Surgically induced diffuse scleritis: comparison of incidence in phacoemulsification and conventional extracapsular cataract extraction

Surgically induced diffuse scleritis (SIDS) is a recognised but less well reported cause of pain and may be failure to recognise it which explains its absence from the national statistics.

Patients who had undergone previous ipsilateral iris surgery (trabeculectomy, Scheie's procedure, or YAG iridotomy) appeared to be at increased risk, although this only attained statistical significance in the phacoemulsification group (p=0.009). General anaesthesia was associated with higher statistical risk for ECCE patients only (p=0.009). It is unclear why this should have been the case. Intraoperative complications did not increase the risk of developing SIDS, and there was no association with concurrent ocular or systemic disease. The mean age of the SIDS patients in the ECCE group was approximately 11 years younger than that in the phaco group (62.5 ± 73.6).

It is our practice to take a thorough history from the postoperative patient. Severe pain, especially that waking the patient from sleep, is a common feature. Examination of the patient both on the slit lamp and in daylight is conducted, the latter to recognise the characteristic violaceous injection of the scleral vasculature. B-scan ultrasonography is performed to measure scleral thickness. Relative thickening compared to the contralateral eye or absolute thickness greater than 1.8 mm supports the diagnosis. Affected patients usually show a favourable response to a combination therapy with oral non-steroidal anti-inflammatory agents, topical steroids, and a topical cycloplegic. SIDS should be considered in the differential diagnosis of a painful red eye postoperatively. Prompt diagnosis and appropriate therapy lead to early resolution of SIDS and improved visual outcome.

Table 1

<table>
<thead>
<tr>
<th>Complication</th>
<th>Phako (%)</th>
<th>ECCE (%)</th>
<th>NCSS (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>Astigmatism*</td>
<td>9.5</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Cystoid macular oedema</td>
<td>1.95</td>
<td>3.1</td>
<td>0.05</td>
</tr>
<tr>
<td>SIDS</td>
<td>1.35</td>
<td>2.3</td>
<td>5.6</td>
</tr>
<tr>
<td>Uveitis</td>
<td>0.75</td>
<td>1.9</td>
<td>0.4</td>
</tr>
<tr>
<td>Posterior capsule opacification</td>
<td>0.6</td>
<td>&lt;1</td>
<td>0.03</td>
</tr>
<tr>
<td>Endophthalmitis</td>
<td>0.3</td>
<td>3.1</td>
<td>0.1</td>
</tr>
<tr>
<td>Corneal decompensation</td>
<td>0.15</td>
<td>&lt;1</td>
<td>1.2</td>
</tr>
<tr>
<td>Subluxed IOL</td>
<td>0.15</td>
<td>&lt;1</td>
<td>1.1</td>
</tr>
</tbody>
</table>

*Astigmatism requiring suture removal.

References
1. Scott JA, Clearkin LG. Surgically induced scleritis following cataract extraction. Eye 1994;8:292-7

Total exudative detachment as a first presentation of von Hippel Lindau disease

Von Hippel Lindau disease is a rare condition characterised by retinal and central nervous system haemangioblastomas. It is also associated with renal cell carcinoma, phaeochromocytoma, and renal, pancreatic, and epididymal cysts.

The disease usually presents with neurological symptoms and/or visual disturbance; angiomata if seen in the retina can often be treated in an attempt to prevent progression to retinal detachment.

We report a young girl with von Hippel Lindau disease whose initial presentation was with a total exudative retinal detachment.

Case report
A 14 year old girl presented to the eye casualty department with no perception of light in her left eye. She gave a 2 month history of gradual, painless loss of vision, but had delayed previously mentioning her symptoms. Past ocular history was unremarkable; an optometrist visit a year previously was normal. She was otherwise well.

Slit lamp examination on the left revealed a quiet anterior segment, with retinal folds visible abutting the posterior lens capsule (Fig 1A). The right eye was entirely normal. An ultrasound B-scan showed a left total funnel retinal detachment (Fig 1B). Computed tomography scan demonstrated normal orbits.

The patient's identical twin was also present and was examined: both her eyes were normal.

