte subsets. Other benefits include easier cell preparation and multiparameter analyses of specimens.9 Earlier shortcomings including blood contamination, errors introduced by non-viable cells, difficulty in identifying monoclonality, and slow, single cell suspension analysis, have been overcome.

Figure 1 Photograph of MRI with contrast demonstrating multiple well defined, enhancing lesions of varying size located at the grey-white matter junction region involving both frontal lobes (A), and the left parietal lobe (B).

References

However, correlation of flow cytometry analysis with morphological examination of the tissue section or cytology preparation is still required. Although two early reports did not find flow cytometry helpful in diagnosing intraocular lymphoma, three recent cytology studies detected malignancy in 100% of 13 cases with 70% of 10 cases usually with one vitrectomy.

While primary CNS-NHL is initially extremely sensitive to radiotherapy or corticosteroids, tumours will recur in 90% of patients within 1 year. Combined treatment with radiotherapy, corticosteroid and chemotherapy may double median survival time, but the 5 year survival rate is only 4% to 30%. In the minority of patients with this rare disease who have eye involvement, early vitrectomy using cytology and flow cytometry can provide a prompt diagnosis.

Acknowledgments

Dr Arroyo is a recipient of an NIH K-23 physician training award.

None of the authors has any proprietary interest in any of the products in this paper.

Mirela Stancu
Department of Hematology, Beth Israel Deaconess Medical Center, Harvard Medical School, Brookline Avenue, Boston MA, USA

Jorge G Arroyo
Division of Ophthalmology, Beth Israel Deaconess Medical Center, Division of Ophthalmology, Brigham and Women’s Hospital, and the Department of Ophthalmology, Massachusetts Eye and Ear Infirmary, Harvard Medical School, Boston MA, USA

Correspondence to: Jorge G Arroyo, MD, Department of Ophthalmology, Retina Service, 12th Floor, Massachusetts Eye and Ear Infirmary, 243 Charles Street, Boston, MA 02114, USA; jarroyo@caregroup.harvard.edu

Accepted for publication 18 July 2001

References


Intravitreal triamcinolone acetonide for macular oedema due to central retinal vein occlusion

No proved treatment exists for macular oedema secondary to central retinal vein occlusion (CRVO) despite the potential for significant visual loss in affected eyes. We report a patient with bilateral non-ischaemic CRVOs and macular oedema treated with intravitreal triamcinolone acetonide.

An 80 year old woman presented with a 2 week history of acute visual loss in her left eye (LE). She had a 9 month history of poor vision in her right eye (RE). On examination, visual acuity was counting fingers RE and 20/400 LE. Intraocular pressures were 12 and 14 mm Hg, respectively. Anterior segment examination showed bilateral pseudophakia with no evidence of neovascularisation. Dilated fundus examination revealed bilateral CRVOs with macular oedema. Collateral disc vessels were present in the RE. Fluorescin angiography showed diffuse leakage and blockage from intraretinal haemorrhages in both eyes with no areas of non-perfusion (Fig 1). OCT measured a central macular thickness of 775 µm in the RE and 589 µm in the LE (Fig 2A). Owing to the poor visual outcome in the RE with observation, the patient was offered an intravitreal injection of triamcinolone acetonide (Apothecon, Princeton, NJ, USA) in the LE. Topical 0.5% amethocaine (tetracaine, Novartis, Atlanta, GA, USA) was used for anaesthesia. Two drops were applied three times, 5 minutes apart. Betadine (povidone-iodine) 5% (Alcon, Ft Worth, TX, USA) was then applied to the conjunctiva. An injection of 4 mg (0.1 ml) triamcinolone was performed using a sharp 27 gauge, half inch needle through the inferior pars plana, 3.5 mm from the corneal limbus appropriate for her pseudophakic status.

Six weeks post-injection, visual acuity was 20/80 in the LE. Central macular thickness measured 160 µm with a normal foveal contour (Fig 2B). Visual acuity was 20/30 after 3 months. Central macular thickness remained normal by OCT. Six months post-injection, visual acuity declined to 20/400 because of recurrence of macular oedema.
References


Should active CMV retinitis in non-immunocompromised newborn babies be treated?

Cytomegalovirus (CMV) retinitis in children with congenital CMV infection is known to differ from that described in immunocompromised subjects, because it is present at birth and does not progress postnatally if the child is otherwise immunocompetent. Regarding treatment, there are limited data in infants with symptomatic congenital CMV infection and it is not known whether antiviral drugs alter the prognosis. We describe a case of congenital CMV retinitis that progressed postnatally and our treatment regimen.

Case report

A 9 day old baby was referred to the eye clinic after having been diagnosed with congenital syphilitic CMV infection.

During pregnancy, at 34 weeks, poor growth and ventriculomegaly were noted and a scan at 35 weeks showed bilateral cysts in the occipital horn of the lateral ventricles. He was born at 40 weeks by emergency caesarean section. The mother was a healthy 18 year old woman. His birth weight was 2.1 kg and his head circumference 31.5 cm, both of which were below the 0.4th centile.

Brain magnetic resonance imaging confirmed the presence of bilateral occipital cysts and periventricular echodensities.

Electrodiagnostic tests were performed at 4 months and suggested moderate vision in the right eye and poor vision in the left.

On last examination at 5 months the baby would not fixate with the left eye and had developed a divergent squint while he was fixing on bright objects with his right eye. Fundus examination revealed a pale optic disc on the left, vascular attenuation in the left eye, pigmented scars superiorly in both eyes, and right macular gliosis. There was no gross refractive error.

His growth followed a centile just below the 0.4th with delayed development and microcephaly.

Comment

Cytomegalovirus is the most common cause of congenital and perinatal viral infections throughout the world. The incidence of congenital infection ranges from 0.2–2.4% of all live births, with higher rates in populations with a lower standard of living. The fetus may become infected as a consequence of primary and recurrent maternal infection. The risk of fetal infection is greatest with maternal primary CMV infection (40%) and much less likely with recurrent infection (1%). Only 3% of all congenitally infected children have severe disease, another 5% have mild involvement, and 90% are born with subclinical but chronic CMV infection. The most characteristic signs include intrauterine growth retardation, prematurity, hepatosplenomegaly and
jaundice, thrombocytopenia and purpura, and microcephaly and intracranial calcifications. Neurological problems include mild increase in cerebrospinal fluid protein, sensorineural hearing loss, and chorioretinitis. Among the most severely affected infants, mortality may be as high as 40%, and death may occur in the neonatal period or months later. The likelihood that infants who survive symptomatic congenital CMV infection will have normal hearing and intellectual development is small. In infants with subclinical infection, the outlook is much better.

Active CMV infection is best demonstrated by virus isolation from urine, saliva, bronchoalveolar lavages, breast milk, cervical secretions, and tissues obtained with biopsy. The definitive method for diagnosis of congenital CMV infection is isolation of the virus in urine or saliva. This must be done shortly after birth. Intrauterine, perinatal, or early postnatal acquisition of CMV characteristically causes a more chronic infection with regard to virus excretion than does infection acquired later in life. The virus is shed into the urine for approximately 5 years and frequently into the nasopharynx for 2–4 years. The quantity of virus excreted in these sites is much greater than that found in infected older children and adults. Immaturity of the immune response of the fetus and of young infants is believed to be the major determinant of virulence of the congenital and early postnatally acquired infections. Still, the extent and exact nature of the immune defect in congenitally infected infants have yet to be resolved. CMV can also be diagnosed in utero by isolation of the virus in the amniotic fluid.

CMV retinitis is known to occur in immunosuppressed adults and its incidence has increased in the last years as a result of acquired immunodeficiency syndrome (AIDS). It is estimated that the occurrence of CMV retinitis in immunocompromised children is unusual and only few cases have been reported in the literature.

Congenital CMV infection may also be associated with retinitis. The precise incidence of CMV retinitis has been reported to be up to 25% of infants with severely symptomatic congenital CMV and in approximately 1% of infants who are asymptomatic and congenitally infected. The retinitis in congenitally infected children differs from that described in immunocompromised subjects because it usually does not progress after birth if the child is otherwise immunocompetent.

The baby we treated presented to us at 9 days of age with active progressive bilateral CMV retinitis and congenital symptomatic CMV infection. He was not immunosuppressed. Diagnosis was made by the clinical picture and blood and urine cultures, from which the virus was isolated. The optimal treatment for CMV retinitis in children has not yet been established. There are limited data regarding ganciclovir treatment in infants with symptomatic congenital CMV infection and it is not known whether antiviral drugs alter the prognosis. We prescribed the currently recommended therapeutic regimens of ganciclovir which are identical to those used in adults, with drug doses adjusted for body weight. In our case dramatic improvement of the retinitis occurred within 1 week of treatment and ganciclovir was stopped and the child was otherwise immunocompetent. We cannot be sure to what extent the treatment contributed to the resolution of the retinitis as the natural history of the disease is not known. However, despite the rapid resolution of the retinitis, the visual outcome was not ideal. Based on current evidence it would seem appropriate to recommend early treatment of active retinitis. It is important to be aware that active and progressive CMV retinitis can occur in children with symptomatic congenital CMV infection who are not otherwise immunocompromised. The associated retinal vasculitis can result in significant visual impairment.

Urgent assessment and prompt initiation of treatment is recommended as it is likely to improve the visual prognosis by arresting the necrotising retinitis from spreading to the posterior pole, or the contralateral eye. Reporting such rare cases should raise awareness of this serious condition with potentially devastating ocular sequelae. It may in time clarify the spectrum of the disease and help in establishing the best way to manage its complications.

References

reported cases. Nevertheless, even in the pediatric age group, it is important to consider the diagnosis in the presence of a painless, gradually progressive lacrimal gland mass. A published system for evaluation of lacrimal gland masses scores the duration of symptoms, presence of persistent pain or sensory loss, as well as radiological features, in order to determine the appropriate surgical management. The case described scored the maximum 8 points, where a score higher than 3 suggests pleomorphic adenoma requiring excision without previous incisonal biopsy. Complete excision with an intact pseudocapsule has prognostic importance in avoiding later pervasive recurrence and malignant transformation.

Carolyn A Cates, Ruth M Manners Southmead Eye Unit, Southampton SO16 6YD, UK

Geoffrey E Rose Moorfields Eye Hospital, London EC1V 2PD, UK

Correspondence to: Mrs C A Cates; cate@doctors.org.uk

Accepted for publication 15 August 2001

References

Surgical performance for specialties undertaking temporal artery biopsies: who should perform them?

The management of temporal arteritis carries a large burden on resources and commits the patient to long term anti-inflammatory medication with its concomitant side effects. We undertook a retrospective survey of the recent surgical practice at King’s College Hospital, London. This was to compare performance for the specialties involved in biopsying temporal arteries. We are unaware of any previous publication specifically addressing surgical efficiency. Biopsies of long specimens could help reduce morbidity and mortality associated with false negative results and avoid further unnecessary and expensive investigation.

Information was obtained by cross referencing clinical, surgical, and histopathological records for all the temporal artery biopsies done during a 2 year period from March 1998 to March 2000. Arterial specimen length was taken as the performance indicator. Analysis of the biopsy rates for years 1 and 2 was done and then individual lengths were compared.

Table 1 Annual biopsy rates per department

<table>
<thead>
<tr>
<th>Year</th>
<th>Year 2</th>
<th>Total</th>
<th>Percentage</th>
</tr>
</thead>
<tbody>
<tr>
<td>Ophthalmology</td>
<td>11</td>
<td>14</td>
<td>34.15</td>
</tr>
<tr>
<td>Neurosurgery</td>
<td>9</td>
<td>14</td>
<td>35.15</td>
</tr>
<tr>
<td>Maxillofacial</td>
<td>6</td>
<td>16.1</td>
<td></td>
</tr>
<tr>
<td>General surgery</td>
<td>4</td>
<td>5</td>
<td>12.2</td>
</tr>
<tr>
<td>Vascular surgery</td>
<td>2</td>
<td>4.9</td>
<td></td>
</tr>
<tr>
<td>Total</td>
<td>27</td>
<td>41</td>
<td></td>
</tr>
</tbody>
</table>

Results
Thirty nine patients underwent 41 biopsies, yielding an average of 19.5 per year or 1.6 per month. Of these, six were positive. This implies a 14.6% positive biopsy rate. Of the two re-biopsies, one unveiled a repeat negative result and the other a positive result. The five departments performing surgery were: ophthalmology—14 biopsies (34%) from 13 patients; neurosurgery—14 biopsies (34%); maxillofacial surgery (MFS)—six biopsies (14.6%) from five patients; general surgery (GS)—five biopsies (12.2%); and vascular surgery (VS)—two biopsies (4.9%). For analysis MFS, GS, and VS are grouped together as they performed relatively few biopsies. Analysis of biopsy rates is depicted in Table 1. Overall, 14 biopsies were done in year 1, with an average length of 13.8 mm, and 27 in year 2, with an average of 13.1 mm. Regarding specimen lengths, ophthalmology averaged 15 mm in year 1 and 16.4 mm in year 2. Total average was 16.1 mm. This ranged from 5 mm to 30 mm. The 5 mm specimen was deemed too short and re-biopsy yielded a specimen of 23 mm. There were no failed biopsies. Neurosurgeons averaged 10.8 mm in year 1 and 9 mm in year 2, with a total average of 9.6 mm. Specimens ranged from no artery to 20 mm. No re-biopsy of the unsuccessful sample took place. The last group averaged 15.7 mm in year 1 with one specimen containing no artery and 13.1 mm in year 2, also with one unsuccessful re-biopsy procedure. Total average length was 14.3 mm with a range of no artery to 28 mm. The average length of the 35 negative results was 12.9 mm.

