Serpiginous choroidopathy presenting as choroidal neovascularisation

Serpiginous choroidopathy is an insidious, relentlessly progressive, idiopathic inflammatory disease affecting the retinal pigment epithelium and inner choroid. Choroidal neovascularisation (CNV) is a well recognised late complication of serpiginous choroidopathy in 10–25% of affected patients. In all previously reported cases CNV was recognised at the time of or after the diagnosis of serpiginous choroidopathy was established. We report a patient presenting with CNV who subsequently developed clinical findings characteristic of serpiginous choroidopathy.

Case report

A 31 year old man presented with decreased vision in his right eye in July 1997. Examination revealed acuities of 20/40 right eye and 20/20 left eye with normal anterior segments. The right fundus showed subretinal fluid and haemorrhage adjacent to the disc (Fig 1A). The left eye showed an irregularity superior to the optic disc (Fig 1B). The vitreous and fundi were otherwise normal bilaterally. Fluorescein angiography (Fig 2A, B) revealed peripapillary choroidal neovascular membranes in both eyes that were treated with argon laser photocoagulation. In April 1998 and February 1999 the left eye required photocoagulation for recurrent peripapillary CNV. Evaluation for floaters in February 2000 revealed 1+ vitreous cells and new lesions in the left eye.

Examination at the National Eye Institute in April 2000 revealed acuities of 20/40 right eye and 20/16 left eye with normal anterior segments. The vitreous contained trace cells without haze bilaterally. The right fundus showed a large peripapillary chorioretinal scar. The left fundus revealed a chorioretinal scar superior to the disc and two yellow, irregularly circumscribed, deep macular lesions (Fig 3A, B). The retinal vessels and discs were normal and no subretinal fluid, haemorrhage, or macular oedema was noted in either eye.

Fluorescein angiography revealed early hypofluorescence and late hyperfluorescence corresponding to the macular lesions in the left eye (Fig 3C, D) with no evidence of CNV in either eye. Laboratory studies were non-diagnostic. A diagnosis of serpiginous choroidopathy was made based on the clinical and fluorescein characteristics of the macular lesions in the left eye.

Comment

CNV in serpiginous choroidopathy is associated with a poor visual prognosis. In a small study CNV was reported to develop within 16 months of the serpiginous diagnosis. In a larger retrospective study of 53 serpiginous patients active CNV was found in three patients at the time of initial diagnosis and in three others within 2–17 months. Our patient differs from those previously reported in that he was diagnosed and treated for idiopathic CNV before the recognition of clinical findings.
diagnostic of serpiginous choroidopathy. Other causes of posterior uveitis associated with CNV and choriotelial lesions similar to those seen in our patient include acute posterior multilocular plaqueoid epiphielopathy (APMPE), presumed ocular histoplasmosis syndrome (POHS), sarcoidosis, multifocal choroiditis, birdshot chorioretinopathy, and toxoplasmosis. As with most cases of serpiginous choroidopathy, the CNV in these entities typically occurs late in the disease course.

The exact pathogenesis of idiopathic CNV is unknown. CNV in eyes with uveitis, however, is believed to develop in direct response to the intraocular inflammation which may alter the balance between vascular growth factors, such as vascular endothelial growth factor (VEGF), and inhibitors. In the early stages of development active serpiginous lesions and CNV may appear as poorly defined subretinal lesions difficult to differentiate by ophthalmoscopy. Typically with fluorescein angiography classic CNV and serpiginous lesions are readily distinguished as the former shows early hyperfluorescence while the latter characteristically shows early blockage.Occult CNV, which may show subtle or less pronounced early hyperfluorescence with late leakage, however, may be more difficult to distinguish from an early serpiginous lesion.

This case illustrates that serpiginous choroidopathy may present with CNV. In contrast to idiopathic CNV, optimal treatment of CNV in patients with uveitis may require immunosuppressive treatment that addresses the underlying ocular inflammation with or without adjunctive laser therapy. Further investigation is needed to better define the role of emerging therapies for CNV such as photodynamic therapy which may offer promise for the treatment of CNV in uveitis patients.

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References


Optic neuritis in anti-GQ1b positive recurrent Miller Fisher syndrome

Only five cases of optic nerve involvement in Miller Fisher syndrome (MFS) have been documented in the literature. This report further confirms that optic neuritis may be seen in anti-GQ1b positive MFS.

Case report

This 23 year old woman presented with acute blurring vision, diplopia, and pain with eye movement. Her visual acuity was 20/20 in both eyes and 20/200 left eye with left relative afferent pupillary defect (RAPD). She had left red colour desaturation. Her visual field on tangent screen revealed an enlarged left blind spot and a left upper quadrant temporal peripheral field constriction. She had bilateral sixth nerve palsies, nystagmus in all gazes, and left optic disc oedema. After 1 week her visual acuity improved to 20/20 in both eyes, but her left disc remained oedematous. She then developed progressive gait ataxia to such a degree that she was unable to walk. Dysmetria and dysdiadochokinesia were more marked in her left upper extremity. She had left sensory, and absent lower extremity deep tendon reflexes, and bilateral Babinski’s. She also had tingling in her hands and feet and decreased lower extremity vibratory sensation. Her mental status was normal throughout her illness. She was not taking any drugs. A magnetic resonance image (MRI) of the brain and entire spine and MR venogram were all normal. Her cerebrospinal fluid (CSF) opening pressure was 150 mm H2O. Her CSF protein was elevated at 70 mg/dl, but CSF glucose and cell count were normal; CSF VDRL, Gram stain, routine bacterial, viral, and fungal cultures were negative, and the low oligoclonal bands were seen on CSF electrophoresis. Her visual evoked potential (VEP) revealed a delayed left P100 latency at 131 ms and her brainstem auditory evoked potential (BAEP) was normal. Electromyogram/nerve conduction study (EMG/NCV) study revealed mildly prolonged median and peroneal F-waves, normal distal motor latencies in her extremities and a reduced left median sensory nerve action potential (SNAP), anti-GD1a antibody (162 EIA U (normal = 100) = 100) Antibodies to GQ1b were present. Antibody to GQ1b gangliosides are known to be present in the human optic nerve and anti-GQ1b antibodies can cross the blood-brain barrier, the optic disc oedema in this patient could represent an initial anti-GQ1b optic neuritis presenting as optic neuritis. Since high concentrations of anti-GQ1b positive MFS, two other cases of presumed optic neuritis were associated with anti-GQ1b positive MFS. The patient presented here had markedly decreased visual acuity, pain with eye movement, dyschromatopsia, and optic disc oedema that resulted in good visual recovery are all indicative of the diagnosis of optic neuritis. Since high concentrations of anti-GQ1b antibodies were present in this patient, there was no indication of the presence of GBS. Therefore, the possibility of a central nervous system feature of anti-GQ1b positive recurrent MFS is high. The presence of visual impairment in MFS, visual evoked potentials were either absent or suggestive of pre-chiasmal and post-chiasmal visual pathway dysfunction. Demyelinating optic neuropathies confirmed by VEP were reported in one patient with possible MFS. Two other cases of presumed optic neuritis were associated with anti-GM1 antibody positive MFS. The patient presented here had markedly decreased visual acuity, pain with eye movement, dyschromatopsia, and optic disc oedema that resulted in good visual recovery are all indicative of the diagnosis of optic neuritis. Since high concentrations of anti-GM1 antibodies are present in 80% to 100% of patients with MFS, MFS may be immunologically differentiated from GBS by the presence of anti-GQ1b and anti-GM1 antibodies. Although both anti-GD1a IgG and anti-GM1 IgG are associated with GBS, anti-GM1 IgG is present in patients with typical MFS who have limb weakness, as in this patient. As further evidence linking this antibody to MFS, the decrease in anti-GQ1b antibody levels after plasmapheresis correlated with the clinical recovery in this patient. Therefore, the elevated titres of anti-GQ1b and anti-GM1 antibodies, along with the clinical triad of ophthalmoplegia, areflexia, and ataxia in this patient all support the diagnosis of MFS, and not GBS.

In rare cases, MFS has been known to recur. This patient presented with a relapse of similar clinical features to the initial episode from her initial episode. In the study done by Chida et al, patients with recurrent MFS appeared to have similar HLA typing characteristics as the non-recurring ones. Both types of recurrent MFS, HLA-DR2 and Cw3 alleles, but the frequency of HLA-DR2 was slightly higher in the patients with recurrent MFS. Therefore, this patient’s HLA-DR2-positive status may have been a risk factor for her recurrence of MFS. This case report emphasizes that optic neuritis may be a central nervous system feature that should be recognised as part of the Miller Fisher syndrome. The presence of both anti-GQ1b IgG and anti-GM1 IgG in this patient provides immunological evidence supportive
of typical MFS. The delayed P100 latency in her VEP also provides electrophysiological evidence that the optic nerve is affected in anti-GQ1b antibody positive MFS. Furthermore, this is the first documented case known to the author of optic neuritis in the recurrent subtype of MFS which is associated with a higher frequency of the HLA-DR2 allele.

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References

Ocular myasthenia gravis and inflammatory bowel disease: a case report and literature review

Myasthenia gravis has been reported to be associated with both ulcerative colitis (UC) and Crohn’s disease (CD). The link between inflammatory bowel disease (IBD) and myasthenia gravis (MG) is thought to be related to the production of autoantibodies. Myasthenia gravis is also associated with other autoimmune diseases including alopecia, lichen planus, vitiligo, and systemic lupus erythematosus.

Similarly, IBD frequently presents with other autoimmune disorders. One study demonstrated a 9.4% prevalence of autoimmune disorders in patients with UC including sclerosing cholangitis, thyroid disorders, vitiligo, insulin dependent diabetes mellitus, thyroid disease, pernicious anaemia, scleroderma, and seropositive rheumatoid arthritis. Despite the association between MG and other autoimmune disorders, there are relatively few reports of ocular findings as the presenting sign of MG in patients with IBD.

Case report
A 21 year old African-American male, with a medical history of biopsy proved ulcerative colitis diagnosed in 1995, focal segmental glomerular sclerosis determined by renal biopsy in 1995, and primary sclerosing cholangitis determined by liver biopsy in 2000 presented to the neuro-ophthalmology service with complaints of binocular diplopia and ptosis of the left upper eyelid. Both the diplopia and the ptosis were better in the morning and worsened during the course of the day. His ulcerative colitis had been in remission for the past 5 years without medication.

Best corrected visual acuity was 20/25 in each eye. The external examination revealed ptosis of the left upper eyelid that worsened in sustained upgaze. He had limited extraocular motility in all fields of gaze (Fig 1). The remainder of the neuro-ophthalmic examination was normal and he had no difficulty with speech or swallowing.

Laboratory evaluation revealed a positive acetylcholine receptor antibody and normal thyroid function studies. There was no evidence of a thymic mass on magnetic resonance imaging of the chest.

The patient returned to the emergency room 1 week later with difficulty swallowing and shortness of breath. He was hospitalised for plasmapheresis and upon discharge treated with imuran, prednisone, and mestinon. One month later his ptosis resolved and his extraocular motility was normal.

Comment
Autoimmune disorders, including MG, occur more frequently in UC than in CD. It is not clear how many other cases of IBD manifested with ocular presentations as the initial finding of MG as in our case report. Our literature review revealed only one other purely ocular presentation of myasthenia associated with ulcerative colitis; however, details of the ocular examination were not included. Another report, of a 21 year old woman with a 3 year history of Crohn’s disease, documented diplopia and unilateral ptosis as the initial findings of MG. She was found to have acetylcholine receptor antibodies and her ocular findings improved with pyridostigmine.

Because of the relatively few reports of ocular myasthenia in patients with IBD we reviewed the English literature and found four additional reports of MG in patients with IBD. Based on these four reports and the three (including the present report) with ulcerative colitis in patients with IBD (Table 1), the mean duration of IBD before the diagnosis of MG was 10 years.

Autoimmune dysregulation is the central defect in both MG and IBD. Both IBD and MG may be associated with an elevated carcinoembryonic antigen (CEA) and decreased peripheral lymphocyte counts that subsequently normalise following thymectomy. Some studies have shown abnormal thymic involution and the presence of an abnormal ratio of T suppressor to T helper cells in both MG and UC, while others have noted a decline in suppressor T cells and an increase in

<table>
<thead>
<tr>
<th>Reference</th>
<th>Age (years)</th>
<th>Sex</th>
<th>IBD</th>
<th>Duration of IBD before diagnosis of MG (years)</th>
<th>AchR antibody reactivity</th>
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<tr>
<td>Miller 1971</td>
<td>35 Male</td>
<td>UC</td>
<td>13</td>
<td>Unknown</td>
<td></td>
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<tr>
<td>Tan 1974</td>
<td>38 Male</td>
<td>UC</td>
<td>12</td>
<td>Unknown</td>
<td></td>
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<tr>
<td>Martin et al, 1991</td>
<td>63 Male</td>
<td>CD</td>
<td>15</td>
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<tr>
<td>Gowen-Rousseau et al, 1993</td>
<td>27 Female</td>
<td>UC</td>
<td>10</td>
<td>Positive</td>
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<tr>
<td>Finnie et al, 1994</td>
<td>21 Female</td>
<td>CD</td>
<td>3</td>
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<tr>
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<td>11 Male</td>
<td>CD</td>
<td>9</td>
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<tr>
<td>Present report</td>
<td>21 Male</td>
<td>UC</td>
<td>7</td>
<td>Positive</td>
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</tbody>
</table>

IBD = inflammatory bowel disease, MG = myasthenia gravis, AchR = acetylcholine receptor, UC = ulcerative colitis, CD = Crohn’s disease.
immature helper T cells suggesting migration without normal maturation. The immunological link between MG and IBD is highlighted by two reports of patients undergoing surgical treatment. One report of a patient with both MG and CD documented improvement in perineal and perianal disease following thymectomy for severe uncontrolled MG. Another patient with both MG and UC demonstrated regression of the myasthenia following proctectomy.

Although the simultaneous occurrence of these two autoimmune disorders is uncommon, it is important to understand that ocular findings may be the initial manifestation of MG in patients with IBD.

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References

Magnetic resonance imaging findings in malignant melanoma of the lacrimal sac

A case of primary malignant melanoma of the lacrimal sac is presented. This is the first report of the preoperative magnetic resonance imaging (MRI) findings of malignant melanoma of the lacrimal sac.

Case report
A 54 year old Chinese woman was referred to an ophthalmologist complaining of a 6 month history of left sided bloody tears and epistaxis. She had a firm, non-tender left medial canthal swelling, and syringing revealed left nasolacrimal duct (NLD) obstruction. Ocular and peri orbital examination was otherwise normal. A dacryocystogram (DCG) demonstrated a filling defect in the lacrimal sac with NLD obstruction.