At this stage the family history proved helpful. The patient's father had recently been diagnosed with renal cell carcinoma and pituitary vascular tumours. The paternal line had a cluster of various carcinomas, including three relatives with cerebellar tumours. Further investigation revealed at least one of these to be histologically similar.
confirmed haemangioblastoma. The possibility of von Hippel Lindau disease was raised and the patient's father was screened: the diagnosis was confirmed with polymerase chain reaction analysis, showing a frame shift mutation (single base pair deletion [G]) in codon 195 (exon 3) of the von Hippel Lindau gene. Subsequent extended family screening has revealed the same mutation in the patient and nine other living relatives (Fig 2), including the patient's identical twin. Five of these relatives have clinical manifestations of the disease, and in one case an asymptomatic stage 1 renal cell carcinoma was diagnosed and successfully resected.

Miss AB's left eye has subsequently developed pupil block glaucoma which is currently well controlled medically. It remains blind with a rigid total retinal detachment. The cause of this detachment is most likely to be an optic nerve or retinal haemangioblastoma; however, no causal lesion was identified on ultrasound or computed tomography (CT). The few retinal vessels that can be seen (Fig 1A) do not show any abnormality. Her other eye remains normal and screening for signs of the disease elsewhere has proved negative.

Comment

This case highlights the importance of considering von Hippel Lindau disease as a diagnosis when confronted with a patient with an unexplained exudative retinal detachment.

Von Hippel Lindau disease has an autosomal dominant inheritance pattern with an incidence of one in 36,000; with a 70% prevalence of ocular haemangioblastomas. Von Hippel Lindau gene mutation carriers have a 35% probability of visual loss by age 50.1

Ocular angiomas occur commonly in the superotemporal mid-pterinal retina, less commonly on the optic disc (8%), and at the posterior pole (1%).1

If left untreated, lesions can cause vitreous haemorrhage, macular oedema, epiretinal membrane formation, and exudative or tractional retinal detachment. This last complication is well demonstrated in our patient's case, with a 2 month delay between onset of symptoms and first presentation. Treatment comprises laser photocoagulation of angiomas less than one disc diameter.2 Cryotherapy, or vitreoretinal surgery with trans-scleral drainage and endolaser may be indicated in larger lesions,3 especially if vitreous traction is present.4 Early treatment necessarily offers a better prognosis.

This case underscores the value of taking a good family history in order to detect hereditary diseases, which can then be confirmed with genetic molecular analysis. The importance of identification and subsequent screening of von Hippel Lindau gene mutation carriers (both affected and unaffected) is exemplified in our patient's family—with the potential benefit of early diagnosis and treatment of asymptomatic retinal and cerebellar haemangioblastomas, as well as potentially fatal tumours. The screening should be maintained regularly, as the protein manifestations of von Hippel Lindau disease may occur de novo at any age.5

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References


Figure 1 CT scan of the orbits showing dilated superior ophthalmic vein. The majority of the venous outflow from the cavernous sinus is via the superior ophthalmic vein which is markedly diluted as seen in this scan.
a consequence there was a relative hold up to the filling of the sigmoid sinus. The AVM therefore now drained anteriorly via a collateral circulation into the cavernous sinus. The later phases of the angiogram revealed that there was relatively little downward venous outflow from the cavernous sinus and the majority of this venous outflow was therefore shunted via the superior ophthalmic vein which was markedly dilated as a consequence. The patient’s symptoms of unilateral proptosis and venous engorgement were therefore a manifestation of increased blood flow through collateral shunts that had developed as a consequence of a stricture in the principal drainage channel of the existing AVM, which itself had not changed significantly in size.

**Comment**

Arteriovenous malformations (AVM) are lesions which consist of racemose networks of arterial and venous channels which communicate directly rather than through a normal capillary bed. These communications are of two types: fistulous and plexiform. In the fistulous type an arterial channel empties directly into a venous channel, while in the plexiform type one or more arterial channels feed a vascular conglomerate that comprises multiple arteriovenous communications from which one or more venous channels emerge as draining veins. The fistulous types are usually supplied by meningeal branches of the external carotid artery and therefore they are also known as dural AVMs. The plexiform type, in contrast, are supplied by branches of the cerebral or cerebellar arteries and hence are also known as pial AVMs. The AVM most commonly encountered by the ophthalmologist is the acquired carotocavernous sinus fistula, a dural AVM whose characteristic neuro-ophthalmic presentation results from the arterialisation of the orbital venous system. Proptosis and orbital venous engorgement secondary to direct arterialisation of the venous system has also been reported in dural AVMs involving the torcular herophili and the Galenic system. The more common plexiform (pial) AVMs typically present with complications that arise from the massive venous run off that is generated by the associated arteriovenous shunts—namely, intracranial haemorrhage, seizures, and recurrent headache. Symptomatic orbital drainage of plexiform (pial) AVMs is rare and when it does occur the AVM is usually in an anterior location.