Comment
It would appear that our ophthalmology unit is best suited to perform the majority of biopsies. They already perform one third of all biopsies and achieve the longest average length. They also re-biopsy when specimen length is best suited to perform the majority of biopsies. This would increase our diagnostic efficiency and reduce the impact that temporal arteritis has on our resources.

Gavin D Galloway
Ophthalmology Department, West Suffolk Hospital, Bury St Edmunds, IP33 9ZQ, UK

Bernhard Klebe
Department of Internal Medicine, Kent and Canterbury Hospital, Canterbury CT1 3NG, UK

Paul Riordan-Eva
Department of Ophthalmology, King’s College Hospital, London SE5 9RS, UK

Correspondence to: Mr Gavin Galloway; ggvgl@dans.org.uk

Accepted for publication 15 August 2001

References

Table 2 Performance indicator—average arterial length (mm)

<table>
<thead>
<tr>
<th>Year</th>
<th>Year 2</th>
<th>Total</th>
<th>Range</th>
</tr>
</thead>
<tbody>
<tr>
<td>Ophthalmology</td>
<td>16</td>
<td>16.4</td>
<td>16.1</td>
</tr>
<tr>
<td>Neurosurgery</td>
<td>9</td>
<td>9</td>
<td>9.6</td>
</tr>
<tr>
<td>Maxillofacial</td>
<td>16</td>
<td>13.2</td>
<td>no artery–20</td>
</tr>
<tr>
<td>General surgery</td>
<td>11</td>
<td>12.4</td>
<td>no artery–28</td>
</tr>
<tr>
<td>Vascular surgery</td>
<td>22.5</td>
<td>22.3</td>
<td>no artery–30</td>
</tr>
<tr>
<td>Total</td>
<td>13.8</td>
<td>13.1</td>
<td>13.3</td>
</tr>
</tbody>
</table>

www.bjophthalmol.com

For quite some time there has been a plethora of problem oriented approach learning texts available in medicine. They are liked enormously because of their practical approach, often excellent illustrations, readability, and because they can be a great text for quick reference and, indispensably, revision for membership examinations. Ophthalmology has also earned similar textbook approaches except that they have been more didactic and nearly always skewed to surgical conditions. John Ferris is to be congratulated in providing a first class book, which I am sure, is eagerly awaited.

I can imagine the book could become essential for the ophthalmology exams as well as being very useful for the MRCP short cases. The book bravely, and in the main successfully, covers the broad spectrum of medical ophthalmology, including neurology. What is exciting is that he brings in systemic features—for example, the hands and face, into ophthalmic learning, further emphasising that ophthalmologists need to look outside the eye. I particularly like the chapters on laboratory tests and radiology, essential for problem oriented approach to learning.

In general, the illustrations are good and the text apt for this stage in learning, although by no means complete, as I am sure each subspecialist may argue unnecessarily. Nevertheless, for exam purposes, I find that this book fulfils its requirements and I would recommend it to all trainees for their exams.

Andrew D Dick
University of Bristol

BOOK REVIEW

International Centre for Eye Health

The International Centre for Eye Health has published a new edition of the Standard List of Medicines, Equipment, Instruments and Optical Supplies (2001) for eye care services in developing countries. It is compiled by the Task Force of the International Agency for the Prevention of Blindness. Further details: Sue Stevens, International Centre for Eye Health, 11–43 Bath Street, London EC IV 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 blindness in India by the year 2020 and to establish strong links between Indian and British ophthalmologists, is regularly sending volunteer surgeons to India. Details can be found at the charity website (www.secondsight.org.uk) or by contacting Dr Lucy Mathen (lucymathen@yahoo.com).

Specific Eye Conditions (SPECS)

Specific Eye Conditions (SPECS) is a not for profit organisation which acts as an umbrella organisation for support groups of any condition or syndrome with an integral eye disorder. SPECS represents over fifty different organisations related to eye disorders ranging from conditions that are relatively common to very rare syndromes. We also include groups who offer support of a more general nature to visually impaired and blind people. Support groups meet regularly in the Boardroom at Moorfields Eye Hospital to offer support to each other, share experiences and explore new ways of working together. The website www.eyeconditions.org.uk acts as a portal giving direct access to support groups own sites. The SPECS web page is a valuable resource for professionals and may also be of interest to people with a visual impairment or who are blind. For further details about SPECS contact: Kay Parkinson, SPECS Development Officer (tel: +44 (0)1803 254 238; email: k@eyeconditions.org.uk; website: www.eyeconditions.org.uk).

EUPO 2002 Course Retina

A course on retina will be held on 15–17 March 2002 at Erlangen, Germany, where European professors will teach European residents. Further details: Priv Doz Dr Ulrich Schonherr, Friedrich-Alexander-University of Erlangen-Nuernberg, Department of Ophthalmology, Schwabachanlage 6 (Kopfkinikum), D-91054 Erlangen, Germany (tel: +49 9131 853 4379; fax: +49 9131 853 4332; email: ulrich-schoenherr@augen.imed.uni-erlangen.de).

XXIXth International Congress of Ophthalmology

The XXIXth International Congress of Ophthalmology will be held on 21–25 April 2002 in Sydney, Australia. Further details: Congress Secretariat, C/- ICMS Australia Pty Ltd, GPO Box 2609, Sydney, NSW 2001, Australia (tel: +61 2 9241 1478; fax: +61 2 9251 3552; email: ophthalm@icmsaust.com.au; website: www.opthalmology.aust.com).

12th Meeting of the European Association for the Study of Diabetic Eye Complications (EASDEC)

The 12th meeting of the EASDEC will be held on 24–26 May 2002 in Udine, Italy. The deadline for abstracts is 15 February 2002. Three travel grants for young members (less than 35 years of age at the time of the meeting) are available. For information on the travel grants, please contact Pr CD Agardh, President of EASDEC, Malmö University Hospital, SE-205 02 Malmö, Sweden (tel +46 40 33 10 16; fax: +46 40 33 73 66; email: carl-david.agardh@endo.mas.lu.se). Further details: NORD EST CONGRESSII, Via Aquilea, 21–33100 Udine, Italy (tel: +39 0432 21391; fax: +39 0432 50687; email: nordest.congressii@ud.netuno.it).

International Society for Behçet's Disease

The 10th International Congress on Behçet's Disease will be held in Berlin 27–29 June 2002. Further details: Professor Ch Zouboulis (email: zoubbere@zedat.fu-berlin.de).

Singapore National Eye Centre 5th International Meeting

The Singapore National Eye Centre 5th International Meeting will be held on 3–5 August 2002 in Singapore. Further details: Ms Amy Lim, Organising Secretariat, Singapore National Eye Centre, 13 Third Hospital Avenue, Singapore 168751 (tel: (65) 322 8374; fax: (65) 227 7290; email: amy_lim@snec.com.sg).

BEAVRS Meeting

The next BEAVRS meeting will be held in the Dalmahoy Hotel near Edinburgh on 31 October to 1 November 2002. Further details: Susan Campbell, Medical Secretary, Gartnavel General Hospital (email: susan.j.campbell@northglasgow.scot.nhs.uk).

NOTICES

Glucoma

The latest issue of Community Eye Health (No 39) discusses the glaucomas, with an editorial by Professor Gordon J Johnson, director of the International Centre for Eye Health. For further information please contact: Journal of Community Eye Health, International Centre for Eye Health, Institute of Ophthalmology, 11–43 Bath Street, London EC IV 9EL, UK (tel: +44 (0)20 7608 6910; fax: +44 (0)20 7250 3207; email: eyeresource@ucl.ac.uk; website: www.jech.co.uk). Annual subscription (4 issues) £25.50/US$40. Free to workers in developing countries.

www.bjophthalmol.com
Recurrent corneal ulceration as late complication of toxic keratitis

Barry N Wasserman and Robert P Liss

Br J Ophthalmol 2002 86: 245-246
doi: 10.1136/bjo.86.2.245

Updated information and services can be found at:
http://bjo.bmj.com/content/86/2/245

These include:

References
This article cites 6 articles, 2 of which you can access for free at:
http://bjo.bmj.com/content/86/2/245#BIBL

Email alerting service
Receive free email alerts when new articles cite this article. Sign up in the box at the top right corner of the online article.

Errata
An erratum has been published regarding this article. Please see next page or:
/content/86/4/487.2.full.pdf

Notes

To request permissions go to:
http://group.bmj.com/group/rights-licensing/permissions

To order reprints go to:
http://journals.bmj.com/cgi/reprintform

To subscribe to BMJ go to:
http://group.bmj.com/subscribe/
TREATMENT

by minimal amniotic fluid. Fetus B was alive with several anomalies including bilateral infraorbital cystic masses, protruding tongue, and large median facial cleft. Amniocentesis revealed a 46XY karyotype without numerical or structural chromosomal abnormalities. The parents denied any significant medical history, family history, of congenital anomalies, or use of drugs or alcohol. Their twin other children, ages 17 and 33 months, were normal. No family members exhibited features of TCS on visual examination.

Necropsy measurements were consistent with 16 weeks of gestational age for twin A and 21 weeks for twin B. Both twins had bilaterally hypoplastic zygomas, maxillae, and related muscles; severely hypoplastic and misplaced pinnae; bilaterally agenetic external auditory canals; bilateral lateral facial clefts, and severe micrognathia (Fig 1). Twin A exhibited left sided choanal atresia, agenesis of the hard and soft palate, and multiple visceral anomalies, including dual superior vena cava, bilobed right and unilobed left lungs, bilateral renal and urogenital agenesis, rudimentary urinary bladder, and absent epididymides. Twin B was noted to have right sided choanal atresia, soft palate aplasia, hard palate hypoplasia, and a left sided cleft; twin B had no visceral anomalies. Ocular pathologica l dissection of twin A illustrated bilateral microphthalmia; corneal scleralisation; and maldevelopment of the uvea, lens, and retina. Ocular dissection of twin B revealed microphthalmia, aniridia, congenital cataracts, and bilateral vascularisation of the corneas.

Comment

These twins had multiple features characteristic of TCS, most notably hypoplastic zygomas, maxillae, and related muscles—perhaps the most characteristic features of TCS. In contrast, visceral anomalies, such as those of twin A, are rare. Only two cases have been previously reported: one with tracheoesophageal fistula, rectovaginal fistula, and anal atresia; another with achalasia.6 Renal agen esis, found in twin A, has not been previously described in association with TCS. While ophthalmological features in TCS are often extensive, they seldom involve the intraocular structures. Common findings include a defective inferior lateral angle of the orbit, caudal displacement of the superolateral orbit, true and pseudocolumbomas of the lids, lateral canthal dystopia, orbital lipodermoids, corneoscleral dermoids, and microphthalmos.4 Cataracts, lacrimal duct atresia, pupillary ectopia, distichiasis, and uveal colobomas have been reported less commonly. Ocular involvement in these twins is rare in TCS. Furthermore, aniridia, corneal scleralisation, and uvea, lens, and retinal maldevelopment are previously unreported.

TCS is an autosomal dominant disorder affecting one in 50 000 live births. The disorder appears to have arisen in these twins with no relevant family history, as occurs in 60% of cases.5 Expressivity is highly variable, ranging from clinically apparent perinatal death secondary to airway compromise;4 the disease severity that resulted in the fetal death of twin A is highly unusual. The responsible gene, TCOF1, has been mapped to 5q32–33.2 and the structure of its protein product, treacle, elucidated.4 To date, 51 disease causing mutations have been identified, nearly all resulting in a premature termination codon.5 Significantly, no relationship has been found between any single mutation and phenotype severity. The ophthalmic pathology observed in these twins may have resulted from a focal TCOF1 mutation and a yet to be defined role treacle may have in eye (and renal) development. Considering the number and severity of abnormalities, the failure to identify a single “genetic hot spot,” and the novel ophthalmic features, an alternative and perhaps more likely explanation is that a second gene, itself involved in ophthalmic embryology, was affected along with TCOF1. This unidentified gene may have been disrupted from a translocation involving 5q32–33.2 or from a deletion large enough to result in a contiguous gene syndrome. A final consideration is that these twins may not have had TCS but rather a new though closely related syndrome. Further genetic investigation may shed light upon these speculations.

Acknowledgements

JLP and GB contributed equally to this work.