An ENT opinion was sought, and nasal examination revealed left sided septal deviation, with no obvious cause for the epistaxis. Computed tomography (CT) of the head and orbits demonstrated a left lacrimal sac lesion extending into the NLD with proximal dilation of the duct and no apparent bone erosion (Fig 1A) MRI confirmed the presence of a lacrimal sac lesion with intermediate signal intensity on T1 and T2 weighted images (Fig 2A, B). The lesion enhanced with intravenous gadolinium.

An incisional biopsy of the lacrimal sac (Fig 1B) under frozen section control, and paraffin sections, confirmed malignant melanoma. A full medical review, including MRI of the chest and abdomen, and liver function tests, excluded tumour elsewhere. However, abdominal MRI and ultrasound revealed a co- incidental polycystic liver.

Three weeks after biopsy, a wide local excision including the medial upper and lower eyelids, dacryocystectomy and medial maxillectomy was performed. A tumour, confined to the sac, and invasion through the medial wall of the upper NLD, into the lateral wall of the nose, and apposing nasal septal mucosa, was seen per operatively and confirmed histologically.

She underwent postoperative adjuvant radiotherapy (55 grays) and to date, 4 months later, remains well.

Comment
Malignant melanoma of the lacrimal sac is rare accounting for 5% of lacrimal sac tumours. It has an unfavourable prognosis compared with other causes of lacrimal sac tumour, and is considered more aggressive than cutaneous malignant melanoma. Treatment is generally poor, with up to 80% of cases recurring within 2 years.

Radiological features of lacrimal sac tumours include filling defects on DCG and mass lesions on CT. However, to the authors’ knowledge, this is the first report of the MRI findings of malignant melanoma of the lacrimal sac.

Owing to the paramagnetic properties of melanin, malignant melanoma appears hyperintense on T1 weighted imaging, and hypointense on T2 weighted imaging. A study of six mucosal melanomas of the head and neck found that on T1, five lesions were hyperintense and one was isointense. On T2, five were of mixed intensity and one was iso-intense. They concluded that hyperintensity on T1 of mucosal melanomas was characteristic but not universal.

The majority of malignant lacrimal sac tumours are epithelial in origin. Imaging features suggesting malignancy include invasion of bone, rapid growth, and irregular margins with skin fixation. On MRI, the majority of epithelial tumours have intermediate signal intensity on T1 and high T2 signal intensity. High tumour cellularity is associated with intermediate to low T2 signal intensity. High signal intensity on T1 is not specific for malignant melanoma. Subacute haemorrhage caused by the presence of methaemoglobin is more likely and although melanoma may undergo intratumoral haemorrhage, other tumours with a tendency to bleed include small cell lung carcinoma, choriocarcinoma, and renal cell carcinoma metastases.

Less likely causes include fat containing tumours (lipoma, dermoid, and teratoma)
reinforced MRI fat suppression methods, paramagnetic material (manganese, iron, and copper), and very high (non-paramagnetic) intratumoral protein concentration.

MRI has been reported as a useful investigative tool in the assessment of lacrimal disease owing to its ability to delineate soft tissues. Intravenous and intracanalicular gadolinium adds useful information on lesion enhancement and lacrimal apparatus structure and function. The predictive value of MRI for lacrimal sac melanoma, however, appears to be variable. Hyperintensity on T1 relies on the paramagnetic properties of melanin, the presence of which is variable in anaplastic melanoma. This is supported by our case, where only moderate T1 hyperintensity with contrast enhancement was demonstrated.

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Photodynamic therapy for recurrent myopic choroidal neovascularisation after limited macular translocation surgery

Limited macular translocation (LMT) is one of the treatment options for subfoveal choroidal neovascularisation (CNV) resulting from pathological myopia. The fundamental surgical principle involves the transposition of the foveal neurosensory retina to a new site with more healthy underlying retinal pigment epithelium. Direct laser photocoagulation is usually employed as an adjunct measure in eradicating the original CNV after the surgery. It has been observed that geometrically sizeable translocation is a prerequisite for a long term surgical success. The degree of translocation is, however, not often predictable and any ineffective displacement may render the subsequent laser photocoagulation extremely difficult or even impossible to perform. As a result, the recurrent or persistent CNV intruding the newly relocated fovea may jeopardise the final visual outcomes. Photodynamic therapy (PDT) may be considered a viable adjunct treatment option in such circumstance.

Case report

A 41 year old woman with pathological myopia of –1.10 dioptres in both eyes presented with a subfoveal CNV and subretinal haemorrhage in her right eye in July 2000. The corrected visual acuity (BCVA) was 5/200 in her right eye and 20/30 in her left eye. LMT with superotemporal 6 mm scleral imbrication was performed in July 2000. The operation was uneventful and an inferior displacement of the fovea by 600 µm was achieved. The CNV, however, was still located in the vicinity of the juxtafoveal area and therefore laser photocoagulation, bearing the potential risk of late creeping scar, was not suggested. At the 4 months postoperative visit, her left BCVA was 20/200 and the original CNV became more fibrotic with minimal leakage upon fluorescein angiogram. Nevertheless, she came back at 5 months with a return of metamorphopsia and a drop in her right vision from 20/200 to 10/20. Dilated fundus examination showed a tiny patch of submacular haemorrhage in direct continuity with the old fibrotic scar (Fig 1A). Fluorescein angiogram of the early phase demonstrated a fresh recurrent CNV budding out from the original fibrotic CNV and extending to the centre of the foveal avascular zone (Fig 1B). Moderate fluorescein leakage could be seen in the late phase (Fig 1C). Treatment comprising revision macular translocation surgery, submacular surgery, photodynamic therapy, and observation had been thoroughly discussed with the patient. In view of minimal invasiveness and comparatively better preservation of surrounding neurosensory retinal tissue, PDT was adopted in treating the CNV recurrence. PDT with verteporfin infusion and laser delivery was performed in accordance with the standard protocol. As a result, the blood clot in the fovea was gradually reabsorbed and the vision improved to 20/200 at 3 months of follow up. Complete regression of the recurrent CNV at the fovea without angiographic leakage was documented over the follow up angiogram at 3 months and subsequently (Fig 1D). The vision remained stable at 20/200 in the latest visit at 24 months after the PDT.

Comment

It has been shown that significant visual improvement may be achieved by LMT for the treatment of subfoveal CNV associated with age related macular degeneration (AMD) or pathological myopia. However, the surgical techniques are demanding and the potential complications are not unusual. One of the late postoperative visually important complications is recurrence of the CNV and this is partially caused by an ineffective translocation of the fovea or a large lesion size of CNV. The incidence of persistent or recurrent CNV after limited LMT has been reported to be 40% and 35% respectively in age related macular translocation and being 21% and 14% respectively in pathological myopia. Not many treatment options are available once the fovea is involved. Viable surgical options including repeated LMT, full 360 degree retinotomy MT, or submacular surgery may be considered but the surgical risk may be inadvertently higher in the redetracement of the neurosensory retina. PDT induces a selective thrombosis of the abnormal CNV and has been proved to be an effective treatment in preventing a significant loss of vision in patients with CNV secondary to AMD or pathological myopia.

Figure 1 Right eye with recurrent myopic CNV after LMT. (A) Fundus photograph of the patient showing the recurrent part of CNV budding from the original one with haemorrhage involving the subfoveal area. (B) Early phase fluorescein (FA), demonstrating the filling of choroidal vascular complex with early hyperfluorescence. (C) Late phase FA showing moderate fluorescence leakage from the CNV. Photodynamic therapy (PDT) with the size of the laser spot as marked was delivered. (D) Late phase FA at 12 months revealing a complete regression of the recurrent CNV and late scar staining of the original CNV.
Acquired Glanzmann’s thrombasthenia causing prolonged bleeding following phacoemulsification

Phacoemulsification under topical anaesthesia has not been used in ophthalmic surgery before. Extensive haematological evaluation revealed the underlying cause to be an acquired form of Glanzmann’s thrombasthenia, a very rare condition.³

Case report

A 79 year old woman underwent left phacoemulsification with intraocular lens implantation under topical anaesthesia through a clear corneal temporal incision. The procedure was uneventful but she was seen to bleed from the operated eye in the recovery room. The eye was patched but the bleeding continued soaking the pads. When re-examined 2 hours later, as there was continuous bleeding, the eye was patched with gentle pressure. Examination the next day showed that the bleeding was persistent. Pressure bandage was reapplied. Examination in the operating theatre confirmed the conjunctival origin of the bleeding from the site where the left hand surgeon had conjunctiva during surgery. Cauterisation and an attempt to suture the conjunctiva were unsuccessful. It was decided that the safest option was to use a small piece of oxidised regenerated cellulose (Surgicel, Ethicon) on the bleeding site and patch the eye.

The piece of Surgicel with clotted blood that was lying loose on the conjunctiva was removed at review 24 hours later. The conjunctival site had stopped bleeding with evidence of altered blood on the surface where Surgicel had been applied (Fig 1A). At her last review 8 weeks later, she was found to have a corrected visual acuity of 6/18 due to pre-existent macular changes secondary to retinal detachment that was reattached in 1976. The conjunctiva had healed well (Fig 1B). The patient had previously undergone an uneventful phacoemulsification and intraocular lens implantation in her right eye under sub-Tenon’s anaesthesia.

The patient’s recent medical history was significant for recurrent admissions elsewhere for investigation of severe anaemia following gastrointestinal bleeding. Platelet count and clotting screen had been normal. Angiodyplasia of stomach and duodenum were treated with laser and angiodyplasia of colon was treated by hemicolecystomy. Three episodes of epistaxis and an episode of vaginal bleeding were managed conservatively. She had received 60 units of blood transfusion over a period of 1 year. Interestingly, she had appendicectomy and multiple dental extractions elsewhere many years previously without any significant bleeding. She has not been on any antiplatelet agents or anticoagulants. There was no family history of bleeding disorders.

A defect in the platelet function was suspected, as her coagulation screen including the platelet count was normal. Platelet aggregation tests showed no aggregation against any agonists other than ristocetin, which is dependent on platelet glycoprotein Ib. The platelets showed normal normal level of glycoprotein antibodies. The patient’s serum showed presence of inhibitory antibody against glycoprotein Ib/IIa. This led to a diagnosis of acquired Glanzmann’s syndrome, an extremely rare condition of autoimmun thrombasthenia. No underlying malignant, autoimmune, or lymphoproliferative disorder had been identified as a cause for this patient’s acquired Glanzmann’s thrombasthenia.

Comment

The patient described had uncontrollable bleeding for 36 hours following a procedure, which is generally considered safe for a patient with a bleeding disorder. She developed bleeding from the conjunctival site where the surgeon grasped the conjunctiva during certain stages of the procedure. One would usually not expect any significant bleeding from this site; however, in a patient with compromised haemostasis the bleeding may be prolonged. Although the bleeding was no more than a gentle ooze at any point in time it was persistent enough for 36 hours before the topical haemostatic material Surgicel had been put to use. The consequences of an intraocular bleed may have seriously threatened her sight.

We are not aware of any reports of the use of Surgicel in ophthalmic surgery. All reports of its use are in other fields of surgery.†† This material is supposed to swell up with blood and form a gelatinous mass that aids in the formation of clot. It acts as a haemostatic adjunct. The exact mode of its action in this patient with antiplatelet antibodies is unclear. Our experience shows that oxidised regenerated cellulose (Surgicel) may have a role in ophthalmic surgery especially in lacrimal and orbital surgery, when faced with bleeding that is difficult to stop. Various cautionary tales associated with use of Surgicel have been reported.‡‡

Our report suggests that in the presence of a severe bleeding disorder, clear corneal phacoemulsification under topical anaesthesia may not be totally safe. When performing such a procedure in a patient with known bleeding disorder it may be safe to take all the necessary precautions in consultation with a haematologist to avoid a serious bleed that may be sight and life threatening. There may be a role for haemostatic agents like Surgicel.

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REFERENCES


Acquired Glanzmann’s thrombasthenia causing prolonged bleeding following phacoemulsification

Phacoemulsification under topical anaesthesia using clear corneal incision is not a challenging procedure for the haemostatic system. In patients with known bleeding diathesis, this may be the procedure of choice to remove cataract. We report a patient who bled continuously for 36 hours following phacoemulsification under topical anaesthesia through a clear corneal incision. This was managed by using a topical haemostatic agent that has not been used in ophthalmic surgery before. Extensive haematological evaluation revealed the underlying cause to be an
Institute for further evaluation. Vitreal cytology slides were sent to the National Eye Institute unresponsive to corticosteroid treatment.

Although Propionibacterium acnes, a Gram positive anaerobic bacillus, is the most commonly identified cause of delayed onset postoperative endophthalmitis, routine vitreous cultures are frequently inadequate for its diagnosis. This case describes the utility of the histopathological technique of microdissection and polymerase chain reaction (PCR) for the diagnosis of delayed postoperative endophthalmitis.

**Case report**

A 78 year old man with a history of vitreous floaters, a coronary bypass, and aortic valve replacement underwent an uncomplicated cataract extraction with intraocular lens (IOL) implantation in the right eye. Three months later, he developed increasing floaters in the right eye and was diagnosed with vitritis unresponsive to corticosteroid treatment. Examination revealed acuities of 20/25 in the right eye and 20/20 in the left with normal intraocular pressures. The right eye was significant for no anterior chamber cells or flare, dilated iris vessels, and peripheral pigmentary degeneration. The left eye was normal with the exception of trace vitreous cells and a choroidal naevus. A diagnostic vitrectomy was performed in the right eye. A portion of the vitreous specimen was cultured for fungi, aerobic and anaerobic bacteria, and the remainder was processed for cytopathological examination. All cultures for micro-organisms were negative.

The vitreous supernatant and unstained cytology slides were sent to the National Eye Institute for further evaluation. Vitreal analysis for interleukin 2 (IL-2), IL-4, IL-6, IL-10, IFN-γ, and TNF-α using ELISA (Endogen, Woburn, MA, USA) revealed undetectable cytokine levels. The vitreous slides were stained with Giemsa, Gram, and immunohistochemical stains for T cells, B cells, and macrophages. Cytopathological examination showed clusters of macrophages admixed with CD4+ and CD8+ T cells and B cells (Fig 1A). Gram positive bacilli were seen in the cytoplasm of a few macrophages (Fig 1B). The engulfed bacilli were microdissected under a microscope with a 30 gauge needle and submitted for PCR. Nested PCR with P. acnes specific oligodeoxynucleotide primers complementary to regions of 16S rDNA was used. The primers were Pa1, AAG GCC CTG CTT TTG TGG; Pa2, TCC ATC CGC AAC CGC CGA A; and Pa3, ACT CAC GCT TCG TCA CAG. Nested-PCR analysis revealed P. acnes (Fig 2). A diagnosis of delayed postoperative endophthalmitis was made.