We describe a patient with a posteriorly located plexiform (pial) AVM who presented with proptosis and vascular engorgement of the left globe as a result of the venous outflow of the AVM being shunted anteriorly through newly opened collateral channels. Unilateral proptosis associated with anterior shunting has previously been described in a child with bilateral sigmoid sinus hypoplasia, but has never been described in a patient with a posteriorly located plexiform AVM. Our case is also extremely unusual in that we have angiographic evidence which shows that these collateral channels developed as a result of an acquired stenosis at the junction of the transverse and sigmoid sinus, previously its dominant route of venous drainage. The aetiology of this focal stenosis is unknown. It is unlikely to be a consequence of the stereotactic radiosurgery as the site of the stenosis is remote to the previously treated area. It is more likely that the stenosis represents a response to the chronic endothelial damage that is known to accompany the hyperdynamic circulation of AVMs.

This case serves to illustrate that not all adults who present with proptosis and venous engorgement of the globe have a carotocavernous sinus fistula and other anomalies of venous drainage must occasionally be considered.

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Case report
We examined a 10 month old male child who presented to us with complaints of ptosis and ophthalmoplegia of the right eye for a month before presentation. The child was born normally at full term to a healthy young mother with an uneventful perinatal and postnatal period. The milestones of the child were recorded as normal. The child had an older sister aged 3 years who gave no history of similar complaints. There was no history of fever, vomiting, pain, trauma, seizures, difficulty in swallowing or chewing, or any history of excessive lassitude or listlessness given by the parents with regard to the child.

General physical examination revealed a child with normal physical and mental development. Systemic examination revealed no presence of heart block or any other cardiovascular abnormality. Neurological examination revealed no motor weakness, incoordination, ataxia, or areflexia. On ocular examination, the patient had moderate ptosis of the right upper lid with absence of any voluntary movement of the right eyelid. Ocular movements of the left eye appear to be normal. The child was attentive to both visual and auditory stimuli and frequently changed his head posture to look in the direction of origin of the visual or auditory stimuli. The pupillary reactions were normal in both eyes. In the primary gaze position, the visual axes of both eyes were parallel (Fig 1). The child was examined twice after a period of sleep and similar findings were recorded again.

Examination of the anterior and posterior segment of both eyes under general anaesthesia was unremarkable. The forced duction test was negative. Based on the above findings, a tentative diagnosis of an intracranial space occupying lesion was made. However, a contrast enhanced computed tomograph (CT) scan of the head and orbit revealed no abnormalities. Cerebrospinal fluid examination was also normal.

The child was subsequently subjected to an edrophonium test (0.15 mg/kg body weight intravenously), which was unequivocally positive with complete resolution of the ptosis and ophthalmoplegia (Fig 2). A repetitive stimulation of the left median nerve demonstrated a significant decremental response (22%). Subsequently, a serum analysis of anti-acetylcholine receptor (AChR) antibody binding antibodies was performed. Abnormal titres of anti-acetylcholine receptor binding antibodies (>0.8 nmol/l) were present which confirmed the diagnosis of childhood myasthenia gravis. A contrast enhanced CT scan of the thorax was normal. The child was started on tablet neostigmine 3 mg once daily along with oral prednisolone (0.5 mg/kg body weight) and followed up regularly. Marked improvement of the ptosis and ophthalmoplegia was observed which persisted at 1 year of follow up. Systemic steroids were gradually tapered off after 6 months.

Comment
Our patient was diagnosed to have juvenile myasthenia gravis, which comprises approximately 1% of all cases of myasthenia gravis. It

References
Phototoxic maculopathy following uneventful cataract surgery in a predisposed patient

Operating microscope light induced foveal damage is a well recognised occurrence following ocular surgery including complicated cataract extraction, complex anterior segment procedures, and vitrectomy surgery. An increased risk of phototoxicity is associated with an operating time greater than 100 minutes, increased body temperature, unfiltered blue light, and hyperoxaemia. We report a patient who developed a phototoxic lesion during routine cataract surgery possibly related to underlying systemic lupus erythematosus (SLE) and hydroxychloroquine treatment. We examine the measures taken to prevent recurrence with second eye surgery and the implications for routine cataract surgery are discussed.