J L Prenner, G Binenbaum, D F Carpentieri, S M Goldstein, R S Douglas, E Ruchelli, J A Katowitz
Scheie Eye Institute, Department of Ophthalmology, University of Pennsylvania; Departments of Pediatric Ophthalmology and Pathology, Children’s Hospital of Philadelphia, Philadelphia, Pennsylvania, USA

R W Hertle
The Laboratory of Sensorimotor Research, The National Eye Institute, The National Institutes of Health; Bethesda, Maryland, USA

Correspondence to: J L Prenner, MD, Department of Ophthalmology, Scheie Eye Institute, University of Pennsylvania, 51 North 39th Street, Philadelphia, PA 19106, USA; jlprenner@excite.com

Accepted for publication 7 September 2001

References


Acute postural drop in optic nerve perfusion after vitrectomy and gas in a patient with diabetic autonomic neuropathy

We report the case of an insulin-dependent diabetic who suffered transient blindness as a result of a change of posture following vitrectomy surgery with injection of gas.

This 34 year old woman with longstanding diabetic disease, postural hypotension, and proliferative retinopathy had undergone argon laser photocoagulation (PRP) to both eyes. Her right eye was amblyopic and had visual acuity of counting fingers (CF), whereas the left eye, at best, had vision of 6/12 that was frequently impaired by recurrent vitreous haemorrhage. She underwent left vitrectomy and required 30% sulphur hexafluoride (SF6) gas to support the incidental, small, inferior detachment related to a round hole. The patient postured overnight and remained under the care of the physicians.

The following day her vision dropped suddenly and severely on her way to the eye clinic. Visual acuity was no perception of light (NPL) in the operated eye and CF in the right eye. The left pupil was already dilated and anterior segment examination was satisfactory. The view of the fundus was limited owing to the presence of gas and residual haemorrhage but the retina was flat. The intraocular pressure (IOP) was 42 mm Hg and the vision improved to light perception. Her blood pressure improved in the supine position to 150/80. Following removal of gas the IOP fell to 20 mm Hg and the vision improved to counting fingers. Four weeks later, with disappearance of the gas her best corrected vision had improved to 6/18 and the retina remained flat.

Comment

Judging by the severity of the visual loss we believe that it was due to obstruction of both the central retinal artery and optic nerve perfusion. The precipitating factor in this case would appear to have been the change in posture from supine to sitting. The presence of gas in the operated eye had raised the intraocular pressure to a level such that the perfusion pressure to the eye was effectively eliminated resulting in a period of no light perception.

Restoring the patient to the supine position was sufficient alone to reverse the condition and improve the perfusion of the eye. When the IOP was reduced to normal she was out of this critical situation and was able to maintain the ocular perfusion, even when sitting.

Collapse of the central retinal artery is known to occur in conditions of high IOP but vision does not fluctuate with posture. However, it is known that raised IOP also reduces orthograde axonal transport and compromises nerve head perfusion. Postural hypotension in diabetics is secondary to autonomic neuropathy. The pathophysiology is not clear but it seems the main factor is blunted catecholamine response to standing and failure of the lower limb vascular resistance to increase adequately. Systemic hypotension can cause a drop in optic nerve blood flow as illustrated by numerous reports in the literature of non-arteritic anterior ischaemic optic neuropathy (NA-AION) occurring following acute blood loss. Patients with NA-AION have a significantly greater postural pressure change in IOP compared with healthy subjects as shown by James and Smith. In the same study they demonstrated a higher pulsatile ocular blood flow (POBF) on standing compared with supine. In this patient with autonomic neuropathy it is likely that the normal increase in POBF on standing was inadequate.

We advise particular caution in diabetics undergoing ocular surgery in which there may be a significant postoperative pressure rise. Similar problems can occur during surgery if hypotensive anaesthesia is either deliberately or inadvertently employed in diabetics. Beware of assuming poor postoperative vision to be purely the result of the presence of haemorrhage or gas; patients may have quite poor vision immediately after vitreoretinal surgery but will rarely have NPL unless there is also optic nerve compromise. We also suggest one should aim for a lower postoperative IOP in diabetics.

A Al Khaier, J C Khan, R L Burton
Department of Ophthalmology, The West Norwich Hospital, Bawtare Road, Norwich NR2 3TQ, UK

Correspondence to: Mr Burton; ted.burton@norfolk.norwich.nhs.com

Accepted for publication 12 September 2001

References


Optical coherence tomography imaging of severe commotio retinae and associated macular hole

Commotio retinae results in retinal opacification following blunt trauma. Mild commotio retinae usually settles spontaneously with minimal sequelae but more severe cases are associated with visual loss. We are not aware of any previous reports describing optical coherence tomography (OCT) imaging of severe commotio retinae with an associated full thickness macular hole (FTMH).

Case report

A 15 year old boy presented 24 hours after blunt trauma from a football striking his right eye. On examination his best corrected visual acuity was counting fingers right eye and 6/6 left. Biomicroscopic examination revealed extensive commotio retinae over the posterior pole, no posterior vitreous detachment (PVD), and a FTMH. Colour photography and OCT imaging (OCT 2000 scanner, Zeiss-Humphrey) were performed (Fig 1). OCT confirms a FTMH and demonstrates extensive disruption of photoreceptor outer segments and retinal pigment epithelium (RPE).

Figure 1 (A) Right macula of 15 year old boy with extensive commotio retinae over posterior pole and an associated macular hole at 1 day after blunt injury. (B) Horizontal OCT scan through centre of macula confirms a full thickness macular hole and demonstrates extensive disruption of photoreceptor outer segment/retinal pigment epithelium layer. The optic disc is seen at the nasal edge of the scan.
He was treated conservatively with a short course of topical steroids. The colour fundus and OCT appearance at 1 month are shown in Figure 2. Despite spontaneous macular hole closure, visual acuity remained at counting fingers at 1 year follow up.

**Comment**

The major site of retinal trauma appeared on OCT to be at the level of the photoreceptor outer segment/RPE interface. The OCT images are consistent with fragmentation of photoreceptor outer segments and damaged cell bodies, as suggested by Sipperley et al in their study of the histological changes in commotio retinae in primates.

The exact pathogenesis of macular holes remains uncertain. Ho et al outlined the three basic historical theories regarding aetiology—the traumatic theory, the cystic degeneration and vascular theory, and the vitreous theory. Of these, the latter has gathered the most support in the context of idiopathic macular holes.

In our case, the OCT imaging reveals that the edges of the macular hole are elliptical and irregular with no associated PVD, cortical vitreous condensation, or overlying prefoveal opacity. The characteristics suggest a different mechanism of hole formation from that proposed in idiopathic senile macular holes. We believe that mechanical distortion of the retina, relative to the vitreous and underlying sclera, created disruption of the photoreceptor outer segment level that the retina has the least support from Müller cells and is therefore likely to undergo greatest deformation.

In the only previous report of OCT imaging in traumatic macular hole, a case with mild commotio retinae was described in which extensive outer retinal disruption was not observed. There have been some encouraging reports suggesting that vitrectomy can successfully close traumatic macular holes as well as improve visual function in many cases. However, it seems unlikely that cases with severe commotio retinae, and associated photoreceptor/RPE damage, as demonstrated in our cases, would gain any benefit from surgical as opposed to spontaneous closure of a traumatic FTMH. The final visual prognosis is severely limited by the extent of initial photoreceptor damage, and the excessive pigment atrophy and clumping that follows. We believe OCT imaging provides additional information both on the pathogenesis of commotio retinae and in the assessment of outer retina disruption following ocular trauma. This information may help in the selection of patients likely to benefit from surgical intervention.

**References**


**Acute ocular ischaemia and orbital inflammation associated with systemic lupus erythematosus**

We report a patient with systemic lupus erythematosus (SLE) who developed bilateral ocular ischaemic syndrome in association with orbital inflammation leading to devastating visual loss.

**Case report**

A 73 year old white woman presented with unilateral acute anterior uveitis and polyarthritis of the hands, knees, and neck. Over the next 4 years she suffered recurrent anterior uveitis, marginal keratitis, and episcleritis, which were treated with topical steroids. She had bilateral age related cataract and underwent left cataract surgery. Postoperatively, her visual acuities were 6/60 right eye and 6/9 left eye.

She was admitted 7 months later with extensive erythematous, scaly plaques on sun exposed areas and a purpuric eruption on the abdomen, buttocks, and legs. Biopsy of a plaque showed changes consistent with SLE including follicular keratin plugging, epidermal atrophy, degeneration of the basal layer with collagen body formation, basement membrane thickening, myxoid change in the dermis, and peripapillary lymphocyte infiltrates. Biopsy of the purpuric eruption was consistent with a leucocytoclastic vasculitis. Further investigations revealed positive rheumatoid factor (1/100), ANA (1/25), pANCA (1/100), and anti-EN antibodies (1/100). There was reduced C3 and C4 polyclonal increase in IgA and IgM, lymphopenia, thrombocytopenia, and reduced creatinine clearance but no proteinuria. Anticardiolipin antibodies were negative. A diagnosis of SLE was made and oral prednisolone (0.8 mg/kg) was commenced.

Three weeks later she developed a rapid onset of marked bilateral conjunctival and lid oedema, subconjunctival haemorrhage, and proptosis. Visual acuity deteriorated to perception of light in each eye. There was gross restriction of ocular movements. Signs of ocular ischaemia developed in both eyes with corneal oedema, iris neovascularisation, fibrinous anterior uveitis, hyphaema, and raised intraocular pressure (Fig 1). Computed tomography and B-scan ultrasonography of the orbits showed posterior scleritis, choroidal detachment, vitreous haemorrhage, and thickening of the extraocular muscles (Fig 2).

She was treated with topical dexamethasone 0.1% and cortecol 1%, oral acetazolamide 500 mg daily, intravenous cyclophosphamide (10 mg/kg) and subsequently intravenous immunoglobulin (0.5 g/kg/day for 5 days). Over the following month there was regression of the proptosis, conjunctival and lid oedema, corneal oedema, and fibrinous uveitis. The intraocular pressure was controlled and the visual acuity remained perception of light in each eye. Subsequently, the patient developed pneumonia, VIIth nerve palsy, left sided hemiparesis, and died 7 weeks after admission. Postmortem examination revealed bronchopneumonia, lung abscess, pleural effusions, fibrinous pericarditis, cardio-megaly, and jejunal mucosal haemorrhages.

**Figure 1** Anterior segment of right eye showing conjunctival injection, corneal oedema, hyphaema, rubecois iridis, and cataract.

**Figure 2** B-scan ultrasound of left eye showing marked posterior scleritis (crosses) and choroidal detachment (arrow).
The family did not consent to histopathological examination of the eye.

Comment
Acute orbital inflammation and orbital myositis leading to proptosis are rare manifestations of SLE. To our knowledge acute ocular ischaemic syndrome with orbital inflammation is a novel complication of SLE. The ophthalmic manifestations may have resulted from a vasculitis involving the vessels supplying the globe and extraocular muscles. The limitation of ocular movements was most probably the result of mechanical restriction. Although the proptosis, lid and conjunctival oedema improved with immunosuppression, the visual acuity did not recover. This may have been because of irreversible visual loss from optic nerve dysfunction due to ischaemia, compression from the acute orbital inflammation, or secondary to raised intraocular pressure. Close cooperation between ophthalmologist and rheumatologist is essential in the management of these patients, in order to try and prevent blindness.

P Stavrou, P I Murray
Birmingham and Midland Eye Centre, City Hospital NHS Trust, Birmingham, UK
K Batta
Birmingham Skin Centre, City Hospital NHS Trust, Birmingham, UK
C Gordon
Department of Rheumatology, City Hospital NHS Trust, Birmingham, UK

Correspondence to: Professor P I Murray, Academic Unit of Ophthalmology, Division of Immunology and Infection, Birmingham and Midland Eye Centre, City Hospital NHS Trust, Birmingham B18 7QY, UK. P.I.Murray@bham.ac.uk

Accepted for publication 10 October 2001

References

Chickenpox neuroretinitis in a 9 year old child

Chickenpox in children is usually thought of as a benign infectious disease with few ocular complications. Posterior segment involvement from primary varicella zoster infection has rarely been reported in children. We describe the clinical features and visual outcome of an unusual case of neuroretinitis presenting in a 9 year old child.

Case report
An immunocompetent 9 year old boy acquired primary varicella zoster virus (VZV) infection from his sibling and developed the characteristic exanthematous vesicular rash. Four days after the onset of the rash he woke with discomfort in his right eye and described his vision as being “all grey” on that side. He presented to the emergency department the same day and was found to have a visual acuity of 3/6 on the right and 3/3 on the left (logMAR). A relative afferent pupillary defect (RAPD) was present on the right. His anterior segment was quiet with no vitritis; however, he had slight macular thickening and a subtle cherry red spot on funduscopv, along with some mild peripapillary swelling and disc haemorrhage.