**Comment**

The most common causes of vitritis in elderly patients are acquired or postoperative infections, sarcoidosis, and intraocular malignancies masquerading as uveitis. An early diagnostic procedure is indicated if postoperative endophthalmitis is suspected. In this case, although the chronic inflammation and intracytoplasmic Gram positive bacilli in a few macrophages suggested an infectious process, the negative cultures precluded the diagnosis of an infectious endophthalmitis. To further investigate the possibility of a bacterial infection nested PCR was performed on the micro-dissected bacilli. Molecular analysis verified the presence of P. acnes and a diagnosis of delayed postoperative endophthalmitis was confirmed.

Vitreous cultures are positive in less than 50% of postoperative endophthalmitis cases. In a study of 23 patients with delayed onset endophthalmitis aqueous culture and microscopy were diagnostic in 0% of cases, vitreous culture was positive in 24% and PCR from the aqueous and vitreous yielded a positive diagnosis in 84% and 92%, respectively. Treatment of P. acnes endophthalmitis includes intravitreal vancomycin plus consideration of pars plana vitrectomy with or without capsulotomy with or without IOL removal. Although aggressive surgical intervention eradicates the infection similar visual outcomes are reported with more limited surgical treatment.

In our case the intracytoplasmic bacteria in the macrophages were the only evidence of a bacterial infection. To detect the presence of P. acnes we referenced the PCR method described by Hykin that used 150 µl of the vitreous for culture and 100 µl for PCR. Using the technique of microdissection and PCR with a similar volume of vitreous we additionally performed cytology and cytokine analysis which are helpful in the diagnosis of other causes of vitritis.

This case further illustrates the benefits of molecular analysis for the diagnosis of culture...
negative delayed onset endophthalmitis. It also describes for the first time microdissection and PCR for the evaluation of endophthalmitis. Advantages of this technique are that it allows for a more comprehensive pathological examination on a limited specimen and provides the option of having the molecular studies being performed elsewhere.

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Interferon treatment of childhood conjunctival lymphoma

Mucosa associated lymphoid tissue (MALT) lymphoma is the most common ocular adnexal neoplasm. The disease is usually limited to localised (stage I) disease at presentation, and radiotherapy and chemotherapy have been the mainstay of treatment.1

Case report

A 15 year old male was referred by an ophthalmologist after an 8 month history of unusual painless follicles at both nasal corners (Fig 1A). There were no visual symptoms and, based on a working diagnosis of an atypical vernal reaction, topical steroid treatment had resulted in mild size reduction of the lesions. Incisional biopsy was performed after the lesions remained static for 3–4 months.

The patient’s visual acuity was 6/4 in both eyes and intraocular pressures measured 15 mm Hg in each eye. Slit lamp examination demonstrated small follicular deposits in both nasal fornices and nasal palpebral conjunctiva. The rest of the ocular examination was unremarkable. Review of systems was negative and the patient’s past medical history and family medical history did not reveal the presence of lymphoproliferative or autoimmune diseases. There were no findings suggestive of Sjögren’s syndrome and physical examination was normal.

The limited amount of biopsy tissue was divided for routine processing and flow cytometry; frozen tissue was therefore unavailable. Histologically a dense lymphoid infiltrate including benign appearing lymphoid follicles was identified (Fig 1B). Lymphoid follicles were surrounded by centrocytic-like cells and small lymphocytes, some of which infiltrated the conjunctival epithelium. Flow cytometry identified a monoclonal B cell population with a CD5−, CD20−, CD10 equivocal phenotype. The histopathological findings in isolation may have represented either an early marginal zone lymphoma or a benign B cell follicular hyperplasia. Absolute distinction on the small amount of tissue was not possible. However, in conjunction with the flow cytometric finding of a monoclonal B cell population, a diagnosis of low grade B cell lymphoma (probably of MALT type) could be made.

Systemic disease was excluded after the following investigations: lumbar puncture; bone marrow aspirate and trephine; CT chest, abdomen, pelvis and sinuses; gallium scan. The patient was subsequently treated with 10 intralymphatic injections of 10 × 10^6 IU of interferon alfa (IFN-α) over a 4 week period; no side effects were noted during this time. Complete resolution was achieved at 2 months, with no sign of recurrence after 18 months’ follow up.

Comment

Conjunctival lymphoma is mostly a disease of the elderly, with Shields et al reporting a mean age of diagnosis of 61 years.1 While not a common disease, Akpek et al suggest that its prevalence is higher than previously recognised, and that vigilance is required in patients with chronic ocular irritation and conjunctivitis who do not respond to conventional therapy.1 This is the youngest case of conjunctival lymphoma that we know of in the literature; hence conjunctival lymphoma should be considered in the differential diagnosis of atypical conjunctival lesions in younger patients.

Treatments outlined by Shields et al included radiotherapy (44%), complete excisional biopsy (36%), observation (9%), chemotherapy (6%), and cryotherapy (4%).1,2

Radiotherapy has been widely used with successful results but ocular morbidity in the form of corneal ulcer, radiation induced cataract and ocular lubrication disorders have been reported.1,4 Intralesional IFN-α is a relatively new therapy which has been shown to be both effective and safe in a small number of cases.1,4,5 Non-sight threatening ocular complications such as subconjunctival haemorrhage and local chemoresistance have been reported, as well as minor transient systemic effects including headaches, nausea, fevers, chills, and myalgia.1 Administration of intralesional IFN-α is also a relatively simply and quick procedure. It shows great promise as a first line agent to treat conjunctival lymphoma, but long term follow up is needed.

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References


Unilateral corneal anaesthesia and ulceration following squint surgery in a child with Pendred syndrome and bilateral sixth nerve palsy

We present a 4 year old child with Pendred syndrome and bilateral sixth nerve palsy. To our knowledge this association has not been previously reported. In addition, this patient developed unilateral corneal ulceration with associated corneal anaesthesia following squint surgery. We will discuss the pathophysiology of this unusual complication following squint surgery.

Case report

This patient presented when he was 6 months old with right congenital squint. He was diagnosed with Pendred syndrome (senso-neural hearing loss and thyroid dysfunction) by the paediatricians and the otolaryngologists following abnormal thyroid function tests and a computed tomograph (CT) scan of the temporal bones showing Mondini malformations of both cochleas. At presentation his visual acuities were 6/60 right eye and 6/36 left eye using the Cardiff acuity cards. He had bilateral alternating esotropia with an inability to abduct either eye. There was no globe retraction or abnormal lid movements and a magnetic resonance imaging (MRI) scan had shown congenital absence of the auditory nerves but no other abnormality. A diagnosis of bilateral sixth nerve palsy was made. The squint was cosmetically poor and measured at 45 prism dioptries in the distance and near. He had low hypermetropia with no significant anisometria. Funduscopy was normal. He was reviewed regularly in the paediatric eye clinic over the next 3 years during which time his visual acuities were within normal limits, the best recorded acuity being 6/9 right eye and 6/9 left eye using singles.

When he was 4 years old, he underwent bilateral superior rectus and inferior rectus lateral transpositions under general anaesthesia, which was eventful with no immediate postoperative complications, and a cosmetic improvement with alternating convergent squint of 15 prism dioptries for distance and near.

Two months later he developed a left inferior corneal ulcer (Fig 1) with surrounding punctate epitheliopathy which surprisingly did not seem to cause him as much distress as expected. The left corneal sensation was definitely reduced compared to the right which appeared normal. Sensation was assessed clinically (an anaesthesiometer was not available), and was consistently reproducible by different ophthalmologists. There was no exophthalmos or any other sign of thyroid orbitopathy. The right eye remained asymptomatic. Empirical therapy with topical ofloxacin and lubricants was unhelpful. He proceeded to have glue tarsorrhaphy which transiently aided the healing of the corneal ulcer. However, the ulcer quickly recurred when the tarsorrhaphy reversed. He subsequently had left inferior lid shortening with a cantorial sling to elevate the lower lid to protect the corneal epithelium. The ulcer resolved leaving an area of corneal scarring. He is being reviewed regularly in the eye clinic.

Comment

Pendred syndrome is an autosomal recessive disorder characterised by congenital deafness and thyroid gland goitre, although goitre is usually severe and is present at birth, and the goitre generally appears at puberty or later but may be present in early childhood with an associated euthyroid or hypothyroid state. The affected individuals are reported to be otherwise normal.

The pathophysiology of the corneal anaesthesia and ulceration in this patient is uncer- tain. There are several possible reasons for the corneal anaesthesia. They include herps simplex keratitis, postoperative anterior segment ischemia, surgical trauma to the poste- rior ciliary nerves or ciliary ganglion, congenital absence of sensation, and surgery reducing Bell’s phenomenon.

The clinical course was not typical of herps simplex and there was no previous history of corneal pathology. Postoperative anterior ischaemic syndrome was unlikely as only two recti muscles were operated on and no anterior uveitis was observed. To our knowl- edge there are no reported cases of corneal anaesthesia after squint surgery. There was no evidence of prior ocular involvement, which one may expect with trauma to the long posterior ciliary nerves or ciliary ganglion.

Congenital absence of corneal sensation was the most likely cause, especially in view of his unusual cranial nerve anomalies, and we believe he had pre-existing corneal anaesthesi- a before squint surgery despite the absence of any other fifth cranial nerve signs. Follow- ing the lateral transposition of the superior rectus his Bell’s phenomenon was noted to be absent thereby compromising his corneal protection. In addition, he was observed to have significant lagophthalmos while asleep. We believe that the combination of corneal anaesthesia, abolished Bell’s phenomenon, and lagophthalmos compromised his corneal integrity resulting in corneal ulceration.

This case highlights the importance of determining corneal sensation before trans- position surgery on the superior rectus as Bell’s phenomenon may be abolished there- fore compromising corneal protection. This is especially relevant in patients with unusual cranial neuropathy and lagophthalmos.

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Gemella haemolysans acute postoperative endophthalmitis

Endophthalmitis is perhaps the most feared complication of cataract surgery, with a reported incidence between 0.13% and 0.19%. The most common organisms reported in previous studies are Gram positive staphylococci and streptococci. We report a case of severe endophthalmitis with an unusual Gram positive organism, after uncomplicated phacoemulsification, with foldable intraocular lens implantation.

Case report

A 66 year old white man underwent routine phacoemulsification cataract extraction with posterior chamber lens implantation (Acrylic, Model H60M, Bausch & Lomb) to the right eye in January 2002. The left eye had previously undergone similar surgery in September 2001. He was generally in good health, and on no medi- cation. There was a past medical history of sarcoidosis treated with oral prednisolone in 1970, which has since been in remission, and an episode of staphylococcal septicaemia in 1987, without sequelae.

On the first postoperative day, visual acuity measured 6/9 unaided and ocular examination was unremarkable. That same afternoon the patient developed ocular pain, initially relieved by paracetamol (acetaminophen), which however, worsened during the night with progressive deterioration of vision. He presented to the ophthalmic emergency de- partment the following morning with the aforementioned symptoms. Visual acuity was reduced to hand movements right eye and 6/9 left eye. Slit lamp examination revealed an oedematous cornea with Descemet’s folds. The anterior chamber was hazy, with 1 mm hypopyon and a intraocular pressure measured 38 mm Hg.

There was no red reflex. B-scan ultrasound examination showed extensive vitreous debris with attached retina. The left eye was pseudo- phakic with no abnormalities of note. A diag- nosis of acute postoperative endophthalmitis was made. Anterior chamber and vitreous samples were obtained for aerobic and anaerobic culture/sensitivity and Gram stain- ing. Intravitreal vancomycin 2 mg and ami- kakcin 300 µg, each in 0.1 ml of balanced salt solution and subconjunctival ceftroxime 125 mg were administered. Oral ciprofloxacin 500 mg once daily, prednisolone 1 mg once a day, topical gentamicin hourly, ofloxacin hourly, and atropine 1% twice a day were commenced.

Preliminary Gram staining suggested a Gram positive coccus. Therapy was started with ciprofloxacin—oral and topical antibiotics were therefore continued. Owing to difficulty in identifying the nature of the organism, the samples were sent to a regional reference laboratory, which identified *Gemella haemolysans* from both anterior chamber and vitreous aspirates. The organism was reported to be sensitive to gentamicin, ciprofloxacin, laevo- floxacin, amoxicillin/clavulanate, chloramycetin, and resistant to trimethoprim.
The patient continued to make steady progress; 2 months later vision had improved to 6/9 unaided. The patient at that time was troubled by floaters secondary to considerable vitreous debris. At last review in September 2002, visual acuity had further improved to 6/4 with −0.75=Dph correction.

Comment
Gemella haemolytica is an aerobic or facultative anaerobic, Gram positive coccus, a normal commensal of the oral cavity and upper respiratory tract of low virulence.1 2 Systemic infection may lead to septic shock, meningitis, arthritis, or pneumonia, all of which are rare. Identification is difficult. Though Gram positive, the cocci are easily decolourised and hence may appear Gram variable or even negative. Initially Gemella was included under the genus Neisseria but is now classified as a separate genus within the family Streptococcaceae.3 No studies on susceptibility to antiseptics have been published, though there is no reason to believe that it may be resistant to povidone-iodine preparations. The organism is susceptible in vitro to penicillin, streptomycin, vancomycin, chloramphenicol, and tetracyclines. A literature search revealed only one previously reported case of infection by Gemella haemolysana, with keratitis and consecutive cataract surgery.4 5


Does topical brimonidine tartrate help NAION? There is no proved treatment for non-arteritic anterior ischaemic optic neuropathy (NAION). Topical brimonidine tartrate has been reported to have a neuroprotective benefit for retinal ganglion cells following experimental elevation of intraocular pressure and optic nerve injury in the rat, which is blocked with coadminstration of the α2 antagonist, rauwolscine.6 Increased retinal ganglion cell survival has also been shown to occur following oral administration of brimonidine in monkeys with experimental glaucoma.7 These results were the basis of the recently aborted clinical trial of topical brimonidine for acute NAION and our retrospective study of 31 patients with NAION, who were evaluated within 3 weeks of the onset of visual loss and follow up for a minimum of 8 weeks. During 2001–2, we treated all (14) patients with brimonidine tartrate within 14 days (mean 3.5, SD 5.52) of the onset of visual loss. Five patients were treated after 1 day of symptoms. Brimonidine was taken 8.3 times a day in one, 12.3 times a day in another and twice a day in two patients. All (17) untreated patients were evaluated the year before and were matched to the treated group for age, sex, cardiovascular risk factors, previous aspirin use, and previous first eye NAION.

Snellen visual acuity and colour vision, using the Ishihara colour plates, were documented and expressed as a decimal equivalent (for acuity: 20/60 = 0.33 and light perception = 0.001; for colour vision: the number of correctly identified plates/the total number of plates). The virostat (Humphrey or tangent perimetry) were analysed and defects were graded according to the following scale: 0 = normal, 1 = acute nerve fibre bundle defects, 2 = relative central (<6 degrees), 3 = macular defects, 4 = no light perception. A third examiner, who was unaware of the dates of the visual fields and the patients' treatment status, also evaluated all visual fields and determined, in each patient, whether the field was better or worse than or equivalent to the other field. The intraocular pressure was measured by Goldmann tonometry on except two patients. The pressure was 25 mm Hg in one patient in the treated group and 24 mm Hg in one patient in the untreated group.