Case report

A 39 year old woman presented with blurred left vision and glare. One year previously she had been diagnosed with SLE for which she had been taking hydroxychloroquine 200 mg and prednisolone 5 mg daily for 18 months. She had bilateral subcapsular cataracts and underwent routine left phacoemulsification and lens implant surgery under general anaesthesia.

Directly after surgery, the patient noted two confluent blind spots immediately below fixation in the left eye. When first seen by us the left visual acuity was 6/9, intraocular pressure was 16 mm Hg, and the eye was quiet with a well centred posterior chamber lens implant. Fundal examination of the left eye showed two well circumscribed areas of coarse mottled retinal pigment epithelial (RPE) disturbance, approximately one and a half disc diameters in size, superotemporal to the fovea (Fig 1). There was no subretinal fluid or haemorrhage associated with these lesions. A fluorescein angiogram supported these findings (Fig 2) and the lesions were felt to be consistent with photic injury.

The patient was keen to proceed with right phacoemulsification and lens implantation in view of anisometropia. She had stopped her hydroxychloroquine immediately after the first operation and the following precautions were agreed with the patient before surgery in the second eye: the use of microscope filtration throughout surgery, switching off the microscope light when instruments were not in the eye, tilting of the eye into downgaze (so that any phototoxic lesion would be superior to fixation and not result in a homonymous scotoma), and the maintenance of low body temperature and low inspired oxygen concentration during surgery.

An uncomplicated right phacoemulsification and lens implant was subsequently performed. Two weeks after surgery, the right visual acuity was 6/5, anisometropia was abolished, and funduscopy was normal.

Comment

Phototoxic lesions are defined as the retinal lesions produced after a relatively short exposure to an intense light source such as the operating microscope. Historically, these lesions were typically located inferior to the fovea as a result of the slight downgaze of the eye during extracapsular cataract surgery and a coaxial microscope beam directed inferior to the fovea. The incidence of retinal phototoxicity from the operating microscope has been reported at between 0% and 28%, with large series reporting angiographic evidence of retinal phototoxicity in 3–7% of cases.

Retinal phototoxic lesions first appear as well circumscribed outer retinal whitening (oedema) with mild disturbances of the RPE often with a light border visible after a few days. Ophthalmoscopically, a subtle discrete margin can be seen. After the first week, lesions are characterised by coarse alterations of the RPE layer with a fluorescence angiography demonstrating characteristic early discrete mottled hyperfluorescence with late staining but no leakage.

Certain photosensitising drugs, such as hydroxychloroquine, have been reported to predispose to the development of retinal phototoxic lesions. However, there are no published reports of a similar association with systemic photosensitive conditions such as SLE. It has been suggested that patients who have SLE have increased numbers of chromosome breaks and rearrangements correlated with a low molecular weight chromosomal damaging agent (clastogenic factor) present in lymphocytes that sensitises them to near ultraviolet light (360–400 nm) light. The presence of this photoactivated agent explains why SLE patients show an aggravation of their characteristic skin condition after exposure to sunlight. Certain pharmacological agents, such as hydroxychloroquine, which is used in the treatment of SLE, can also predispose to the development of phototoxicity. The mechanisms behind the predisposition to phototoxicity in humans of chloroquine and its derivatives are less clear. However, it is known that these drugs accumulate in the RPE layer of the retina and can cause drug induced lysosomal abnormalities including diminished vesicle fusion, diminished exocytosis, and reversible “lysosomal storage disease.” These two different predisposing mechanisms to phototoxicity presumably coexisted in this patient and may have significantly increased her risk.

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References

Figure 1 Left eye showing two well circumscribed areas of coarse mottled retinal pigment epithelial disturbance.