On review in the ophthalmology clinic 2 days later his vision had reduced to 1/60 (Sheridan Gardiner singles) on the right. He had no new skin lesions and all those present had crusted. No lid lesions were present. He had a marked RAPD, red desaturation, and mild conjunctival injection. His anterior segment and vitreous remained clear. The right disc was hyperaemic with peripapillary swelling and haemorrhage. The macular area was pale and oedematous (Fig 1). Examination of the left eye was completely normal.

Considering the onset of ocular symptoms and signs following the appearance of the typical VZV skin lesions, a presumptive diagnosis of chickenpox neuroretinitis was made. He was admitted and commenced on intravenous aciclovir (250 mg × 3 per day). Confirmatory IgM titres for VZV were unfortunately not performed. No change in his acuity was observed over the next few days; however, his right disc was noted to become slightly pale after 2 days of treatment. At this point intravenous methyl prednisolone was instituted at a dose of 5 mg/kg per day. Despite a gradual resolution of the macular and peripapillary oedema over the next 5 days, his disc remained pale (Fig 2) and his acuity measured as 3/30 (logMAR) after 7 days of intravenous aciclovir and 5 days of methyl prednisolone. Systemically he remained completely well and afebrile on treatment. He was discharged with a further 3 day course of oral aciclovir and a 6 day reducing course of oral prednisolone.

Over 5 months of follow up his acuity has not improved beyond 3/30 (logMAR). The right optic disc is pale and a yellow lipid deposit is present at the macula with some reticular macular pigmentation. The left eye has been normal throughout.

Comment
Posterior segment involvement as part of primary VZV infection in children has only been reported twice to our knowledge. Copenhagen1 reported a 3 year old with bilateral papillitis and a unilateral macular lesion associated with encephalitis following VZV infection. This was a complete recovery of vision and resolution of the macular lesion within 3 weeks of presentation. Capone and Meredith1 describe a case of unilateral central visual loss in a 2 year old child caused by chickenpox retinitis with optic neuritis resulting in a poor visual outcome. Their patient presented with an acute exotropia 24–48 hours before the onset of cutaneous VZV. Funduscopy revealed papillitis, phlebitis, and a macular oedema which have resulted in irreversible visual loss. The young age of the patient is atypical of ocular VZV infection.2 Adults who contract primary VZV infection tend to run a more severe course than children.3 Ocular complications in children are extremely rare.4 The typical posterior segment involvement of VZV is acute retinal necrosis (ARN),4 The youngest case of ARN in association with chickenpox has been reported in a 4 year old.5 In adults, ARN is described as being less severe when presenting at the time of primary zoster infection than as a result of secondary reactivation of latent, previously acquired VZV.6 The changes typical of ARN were absent in this case. Unilateral papillitis and retinitis confined to the macular area were the main features. Optic neuritis has been reported by several authors in association with primary VZV.7–9 Many of these cases are bilateral and coincident with encephalitis8 or occurring in those who are immunocompromised.6 Unilateral optic neuritis has been described in an 18 year old several weeks following a varicella rash which remitted without sequelae following the administration of corticosteroid.7 The mainstay of treatment of VZV retinitis is with intravenous aciclovir. Whether any advantage is gained in administering systemic steroid with the aciclovir is controversial.7 We do not know if a more positive visual outcome may have been achieved if intravenous therapy had been commenced on presentation.1 It is therefore suggested that prompt treatment of VZV retinitis with intravenous aciclovir be started in patients, particularly in a child, presenting with any posterior segment signs.

J R MacKinnon, T Lim Joon, J E Elder
Department of Ophthalmology, Royal Children’s Hospital, Melbourne, Australia

www.bjophthalmol.com
zymes were slightly abnormal (171 mg/l) were markedly raised. Liver en-
tration. His erythrocyte sedimentation rate in the
(Fig 2). He was admitted for further investiga-
affecting the right elbow and nailfold infarcts
2 month history of general malaise, arthral-
ments in the left. Systemic inquiry revealed a
was 6/60 in the right eye and hand move-
for routine risk factors assessment.

A 58 year old white man presented with sud-
loss occurs in up to 8% of patients with Wegener’s granulomatosis (WG) during the
disease lifetime, although only a small proportion of these cases are the result of central retinal artery occlusion (CRAO). We
describe an unusual case of a patient with bilateral CRAO as a presenting feature of WG and α1-antitrypsin (AAT) deficiency.

Case report
A 58 year old white man presented with sud-
en painless loss of vision to the right eye. Vision was hand movements in the right eye and 6/6 in the left. Funduscopy revealed an acute right CRAO with macular oedema. There were no signs of uveitis or retinal vasculitis. Management consisted of intra-
venous acetazolamide (500 mg), ocular mas-
sage, and anterior chamber paracentesis. He was subsequently sent home with aspirin treatment, and referred to his family doctor for routine risk factors assessment. The following morning, he returned to the eye casualty department with a left CRAO, which was treated in the same way. Vision was 6/60 in the right eye and hand move-
ments in the left. Systemic inquiry revealed a 2 month history of general malaise, arthral-
gia, and myalgia. General examination re-
vealed evidence of vasculitic rash (Fig 1) a-
flecting the right elbow and nailfold infarcts (Fig 2). He was admitted for further investiga-
tion. His erythrocyte sedimentation rate in the
first 128 mm, and C reactive protein (171 mg/l) were markedly raised. Liver en-
zymes were slightly abnormal (γGT, 165 IU/l, ALT, 85, alkaline phosphatase, 153). There were traces of blood and protein in the urine, with a normal creatinine clearance (86 mmol/ l). He was commenced on oral prednisolone (60 mg/day) for presumed systemic vasculitis. Normal echocardiography and blood cultures excluded endocarditis. Ultrasonogram of his carotid arteries was normal. His vision gradu-
ally improved, and was recorded 6 days later as 6/12 in the right eye and 6/6 in the left. Patchy peripheral field defects remained.

Subsequently, his serum was positive for cytoplasmic antineutrophil cytoplasmic anti-
body (cANCA) at a titre of 1/160, confirming the diagnosis of WG. Monthly pulses of intra-
venous cyclophosphamide were employed, with gradual tapering of oral prednisolone dose. There was also an incidental finding of homozygous AAT deficiency (Z allele) from serum protein electrophoresis.

Comment
WG is a rare disease with a reported annual incidence of 8.5 per million population1 in the UK, of which up to 16% present initially with ocular disease.2 CRAO in patients with WG is rare, with only several reported cases3,4 since 1960. Ocular and ocular adnexal involvement is the result of parenchymal necrosis, small vessel vasculitis, and granulomatous inflam-
mation. The orbit may be the site of primary inflam-
mation or become secondarily involved from disease of the paranasal sinuses and nasopharynx. Mechanisms by which visual loss occurs in WG include vascular occlusion, macular oedema, inflammatory destruction of retina, optic nerve, or corneoscleral tissue.5 Systemically, the most commonly affected tis-
sues are the lungs and kidneys. The current therapy for WG is cyclophosphamide (daily or pulse) and corticosteroids. Multicentre trials are in progress to assess the optimum treatment duration for induction, and subse-
quent maintenance of remission.

This is the first reported case of bilateral CRAO in WG and AAT deficiency. AAT is nor-
mal present in serum and inhibits uncon-
trolled enzymic destruction of connective tis-
sue during inflammation. Deficiency of AAT results in progressive emphysema and liver injury.6 Research suggests an increased inci-
dence of WG in patients with AAT deficiency.7 The incidence of AAT deficiency has been reported as being significantly increased in
Fibrous histiocytomas of the corneoscleral limbus are rare tumours. We present a case report and a review of the clinical and histopathological findings from cases presented in the literature.

Case report

A 19 year old white male presented with a 6 month history of a painless growth on the inferior corneoscleral limbus of the left eye (Fig 1A and B). There were no other ocular symptoms. He had no medical history of note. His vision was 6/5 unaided in the right eye, and 6/6 unaided in the left eye. The growth was approximately 5 mm in diameter, vascularised, and yellowish in appearance. There were no other ocular abnormalities. The lesion was excised by conjunctival excision and sent for histopathological opinion.

Histological sections of the lesion showed a stromal hypercellular nodule consisting of spindle cells, small lymphocytes, multinucleated giant cells, and histiocytes. (Fig 1C–F). Some histiocytes had foamy cytoplasm. Many capillary sized blood vessels were present. In areas, the spindle cells were arranged in a storiform pattern. A fine collagenous meshwork extended throughout the lesion. Immunohistochemistry confirmed the cellular composition of the lesion (Fig 1G, H) as rich in inflammatory cells, including foam cells and siderophages. No atypical nuclei or mitotic figures are present. Although some authors regard these tumours as reactive proliferations of fibroblasts, others do not accept this view because the lesions tend not to regress spontaneously. Recurrence is rare, with less than 5% of cutaneous benign fibrous histiocytomas recurring after local excision.

In contrast, malignant fibrous histiocytomas of the corneoscleral limbus characteristically appear in later life, between the ages of 50–70 years, with an equal distribution of males to females. They are highly aggressive tumours, and have been reported to have a local recurrence rate of 100% if a limited excision is performed. Recurrence can occur within a few months of excision. There are seven reported cases of corneoscleral malignant fibrous histiocytoma. Two of the cases had an enucleation and two cases underwent orbital exenteration.

Comment

A positive pathergy test is an important diagnostic sign of Behçet’s disease. Its prevalence varies by geographic region, being less common in patients from Northern Europe. Nevertheless, patients with Behçet’s disease should be made aware of this potential complication if they intend to undergo acupuncture.

P I Murray, N Aboteen
Birmingham and Midland Eye Centre, Birmingham B18 7QU, UK

Correspondence to: Professor P I Murray, Academic Unit of Ophthalmology, Division of Immunology and Infection, Birmingham and Midland Eye Centre, City Hospital NHS Trust, Dudley Road, Birmingham B18 7QU, UK; P.I.Murray@bham.ac.uk

Accepted for publication 22 October 2001

Reference


Corneoscleral fibrous histiocytoma

Fibrous histiocytomas of the corneoscleral limbus are rare tumours. We present a case report and a review of the clinical and histopathological findings from cases presented in the literature.

Case report

A 19 year old white male presented with a 6 month history of a painless growth on the inferior corneoscleral limbus of the left eye (Fig 1A and B). There were no other ocular symptoms. He had no medical history of note. His vision was 6/5 unaided in the right eye, and 6/6 unaided in the left eye. The growth was approximately 5 mm in diameter, vascularised, and yellowish in appearance. There were no other ocular abnormalities. The lesion was excised by conjunctival excision and sent for histopathological opinion.

Histological sections of the lesion showed a stromal hypercellular nodule consisting of spindle cells, small lymphocytes, multinucleated giant cells, and histiocytes. (Fig 1C–F). Some histiocytes had foamy cytoplasm. Many capillary sized blood vessels were present. In areas, the spindle cells were arranged in a storiform pattern. A fine collagenous meshwork extended throughout the lesion. Immunohistochemistry confirmed the cellular composition of the lesion (Fig 1G, H) as rich in inflammatory cells, including foam cells and siderophages. No atypical nuclei or mitotic figures are present. Although some authors regard these tumours as reactive proliferations of fibroblasts, others do not accept this view because the lesions tend not to regress spontaneously. Recurrence is rare, with less than 5% of cutaneous benign fibrous histiocytomas recurring after local excision.

In contrast, malignant fibrous histiocytomas of the corneoscleral limbus characteristically appear in later life, between the ages of 50–70 years, with an equal distribution of males to females. They are highly aggressive tumours, and have been reported to have a local recurrence rate of 100% if a limited excision is performed. Recurrence can occur within a few months of excision. There are seven reported cases of corneoscleral malignant fibrous histiocytoma. Two of the cases had an enucleation and two cases underwent orbital exenteration.

Comment

A positive pathergy test is an important diagnostic sign of Behçet’s disease. Its prevalence varies by geographic region, being less common in patients from Northern Europe. Nevertheless, patients with Behçet’s disease should be made aware of this potential complication if they intend to undergo acupuncture.

P I Murray, N Aboteen
Birmingham and Midland Eye Centre, Birmingham B18 7QU, UK

Correspondence to: Professor P I Murray, Academic Unit of Ophthalmology, Division of Immunology and Infection, Birmingham and Midland Eye Centre, City Hospital NHS Trust, Dudley Road, Birmingham B18 7QU, UK; P.I.Murray@bham.ac.uk

Accepted for publication 22 October 2001

Reference

Follow up of these patients ranged from 18 months to 5 years and all were free from recurrence. The other three patients had a local excision. One patient developed local scleral recurrence 2 months later, (it is not evident from the literature if the borders of excision were clear of tumour.) He was found to have a metastasis in the parotid 4 months later, and even after a parotidectomy and radical neck dissection, the "parotid gland" was found in the specimen. One patient developed a local recurrence. The other two patients had a local excision and cryotherapy. After 1 year he was free of any recurrence. Malignant fibrous histiocytomas have a broad range of histological appearances; storiform-pleomorphic, myxoid, giant cell, and inflammatory. The storiform-pleomorphic type is the most common. The cells are predominantly plump pleomorphic spindle-shaped with occasional large, ovoid histiocyte-like cells. Modest amounts of inflammatory cells, such as lymphocytes and plasma cells may be present. The differential diagnosis of a malignant fibrous histiocytoma includes pleomorphic carcinoma, malignant melanoma, and other sarcomas. For these limbic fibrous histiocytomas with a benign histopathological appearance, the management should be local surgical excision. Malignant fibrous histiocytomas need to be managed cautiously, preferably by widespread local excision and cryotherapy at the earliest opportunity. If necessary, enucleation should be considered to fully excise a limbal malignant fibrous histiocytoma.