Statistical analysis of the data involving comparisons of the treated and untreated groups at baseline and 8–12 weeks was performed using the two tailed t test. The Wilcoxon signed rank test was used to compare the individual visual performance changes from baseline to the 8–12 week examination.8 For visual acuity and colour vision, a positive rank indicated improvement and a negative rank indicated a worse visual outcome. For the visual field grade, a decrease in the grading scale was reversed. Spearman correlation analysis was performed on the time to start therapy and whether worsening in any visual parameter occurred.9 The mean baseline visual field (0.40, SD 0.41; p=0.22) and field (1.9, SD 0.75; p=0.86) for controls. The mean baseline colour vision (0.74, SD 0.44) for the treated group was worse than the colour vision (0.45, SD 0.44) for controls, but the difference was not significant (p=0.07). At the 8–12 week examination, the mean visual acuity was 0.29 (SD 0.30) for treated and 0.49 (SD 0.39; p=0.12) for untreated patients. The mean visual field grade was 2.2 (SD 0.81) for treated and 1.0 (SD 0.70; p=0.04) for untreated patients. The mean colour vision was 0.42 (SD 0.41) for treated and 0.55 (SD 0.46; p=0.43) for untreated patients. For the masked examiner's evaluation, the mean baseline visual field (2.0, SD 0.91) was similar to the field (1.93, SD 0.96; p=0.85) for controls. At the 8–12 week examination, the mean visual field grade was 2.15 (SD 0.99) for treated and 1.87 (SD 0.92; p=0.43) for untreated patients. This examiner further found that the outcome visual fields for the treated group were improved in two patients, worsened in six patients (50%), and unchanged in four patients. The outcome visual fields for the control group were improved in five patients, worse in two patients (13%), and unchanged in eight patients.

The Wilcoxon signed rank analysis demonstrated that for visual acuity, two patients in the control group and 10 patients in the treated group had negative values or a worse outcome at 8–12 weeks (p=0.046). For colour vision, one patient in the control group and eight patients in the treated group had negative values or a worse outcome (p=0.013). For visual fields, one patient in the control group and four patients in the treated group had positive values or a worse outcome at 8–12 weeks (p=0.046).

The average time to start the drops was 3.5 days from the onset of visual loss in those patients who worsened. There was no correlation with a worse outcome and time to initiate therapy.

For all parameters of vision testing, there was a trend for worse visual performance at 8–12 weeks in the group treated with topical brimonidine. Although there was no significant difference for the colour vision outcome, this might reflect that the baseline colour vision value was better for the untreated group. The outcome visual field grade was significantly worse in the treated group. The masked examiner's visual field evaluations demonstrated that more treated patients worsened than in the untreated group. When the baseline and outcome of all visual parameters for each individual were compared, the treated group had a significantly worse outcome at 8–12 weeks.

Our results are not the first description of worse outcome in patients treated with α2 agonists for central nervous system ischaemic disease. Studies in animal models and clinical studies in humans suggest that α2 agonists, including α2-receptor agonists, may impede recovery following stroke. Clonidine administration caused regression of the neurological deficit in animals who had initially recovered. In a recent clinical study, the level of motor recovery of stroke patients was worse in those treated with α2 agonists than in patients not receiving these agents.10

Although in experimental optic nerve injury in animal models, brimonidine appears to offer neuroprotection, our results demonstrate that brimonidine tartrate, applied topically up to four times daily, does not appear to be a beneficial treatment for acute NAION. It
is possible earlier treatment might have been more effective, although patients who worsened received treatment sooner than those who did not worsen. Increased dosing frequency or using a different preparation of botulinum toxin might be more effective. Additionally, the number of subjects in the study was small and a negative trend could appear more profound.

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References

Chronic eye movement induced pain and a possible role for its treatment with botulinum toxin

Chronic ocular pain may have many causes and can be a frustrating problem for both patient and doctor alike. We describe two patients who had similar symptoms and eye findings who had been unable to relieve their pain with conventional analgesia. We postulate a cause for their pain and describe our experience of a treatment strategy using a standard dose of botulinum toxin injection into an extraocular muscle.

Case 1

A 36 year old white woman presented with what was initially thought to be a right orbital cellulitis but investigations and clinical course subsequently suggested a non-infectious idiopathic inflammatory aetiology. Her history suggested orbital myositis and she described right sided facial weakness, nausea, and right sided ptosis. She had a 9 month course of oral steroids and despite this needed tramadol, paracetamol, and flurbiprofen to control her pain. Her symptoms and examination findings slowly stabilised until she was left with marked limitation of upgaze in her right eye. Her symptoms did not change over the next 3 years, at which point she was referred to our care. When she attempted to look up she described a juddering sensation and severe pain just above the eye. She rarely had pain at night but was still using regular oral ibuprofen for pain relief. Her pain was exacerbated by reading or looking at the computer and she complained of vertical diplopia.

On examination she had limitation of abduction and elevation of her right eye and prisms did not improve her symptoms. A tentative diagnosis of inflammatory spasm was made. She was treated with botulinum toxin injection to her right inferior rectus. Two weeks later there was much less tightness and discomfort in the orbit but she had diplopia in all positions of gaze and was forced to occlude one eye. Three months later the pain was much improved. She still found the diplopia intolerable and declined further treatment.

Case 2

A 46 year old white man presented complaining of chronic constant ocular discomfort which followed strabismus surgery 8 years earlier for an A-pattern exotropia with diplopia on downward gaze. The pain was worsened by prolonged television watching and prisms in his glasses did not help. Pain was much worse on upgaze and right gaze, which were limited. Oral non-steroidal anti-inflammatory agents (NSAIDs) did reduce the pain a little but only when taken in high doses (100 mg three times daily flurbiprofen).

On examination he had a right hyperphoria, with an A-pattern exotropia and an abnormal head posture for distance. He still had diplopia. Botulinum toxin was injected into his left medial rectus muscle, which resulted in a profound reduction in his symptoms, leaving him with a small exophoria. His diplopia resolved completely after 10 weeks. The “pressure sensation” and pain in the right eye recur after about 6 months, this time with no diplopia. He had a further injection of botulinum toxin 8 months after the first which again significantly improved his pain but gave him diplopia for 3 weeks. He continues to take flurbiprofen 50 mg three times daily orally.

Comment

The pain demonstrated by these two patients is typically much worse in certain directions of gaze and particularly during prolonged gaze holding such as when reading or watching television. It had a clear precipitating event and the most remarkable feature is that it had persisted for over 2 years in each case without significant progression or regression. No active disease process could be found to account for the continued pain. The pain is severe and responds only to high doses of analgesics, particularly NSAIDs. None of our patients felt that their pain was satisfactorily controlled by their analgesics.

We believe that there may be a process of chronic low grade inflammation affecting the extraocular muscles and the tissues around them which is exacerbated by continued contraction and relaxation of the same muscles. Muscular spasm perhaps triggered by this inflammatory process may be the cause of the most severe pain and this could account for the exacerbations of pain in certain directions of gaze and on prolonged gaze holding activities. Occular muscle ischaemia, perhaps caused by constraining scar tissue, remains a possibility but the onset of the pain is very fast making this less likely.

The pain relief seen in our patients may simply be the result of paralising an inflamed muscle but there is growing evidence for a separate antinociceptive effect of botulinum toxin. No direct peripheral cutaneous antinociceptive effect could be shown by Biersch et al., however inhibition of release of substance P has been demonstrated in vitro and it can be hypothesised that botulinum toxin treatment may reduce the local release of nociceptive neuropeptides from either cholinergic neurons or from C or A delta fibres in vivo. The mechanisms by which botulinum toxin may relieve pain, including a possible analgesic effect of botulinum toxin metabolites, are reviewed by Guyer.

There is a growing literature on the use of botulinum for painful conditions, particularly those in which muscle spasm plays a part. These include writer's cramp, postoperative pain in spinal cerebral palsy, and perhaps more surprisingly writer’s cramp and painful tic convulsif. Many of the reported uses are single case studies and not all controlled trials have shown a positive effect of treatment.

It is not possible to rule out a powerful placebo effect in our patients but, whatever the mechanism of action, their pain was vastly improved and botulinum toxin treatment is very safe in competent hands.

In these cases described botulinum toxin served a dual purpose in that it had the potential to improve their ocular deviation for which it is well known and it also reduced the severe ocular discomfort. Unfortunately, the resulting diplopia limited its usefulness in one case but we feel that this treatment should be considered in this unusual group of patients who present a difficult management problem even to the most experienced ophthalmologists.

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www.bjophthalmol.com
Intrastromal lamellar femtosecond laser keratoplasty with superficial flap

Lamellar keratoplasty has usually been performed taking a trephine to delineate the extent of the tissue to be excised, and a knife or similar instrument to remove the lamellar corneal tissue from the underlying deep corneal bed. In a simple way, the lamellar donor tissue was prepared and inserted into the recipient bed. The depth of the lamellar excision from the corneal epithelial surface to the deep corneal stroma. Marked disadvantages of the technique have been pronounced corneal astigmatism and optical insufficiencies between the interface of the lamellar graft and the recipient corneal bed caused by irregularities of both surfaces. The purpose of the present report was to describe the new femtosecond laser technology, which may allow us to perform a new type of intrastromal lamellar keratoplasty with preservation of an intact Bowman’s layer and regular corneal epithelium.

Case report

Using a corneal contact lens and a femtosecond laser (20/10 Perfect Vision, AmoBensfeld 21/1, D-69123 Heidelberg, Germany) with a wavelength of 1060 nm, a spot size of about 10 μm, and a laser pulse duration of several hundred femtoseconds, a pre-descentmal incision running parallel to the corneal surface was created in five postmortem eyes of slaughterhouse pigs. The diameter of the deep stromal incision was 7 mm. In a second step, a circular sagittal incision was performed starting from the peripheral edge of the already existing incision in the pre-descentmal level to the superficial layer of the corneal stroma. In continuation of the latter sagittal incision, the corneal flap was prepared with a diameter of 7 mm, a thickness of about 100 μm, a hinge, and three positional pikes. The pikes in the flap with the corresponding nucleus in the recipient bed of the flap were formed to increase the rotational stability of the flap after repositioning. The height of the peaks was about 0.40 mm. After opening of the flap the intrastromal segment situated between the pre-descentmal incision and the incision in the superficial stromal level was removed with removal of a mid-stromal segment and preservation of an intact Bowman’s membrane. Considering the decreased amount of allogenic corneal tissue transplanted, and regarding the preservation of the original corneal surface, lamellar intrastromal femtosecond laser keratoplasty may be associated with a smaller rate of immunological graft reaction and with a lower postoperative corneal astigmatism in some eyes. Future clinical studies may show whether positional edges in the superficial flap increase its postoperative rotational stability.

Proprietary interest: none

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Demographic study of paediatric allergic conjunctivitis within a multiethnic patient population

From October 1999, all patients referred to the paediatric ophthalmology service in Bradford have been added to a computerised database. This is the only paediatric ophthalmology service within the city of Bradford and receives all GP referrals of this type. Patients with a clinical diagnosis of chronic allergic conjunctivitis were identified from October 1999 to July 2001. We compared the relative prevalence of chronic allergic conjunctivitis in Asians and white children. Considering the decreased amount of ocular allergy is multifactorial but perhaps with a greater genetic predisposition in certain ethnic communities. We could not comment on the prevalence of chronic allergic conjunctivitis in the community because of referral bias since we only see patients referred by GPs. The extent of which milder cases are treated in the community is not known but we feel that the more severe cases are the ones referred to our department. Our findings highlight that allergic eye disease appears to be more common in Asian and black patients. This may be due to genetic and environmental factors.

We found allergic eye disease to be more common in Asians than white children. It is possible that ocular allergy is multifactorial but perhaps with a greater genetic predisposition in certain ethnic communities. We could not comment on the prevalence of chronic allergic conjunctivitis in the community because of referral bias since we only see patients referred by GPs. The extent to which milder cases are treated in the community is not known but we feel that the more severe cases are the ones referred to our department. Our findings highlight that allergic eye disease appears to be more common and complicated in Asian patients in the Bradford population. This potential risk of sight threatening disease means that they are more likely to require topical steroid treatment. This has led us to recognise that appropriately aggressive treatment is essential in these patients.

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We wish to apologise for an error in the extended report by Barry and König (www.bjophthalmol.com). The latest issue of *Br J Ophthalmol* (1997; 81: 124–8), which is a useful 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; website: www.eyeconditions.org.uk).

The British Retinitis Pigmentosa Society

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

### Surgical Eye Expeditions International

Volunteer ophthalmologists in active surgical practice are needed to participate in short-term, sight restoring eye surgery clinics around the world. Contact: Harry S Brown, Surgical Eye Expeditions International, 27 East De La Guerra, C-2, Santa Barbara, CA 93101–9588, USA (tel: +805 963 3303; fax: +805 965 3564; email: hsbrown.m@cox.net or seeinl@seeinl.org; website: www.seeinl.org).

### Rise in organ transplant numbers

According to UK Transplant, the UK has seen the highest number of organ transplants in the year to 31 March 2002. This equated to a 6% increase on the highest number of organ transplants in 2001 to 31 March 2002. Furthermore during 2002–3, the highest number of people benefited from a cornea transplant for five years (1997–98) and 240 more people had their sight restored than the previous year. For further information see UK Transplant’s website (www.uktransplant.org.uk).

### Elimination of avoidable blindness

The 56th World Health Assembly (WHA) considered the report on the elimination of avoidable blindness (doc A56/26) and urged Member States to: (1) Commit themselves to supporting the Global Initiative for the Elimination of Avoidable Blindness by setting up a national Vision 2020 plan by 2005; (2) Establish a national coordinating committee for Vision 2020, or a national blindness prevention committee to help implement the plan; (3) Implement the plan by 2007; (4) Include effective monitoring and evaluation of the plan with the aim of showing a reduction in the magnitude of avoidable blindness by 2010; (5) To support the mobilisation of resources for eliminating avoidable blindness. The WHA also urged the Director-General to maintain and strengthen WHO’s collaboration with Member States and the partners of the Global Initiative for the Elimination of Avoidable Blindness as well as aid in the coordination and support of national capability.

### MSc course in Community Eye Health

The International Centre for Eye Health is offering a full time MSc course in Community Eye Health from 29 September 2003 to 19 September 2004. The course is not clinical and is specifically for eye health professionals wanting to work in the field of community eye health. The course is designed in keeping with the aims, priorities, and strategies of Vision 2020—the Right to Sight. The course costs £399 for home students and £14110 for overseas students. Further information is available from the Registry, 50 Bedford Square, London WC1B 3DP, UK (tel: +44 (0)20 7927 2239; fax: +44 (0)20 7323 0638; email: Adrienne.Burrough@lshtm.ac.uk; website: www.lshtm.ac.uk).

### Ophthalmic Anesthesia Society (OAS)—17th Scientific Meeting

The 17th Scientific Meeting of the Ophthalmic Anesthesia Society (OAS) will be held 3–5 October 2003 at the Westin Michigan Avenue Chicago, Chicago, USA. Programme co-chairs: Marc Allen Feldman MD MHS and Steven T Charles MD. The CME joint sponsor is the Cleveland Clinic Foundation; CME hours are pending. Fees for OAS members are $300; non-members $475; students $50. Further details: OAS, 793-A Foothill Blvd, PMB 119, San Luis Obispo, CA 93405 USA (tel: +1 805 534 0300; fax: +1 805 534 9300; email: info@eyeanaesthesia.org; website: www.eyeanaesthesia.org).

### Glaucoma Society 24th Annual Meeting and Dinner

The Glaucoma Society 24th Annual Meeting and Dinner will take place on 20 November 2003 from 8.30 am on Tuesday to 9.00 pm on Wednesday in the Royal College of Physicians, London, UK. Further details: Ms Janet Flowers (email: glauoc@ukiere.freeserve.co.uk).

### Detachment Course with international faculty on: Retinal and Vitreous Surgery with Case Presentations preceding the Annual Meeting of Iranian Society of Ophthalmology

The detachment course with international faculty on: Retinal and Vitreous Surgery with Case Presentations preceding Annual Meeting of Iranian Society of Ophthalmology will be held 1196 PostScript
on 29–30 November 2003 and 1–4 December 2003 respectively, at the Razi Conference Center, Hemmat Hyw, Tehran, Iran. Further details: Scientific programme: Prof Ingrid Kreissig, University of Tuebingen, Schleichstr. 12, Breuningerbau, 72076 Tuebingen, Germany (tel: +49 7071 295209; email: ingrid.kreissig@med.uni-tuebingen.de). Local organisation: Dr Arman Masheyekeh, Dr Siamak Moradian, Dept of Ophthalmology, Labbafinejad Medical Center, Pasdaran Ave, Boostan 9, Tehran, 16666, Iran (fax: +98 21 254 9039; email: labbafi@hotmail.com).

5th International Symposium on Ocular Pharmacology and Therapeutics (ISOPT)

The 5th International Symposium on Ocular Pharmacology and Therapeutics (ISOPT) will take place 11–14 March 2004, in Monte Carlo, Monaco. Please visit our website for details of the scientific programme, registration, and accommodation. To receive a copy of the Call for Abstracts and registration brochure please submit your full mailing details to http://www.kenes.com/isopt/interest.htm.


XVth Meeting of the International Neuro-Ophthalmology Society

Unusual case of residual cortical lens matter in anterior chamber

Modern cataract surgery does not allow for any residual cortical matter in the anterior or posterior chamber, not even in the capsular bag. But for a beginner, a residual cortex in the eye is preferable to a ruptured posterior capsule and its associated complications. Therefore, the surgeon can stop and allow a minor complication (retained cortical material) to prevent posterior capsular rupture.

Case report

A 52 year old female patient was operated for posterior subcapsular cataract in her right eye. Her left eye had previously gone into phthisis bulbii 5 years earlier; she underwent a 360° buckling with vitreoretinal surgery in her left eye for traumatic total retinal detachment 6 years earlier.

The surgery was performed by a resident eye surgeon who was in the learning stage of phacoemulsification. During cortical aspiration, the matter at the 12 o’clock position was provoking difficult to handle for the surgeon. The surgeon therefore thought, in the best interest of the patient, that the amount of sub-incisional cortical matter (approximately 2 clock hours, extending up to the capsul-hexis margin towards the centre) would absorb over time. He did not take the risk of further manipulations and getting a posterior capsular tear.

The surgeon increased the size of the corneal incision and implanted the all-PMMMA (Single-piece, Biconvex, Mod C Step Vault, from AI Optics Ltd, India) intraocular lens. (The patient could not afford any other lens and the above lens is available free of cost for deserving patients in our centre.) The wound was closed with 10/0 monofilament Nylon sutures.

The first postoperative day did not reveal any unusual inflammation. The eye was quiet on third postoperative day (first follow up). At second follow up (10th postoperative day), the operated eye revealed a white, fluffy mass (Fig 1) in the anterior chamber. This cotton wool ball-like mass was diagnosed to be retained sub-incisional cortical lens matter based on normal anterior segment and fundus findings. The IOP was 28 mm Hg in her right eye. Because of the raised IOP and the one eyed status of the patient, immediate removal of cortical lens matter was planned. The side port was used to aspirate cortical matter with topical 0.5% oxybuprocaine (proparacaine) eye drops, using a 23 gauge canula. Postoperatively there was normalisation (off oral medications) of raised IOP within 48 hours. All sutures were removed after 6 weeks (Fig 2). The final best corrected visual acuity was 6/6.

Comment

The case is reported to highlight the importance of complete removal of cortical matter. The reason for difficulty in aspirating sub-incisional cortex in our case was inferiorly decentralised capsulorhexis and corneal oedema at the incision site. Other reasons that can hamper the removal of such cortex could be positive vitreous pressure, long tunnel, fluid leakage due to divarication of incision lips, small capsulorhexis, probable miosis and corneal oedema. The raised IOP in our case could be due to obstruction of trabecular meshwork by lens debris and inflammatory components in the form of foamy macrophages and lens particles and reduction of outflow facility of the anterior chamber angle. Lens debris was seen as a fluffy pseudohyppyony layer in the inferior anterior chamber; this can cause a mistaken diagnosis of postoperative endophthalmitis if associated with anterior uveitis.

The full visual recovery seen in our case could be attributed to immediate surgical intervention. The lens cortex retained in the eye after cataract extractions usually undergoes lyses by aqueous but may persist. The techniques that can be used to aspirate such sub-incisional cortex could be widening of the incision, mobilisation of the mass with IOL, verticalisation of irrigation/aspiration tip, using 180° bent canula by Binkhorst, bent and angled coaxial cannulas, and bimanual (one for irrigation and one for aspiration) technique.

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References


Posterior segment complications of graft versus host disease after bone marrow transplantation

The efficacy of bone marrow transplantation (BMT) for the treatment of selected diseases of the haemopoietic system such as chronic myeloid leukaemia (CML) is well recognised. Graft versus host disease (GVHD) is however a common and potentially life threatening complication of this treatment, occurring in up to 75% of cases. It is thought to arise when immunocompetent donor T lymphocytes mount an immune response against host tissues. GVHD is characterised by a triad of enteritis, dermatitis, and hepatitis, but almost all organs may be targeted. Ocular involvement is frequently seen but is usually limited to the anterior segment. Posterior segment manifestations are rare.

This report describes two cases of GVHD with unusual posterior segment involvement that highlight the diversity of presentations in this condition.

Case 1

A 45 year old white male presented with progressive bilateral blurred vision and floaters 10 months post-BMT for CML. He had no history of ocular disease. His symptoms started 10 days after discontinuation of cyclosporin A as a routine protocol and were accompanied by alopecia, poliosis, vitiligo, and oral mucositis. A presumptive diagnosis of acute GVHD was made and cyclosporin restarted together with prednisolone. His other medications were azathioprine, aciclovir, fluconazole, and ranitidine.

Best corrected visual acuity was 6/9 bilaterally, with no afferent pupillary defect. There was no evidence of inflammation in anterior segments or vitreous and normal intraocular pressures. There was mild disc pallor with swelling and surrounding radial choroidal folds and several pale subretinal perifoveal lesions on the left. Systemic examination revealed widespread patchy alopecia with poliosis, vitiligo of the arms, and oral mucositis. Fluorescein angio- graphy showed mild dilatation of the disc capillaries and extensive focal leakage from the retinal pigment epithelium, but no evidence of cystoid macular oedema. Ultra-
sonography showed a thickened posterior sclera. Optical coherence topography (OCT) showed subretinal fluid bilaterally. Cerebral magnetic resonance imaging (MRI) and lumbar puncture revealed no abnormalities.

The clinical appearances were consistent with posterior scleritis together with a diffuse retinal pigment epitheliopathy. A reducing regimen of high dose steroids in combination with acetazolamide resulted in clinical improvement and visual stabilisation.

Case 2
A 31 year old white female underwent total body irradiation and BMT for \(\gamma \beta\)-T cell splenic lymphoma. One month later she developed acute GVHD related erosive enteropathy resulting in life threatening exsanguination. Following successful resuscitation (which precipitated admission to intensive care for 6 weeks), she noted blurred left eye vision and described difficulty in dark adaptation and differentiating between shades of grey; there was right strabismic amblyopia. The patient’s medication comprised aciclovir, cyclosporin, penicillin, propranolol, and lansoprazole.

Visual acuity was 6/18 and N14 with the right eye, and 6/12 and N5 with the left. Colour vision was normal and visual fields were full. There was no afferent pupillary defect. The anterior chambers and vitreous were quiet. The optic discs were normal. At both maculas (Fig 1), there were deep subretinal cream coloured spots and retinal thickening and OCT evidence of subretinal fluid without cystoid changes. Fluorescein angiography showed a few hyperfluorescent spots consistent with focal retinal pigment epithelium dysfunction. Electrodiagnostic tests identified diffuse rod dysfunction.

Since there was biochemical evidence of renal impairment, acetazolamide was considered to be contraindicated to treat the subretinal fluid. By 4 months, the best corrected visual acuities were 6/12 right; 6/6+2 left. Repeat electrophysiology was unchanged, however by ten months the full field electroretinograms had improved to normal limits.

Comment
GVHD is presumed to be caused by donor T lymphocytes recognising minor histocompatibility antigens on recipient tissues that are then subjected to CD8-T lymphocyte mediated attack. Commonly reported ocular manifestations include pseudomembranous conjunctivitis, keratoconjunctivitis sicca, corneal epitheliopathy, and cataract. Posterior segment involvement is rare and includes cotton wool spots as well as central serous chorioretinopathy.

In both of our cases, there seems to be a striking temporal association between the onset of visual symptoms and an adverse event in the course of the disease. In case 1, in whom the cessation of cyclosporin resulted in acute GVHD, the ocular findings were consistent with scleritis, a feature only once previously reported. Postmortem studies have demonstrated choroidal infiltrates in GVHD patients containing histiocyte-like large eosinophils and clinically these may be represented by the pale perifoveal lesions observed in the left eye. Subsequently this patient was shown to be HLA-DR4 positive, a finding common in individuals with Harada’s disease and frequently associated with chronic GVHD.

By contrast, the ocular findings in the second case were not a result of acute but a consequence of previous life threatening GVHD during which exsanguination occurred. While interruption of blood flow to the optic nerve or visual cortex can account for visual loss following extreme haemorrhage, retinal ischaemia has also been documented. The rod photoreceptor system appears most vulnerable, a feature consis-
tent with the electrophysiological findings in this case, and the patient's difficulty with dark adaptation is in keeping with rod dysfunction. Of interest was the subsequent improvement in acuity and electrophysiological responses. Such a pattern parallels electrophysiological studies of children after respiratory or circulatory arrest where initially subnormal ERG responses return to normal levels within 8 months. The mechanism that mediates this recovery is not known.

Graft versus host disease is a common complication of bone marrow transplantation that usually presents to the ophthalmologist with anterior segment signs. However, GVHD may also present with posterior segment presentations of the types described here.

**References**

**Anterior pathway vision loss due to subdural haematoma**

Patients with vision loss associated with subdural haematomas typically present with homonymous hemianopias secondary to compression of the posterior cerebral artery during trans-tentorial herniation. In these cases, necropsy studies have demonstrated pregeniculate involvement in addition to occipital lobe lesions. We present a case illustrating a rarely reported phenomenon of anterior pathway vision loss associated with a subdural haematoma without any evidence of optic disc swelling, occipital lobe disease, or radiographic signs of chiasmal or optic nerve compression.

**Case report**
A 51 year old man, who had previously undergone two craniotomies (October 1992 and November 2000) for resection of an epidermoid tumour at the cerebellopontine angle, developed hydrocephalus and had a
Magnetic resonance imaging (MRI) of the brain showed that the right subdural haematoma was still present but was decreased in size to 2.1 cm on coronal section (Fig 2A) compared to the study performed 1 month earlier. No intraorbital abnormalities were present, and the optic nerves and chiasm appeared free of direct compression by the haematoma. The blood did not appear to surround the optic nerves (Fig 2B). The patient's haemocrit and blood pressure remained within normal limits during the initial presentation and subsequent treatment. The patient underwent craniootomy with further drainage of the subdural haematoma. After 6 months, the patient's vision in the left eye improved to 20/200. Follow up perimetry showed less constriction on the right and improved performance on the left (Fig 1D). Funduscopic examination revealed mild optic nerve pallor in the left eye and a normal appearing right optic nerve.

Figure 2  Magnetic resonance imaging (T1 weighted coronal) of (A) right subdural haematoma with significant right to left midline shift but without direct compression of the prechiasmal optic nerves and (B) without compression of the intraorbital optic nerves.

ventriculoperitoneal shunt placed in January 2001. Eleven months following placement of the shunt, the patient presented with ataxia and headaches but no visual complaints. A computed tomograph (CT) scan showed a right subdural haematoma measuring 2.8 cm on coronal section, and 150 ml of blood were drained via a burr hole.

One month following the drainage procedure, the patient presented to the hospital with a complaint of sudden, painless loss of vision in his left eye occurring 24 hours earlier. The patient had a left afferent pupillary defect with best corrected vision of 20/30 in his right eye and hand movements in his left eye. Extraocular movements were full, and the patient had normal colour vision in his right eye. Slit lamp examination showed only trace nuclear sclerosis cataracts bilaterally. Fundus examination was also normal, with no evidence of optic disc oedema or pallor in either eye (Figs 1A, B). Automated perimetry revealed a constricted field in the right eye and severe, global depression in the left eye (Fig 1C).

This case represents a rare example of anterior pathway vision loss due to subdural haematoma. Most cases of vision loss with subdural haematoma affect the posterior visual pathway, with mechanisms including ocipital infarct and compression of the posterior cerebral artery during trans-tentorial herniation. 1,2 Posterior lesions may present with anterior signs—for example, optic atrophy was seen in three patients with occipital infarcts, two of whom initially presented with severe disc oedema. 3 Necropsy studies have shown that trans-meningeal herniation can result in damage at the level of the optic tract, chiasm, or optic nerves. 4

The anterior visual pathway can be compromised directly by gyrus herniation into the suprasellar cistern, a mechanism associated with meningomas. 3 Prechiasmal vision loss due to intracranial optic nerve infarction has also been reported in the setting of subdural haematoma; in this case the mechanism was presumably due to direct compression of the nerve against basal skull structures, although this specific radiographic finding was not described. 5

The precise mechanism of anterior pathway vision loss due to subdural haematoma in our patient, as well as in the few previous reports, remains poorly understood. 1,4 MRI showed no signs of blood in the orbits, direct compression of the optic nerves or chiasm, or gyrus herniation into the suprasellar cistern. The occipital lobes also appeared normal. Right to left midline shift due to the right sided haematoma was present, probably leading to vascular compromise or nerve compression that could not be visualised on MRI. Visual impairment following chiasmal decompression of mass lesions has been reported, and this mechanism may explain our patient's improved visual acuity and peripheral fields following the drainage of the haematoma. 6 Surprisingly, the subdural haematoma in our patient was smaller at the time of onset of visual symptoms than it had been 1 month earlier.