C S Daniel, B J Clark, S J Tuft
Moorfields Eye Hospital, 162 City Road, London, EC1V 2PD, UK

Correspondence to: Miss Daniel, Department of Pathology, Institute of Ophthalmology, University College London, Bath Street, London EC1V 9EL, UK

Accepted for publication 22 October 2001

References

Haemophilus influenzae corneal ulcer associated with atopic keratoconjunctivitis and herpes simplex keratitis

Haemophilus influenzae is a rare cause of corneal ulceration, usually associated with previous corneal damage. To ensure appropriate treatment, the organism must first be identified by cultures in enriched media. We describe a case of keratitis caused by H influenzae associated with two risk factors—herpetic keratitis and atopic keratoconjunctivitis.

Case report

A 43 year old Hispanic male presented with 1 week of redness, blurred vision, pain, burning, and itching in the left eye. Past ocular history was remarkable for bilateral epithelial and interstitial herpetic keratitis and atopic keratoconjunctivitis with a previous shield ulcer in the left cornea. The patient was not using any medications at the onset of the current episode. He had a history of anhidrotic ectodermal dysplasia and atopic dermatitis since birth. On examination, uncorrected visual acuity was 20/100 in the left eye. The skin of the eyelids was wrinkled, thickened, and hyperpigmented bilaterally with madarosis of the lashes. The conjunctiva had a mild papillary reaction on the right and a severe reaction on the left, with giant papillae in the superior tarsal area. In the left eye, corneal sensation was markedly decreased and a central corneal epithelial defect of 1.0 × 0.5 mm was present with an underlying anterior stromal infiltrate measuring 2.5 × 1.5 mm (Fig 1). Scant keratic precipitates and a small hypopyon were also present.

Scrapings of the corneal ulcer were taken and inoculated on blood and chocolate agar as well as in thiglycollate broth. Viral cultures and polymerase chain reaction (PCR) were also performed to rule out the possibility of a recurrent herpes simplex infection. They were subsequently negative. The patient was started on olofoxacin and fortified cefazolin (50 mg/ml) drops every hour. He was also started on oral aciclovir 800 mg five times a day. On the fourth day, corneal cultures were positive with a heavy growth of H influenzae sensitive to olofoxacin (Fig 2). On the sixth day of treatment, uncorrected visual acuity improved to 20/30, the stromal infiltrate decreased to 1 × 1 mm, with a small epithelial defect, and there was no hypopyon. A supratarsal injection of dexamethasone was given and topical prednisolone acetate 1% three times a day was added. The papillae regressed and the corneal lesion healed completely, leaving a central scar with a deep stromal vessel.

Comment

H influenzae is a tiny Gram negative cocco-bacillus that is an uncommon cause of corneal ulceration. In most series it accounts for less than 2% of all corneal ulcers. It is a common cause of acute bacterial conjunctivitis, especially in children. Unlike Staphylococcus aureus, Streptococcus pneumoniae, and other bacterial causes of conjunctivitis, H influenzae seldom produces corneal ulceration. This is in marked contrast with H pyogenes ocular infections where peripheral ulcers, infiltrates, and pterygienes are commonly seen as complications of conjunctivitis. For H influenzae to infect the cornea, there must be an epithelial defect. Most of the reported cases of H influenzae keratitis have been associated with specific risk factors such as contact lens wear, application of cyanoacrylate glue, or systemic debilitating diseases. In this case, the patient had two significant risk factors—severe atopic keratoconjunctivitis and herpetic keratitis. Despite the negative results of the viral cultures and HSV-DNA PCR, we cannot rule out the possibility that a viral aetiology played a part in this case. Also, it seems likely that complications from the patient’s atopic disease, delayed the healing of the ulcer.

H influenzae is a fastidious organism that needs media enriched with nicotinamide adenine dinucleotide (NAD), such as chocolate agar for growth. It will not grow in blood agar, unless there are also colonies of Staphylococcus aureus, which provide NAD. In that situation, H influenzae will then grow as satellite colonies around the Staphylococcus aureus. This case illustrates the importance of utilising chocolate agar as well as blood agar to make an aetiological diagnosis.

In conclusion, H influenzae is a rare cause of corneal ulceration, which can occur in patients with previous corneal damage from...
diseases such as herpetic keratitis and atopic keratoconjunctivitis. To ensure appropriate treatment the organism must first be identified by cultures in enriched media.

C D Siverio Jr, J P Whitcher
The Francis I Proctor Foundation, UCSF, San Francisco, CA 94143-0944, USA

Correspondence to: Dr John P Whitcher; nepali@itsa.ucsf.edu
Accepted for publication 31 October 2001

References

Blinking and operating: cognition versus vision

The difference in the refractive indices between the air and the tear film results in the tear film having the greatest optical power of any part of the eye. Eyelid blinks are important for maintenance of the tear film. Between blinks there is progressive thinning of the tear film, which becomes non-uniform on the ocular surface and may break up. This produces an irregular air/tear interface, with a reduction in image quality. The longer the period between blinks the greater the effect on the tear film and reduction in vision. Restoration of the tear film occurs immediately following a blink so that the ability to maintain a regular tear film is dependent on the blink rate. A reduction in the blink rate such as, for example, a pause between blinks of 15 seconds has been associated with a change in the shape of the profile of the corneal tear film and up to a 6% reduction in visual acuity. More importantly, however, a reduction in blink rate leads to a reduction in contrast visual acuity.

The ability to distinguish between different layers during surgery, such as the posterior capsule of the lens and anterior vitreous face, or peeling a layer off the retinal surface in vitreoretinal surgery, is dependent on the surgeon having and maintaining good contrast acuity. Blink rates and blink amplitude vary according to vision related behavior and a reduction in the blink rate occurs with tasks of increasing visual difficulty. For example, visual tasks requiring concentration, such as video display terminal use, result in a decrease in average blink rate from 18.4/mm to 3.6 blinks/min.

In order to determine whether the blink rate of ophthalmic surgeons alters during intraocular surgery, the blink rate patterns of nine ophthalmic surgeons were recorded. Two observers recorded the blink rate during casual conversation and when the surgeons were using the operating microscope. None of the surgeons were aware that their blink rates were being recorded, which was done by two medical students during their ophthalmic attachment—that is, their presence in the theatre was accepted as part of their ophthalmic training. The blink rate for each surgeon was recorded in each condition between four and 10 times. The mean blink rate for each surgeon during casual conversation and while operating are presented in Table 1. There was a significant reduction in the average blink rate between both conditions (16.69/min and 4.75/min, p = 0.0001, Student t-test), on average a third and a half fold decrease occurred while operating. It was also noted that the onset of such conversation as the request for an instrument or demonstration of an intraocular structure, was associated with the onset of a blink response.

The reduction in blink rates observed in this study—that is, 16.69 to 4.75, are similar to that found by Patel et al for visual tasks such as video display terminal use. A reduction in blink rate to 4.75 translates to a reduction in contrast acuity of approximately 10% with four surgeons having an expected reduction of more than 60%. Such a reduction in contrast acuity is likely to have an effect on the ability to differentiate between different intraocular surgical layers. Blinking occurs between visual fixations and may be timed so as not to interfere with significant visual input. Blink rate is low when information memory is operating, and cognitive processes utilising display areas accessible to visual input are disrupted during the blackout period of a blink. Blinking is thus suspended during certain cognitive activities to avoid disrupting these processes.

Tear evaporation is dependent on the width of the palpebral fissure and the surrounding humidity. Evaporation increases with widening of the palpebral aperture leading to a lower tear film stability. This has led to a recommendation that video display unit screens be kept below eye level. During ophthalmic surgery the eye pieces are roughly perpendicularly to the face, so that it might be reasonable to lower the height of the eye pieces to reduce the width of the palpebral aperture. Frequent instillation of artificial tears during surgery is not practicable and might also create an abnormal tear film surface in contrast. Likewise, increasing the humidity of the theatre may not be feasible. It might be reasonable to make surgeons, and anyone who uses the microscope, aware of this potential problem so that they can train themselves to blink during parts of the operation or investigation where good contrast acuity is not essential and where disruption of the cognitive processes is likely to have a minimal effect.

K K W Wong, W Y Wan, S B Kaye
St Paul's Eye Unit, 8Z Link, Royal Liverpool University Hospital, Prescot Street, Liverpool L7 8XP, UK

Correspondence to: Mr Kaye; stephen.kaye@rlht.nwest.nhs.uk
Accepted for publication 21 November 2001

References

Table 1 Average blink rates [No/min] for ophthalmic surgeons during periods of casual conversation and while operating using the microscope

<table>
<thead>
<tr>
<th>Surgeon</th>
<th>1</th>
<th>2</th>
<th>3</th>
<th>4</th>
<th>5</th>
<th>6</th>
<th>7</th>
<th>8</th>
<th>9</th>
<th>Mean (SD)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Casual</td>
<td>17.29</td>
<td>17.75</td>
<td>7.75</td>
<td>17.60</td>
<td>27.44</td>
<td>24.67</td>
<td>11.50</td>
<td>13.86</td>
<td>12.33</td>
<td>16.69</td>
</tr>
<tr>
<td>Operating</td>
<td>9.71</td>
<td>9.00</td>
<td>0.29</td>
<td>7.40</td>
<td>1.68</td>
<td>8.67</td>
<td>1.59</td>
<td>3.86</td>
<td>0.54</td>
<td>4.75</td>
</tr>
<tr>
<td>P Value</td>
<td>0.000002</td>
<td>0.002</td>
<td>0.002</td>
<td>0.002</td>
<td>0.0001</td>
<td>0.0001</td>
<td>0.0003</td>
<td>0.0004</td>
<td>0.0002</td>
<td>0.0003</td>
</tr>
</tbody>
</table>

www.bjophthalmol.com
5-fluorouracil may be given. This is known as an augmented trabeculectomy. Infective endophthalmitis is a recognised complication of glaucoma filtering surgery. It may occur in the early postoperative period or it may happen years after surgery. Another entity, probably a precursor to endophthalmitis, has been described as blebitis. Blebitis is an infection of the trabeculectomy bleb without vitreous involvement.

Clinical features of blebitis include pain, photophobia, conjunctival discharge, and severe conjunctival infection centred on an ophalescent filtering bleb. A Siedel test may be positive (this indicates aqueous leakage from the bleb). There may be an anterior chamber reaction. There is no viritis. As many as one per 100 patients/year may develop infection of the bleb. Factors associated with increased risk of bleb related endophthalmitis include increased axial length, thin leaky bleb, conjunctivitis, upper respiratory tract infection, hibernal occurrence, and vitreous wicks. With the increased use of antimetabolites in glaucoma surgery, the incidence of thin walled cystic blebs seems to be increasing. These blebs are more prone to leakage. Some studies conclude that bleb related endophthalmitis is higher when antimetabolites are used. This is more common with inferior limbal trabeculectomy. However, some studies show equal incidence in augmented trabectomey and trabeculectomy without antimetabolite augmentation.

There are few data available for the incidence of blebitis. In most reported cases conjunctival swabs were performed for culture and sensitivity, but organisms causing endophthalmitis may only be present transiently on the ocular surface. In many reports, ocular surface cultures came back positive for Staphylococcus epidermidis and Staphylococcus aureus, which may both be found on healthy normal eyes.

The microbiology of bleb associated endophthalmitis is different from other causes of endophthalmitis. Clinicians should not extrapolate the results of the Endophthalmitis Vitrectomy study to the post-filtration surgery endophthalmitis given the differing pathogenesis and unique spectrum of organisms. The most common organisms are Streptococcus species. The second most common is Haemopo- phillus influenza type b at over 23%. Of the streptococci, S pneumoniae may account for approximately 12%. Between them H influenzae type b and S pneumoniae probably cause more than 35% of blebitis and bleb associated endophthalmitis.

The treatment of endophthalmitis is expensive. It usually involves admission of the patient and frequent use of expensive drops as well as surgical intervention. Inpatient treatment for blebitis has been priced at US$592 (approximately £340) per 24 hours. This can work out to more than £3000 for a 10 day stay in hospital. The admission is cost of four up visits and the morbidity that is involved need to be taken into account. Frequently these patients have pre-existing visual compromise and an episode of endophthalmitis may result in a visually significant loss of vision, an individual disaster with wider social implications.

H influenzae type b vaccine is licensed for use in infants and children. Its use is advised in those patients considered to be at risk for invasive H influenzae type b disease such as sickle cell disease and those receiving treatment for malignancy. After the age of 13 months the vaccine is effective after a single dose. This vaccine has already shown benefit in opthalmology by the dramatic decrease in the incidence of orbital cellulitis in immunised children. It consists of a capsular polysaccharide of H influenzae type b conjugated to a protein carrier. Side effects of the vaccine include fever, headache, malaise, irritability, loss of appetite, vomiting, diarhoea, rash, urticaria, convulsions, erythema multiforme, and transient cyanosis of the lower limbs. Its cost to the NHS is as low as £8.83 for a single dose of 0.5 ml. Pneumococcal vaccine is available. It is a polyvalent pneumococcal polysaccharide from each of 23 capsular types of S pneumoniae. The vaccine is recommended from the age of 2 for people with at least one of the following conditions: haemo- zygous sickle cell disease, asplenia or dysfunc- tion of the spleen, chronic renal disease, nephrotic syndrome, immunodeficiency, immuno- suppression, chronic heart disease, chronic lung disease, chronic liver disease, and diabetes mellitus. It is effective after a single dose if the strains of S pneumoniae prevalent in the community are reflected in the polysaccharides contained in the vaccine. Its cost to the NHS is around £0.5 ml via.

There have been no reports of epidemics of infective blebitis. If it were contagious, there would have been epidemics or clustering in our glaucoma clinics. We can find no evidence of case to case transmission. In fact, all reported cases and series appear sporadic. The association with upper respiratory tract infec- tions and hibernal occurrence are strongly suggestive of respiratory infection with consequent spread to the predisposed eye. We are uncertain whether this is systemic or droplet spread. However, the need for topical antibody protection is negated by the presence of systemic antibody protection to specific bacte- ria. We believe that by minimising the possibility of systemic infections with these agents we diminish the likelihood of blebitis.

It is possible that these vaccines could be given to patients who are destined for trabeculectomy. The cost for both vaccines would be less than £20.00. Two hundred and fifty vaccinations could be paid for by the price of a single episode of bleb associated endophthalmitis. Assuming a long term infection rate of 2%, these vaccines could possibly prevent two cases of bleb asso- ciated endophthalmitis, representing a saving of £5000 to the NHS.

Apart from the cost, vaccination has the potential to prevent significant ocular morbidity. At the very least, these vaccines should be considered in high risk patients undergoing augmented trabeculectomy. We plan to con- duct a prospective study of the effect of these vaccinations upon the incidence of blebitis and bleb related endophthalmitis.

R R Seemongal-Dass, T E James Department of Ophthalmology, Calderdale Royal Hospital, Salterhebble, Halifax HX3 0PW; West Yorkshire, UK Correspondence to: R R Seemongal-Dass Accepted for publication 21 November 2001

References

The wide field multifocal electroretinogram reveals retinal dysfunction in early retinitis pigmentosa

The diagnosis of retinitis pigmentosa (RP) is made on the basis of characteristic retinal pigmentation changes, visual field defects, and reduction in amplitude or loss of rod scotopic standard electroretinogram (ERG) responses, with a possible history of night blindness and a positive family history of RP. Multifocal electrophysiology (mERG), is a new technique that constructs a topographi- cal image of retinal retinal function.7 Reports have suggested that the spatial resolution of mERG is sufficient to detect focal changes in retinal function as RP progresses.8

We describe a case of early RP in which the amplitude and implicit times of the patient's standard ERG rod and maximal responses were normal in the right eye and equivocal in the left eye. However, the peripheral retinal mERG amplitude and implicit times were reduced and delayed. These abnormalities were obtained using a custom built wide field mERG, which facilitates assessment of a 90 degree retinal field.

Case report
A 29 year old woman was referred to the eye clinic by her optometrist. Abnormal retinal pigmentation was found in both funds on routine examination. She had no visual problems and was otherwise systemically well. Her 51 year old mother is known to have RP.

On examination, she had 0.50 dioptres of hypermetropia in both eyes. Her best corrected logMAR visual acuities were −0.075 in the right eye and −0.025 in the left. Her colour vision, anterior segment examination, and intraocular pressures were normal in both eyes. Fundal examination revealed semicircu- lar arcs of intraretinal “bone spicule” pigmen- tation in the inferior mid-periphery of each retina. Her optic discs appeared normal and there was no evidence of attenuation of the retinal vasculature.

www.bjophthalmol.com
A Humphrey 120 point threshold related perimetry test was performed and the patient maintained fixation throughout the test. There was an arc of absolute visual field defect maintained fixation throughout the test. The perimetry test was performed and the patient's averaged central mfERG responses grouped and averaged. The central responses are illustrated in Figure 1. The rod mfERG response is normal (normal range 74–122 nV) (A); the maximal cone mfERG response is normal (normal range of maximal b-wave amplitude 241–709 µV) (B); the maximal cone mfERG response of patient's right and left eye compared with controls (normal range of maximal b-wave amplitude 74–122 nV) (Fig 2A). The average implicit times and amplitude of the scotopic rod, photopic cone and flicker responses of the retinal field. (B) Multifocal responses of a normal eye compared with the patient's eyes. The Ganzfeld ERG responses of a normal eye compared with the patient's eyes. (C) Multifocal responses of patient's right eye with the central 60 degrees of the retina compared with controls (normal range of cone b-wave amplitude 68–222 µV) (C). The global nature of the Ganzfeld ERG response may have inherent limitations in particular circumstances and may require evaluation for effective application in clinical settings. Staffing and financial resource benefits. While we applaud the dissemination of practice guidelines to aid clinical decision making. They suggest that it was unsafe to abandon this practice unless raised in-traocular pressures (IOP) were controlled. The numbers of patients included ranged from 100 to 387. The results of these studies are shown in Table 1. The global nature of the Ganzfeld ERG technique. wide field functional imaging of the retina. I MARMOR MF, KEATING D, EVANS AL. Wide field multifocal electroretinography in retinitis pigmentosa. Am J Ophthalmol 1998; 125:214–26.}

**References**


**Day 1 review following cataract surgery: are we seeing the precise details?**

The Royal College of Ophthalmologists published cataract surgery guidelines1 in February 2001. This document includes protocols relating to postoperative visits suggesting that there are no additional risks to patients who are not reviewed on the first postoperative day. This is a change in recommendation from previous college guidelines in 1995 suggesting a review within 48 hours. There may follow a growing impetus for ophthalmologists to dispense with the first day review, given the reduced demand on clinicians time and the corresponding accrual of staffing and financial resource benefits. While we applaud the dissemination of practice guidelines, they constitute “merely tools, not rules” to aid clinical decision making. They may have inherent limitations in particular circumstances and may require evaluation for effective application in clinical settings. Four studies were quoted by the guideline authors, three of which advocated the omission of day after review2 and one of which was equivocal, suggesting that it was unsafe to abandon this practice unless raised in-traocular pressures (IOP) were controlled. The numbers of patients included ranged from 100 to 387. The results of these studies are shown in Table 1.

**Comment**

In our view, deriving meaningful conclusions that may underpin clinical practice are difficult, owing to the varying methodological...
approaches used in these studies. In Tufail’s study, extracapsular cataract extraction was the predominant surgical technique used. Cohen et al excluded more than 50% of patients with complicated ocular histories or complicated surgery and Whitefield et al had similar extensive exclusion criteria, although the number excluded was not mentioned.

We would draw attention to a recently published study by McKellar and Elder,14 which to our knowledge is one of the largest cohort studies, aside from national cataract surveys, reporting on first and seventh day complications of cataract surgery. Of 1000 patients, the study found that on the first postoperative day complications were observed in 10% of eyes, of which 88% was raised IOP. Unlike most of the previous studies, all patients with available records were eligible, including those with preoperative risk factors and those with surgical complications. These figures align more closely with our “gold standard” of the National Cataract Surgery Survey than the previous mentioned studies. The events most frequently occurring within 48 hours after surgery in the national survey were corneal oedema (9.5%), raised IOP (7.9%), and uveitis (5.6%). Overall, 23.3% of patients had early postoperative complications ranging from minor to sight threatening conditions. The survey also found that several risk indicators were associated with poorer visual outcomes and complications related to cataract surgery: age, ocular co-morbidity (glaucoma, macular disease, amblyopia, and previous ocular surgery), diabetes mellitus, stroke, type of surgical procedure, and grade of surgeon.

In summary, up to 20 000 patients a year in the United Kingdom (10%) may have an uncomplicated early postoperative complication such as corneal oedema or raised IOP if first day review was abandoned. And if McKellar’s study is representative, then 5% of patients would have raised pressure without any previous history or surgical complication and 0.9% of patients could have other potentially serious early complications. Nationally, that equates to almost 12 000 patients annually. It is worth noting that the American Academy of Ophthalmology in its white paper,1 confirms that there are enough significant early postoperative complications to warrant first day review. Are we sufficiently confident in our own practices to diverge?

Credit should be apportioned to the distinguished authors of the cataract surgery guidelines suggesting 24 hour follow up of patients who had undergone complicated surgery, had coexisting eye disease, or had large incision cataract surgery. We would like to reiterate the importance of explicit criteria as part of any review policy and suggest that clinical interpretation of individual circumstances is paramount.

To reconcile the need for an efficient, cost effective review protocol together with a necessity to give due consideration to the entirety of detrimental post-cataract complications, especially given the NHS resource constraints, is difficult. A pragmatic approach may be for clinicians to be discriminately aware of those patients most at risk of developing early complications and instituting review policies accordingly, together with an open door policy for patients who need or want reassurance on the first day following uncomplicated surgery. Furthermore, a multi-professional management approach involving the extended role of trained ophthalmic nurses in postoperative care may reduce demands on physician time.

At the moment, there is a paucity of a good prospective literature on the subject and a need for future studies to address whether those identified complications would result in a change of management at the first postoperative day visit and whether patients would have a poorer outcome if the changes were not instituted.

D Goh
Royal Surrey County Hospital, Guildford GU2 5XX, UK

N Lim
The Western Eye Hospital, Marylebone Road, London NW1 5YE, UK

Correspondence to: Dr Goh; davidgoh1@aol.com

Accepted for publication 29 November 2001

References

Choroidal abnormalities in neurofibromatosis type 1 with non-invasive infrared imaging

Retinal abnormalities have been reported worldwide in patients with neurofibromatosis type 1.15 However, there have been few reports of choroidal abnormalities.16 We report a patient with choroidal abnormalities, associated with neurofibromatosis type 1, using a scanning laser ophthalmoscope (SLO).

Case report
A 37 year old woman presented for an examination with hard contact lenses. Her best corrected visual acuity was 20/20 in both eyes. A diagnosis of neurofibromatosis type 1 was made on the basis of multiple cafe au lait spots, plexiform neurofibroma, and Sakurai-Lisch nodules in the iris in both eyes. The conventional ophthalmoscopic and biomicroscopic fundus examinations were unremarkable. A general medical examination showed no abnormalities except signs of neurofibromatosis type 1. SLO examination showed no abnormalities with a helium-neon laser (633 nm) and regions of multiple, bright patches with infrared imaging (780 nm) using the direct confocal mode (Fig 1) and dark patches with the indirect mode (Fig 2) at the corresponding regions in the posterior pole in both eyes. There were no scotomas in those regions using SLO microperimetry.

Comment
In this case, we observed choroidal abnormalities in a patient with neurofibromatosis type 1. The conventional fundus examination, including biomicroscopic examination and fundus colour photography, did not show remarkable changes. However, the SLO examination showed regions of bright patches with infrared imaging using the direct confocal mode and dark patches using the indirect mode at both posterior poles. Infrared light penetrates the retina into the choroid more than visible light. Therefore, the bright patchy regions seen with confocal infrared imaging and the absence of such regions under helim-neon light examination indicates that the patchy regions are of choroidal origin, as reported by Tasnari and colleagues. In addition, we observed dark patchy regions in the corresponding area using the indirect mode of infrared imaging (which also can obtain images of the deeper retinal layers non-invasively) instead of using indocyanine green fundus angiography. Tasnari and colleagues reported that choroidal abnormalities (100%) occurred more frequently than plexiform neurofibroma (29%) and Sakurai-Lisch nodules in the irides (76%).16

Table 1 Day 1 postoperative complications noted in clinical studies

<table>
<thead>
<tr>
<th>Study (n)</th>
<th>Patients</th>
<th>Corneal oedema</th>
<th>Raised IOP</th>
<th>Uveits</th>
<th>Others</th>
</tr>
</thead>
<tbody>
<tr>
<td>Tufail et al (387)</td>
<td>4 (1%)</td>
<td>8 (2%)</td>
<td>–</td>
<td>–</td>
<td>12 (3.1%)</td>
</tr>
<tr>
<td>Whitefield et al (100)</td>
<td>10 (10%)</td>
<td>3 (3%)</td>
<td>–</td>
<td>–</td>
<td>–</td>
</tr>
<tr>
<td>Cohen et al (21)</td>
<td>26 (13%)</td>
<td>12 (6%)</td>
<td>–</td>
<td>–</td>
<td>–</td>
</tr>
<tr>
<td>Tan et al (227)</td>
<td>5 (4.4%)</td>
<td>3 (1.3%)</td>
<td>1 (0.4%)</td>
<td>–</td>
<td>3 (1.3%)</td>
</tr>
</tbody>
</table>

Figure 1 SLO image of the right eye using the infrared direct confocal mode.