Subdural haematomas can affect vision through compression or vascular compromise at many points along the visual pathway. This case illustrates that optic neuropathy can occur late in the setting of a subdural haematoma, after the volume of the haematoma has begun to decrease because of the many ways in which patients with subdural haematomas can lose vision, they require close follow up, and a sudden change in vision necessitates immediate radiological testing, ophthalmological examination and, possibly, urgent surgical intervention and drainage.

Surodex in paediatric cataract surgery

Paediatric cataract surgery is associated with a high incidence of postoperative inflammation. 7–9 Intensive topical steroid therapy is still relied upon as the conventional mode of prevention and treatment. 7–9 Frequently, adjuvant systemic 10–13 and/or periocular steroids 14 may be required for further control, particularly if the child has a history of, or is at risk of, uveitis (for example, microphthalmos). 7–9 Non-compliance and missed application of steroid drops into the eye impedes control of the postoperative uveitis.

The Oculux Drug Delivery System (DDS; Oculux Pharmaceuticals, Inc, Sunnyvale, CA, USA) is a biodegradable device that allows sustained drug release after insertion into the anterior chamber (AC). Surodex is a DDS with 60 μg of dexamethasone incorporated into the polymer matrix (poly(lactic-glycolic)-acid, PLGA) with sustained and controlled release of dexamethasone over 7 days, achieving higher intraocular drug levels than with conventional dexamethasone eye drops. 14–16 Randomised controlled trials found Surodex to be as effective as 0.1% dexamethasone in the prevention and treatment of postcataract surgery inflammation. 14–16 Surodex is approved for use in cataract surgery in Singapore.

We reviewed retrospectively all paediatric patients who underwent cataract surgery with the insertion of one pellet of Surodex into the AC at the conclusion of surgery. Eighteen eyes of 13 patients (nine males and four females) were diagnosed with cataracts at a mean age of 57.4 months (range 1 day to 136 months). The mean age at surgery was 66.5 months (range 1 week to 139 months) and follow up period ranged from 6–18 months (mean 7.8 months). Factors predisposing to cataracts included hereditary cataracts (two), microphthalmos (three),

References


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Table 1  Pseudophakic and aphakic groups

<table>
<thead>
<tr>
<th></th>
<th>Pseudophakic group</th>
<th>Aphakic group</th>
</tr>
</thead>
<tbody>
<tr>
<td>No of eyes</td>
<td>11 (61.1%)</td>
<td>7 (38.9%)</td>
</tr>
<tr>
<td>Mean age at surgery</td>
<td>84.8 months</td>
<td>14.3 months</td>
</tr>
<tr>
<td>No of eyes with posterior capsular opening and anterior vitrectomy</td>
<td>7 (63.6%)</td>
<td>7 (100%)</td>
</tr>
<tr>
<td>No of eyes with intact posterior capsule</td>
<td>4 (36.4%)</td>
<td>0</td>
</tr>
<tr>
<td>Complications</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Intraoperative complications</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>Postoperative complications</td>
<td>4 (36.4%)</td>
<td>1 (14.3%)</td>
</tr>
<tr>
<td>1 Malposition of PCIOL</td>
<td>1</td>
<td>0</td>
</tr>
<tr>
<td>2 Vitreous strand in anterior chamber with peaked pupil</td>
<td>1</td>
<td>0</td>
</tr>
<tr>
<td>3 Posterior synechiae</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>4 Fibrinous inflammation</td>
<td>0</td>
<td>0</td>
</tr>
<tr>
<td>5 Raised intraocular pressure</td>
<td>1</td>
<td>0</td>
</tr>
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severe atopic dermatitis (one) and traumatic cataract (one). The types of cataract included total/mature (seven), nuclear (two), lamellar (three), subcapsular (two), posterior lenticu- nus (two), and posterior polar (two).

All eyes underwent lens aspiration through or open anterior capsulotomy or continu- ous curvilinear capsulorhexis (CCC) under general anaesthesia. Fourteen eyes had either posterior capsulotomy (with the vitrectomy cutter) or a posterior CCC (surgeon’s pre- ference). Anterior vitrectomy was done in 13 eyes. Eleven eyes (61.1%) were implanted with a foldable intraocular lens (IOL) (Acrysof lens MA60BM, 10 and MA30BM, 1) (pseudophakic group) (Table 1). This group was older (mean 84.81 months) than the aphakic group (mean age 14.31 months). Only seven eyes (63.6%) of the pseudophakic group underwent posterior CCC/capsulotomy and anterior vitrectomy, compared to all eyes in the aphakic group. Complications were encountered in four eyes in the pseudophakic group (36.4%)—malposition of IOL, vitreous strand in AC, posterior synechiae and raised IOP.

Four eyes (two in the pseudophakic and two in the aphakic group) did not receive additional postoperative topical steroids (predni- solone acetate 1%). This decision was made for patients 4 and 12 as there was minimal manipulation and iris trauma intraoperatively. These children were older (ages 131 and 115 months at surgery), allowing for easier follow up examination. Patient 13 had developmental delay and was difficult to manage. All four eyes were assessed to be quiet by slit lamp examination 2–4 weeks postoperatively. Additional ster- oids were not indicated and there was no glaucoma or endophthalmitis.

One eye (patient 17) required adjunctive periocular dexamethasone (1 mg) for fibri- nous inflammation in the first week. The left eye of patient 6, which had been quiescent and without treatment for 2 months, devel- oped raised IOP (30 mm Hg) at 3 months after surgery. This was controlled with topical beta-blockers. There was no glaucomatous cup- ping and visual fields could not be performed in the young age of the child.

Patient 13 had severe atopic dermatitis requiring systemic prednisolone. When visually significant cataract developed in the left eye, preoperative prednisolone was increased prophylactically and a pellet of Surodex was inserted at the end of surgery. As there was minimal inflammation, the systemic steroid was tapered over 2 weeks and the steroid eye drops were stopped after 3 weeks. This eye achieved a final visual acuity of 20/20.

Comment

Fibrinous anterior uveitis is common after paediatric cataract surgery, occurring in varying severities in up to 10% of cases.15

In our series, only two eyes (11.1%) developed inflammation that required addi- tional steroid therapy. The remaining 16 eyes achieved good control of inflammation, par- ticularly the two eyes that received Surodex without postoperative topical steroids. None experienced rebound uveitis after 1 week, when the pellet had ceased its release of dexamethasone. This suggests that in selected eyes, a Surodex pellet alone may be adequate to control postoperative inflamma- tion. A randomised controlled trial comparing Surodex versus conventional steroid eye drop therapy will be needed to determine the ultimate efficacy of Surodex in paediatric eyes.

The efficacy of eye drops is dependent on compliance and timely application for drug penetration and absorption. In infants and young children, the systemic absorption of the steroid may have potentially serious complications such as hyperglycaemia and immunosuppression. Surodex significantly reduces the problems of compliance and timely application for drug to the target site, potentially eliminating the problems of compliance.

The single complication encountered, which may be related to Surodex insertion, is the late onset of raised IOP (patient 6) despite the lack of marked postoperative inflammation. The fellow eye had also under- gone cataract surgery with insertion of Surodex without complications. Steroid responsive glaucoma is an unlikely cause as the drug has been shown to persist only for 7 days in rabbits, although this has not been demonstrated in human eyes.14 Gonioscopy may reveal focal peripheral anterior synec- chiae (PAS) at the site of residual pellet, but it is unlikely that this minor degree of synechiae may cause angle closure glaucoma, although the pupil may persist for weeks in the angles.15 16 Unfortunately, gonioscopy was not performed in this eye. Glaucoma after paediatric cataract surgery is, however, a complication that increases in frequency with longer durations of follow up (3–22%).13 14 17

We acknowledge that there are several limitations to these findings. Firstly, being a retrospective review, the efficacy of Surodex in preventing posterior capsular opacification, an indicator of postoperative inflammation, could not be assessed. We are also unable to establish if Surodex alone is sufficient for postoperative control of inflammation, this would require a prospective randomised clinical controlled trial. The efficacy of control of postoperative inflammation and safety are incomplete without the assessment of flare and endothelial cell counts but these are difficult in children, although endothelial cell count studies in adult eyes have shown no significant change.14 15 Finally, gonioscopy to visualise the angles to look for PAS was also not done.

Surodex has previously been shown to be safe and effective in uncomplicated cataract surgery in adults. This retrospective review provides preliminary data to suggest that Surodex may be an effective and safe adjunctive anti-inflammatory agent that in some paediatric eyes may eliminate the need for other steroid administration. Further studies will be required to determine the ultimate safety of Surodex in paediatric eyes.

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References
Pars plana ciliary epithelial proliferation in 13q deletion syndrome

The 13q deletion syndrome is an uncommon chromosomal disorder affecting the long arm of chromosome 13 which is deleted to a variable degree. This syndrome is phenotypically characterised by mental retardation, structural malformations, facial dysmorphism, and a predisposition to develop retinoblastoma. This intraocular tumour is diagnosed in approximately 80% of cases with this syndrome. Moreover, the incidence of bilateral retinoblastoma is much higher in individuals with this syndrome. The retinoblastoma susceptibility gene (RB-1), which encodes for the nuclear phosphoprotein of chromosome 13 which is deleted to a variable degree, is deleted to a variable degree. The involvement of the q14 band on the long arm of chromosome 13 causes patients to have this syndrome at an increased risk for developing retinoblastoma. In addition, children with 13q deletion syndrome also show optic nerve hypoplasia and retinal dysplasia. In this case, we report additional findings of pars plana ciliary epithelial proliferation in 13q deletion syndrome.

Case report

A 5 month old Hispanic female infant with a karyotype 46,XX, del (13) (q14.1q21.3) was referred to us for the evaluation of retinoblastoma. She had dysmorphic features, such as craniosynostosis. There was no family history of ocular or systemic disease. While under anaesthesia, the patient’s right eye was examined, revealing a mass that occupied a large area of the inferior nasal portion of the retina, and extending into the vitreous cavity (Fig 1). It obscured the optic nerve head and was associated with retinal detachment. There were also fine vitreous seedings. Ultrasonography disclosed a retinal tumour mass, which was associated with retinal detachment. There was no tumour invasion of the uvea or the posterior optic nerve. In addition, the globe showed multilayered plaquid non-pigmented ciliary epithelial proliferation at the pars plana ciliaries. The optic nerve hypoplasia. The epithelial proliferation revealed benign histological features, unlike the malignant neoplastic proliferation of the retina. The retinoblastoma shows numerous Flexner-Wintersteiner and Homer-Wright rosettes (haematoxylin and eosin; original magnification ×200). Inset: note a large endophytic retinoblastoma arising from the inferior retina and obscuring the optic disc.

Comment

The extent of the deletions affecting the long arm of chromosome 13 may result in various developmental anomalies that constitute 13q syndrome. The proliferation of the pars plana ciliary epithelium, as noted in the present case, and its association with retinoblastoma suggests that the RB-1 gene, or a gene close to the RB-1 locus, may have a role in the proliferation of the other neuroepithelial structures of the eye, including the pars plana ciliary epithelium. In embryological terms, the non-pigmented ciliary epithelium is derived from the inner layer of the optic cup which also gives rise to the neural retina. Proliferation of the pars plana ciliary epithelium in an eye that harbours retinoblastoma suggests that the RB-1 gene may play a part in such epithelial proliferation. However, this epithelial proliferation has not been previously reported in eyes with retinoblastoma. Although the cause of proliferation of the ciliary epithelium is not clear, this case suggests that the non-pigmented ciliary epithelial proliferation at the pars plana ciliaries in an eye that harbours retinoblastoma may be related to 13q deletion syndrome. Such findings may be unique to this syndrome, but previous reports about it have not been mentioned. The lack of previously reported cases with findings of pars plana epithelial proliferation suggests that this syndrome may have variable phenotypic expression.

Acknowledgements

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References


Phacoemulsification of posterior polar cataracts—a surgical challenge

Polar cataracts are relatively uncommon yet they pose a significant chal-
lengte to the cataract surgeon. Cataract surgery in these cases is frequently accompanied by a high incidence of posterior capsule rupture (PCR).

**Morphology**

Posterior polar cataracts are associated with remnants of the hyaloid system or the tunica vasculosa lentis. These cataracts may also occur without any relation to hyaloid remnants and appear as circular or rosette shaped opacities; they are hereditary and usually transmitted as a dominant trait. The gene for this has been mapped to chromosome 16q22.

**Classification**

See Table 1 and Figure 1.

**Methods**

The incidence of posterior polar cataracts in our centre is approximately 5 per 1000. We conducted a retrospective review from 1994 to 1999 and identified 31 patients (36 eyes) who had surgery for posterior polar cataracts.

**Results**

Four eyes had PCR (11.1%) and the other 32 had uncomplicated surgery; 34 eyes achieved a best corrected visual acuity of 6/12 or better (94.4%).

**Comment**

Our series showed a PCR rate of 11.1% in contrast with the 26% incidence reported by Osher et al. and 36% by Vasavada and Singh. No hydrodissection was attempted and only careful controlled hydrodelineation was performed. This was done using small aliquots of balance salt solution (BSS) to loosen the nucleus while simultaneously watching the capsular bag to ensure that the fluid wave passed gently. In some cases, no hydroprocedures were necessary as there was slow separation of the nucleus by the BSS flowing from the phaco tip.

Vasavada and Singh described the use of step by step chop in situ and lateral separation to minimise stress on the capsule-zonule complex. We preferred the use of the "lambda" technique which involved sculpting in the shape of the Greek letter (λ), followed by cracking along both "arms" and removal of the central piece first. The advantage of this is its gentleness in not stretching the capsule while removing the quadrants, especially the first one. We emphasise that this is our preferred technique and other techniques would be equally effective in skilled hands.

We also used low vacuum, low aspiration, and low inflow parameters to ensure a more stable anterior chamber; bottle height was at 50 cm, vacuum at 100 mm Hg, and aspiration flow rate at 20 ml per minute. Optimum power setting was achieved when minimal movement of the nucleus occurred while sculpting.