Figure 2 SLO image of the right eye using the infrared indirect mode.
They also reported that bright patchy choroidal regions should be a new diagnostic criterion for neurofibromatosis type 1. The non-invasive SLO examination with confocal and indirect infrared imaging may be useful in the diagnosis of patients with neurofibromatosis type 1.

F Mori, N Kitaya, T Hikuchi, A Yoshida
Department of Ophthalmology, Asahikawa Medical College, Asahikawa, Japan

Correspondence to: Fumihiko Mori, MD, PhD, Department of Ophthalmology, Asahikawa Medical College, Midorigaoka Higashi 2-1-1, Asahikawa 078-8510, Japan, morinao@d5.dion.ne.jp

The authors have no proprietary interest in any aspect of this report.

Accepted for publication 5 October 2001

References

Corneal melt and perforation secondary to floppy eyelid syndrome in the presence of rheumatoid arthritis

Floppy eyelid syndrome (FES) is an uncommon condition that is often underdiagnosed or misdiagnosed owing to the somewhat trivial and non-specific symptoms with which it often presents. In association with the dry eye of rheumatoid arthritis it can, however, have devastating effects.

Case report

A 60 year old moderately obese man with well controlled rheumatoid arthritis (RA) presented to the eye clinic with recurrent red and gritty eyes. A diagnosis of dry eye syndrome with blepharitis was made. He was also found to have a mucocoele of the left lacrimal sac. Lid hygiene and ocular lubricants yielded an initial response and he was discharged.

He re-presented 5 years later with similar symptoms and reduced visual acuity (VA) of 6/36 in the left eye. A diagnosis of dry eye syndrome with secondary corneal epithelial changes was made. Topical lubricants failed to relieve the condition satisfactorily and he was therefore scheduled for punctal occlusion. However, upon admission for this 8 weeks later he was found to have an asymptomatic perforation of the left cornea, with a VA of 6/60. The perforation was treated with glue and a bandage contact lens, topical antibiotic, steroids, and lubricants. Systemic immunosuppression was considered in view of the history of RA, but was withheld as a heavy growth of staphylococcus had been cultured from the cornea. The right eye demonstrated signs of dryness but was otherwise healthy with VA of 6/9.

Bilateral punctal occlusion was undertaken as planned, and in addition a left dacryocystorhinostomy (DCR) to eliminate the mucocoele as a potential reservoir of infection. The eye, however, continued to slowly deteriorate, with persisting mucopurulent discharge, despite the DCR. Eventually uncontrolled endothelialisation developed requiring evisceration.

He re-presented 8 months later with reduced VA of 6/60 in the right eye secondary to a corneal melt (Fig 1A). A chronic mucopurulent discharge had also developed in the right eye, but no lacrimal sac mucocoele was identifiable. On this occasion, however, it was noted on examination that while evertting the eyelids, all four lids exhibited excessive laxity (Fig 1B). This, together with a florid papillary tarsal conjunctival reaction and the chronic mucus discharge, led to a diagnosis of RA associated dry eye syndrome exacerbated by FES.

All four eyelids were immediately subjected to considerable shortening by pentagonal excision; the corneal melt was treated with a bandage contact lens, with topical antibiotic, steroids and lubricants. The response to surgery was dramatic with complete resolution of discharge and gradual spontaneous repair of the corneal melt (Fig 2). The VA eventually recovered to 6/9.

Comment

FES occurs most frequently in middle aged obese males, although it has been described in young, slim males, females, and one child. Typically, the upper tarsus is rubbery and the upper eyelids everts easily with gentle upward pressure. A florid papillary conjunctivitis and chronic mucus discharge are common. Severe corneal involvement is rare, with only four reports in the literature of ulceration in association with FES and only two cases of perforation.3,4

Although the exact pathophysiology of FES is uncertain, a sequence of events may lead to its development and to the secondary corneal changes has been proposed.1 Unknown predisposing factors, possibly congenital, create a floppy upper tarsus. Whereas examination of post-mortem specimens has revealed normal tarsal collagen, elastin fibres are nearly absent.6 It is unclear whether this finding is causative or secondary. During sleep, a local pressure induced ischaemia may develop in the tarsus that, when relieved, results in a repurification injury which could injure tarsal elastin. In addition, there is a high incidence of obstructive sleep apnoea in patients and nocturnal dip in the PaO2 could further contribute to the local ischaemia and subsequent elastin damage.7

Corneal involvement may occur through one or more mechanisms. Spontaneous nocturnal lid eversions resulting from pressure of the pillow on the upper lid may lead to repeated trauma of the corneal epithelium. Lash posis may contribute to this direct trauma. The cornea, however, may be damaged from a more subtle but important mechanism. Affected lid specimens demonstrate a marked polymorphonuclear infiltrate, which may be the sequela of the repurifcation injury described above; this tarsal infiltrate and the associated papillary response may have direct toxic effects on corneal epithelium and stroma.8 It is perhaps intuitive that the corneal complications found in FES may be more severe when, as in our case, co-existing pathologies are present. Blepharitis and RA associated dry eye may both independently cause significant corneal pathology.

This case serves as a reminder that multiple pathologies may contribute to the clinical picture. If FES is not to be missed, ocular examination must include lid eversion and inspection of the tarsus.

J D Rossiter
Southampton Eye Unit, Southampton General Hospital, Tremona Road, Southampton SO16 6YD, UK

R Ellingham
Bristol Eye Hospital, Lower Maudlin Street, Bristol BS1 2LX, UK

K N Hakim, J M Twomey
Department of Ophthalmology, Taunton and Somerset Hospital, Taunton TA1 5DA, UK

Correspondence to: Mr Rossiter
Accepted for publication 10 October 2001

References
Ocular trauma with small framed spectacles

Penetrating injuries are widely reported with spectacle related eye trauma, particularly in car accidents. The use of high grade plastics and secure frames have been shown to reduce the incidence of spectacle related eye trauma. Spectacle safety may be compromised in the trend for small frames and frameless spectacles and may place patients at risk of serious ocular injury.

We present the case of an aphakic patient who sustained a blunt injury following ocular compression by her spectacles. Her injuries could have been avoided if larger framed spectacles had been worn.

Case report

A 79 year old aphakic woman sustained a non-penetrating injury to her left globe by walking into a door. She noted a sharp pain and sudden loss of vision. The globe was compressed by her spectacles, which were smaller than her orbital rim. Her glasses were not damaged and there was minimal periocular soft tissue injury.

She was aphakic, following bilateral cataract extraction for congenital cataracts. The spectacle refraction was +9.00 with a short back vertex distance of 5 mm (Fig 1).

The pinhole acuity was 6/36, a quiet, deep anterior chamber was noted with no aqueous leak, the intraocular pressure was 0 mm Hg. Funduscopy revealed a light vitreous haemorrhage, peripapillary choroidal ruptures, and a 360 degree suprachoroidal haemorrhage (Fig 2).

Hypotony following traumatic ciliary artery spasm was diagnosed and the patient was treated conservatively, with topical atropine twice daily and dexamethasone four times daily. After 4 days the hypotony resolved and the intraocular pressure returned to 14 mm Hg. The suprachoroidal haemorrhages resolved over 2 weeks and the visual acuity improved to 6/24.

Comment

The potential ocular damage from framed and frameless spectacles has been highlighted in a number of reports. These often result from minor road traffic accidents or inflation of air bags that damage the spectacles. The trauma is usually sufficient to break the lenses in the spectacle and the resultant globe laceration is the main cause of morbidity.

The recent trends towards smaller framed spectacles has not been reported as a potential risk to the patient; however, in this case, with small framed spectacles (with a short back vertex distance), minor trauma was sufficient to cause serious eye injury. The patient’s previous larger framed spectacles would have prevented such an injury as the lenses would have been supported by the orbital margin and not the globe.

This case demonstrates the previously unconsidered risk of small framed spectacles in aphakic patients. The back vertex distance may be short, increasing the risk of blunt injury.

J Clarke, R Newsom, C Canning
Southampton Eye Unit, Southampton, UK

Correspondence to: Mr J C Clarke, Southampton Eye Unit, Southampton General Hospital, Tremona Road, Southampton SO16 6YD, UK

Accepted for publication 10 October 2001

References


Late opacification of SC60B-OUV acrylic intraocular lenses

Optical clarity of the intraocular lens (IOL) is paramount in maintaining visual improvement after cataract extraction. While the advent of newer foldable IOLs have revolutionised cataract surgery by the smaller incisions required, their long term safety will be established only with time. There have been recent reports of two separate groups of hydrophilic IOLs that have shown various degrees of opacification, 6 months to 2 years after implantation. These include the Hydroview lens (Bausch & Lomb Surgical, Claremont, CA, USA) and the model SC60B-OUV (Medical Developmental Research, Clearwater, FL, USA). We report our experiences with two patients who had permanent reduction in visual acuity 1–2 years after implantation of the SC60B-OUV IOL.

Case reports

Case 1

An 82 year old man with visual acuities of 6/60 in both eyes because of a right macular hole and left posterior subcapsular cataract underwent routine left phacoemulsification and intraocular lens implantation in March 1998. A foldable hydrophilic acrylic IOL (Model SC60B-OUV, MDR, Inc) was implanted in the capsular bag. Postoperative recovery was uneventful, with left visual acuity improving to 6/4 with correction, 6 weeks after the surgery.

He was referred back to the clinic in March 2000 with symptoms of intermittent binocular diplopia, which was relieved with fresnel prisms. His left visual acuity was 6/6, but the intraocular lens was found to be uniformly cloudy (Fig 1A). As the patient was not experiencing any symptoms from his cloudy IOL, no intervention was advised. Over the next 6 months the brownish discoloration of the IOL increased in intensity and the patient complained of “foggy vision.” His visual acuity dropped to 6/12 and he was disturbed by the distorted, but clear images from his right eye and hazy images from his left eye. Fundus examination of the left eye has also become progressively difficult. IOL exchange is being considered, bearing in mind the presence of the macular hole in the fellow eye and the risks of removing a posterior chamber IOL 3 years after implantation.

Figure 1 (A) Diffuse opacification of the IOL 2 years after implantation in case 1. (B) The explanted opacified IOL optic from case 2 is compared to a normal acrylic IOL.
but approximately 1 year after the surgery there was gradual deterioration of vision to 6/24 in August 2000. At this stage the IOL was noted to be cloudy; there was also progression of her endothelial dystrophy. She underwent uneventful right penetrating keratoplasty with exchange of posterior chamber IOL in November 2000 and has a current right visual acuity of 6/12. The explanted opacified IOL is compared to a normal clear acrylic IOL in Figure 1B.

Comment

The safety and efficacy of AcrySof polyacrylic IOLs has been reported to be equal to or better than poly(methylmethacrylate) IOLs.1,2 The unexpected late opacification of the acrylic IOL (SC60B-OUV), implanted in the only “good” eye of both our patients, resulted in significant visual disability and clinical dilemma. This model of IOL was first produced in June 1997 by Medical Development Research (MDR, Inc). More than 60,000 of these lenses have been implanted worldwide, but only outside the United States.1 Reports of opacification of the IOL started coming through to the manufacturer in May 1999. (Summary of SC60B-OUV lens opacification investigation, personal communication from MDR, Inc, 20 July 2001). Several theories have been put forward to explain the late clouding of the IOL optic.1,4 Analysis of 23 explanted IOLs of the same model has shown that degeneration of the ultraviolet filtration material and calcium deposits within the optic biomaterial are responsible for the opacification of the IOL.5 Werner et al. analysed nine explanted IOLs of the same model and demonstrated the presence of calcium phosphate salts in the deposits within the optics of the IOL.6 Investigations by the manufacturers identified four lots of polymer biomaterial formulated and prepared by Vista Optics (London) and used by MDR, Inc in the IOL manufacture, that correlated with opacification complaints (Summary of SC60B-OUV lens opacification investigation, personal communication from MDR, Inc, 20 July 2001).

Ninety two of the estimated 60,000 SC60B-OUV IOLs implanted were explanted and returned to the company. MDR, Inc ceased exporting SC60B-OUV lenses in June 2000 and claims that the opacification represents only 0.15% of total SC60B-OUV IOLs implanted. (Summary of SC60B-OUV lens opacification investigation, personal communication from MDR, Inc, 20 July 2001). However, this does not account for those patients who have not yet had their opaque IOLs identified or explanted. The lateness of the onset of opacification and resulting visual disability may mean that we are seeing only the tip of the iceberg.

A Joseph, H S Dua

Division of Ophthalmology and Visual Sciences, University of Nottingham

Correspondence to: Professor H S Dua, Division of Ophthalmology and Visual Sciences, B Floor, Eye Ear Nose and Throat Block, University Hospital, Queen’s Medical Centre, Nottingham NG7 2UH, UK; harminder.dua@nottingham.ac.uk

Accepted for publication 3 December 2001

References


Treatment of superior limbic keratoconjunctivitis with a unilateral bandage contact lens

The typical patient with superior limbic keratoconjunctivitis (SLK) is a woman aged between 20 and 60 years of age with chronic red and irritated eyes.1 Although both eyes are usually affected, the condition maybe asymmetrical.1 After episodes of exacerbation and remission it usually resolves. The patient may also have abnormal thyroid function.2 SLK has been treated with silver nitrate or thermal cauterisation of the superior bulbar conjunctiva, pressure patching, and large diameter bandage contact lenses (BCL), topical trans-retinoic acid 0.1%, and resection or suture of the superior bulbar conjunctiva.1,2 Over 50% of patients with SLK are said to have keratoconjunctivitis sicca and recently upper punctal plugs have been used to treat SLK.

We report two cases in which a unilateral BCL wear ameliorated the symptoms of bilateral SLK and a possible explanation is discussed.

Case reports

Case 1

A 38 year old woman presented with a 3 month history of irritable photophobic eyes that were unresponsive to preserved lubricants. Her right eye was anhydromyxidrosis. She complained of persistent severe discomfort, photophobia, and a burning sensation in both eyes. The ocular examination was unchanged. A silicone hydrogel BCL (Pure Vision, Bausch and Lomb) was inserted into the right eye and within an hour she had symptomatic relief in both eyes. Two months later the BCL was lost and her bilateral ocular discomfort returned and the left BCL was refitted with immediate symptomatic relief in both eyes. The patient was referred to an endocrinologist who commenced carbimazole and β blockers. Her treatment was later changed to propylthiouracil after she suffered from carbimazole induced arthralgia. Thyroid function tests were normal 6 months later and the propylthiouracil was stopped.

Comment

The pathogenesis of SLK is unclear. It may be the result of mechanical irritation from increased pressure of the upper eyelid against the globe and/or increased motility of the upper bulbar conjunctiva from hyperthyroidism or ageing.1 Increased upper eyelid tightness may be the result of thyroid eye disease or chronic inflammation and, in addition, may

Figure 1 Case 2, left eye. (A) The superior bulbar conjunctiva, at presentation, showing hyperaemia and rose Bengal staining. (B) After 3 months of bandage contact lens wear the superior conjunctival hyperaemia had resolved and the patient was asymptomatic.
impair the normal turnover of bulbar conjunctival epithelial cells. This may be aggravated, in some patients, by blepharospasm, which increases the force on the globe. Therapeutic lenses can produce rapid symptomatic relief in SLK. They may be helpful in the treatment of SLK as they help to maintain a larger surface area, and may facilitate healing of punctate epithelial erosions by protecting the ocular surface from the eyelids, reduce upper lid pressure on the globe and alter tear dynamics. During blinking, as the upper lid moves downwards to meet the lower lid significant forces are exerted on the globe. The lens can reduce the force on the superior limbus from blinking as it has a lower mechanical stiffness and elastic modulus. In the first week of contact lens wear tear production increases dramatically and tear tear tonicity rises as evaporation increases. A lens may then aid aqueous tear deficiency, which can accompany SLK, by ensuring a continuous precorneal tear film. It is difficult to be certain of the mechanism of action, and this reduction, which increases with duration of lens wear, would then decrease bilateral reflex blinking. The lens can reduce the force on the globe, and alter tear dynamics. This may be aggravated, in some patients, by blepharospasm in SLK. It should not be forgotten that continuous BCL wear carries risks including microbial keratitis and corneal vascularisation. New extended wear silicone BCL, as used in our patients, increase oxygen transfer and have extended wear silicone BCL, as used in our patients, increase oxygen transfer and have extended wear silicone. In the first week of contact lens wear tear production increases dramatically and tear tear tonicity rises as evaporation increases. A lens may then aid aqueous tear deficiency, which can accompany SLK, by ensuring a continuous precorneal tear film. It is difficult to be certain of the mechanism of action, and this reduction, which increases with duration of lens wear, would then decrease bilateral reflex blinking. The lens can reduce the force on the globe, and alter tear dynamics. This may be aggravated, in some patients, by blepharospasm in SLK. It should not be forgotten that continuous BCL wear carries risks including microbial keratitis and corneal vascularisation. New extended wear silicone BCL, as used in our patients, increase oxygen transfer and have been shown to reduce such risks.

**References**


**MAILBOX**

Keratectasia after PTK

Takahashi and colleagues have elegantly described an interesting and rare complication of phototherapeutic keratectomy (PTK) in their recent report of an unusual case of keratectasia after PTK. The hypothesis that risk of ectasia is proportional to residual stromal base, or depth of ablation, fits with the assumed biomechanical aetiology of this recently reported complication of laser refractive surgery. The generally accepted empirical minimal thickness of 250–300 µm of corneal stroma, excluding flap thickness, remains speculative, as we do not understand the underlying pathophysiology. Indeed, although Holland et al. highlighted the association of thin residual stromal thicknesses, post-PRK and LASIK, with keratectasia, they also described this complication after surface based hyperthermic PRK ablation, where the centre was minimally ablated and residual stromal thickness was greater than 360 µm. The authors suggest, in the reported case, that band-shaped keratopathy (BSK) may have compromised the tensile strength of the cornea. This seems unlikely as this condition generally affects the superficial anterior cornea, and usually does not penetrate deeper than Bowman’s layer. The suitability for treatment by PTK. However, further clinical detail which the authors have not provided might reveal underlying corneal pathology with secondary “rough” BSK rather than “smooth” BSK. However, there are a number of reasons, other than simple biomechanical compromise, for keratectasia following PTK in this case: (1) forme fruste keratoconus—as no preoperative topographic or surface asymmetry values are presented to enable the reader to rule this out; (2) clinical keratoconus, which seems less likely in respect of patient’s age and a preoperative cylindrical error of only –1.5D; (3) idiopathic keratectasia, possibly secondary to widespread deregulated keratocyte apoptosis. The latter has been demonstrated after LASIK, with a considerable and longstanding decrease in keratocytes in the peri-ablation area. Also, Helena et al. demonstrated apoptosis to a depth of at least 50 µm after all of the following procedures: epithelial scrape, corneal scrape PRK, transthepithelial PRK, and LASIK. Epithelial scrape and LASIK demonstrated keratocyte apoptosis to depths of up to 75 µm and 100 µm, respectively. The authors have recently identified a keratocyte free zone 160 µm into the stroma following LASIK, and theoretically more widespread apoptosis as a response to excimer laser photorefractive surgery, may contribute to keratectasia. While it is difficult to ascertain why keratectasia occurs, in this case with a residual stromal thickness of over 500 µm, from the data provided the most likely aetiologies would seem to be either undiagnosed forme fruste keratoconus or idiopathic keratectasia. Currently, recent reviews illustrate the dearth of substantial information available regarding idiopathic keratectasia (iatrogenic kerato- conus), with a little over 60 cases published. At this point, although some are likely to be due to over-ablation, for many cases such as this the exact aetiology remains unknown and is likely to be multifactorial, and one of these factors is residual corneal thickness. The fact that keratectasia can occur, after what would be considered minimal ablation, highlights the unpredictable occurrence, but with over a million cases of LASIK or PRK occurring each year, the stimulus to identify contributing factors is significant.

**References**


**BOOK REVIEWS**

Age Related Macular Degeneration—Current Treatment Concepts.


This book is one in a large series of medical radiology textbooks concentrating on diagnostic imaging and radiation oncology. There are 71 authors in all, the majority of whom work either in the eastern United States or Germany, and the book contains 24 chapters. The text is aimed at specialists in radiotherapy rather than ophthalmology. The majority of the chapters are concerned with radiotherapy of age related macular degeneration (AMD) with contributions from experts in this field. Some of the chapters contain results of controlled studies and are of good value, whereas others have less scientific merit in that they are long term follow up studies without controls. For ophthalmologists wishing to look into the subject of
radiotherapy for ARMD this would be a good source of material and is well referenced.

There are a few chapters on the clinical manifestations, diagnosis, and surgery of ARMD but there is no real mention of laser treatment. Many of these chapters are of limited scope and do not provide a comprehensive overview of the ophthalmic assessment and management of ARMD.

Although not stated in the book it reads as if it is the proceedings of a clinical meeting. The chapters do not read in a coordinated way and essentially present the results of individual units describing their methods and results of radiotherapy. As such it is a useful source of information for the with an interest in this topic but it is of limited value for ophthalmologists wishing to obtain a balanced view of current treatment of ARMD.

R H B Grey
Bristol Eye Hospital, Lower Maudlin Street, Bristol BS1 2LX, UK

Clinical Ophthalmic Pathology.

This is an excellent, easy to read, well illustrated book. It is one of the first of its kind to bring pathology alive by describing diseases via pathogenesis as opposed to anatomy. For the trainee in ophthalmology, optometry, and visual science it, therefore, provides a more logical approach to the understanding of ocular diseases. As the book attempts to cover many subjects it sometimes does not do justice to the subject. It would have been better to have had the reading lists at the end of each chapter. However, as it stands the book is an excellent introduction to pathology complementing clinical textbooks. If read together with clinical texts it certainly will do justice to the basic science. It would have been desirable to have this book illustrated by a variety of figures and diagrams.

The chapters do not read in a coordinated way and essentially present the results of individual units describing their methods and results of radiotherapy. As such it is a useful source of information for the with an interest in this topic but it is of limited value for ophthalmologists wishing to obtain a balanced view of current treatment of ARMD.

A Dick

NOTICES

Childhood blindness

The latest issue of Community Eye Health (No 40) discusses new issues in childhood blindness, with an editorial by Clare Gilbert, senior lecturer at the International Centre for Eye Health. 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; website: www.jchc.co.uk). Annual subscription (4 issues) UK£23/US$40. Free to workers in developing countries.

International Centre for Eye Health

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

Second Sight

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

Specific Eye Conditions (SPECS)

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

XXIXth International Congress of Ophthalmology

The XXIXth International Congress of Ophthalmology will be held on 21–25 April 2002 in Sydney, Australia. Further details: Congress Secretariat, C/- ICMS Australia Pty Ltd, GPO Box 2609, Sydney, NSW 2001, Australia (tel: +61 2 9241 1478; fax: +61 2 9251 3552; email: ophthal@icmsaust.com.au; website: www.ophthalmology.aust.com).

12th Meeting of the European Association for the Study of Diabetic Eye Complications (EASDEC)

The 12th meeting of the EASDEC will be held on 24–26 May 2002 in Udine, Italy. The deadline for abstracts is 15 February 2002. Three travel grants for young members (less than 35 years of age at the time of the meeting) are available. For information on the travel grants, please contact Dr C G Agardh, President of EASDEC, Malmö University Hospital, SE-205 02 Malmö, Sweden (tel: +46 40 33 10 16, fax: +46 40 33 73 66; email: carl-david.agardh@endo.max.lu.se). Further details: NORD EST CONGRESSI, Via Aquileia, 21–33100 Udine, Italy (tel: +39 0432 21399; fax: +39 0432 50687; email: nordest.congressi@ul.net.it).

3rd Interdisciplinary Symposium on the Treatment of Autoimmune Disorders

The 3rd Interdisciplinary Symposium on the Treatment of Autoimmune Disorders will be held in Leipzig, Germany on the 6–8 June 2002. Topics to be covered include: basic aspects of autoimmune diseases, experimental therapeutic concepts, and clinical studies providing novel concepts or novel focus on established therapies. There will also be the presentation of the Nils-Illa-Richter Award (application deadline is April 2002, further details on the web site). Further details: Prof. Dr. med. Michael Sticherling, Department of Dermatology, University of Leipzig (email: stichm@medizin.uni-leipzig.de; website: www.autoimmun.org); Fördergesellschaft zur Therapie von Autoimmunerkrankungen e.V. (email: autoimmun.org@gmx.de).

International Society for Behçet’s Disease

The 10th International Congress on Behçet’s Disease will be held in Berlin 27–29 June 2002. Further details: Professor Ch Zouboulis (email: zoubbere@zedat.fu-berlin.de).

Singapore National Eye Centre 5th International Meeting

The Singapore National Eye Centre 5th International Meeting will be held on 3–5 August 2002 in Singapore. Further details: Ms Amy Lim, Organising Secretariat, Singapore National Eye Centre, 11 Third Hospital Avenue, Singapore 168751 (tel: (65) 322 8374; fax: (65) 227 7290; email: Amy_Lim@sne.com.sg).

BEAVRS Meeting

The next BEAVRS meeting will be held in the Dalmahoy Hotel near Edinburgh on 31 October to 1 November 2002. Further details: Susan Campbell, Medical Secretary, Gartnavel General Hospital (email: susan.j.campbell.wg@northglasgow.scot.nhs.uk).

CORRECTION

The authors of the letter “Recurrent corneal ulceration as late complication of toxic keratitis”, appearing in the February issue of BJO (2002;86:245–6), would like to add an author, SH Santaner.