The epinucleus and cortex were removed using manual dry aspiration with Simcoe cannula. This method is gentler as we believe that the aspiration pressure is more controllable with our "million dollar" hands. There is also no "after aspiration" effect which in the automated unit can continue for several milliseconds even after the foot has been taken off the pedal. The disadvantage of manual aspiration is the increased surgical time.

**Table 1**

<table>
<thead>
<tr>
<th>Type</th>
<th>Classification of posterior polar cataracts</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>Opacity associated with posterior subcapsular cataract.</td>
</tr>
<tr>
<td>2</td>
<td>Opacity with ringed appearance like an onion.</td>
</tr>
<tr>
<td>3</td>
<td>Opacity with dense white spots at the edge often associated with thin or absent posterior capsule.</td>
</tr>
<tr>
<td>4</td>
<td>Combination of the above 3 types with nuclear sclerosis.</td>
</tr>
</tbody>
</table>

The status of the posterior capsule (PC) dictated the action of the surgeon. If the PC was absent or torn but with no vitreous loss, a dispersive viscoelastic was injected over the defect to tamponade and push the vitreous face backwards. A dispersive rather than a cohesive viscoelastic is preferable as it is more adapted to maintaining a space and stabilising the anterior vitreous face. If there was PCR with vitreous loss, a two port anterior vitrectomy was performed. Intraocular lens implantation in these cases would depend on the extent of the PCR and the integrity of the remaining PC.

Surgical management of posterior polar cataracts poses a special challenge to the cataract surgeon. It is important that the surgeon and the patient understand the technical difficulties associated and are aware of potential complications. It may be prudent to address these cases at the end of an operating list or to shorten the list in anticipation of prolonged surgical time. The surgeon should use a technique that he or she is most familiar and comfortable with. With emphasis on gentleness, together with patience and a well practised technique, the incidence of PCR can be minimised with phacoemulsification for posterior polar cataracts.

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**Molluscum contagiosum in an immune reconstituted AIDS patient**

In spite of lower viral loads and increasing T cell counts, AIDS patients receiving highly active antiretroviral therapy (HAART) are not always successful in mounting an immune response to some opportunistic pathogens. In fact, CMV retinitis, which was known to occur in HIV patients with CD4 counts below 50 × 10^3/l, has been described in immune reconstituted patients with CD4 counts above 200 × 10^3/l. It is therefore important to make observations about the clinical spectrum of infectious disease in immune reconstituted AIDS patients. Here we report an isolated
lesion of molluscum contagiosum in an immune reconstituted AIDS patient.

Case report
A 46 year old Hispanic female presented with a history of burning, tearing, and itching of her left eye for 1 month. Three years earlier she had been diagnosed with AIDS during a hospital admission for Pneumocystis carinii pneumonia (PCP). At that time her CD4 count had been 27 x 10^6/l and her viral load 905,000. After 3 months of HAART with combivir, norvir, and fortovase her viral load dropped to 15,000 and her CD4 count rose to 184 x 10^6/l. Her viral load became undetectable 6 months after initiation of therapy and has remained so for 2½ years. Her recent CD4 count was 435 x 10^6/l.

Best corrected visual acuity was 20/25 in each eye. A 2 mm smooth, dome-shaped, translucent papule with central umbilication has remained so for 2½ years.

Histology revealed a primary nodular malignant melanoma, at least 7 mm thick, composed of epithelioid sparsely pigmented melanocytes positive for S100 and vimentin immunostains. Excision biopsy with conjunctival autografting was performed.

The patient was referred for adjuvant treatment with cryotherapy. To date, there has been no sign of local recurrence.

In June 2001, the patient was referred by his general practitioner to the oral surgery service at the same hospital with a 6 month history of pain on the right side of his neck. He had also noticed some right facial swelling. Clinical examination disclosed a diffuse, firm mass over the lower pole of his right parotid gland, measuring 4 cm in diameter. There were no overlying skin changes.

Magnetic resonance imaging (MRI) of the head and neck showed a well defined lesion within the right parotid gland involving the deep lobe and the deeper portion of the parotid gland.

Figure 1 Pedunculated lesion arising from the nasal limbal conjunctiva, right eye.

Case report
A 79 year old white man presented to the eye clinic in September 1999. He had been noted 3 years previously to have a small inclusion cyst of the bulbar conjunctiva in his right eye, which he complained had increased in size and was becoming red and sore. Examination showed an inflamed pedunculated lesion 1 cm in diameter arising from the nasal limbal conjunctiva (Fig 1). The lesion was granulomatous and amelanotic. There was adjacent corneal opacity. Ocular examination was otherwise unremarkable.

The appearance of the lesion was felt to be unusual with a presumptive diagnosis of conjunctival malignancy. Excision biopsy with conjunctival autografting was performed.

Histology revealed a primary nodular malignant melanoma, at least 7 mm thick, composed of epithelioid sparsely pigmented melanocytes positive for S100 and vimentin immunostains. Excision was deemed incomplete.

The patient was referred for adjuvant treatment with cryotherapy. To date, there has been no sign of local recurrence.

In June 2001, the patient was referred by his general practitioner to the oral surgery service at the same hospital with a 6 month history of pain on the right side of his neck. He had also noticed some right facial swelling. Clinical examination disclosed a diffuse, firm mass over the lower pole of his right parotid gland, measuring 4 cm in diameter. There were no overlying skin changes.

Magnetic resonance imaging (MRI) of the head and neck showed a well defined lesion within the right parotid gland involving the deep lobe and the deeper portion of the parotid gland.
superficial lobe. A few small lymph nodes were visible at several sites bilaterally but none appeared enlarged. Computed tomography (CT) of the chest, abdomen and pelvis revealed single nodes measuring 3–5 mm at both lung bases, which may represent lung metastases.

An orthopantomogram was normal, and fine needle aspiration (FNA) cytology was performed. This showed scattered lymphocytes and highly pleomorphic non-lymphoid malignant cells, some containing flecks of pigment (Fig 2). While these appearances alone would not allow definitive diagnosis of melanoma, in the clinical context they were sufficient to conclude that the parotid swelling was likely to be metastatic melanoma. This was confirmed on subsequent parotid excision biopsy, which revealed extensive involvement of the parotid nodes and parotid parenchyma, extending into the external jugular vein.

Comment
Malignant melanoma is a relatively rare tumour in the parotid gland, with most tumours representing metastasis from cutaneous head and neck primaries.1 Very occasionally, as in this case, the primary tumour is non-cutaneous in origin. Conjunctival melanoma metastasising to the parotid has been noted in previous series,2 but remains rare. This case is unusual with respect to the initial size and appearance of the tumour, the previous history of a conjunctival cyst, and that definitive diagnosis of a metastatic lesion from a conjunctival primary was made by FNA. This method has been helpful in the diagnosis of other types of tumour in the parotid,3 and indeed in parotid melanomas of different origin.4 In this case, the patient’s previous ophthalmic history had been unknown to the maxillofacial surgeon managing the case, and the diagnosis only became apparent during reporting of the cytology, when the FNA findings could be compared with previous histology. This illustrates the importance of exhaustive history taking and the value of a cohesive local histopathology service.

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References

Bilateral Aspergillus endophthalmitis in a patient with chronic lymphocytic leukaemia

Aspergillus species are ubiquitous saprophytic moulds, commonly growing in soil, stored hay, and decaying vegetation. Even though exposure to Aspergillus is universal, infection in humans is uncommon.5 Aspergillus infection of ophthalmic interest usually causes keratitis or orbital cellulitis; Aspergillus endophthalmitis is a relatively rare condition that has a devastating course, with blindness as its usual outcome.6 The clinical diagnosis is difficult and the treatment is disappointing. In most cases, ocular involvement results from spread of aspergillosis infection from other organs and typically occurs in injecting drug users and in patients with immune deficiency of various causes. The leucopenia appears to be a predisposing condition for the occurrence of aspergillosis.7

We report an unusual case of bilateral endogenous Aspergillus endophthalmitis in a patient with chronic lymphocytic leukaemia in the absence of any detectable focus of aspergillosis infection elsewhere in the body and that showed a good response to specific systemic therapy.

Case report
A 51 year old white man with a previous diagnosis of chronic lymphocytic leukaemia and use of an immunosuppressive agent was referred to ophthalmological examination because of a red eye, pain, and blurred vision in his right eye. The clinical picture worsened and diagnosis of endophthalmitis was made. Intravitreous amphotericin B injection was performed and did not control the case. Culture of vitreous fluid was positive for Candida. This eye was eviscerated because of increasing pain, progressive infection, and poor response to treatment. Posterior histopathological study was conclusive for Aspergillus endophthalmitis in the right eye (Fig 1B and 2). At the same time, fundus examination of the left eye showed two subretinal exudative lesions located at nasal and inferior retina with retinal oedema associated with superficial haemorrhages (Fig 1A). The vitreous was clean and the central macula remained intact. Visual acuity was 6/6 in this eye.

Vitreous biopsy or culture may yield negative results in some cases of early intraocular Aspergillus endophthalmitis.8 We did not take a vitreous biopsy of the left eye, since we already had the diagnosis in the right eye and the visual acuity was 6/6. This eye was treated with intravenous amphotericin B and oral itraconazole with a good result. The patient remained stable with resolution of the lesions and no focus of systemic aspergillosis was found.

Comment
Fungal endophthalmitis is uncommon. In most of the cases Candida is the causal organism.9 Few cases of Aspergillus endophthalmitis in a patient with chronic lymphocytic leukaemia have been described,4 and according to the literature endogenous Aspergillus endophthalmitis represents a manifestation of disseminated aspergillosis, usually a fatal infection.10 This case is unusual because it is bilateral and no focus of systemic aspergillosis was found.

The cases of intraocular inflammation secondary to Aspergillus are more common in the central macula and have a poor prognosis.11 In our case the localisation of the chorioterinitis in the left eye was out of the posterior area and the patient’s visual acuity remained 6/6.

The major antifungal agent used in aspergillosis is amphotericin B. Without host immune competence, treatment is rarely effective. Penetration of intravenous amphotericin B into the vitreous cavity of the normal or inflamed eye is poor.12 The azole compounds have been used to reduce the significant toxicity and enhance the efficacy
of intravenous amphotericin B; oral fluconazole is the drug of choice because it has excellent penetration in central nervous system and vitreous. Itraconazole may be used. Intravenous amphotericin B and vitrectomy have given the best results in the treatment of these cases.4 Our patient received intravenous amphotericin B and oral itraconazole and this therapy was sufficient to control the infection.

This case shows that Aspergillus endophthalmitis should be considered in all patients with immune deficiency even in the absence of systemic aspergillosis. Treatment with intravenous amphotericin B may be able to control these cases and should be attempted more often.

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Table 1 Demographic details of patients

<table>
<thead>
<tr>
<th>Case no</th>
<th>Age (years)</th>
<th>Sex</th>
<th>Drainage procedure</th>
<th>Delay to needling (years)</th>
<th>No of needlings</th>
<th>No of 5-FU injections</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>72 M</td>
<td>Bilateral Scheie’s</td>
<td>31</td>
<td>2</td>
<td>4</td>
<td></td>
</tr>
<tr>
<td>2</td>
<td>88 F</td>
<td>Left ECCE trabeculectomy</td>
<td>10</td>
<td>4</td>
<td>5</td>
<td></td>
</tr>
<tr>
<td>3</td>
<td>80 F</td>
<td>Right trabeculectomy</td>
<td>12</td>
<td>1</td>
<td>4</td>
<td></td>
</tr>
<tr>
<td>4</td>
<td>35 M</td>
<td>Right trabeculectomy</td>
<td>8</td>
<td>1</td>
<td>32</td>
<td></td>
</tr>
<tr>
<td>5</td>
<td>71 M</td>
<td>Bilateral trabeculectomy</td>
<td>13</td>
<td>1</td>
<td>2</td>
<td></td>
</tr>
</tbody>
</table>

ECCE = extracapsular cataract surgery.

Table 2 Results of pre-bleb and post-bleb needling with a 12 month follow up period

<table>
<thead>
<tr>
<th>Case no</th>
<th>IOP (mm Hg)</th>
<th>No of medications</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>72 M</td>
<td>Pre</td>
</tr>
<tr>
<td>2</td>
<td>88 F</td>
<td>19</td>
</tr>
<tr>
<td>3</td>
<td>80 F</td>
<td>25</td>
</tr>
<tr>
<td>4</td>
<td>35 M</td>
<td>58</td>
</tr>
<tr>
<td>5</td>
<td>71 M</td>
<td>25</td>
</tr>
</tbody>
</table>

Late bleb needling

Surgical manipulation of the trabeculectomy bleb has become a recognised postoperative procedure to increase the success of glaucoma surgery. The first needling revision of a glaucoma drainage bleb was described in 1941 and there are several reports of the successful restoration of failing blebs within the first 3 years following trabeculectomy.5 6 7 8

We report the results of five cases of late bleb needling with 5-fluorouracil (5-FU) where trabeculectomy had been performed between 8 and 30 years earlier.

References


Case reports

The demographic details of all cases are summarised in Table 1.

Glaucoma surgery had taken place between 8 and 31 years before bleb needling and in no case had antimetotics been used at the original surgery. Before bleb needling the average intraocular pressure (IOP) among the patients was 29.4 mm Hg (range 19–58). Each patient showed glaucomatous deterioration despite being on maximum tolerated medical therapy, taking on average three ocular hypotensive agents, and in two cases oral acetazolamide. In all cases an open sclerotomy was confirmed by gonioscopy.

All procedures were performed in the outpatient clinic, by either a consultant or associate specialist, using a slit lamp. The eye was anaesthetised with amethocaine eye drops 1%, and phenylephrine eye drops 2.5% were used for vasoconstriction. After several drops of chloramphenicol the conjunctiva was entered several millimetres from the flap site with a 27 gauge needle mounted on an insulin syringe. In one case aqueous flow was established after perforating scar tissue around an encysted bleb, whereas in the others it was necessary to dissect beneath the scleral flap and enter the anterior chamber. After creating a bleb and confirming a reduction in IOP by applanation tonometry, 5 mg 5-FU (25 mg/ml) were injected into the subconjunctival space around the bleb. After needling, all hypotensive therapies were stopped and replaced by intensive topical steroids and chloramphenicol. The steroid was titrated, and repeat injections of 5-FU with or without needling were given, according to the IOP and appearance of the bleb.

After 12 months follow up from the last needling (Table 2), average IOP was reduced to 14 mm Hg (range 9–17). There was no change in the patients’ visual acuity. Two cases developed a mild corneal epitheliopathy that healed within 8 weeks. There were no other complications from the needling procedure.

Comment

Although trabeculectomy is the preferred glaucoma drainage procedure, only 67% of patients may achieve an adequate target pressure after 1 year.9

In recent years glaucoma surgery has developed with the use of antimetotics and intense postoperative surveillance with bleb manipulation. Reports show that bleb needling used in combination with subconjunctival 5-FU injections can rectify a failing bleb in the early postoperative phase but there are few reports confirming its effect in the late postoperative period.9 10 Some studies have indicated that the success of bleb needling is unrelated to the time lapsed from the original surgery, though in these studies the maximum interim period was less than 4.5 years.

The patients presented in this study had had their original glaucoma surgery at least 8 years previously and bleb needling was carried out before listing the patient for a repeat trabeculectomy with mitomycin C. The only adverse effect noted was a temporary corneal epitheliopathy, probably related to toxicity of the 5-FU. Other reported adverse events after bleb needling include hyphaema, bleb leak, shallow anterior chamber, choroidal effusion and endophthalmitis, but there are no reports of long term hypotony as has been described following mitomycin C trabeculectomy.

These case reports indicate that bleb needling may be successful in achieving a long lasting IOP reduction even several years after the original surgery. The procedure does not require dextrous skills beyond that of a trained general ophthalmologist. It appears at least as safe as trabeculectomy and avoids a formal operation. If it does fail the surgical field is still intact for a “redo” trabeculectomy.

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The success rates for endonasal dacryocystorhinostomy

Tsirbas and Wormald are to be congratulated on their landmark paper in lacrimal surgery, describing endonasal dacryocystorhinostomy (DCR) with mucosal flaps, which is also known as powered endonasal DCR.1 Their results are seemingly impressive, with anatomical success rate of 95% and functional success rate of 89%, closely approaching the same technique, which when they had a minimal follow up of 9 months, had a lower anatomical success rate (91%).

It is important to define success and what this really means, and for lacrimal surgeons to agree consistent outcome criteria. Perhaps lacrimal surgeons should agree the following criteria for DCR surgical success, irrespective of whether it is by an external or endonasal route:

- Assess the outcome a minimum of 6 months after surgery, being at least 3 months after removal of tubes. Or is 1 year after surgery better?
- Assess subjective success based on the patient’s symptoms.
- Assess objective success (anatomical success) based on (i) absence of tear drainage into the nasal space and (ii) presence of a functioning rhinostomy.

Despite these minor quibbles, the authors are to be congratulated on advancing endonasal lacrimal surgery.

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References


Trypan blue stains the epiretinal membrane but not the internal limiting membrane

We read with great interest the paper by Li et al about staining of the internal limiting membrane (ILM) and epiretinal membrane (ERM) with trypan blue (TB).1 We would like to comment on one aspect of this paper, when the authors claimed that a good staining of both the ILM and the ERM was achieved with TB. We disagree that ILM is stained by TB, and propose that TB only stains the ERM, not the ILM.

The authors affirm “ILM staining” with TB as they observed histologically the presence of ILM in four eyes with macular holes at stage III and IV. In one of those eyes, immunohistochemistry was performed, and an epiretinal membrane was present. In the other three cases, immunohistochemistry examination was not performed because of insufficient tissue. Most of the stage III and IV macular holes are known to be associated with an epiretinal membrane,2 and probably an ERM would be seen in addition to the ILM in those three cases if immunohistochemistry for glial elements were performed. Therefore, we believe that TB stained the ERM associated with the macular holes, but not the ILM. In their study, staining with TB of seven patients with idiopathic epiretinal membrane was successfully performed. ERM of proliferative vitreoretinopathy is also reported to be well stained by TB.3 We speculate that TB has binding affinity to some of the glial cell elements of the highly cellular ERM, either those associated with macular holes or not.

Indocyanine green (ICG) is another dye for intracocular staining that has wide acceptance among retina surgeons in the past few years.4 In contrast with the cellular affinity of TB, ICG stains the acellular ILM, because of the fast binding of ICG to collagen proteins of the ILM. ERM tends to be stained negatively by ICG, well because the hydrophilic ICG does not penetrate cell membranes easily.5

TB staining seems to be a good alternative to ICG staining in the surgical management of macular diseases. Further studies are warranted on the intraocular kinetics of that dye.

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accepted for publication 4 April 2003

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4

www.bjophthalmol.com

Thermochemotherapy in hereditary retinoblastoma

Schueler and associates describe their experience with thermochemotherapy (TCT) in bilateral retinoblastoma. The reported results of transpupillary thermotherapy used in combination with chemotherapy are encouraging with 86–96% tumour control. In the current series, however, local recurrence occurred in 38%.

The dosage of carboplatin used in the current series was 10 mg/kg body weight, which is lower than the standard dosage of 18.6 mg/kg body weight. Lower dose of carboplatin, the key drug in the chemotherapy regimen for retinoblastoma, could have influenced the higher recurrence rate.

The authors mention that they treated submacular tumours with TCT. However, in our experience, tumours located in the macular area are better treated initially with chemotherapy for 3–6 cycles in order to achieve maximum possible reduction in tumour size before considering thermochemotherapy. Chemotherapy reduced macular tumours tend to shrink away from the fovea towards one of the major arcades or the optic nerve, thus exposing the foveal region. Regional tumours beyond 3–6 cycles of chemotherapy could be treated with thermochemotherapy. A smaller scar thus produced may optimise residual central vision.

The high mean total duration of thermochemotherapy in the current series is probably because of a smaller spot size of 0.4 mm. The diode laser (Iris Medical Inc, Mountain View, CA, USA) with an operating microscope adapter allows for a spot size of 0.8, 1.2, and 2.0 mm. The relatively newer large spot indirect ophthalmoscope delivery system provides a 1.2 mm spot size. A larger spot size will indeed reduce the duration of thermochemotherapy and allow for a more uniform coverage. Corneal, iris, and lens complications are minimised with better convergent beam optical systems currently available.

We believe that with higher dose of carboplatin, staggered thermotherapy for submacular tumours, use of better optical systems for delivery and a larger spot size for thermochemotherapy, and judicious selection of cases, the tumour regression and vision salvage with TCT could be further optimised.

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Causes of severe visual impairment and blindness in children in Ethiopia

We read with great interest the article by Kello et al. The authors have to be congratulated for the hard hitting and well written article. A current concern for people involved in paediatric eye care is the emergence of what is probably the third epidemic of retinopathy of prematurity (ROP) in developing countries. It is therefore significant that no case of ROP was found in the population screened in this study. Several factors could account for this.

• The very low or nil prevalence of ROP in countries such as Ethiopia, where the study was carried out, is most probably because of lack of intensive care facilities for premature infants and their low survival rates.
• The variation in the incidence of ROP between ethnic groups could also account for this, with the available evidence suggesting that African-American infants are less prone to severe outcome ROP than white infants. However, it is also important to note that the article mentions that children with mental retardation were not examined and that the admission criteria of the blind schools that preclude their admission. This too could have accounted for the gross underestimation of the prevalence of ROP as suggested by Jacobson et al. In addition, these children with mental handicap could be suffering from cerebral palsy and would have been at high risk for ROP because of the higher incidence of retinal vascular anomalies associated with both cerebral ischaemia and prematurity.

An error occurred in the author listings for two letters in the September issue. In the letter by Lee et al (Br J Ophthalmol 2003;87:1184–5) the order should be D K Lee, E B Suhler, W Augustin, R R Buggage. In the letter by Buggage et al (Br J Ophthalmol 2003;87:1190–1) the order should be R R Buggage, D G Callanan, D F Shen, C-C P Chen. The journal apologises for the error.

References
NOTICES

Elimination of avoidable blindness
The latest issue of Community Eye Health (No 46) discusses the resolution of the World Health assembly on the elimination of avoidable blindness. For further information please contact: Journal of Community Eye Health, International Resource Centre, International Centre for Eye Health, Department of Infectious and Tropical Diseases, London School of Hygiene and Tropical Medicine, Keppel Street, London WC1E 7HT, UK. Tel: +44 (0)7948 524238; email: Anita.Shah@lshtm.ac.uk; website: www.jceh.co.uk). Annual subscription (4 issues) UK £28/US$45. Free to developing country applicants.

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's 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 conditions or syndrome with an integral eye disorder. SPECS represents over 50 different organisations related to eye disorders ranging from conditions that are relatively common to very rare syndromes. The website acts as a portal giving direct access to support groups' own websites. 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; website: www.eyeconditions.org.uk).

The British Retinitis Pigmentosa Society
The British Retinitis Pigmentosa Society (BRPS) was formed in 1975 to bring together people with retinitis pigmentosa and their families. The principle aims of BRPS are to raise funds to support the programme of medical research into an eventual cure for this hereditary disease, and through the BRPS welfare service, help members and their families cope with the everyday concerns caused by retinitis pigmentosa. Part of the welfare service is the telephone help line (+44 (0)1280 860 363) for any queries relating to retinitis pigmentosa, especially for those recently diagnosed with retinitis pigmentosa (tel: +44 (0)1280 821 334; email: lynda@brps.demon.co.uk; website: www.brps.demon.co.uk).

Surgical Eye Expeditions International
Volunteer ophthalmologists in active surgical practice are needed to participate in short term, sight restoring eye surgery clinics around the world. Contact: Harry S Brown, Surgical Eye Expeditions International, 27 East De La Guerra, C-2, Santa Barbara, CA 93101-9858, USA (tel: +805 965 3303; fax: +805 965 3564; email: hsbrown.md@cox.net or seeintl@seeintl.org; website: seeintern.org).

Rise in organ transplant numbers
According to UK Transplant, the UK has seen the highest number of organ transplants in six years. Last year (1 April 2002 to 31 March 2003), 2777 patients had their lives saved or dramatically improved through the generosity of 1064 donors. This equated to a 6% increase compared to the previous 12 months (1 April 2001 to 31 March 2002). Furthermore, during 2002–3, the highest number of people benefited from organ transplant for five years (1997–98) and 240 more people had their sight restored than the previous year. For further information see UK Transplant's website (www.uktransplant.org.uk).

Elimination of avoidable blindness
The 56th World Health Assembly (WHA) considered the report on the elimination of avoidable blindness (doc A62/66) and urged Member States to: (1) Commit themselves to supporting the Global Initiative for the Elimination of Avoidable Blindness by setting up a national Vision 2020 plan by 2005; (2) Establish a national coordinating committee for Vision 2020 and a national blindness prevention committee to help implement the plan; (3) Implement the plan by 2007; (4) Include effective monitoring and evaluation of the plan with the aim of showing a reduction in the magnitude of avoidable blindness by 2010; (5) Support the mobilisation of resources for eliminating avoidable blindness. The WHA also urged the Director-General to maintain and strengthen WHO’s collaboration with Member States and the partners of the Global Initiative for the Elimination of Avoidable Blindness as well as aid in the coordination and support of national capability.

Glaucoma Society 24th Annual Meeting and Dinner
The Glaucoma Society 24th Annual Meeting and Dinner will take place on 20 November 2003, from 8:30 am to 3:00 pm at The Royal College of Physicians, London, UK. Further details: Ms Janet Flowers (email: glausoc@ukeire.freeserve.co.uk).

Detachment course with international faculty on: retinal and vitreous surgery with case presentations preceding the annual meeting of Iranian Society of Ophthalmology
The detachment course with international faculty on: Retinal and Vitreous Surgery with Case Presentations preceding the Annual Meeting of the Iranian Society of Ophthalmology will be held on 29–30 November 2003 and 1–4 December 2003 respectively, at the Razi Conference Center, Hemmat Hyw, Tehran, Iran. Further details: Scientific programme: Prof Ingrid Kreissig, University of Tuebingen, Schleichstr. 12, Breuningenbau, 72076 Tuebingen, Germany (tel: +49 7071 295209; email:ingrid.kreissig@med.uni-tuebingen.de). Local organisation: Dr Arman Masheykhchi, Dr Siyamak Moradian, Dept of Ophthalmology, Labbanfinjad Medical Center, Pasdaran Ave, Boostan 9, Tehran, 16666, Iran (fax: +98 21 254 9039; email: labball@hotmail.com).

4th International Congress on Autoimmunity
The 4th International Congress on Autoimmunity will take place 3–7 November 2004 in Budapest, Hungary. The deadline for the receipt of abstracts is 20 June 2004. Further details: Kenes Global Congress Organisers and Association Management Services, 17 Rue du Cendrier, PO Box 1726, CH-1211 Geneva 1, Switzerland (tel: +4122 908 0488; fax: +41 22 732 2850; email: autoim04@kenes.com; website: www.kenes.com/autoim2004).

Wake up call as dream time deadline looms
Scientists have less than a month left to apply for a new Dream Time award from NESTA, the organisation that invests in UK creativity and innovation. Dream Time supports exceptional achievers (with at least 10 years experience in their field) who want time to experiment or follow a passion, but who intend to continue with their career and put what they have discovered to good use. Up to 12 exceptional individuals from the fields of science, technology and the arts will each receive up to £40,000 to pursue their goals and push at the boundaries of knowledge and practice. NESTA is looking for people who can demonstrate evidence of exceptional achievement. This would include a significant body of work collated over at least a decade in their field, the ability to work in new ways and a commitment to the proposed area of exploration. Dream Time is a development of NESTA’s existing Fellowship Programme, which has helped talented and creative individuals to innovate and explore new ideas emerging through periods of personal development. As with all its awards, NESTA is looking for people who demonstrate excellence, promise, creativity, innovation and commitment. Funding can be used on a full- or part-time basis, in tandem with professional careers or temporarily away from the constraints of employment. Offered awards can be for any period of time up to
one year. Dream Time Fellows will be asked to provide 10% in kind support for their plan and will be required to plan ways of disseminating their findings with their professional community. To apply, visit NESTA’s website: www.nesta.org.uk/dreamtime.

14th Meeting of the EASD Eye Complication study group

The 14th Meeting of the EASD Eye Complication (EASDEC) study group will take place on the 21–23 May 2004. There will be key lecture notes on the following topics: Peter Gaede (Denmark)—Results of the Steno 2 study, Hans Peter Hammes (Germany)—Animal models of diabetic retinopathy, Massimo Porta (Italy)—Screening with the London protocols: 12 years after, and Anselm Kampik (Germany)—Surgical options in diabetic retinopathy. There will also be case presentations and oral and poster presentations. The EASDEC board comprises F. Bandello (President), P. J. Guillausseau (Vice President), C-D Agardh (Past President), P. Massin (Secretary), M. Porta (Treasurer). The Scientific and Organizing Committee includes: F. Bandello, P. J. Guillausseau, P. Massin, C-D Agardh, M. Porta, A. Kampik, M. Ulbig, and G. Lang. There are three travel grants available, at 1000 Euro each, for young scientists (less than 35 years at the time of the meeting). Application for the grant should be made together with the submission of the abstract. For further information, contact: Department of Ophthalmology, Ingrid Mannl, Ludwig-Maximilians-University, Mathildenstr. 8, 80336 Munich, Germany (tel: +49–89–5160–3800; fax: +49 89 5160 4778; e-mail: easdec@ak-i.med.uni-muenchen.de. The deadline for abstracts is 2 March 2004.