Figure 2 Fluorescein angiogram of left eye.
Several procedures were undertaken to prevent the occurrence of a phototoxic lesion in the second eye. These included the use of ultraviolet filters on the microscope, although air can also be used in the anterior chamber to defocus the light from the retina as can light barriers over the cornea or in the microscope. The time and power of coaxial illumination from the microscope was minimised by turning the microscope off when in use. Light barriers were not in use in the eye, and the patient’s eye was kept in downgaze so that the operating microscope’s axis was aligned off the patient’s visual axis so that any phototoxic lesion would be extrafoveal and not cause a homogeneous scotoma. Since animal studies have demonstrated a benefit of altering core ocular temperature, the patient’s body temperature was kept low to reduce the temperature within the eye. Alternatively, irrigation solutions may be cooled relative to the eye but ophthalmologists should be aware of the risk of phototoxic retinal lesions in patients taking photosensitising pharmacological agents particularly for underlying potential photosensitising systemic conditions. Consideration should be given to stopping treatment before surgery and taking appropriate surgical precautions.

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References

Bilateral uveitis after intravesical BCG immunotherapy for bladder carcinoma

Intravesical bacille Calmette-Guérin (BCG) is indicated for the treatment and prophylaxis of carcinoma in situ of the urinary bladder and the continous use in the microsce. The time and power of coaxial illumination from the microscope was minimised by turning the microscope off when in use. Light barriers were not in use in the eye, and the patient’s eye was kept in downgaze so that the operating microscope’s axis was aligned off the patient’s visual axis so that any phototoxic lesion would be extrafoveal and not cause a homogeneous scotoma. Since animal studies have demonstrated a benefit of altering core ocular temperature, the patient’s body temperature was kept low to reduce the temperature within the eye. Alternatively, irrigation solutions may be cooled relative to the eye but ophthalmologists should be aware of the risk of phototoxic retinal lesions in patients taking photosensitising pharmacological agents particularly for underlying potential photosensitising systemic conditions. Consideration should be given to stopping treatment before surgery and taking appropriate surgical precautions. It is impossible to definitively prove that intravesimal BCG caused our patient’s bilateral anterior uveitis. The possibility of an infective aetiology by active mycobacterium must also be considered. If this were the case definitive diagnosis could have been established by sending a specimen of aqueous for acid fast staining, culture, and polymerase chain reaction (PCR).

It is plausible that intravesimal BCG can cause acute inflammatory and hypersensitivity reactions in the eye secondary to a systemic inflammatory response. Contrary to previous thought, HLA-B27 need not be a determining factor for uveitis associated with intravesical BCG. The patient presented with case of isolated bilateral anterior uveitis presumed secondary to intravesical BCG in a patient who is HLA-B27 negative.

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References

Childhood blindness

The latest issue of Community Eye Health (No 40) discusses new issues in childhood blindness, with an editorial by Clare Gilbert, senior lecturer at the International Centre for Eye Health. For further information please contact: Journal of Community Eye Health, International Centre for Eye Health, Institute of Ophthalmology, 11–43 Bath Street, London EC1V 9EL, UK (tel: +44 (0)20 7608 6910; fax: +44 (0)20 7250 1307; email: eyecancers@ucl.ac.uk; website: www.jech.co.uk). Annual subscription (4 issues) UK£25/$US40. Free to workers in developing countries.

International Centre for Eye Health

The International Centre for Eye Health has published a new edition of the Standard List of
SPECS represents over fifty different organisations related to eye disorders ranging from conditions that are relatively common to very rare syndromes. We also include groups who offer support of a more general nature to visually impaired and blind people. Support groups meet regularly in the Boardroom at Moorfields Eye Hospital to offer support to each other, share experiences and explore new ways of working together. The web site www.eyeconditions.org.uk acts as a portal giving direct access to support groups own sites. The SPECS web page is a valuable resource for professionals and may also be of interest to people with a visual impairment or who are blind. For further details about SPECS contact: Kay Parkinson, SPECS Development Officer (tel: +44 (0)1803 334; email: k@eyeconditions.org.uk; web site: www.eyeconditions.org.uk).

The 3rd Interdisciplinary Symposium on the Treatment of Autoimmune Disorders

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

BEAVRS Meeting

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

CORRECTIONS

We regret that an error occurred in a paper that appeared in the May 2001 issue of the BJO by Lauande-Pimentel et al (Discrimination between normal and glaucomatous eyes with visual field and scanning laser polarimetry measurements, 2001;85:586–91).

After revision of the paper the authors found that the published cut-off point of the structural linear discriminant formula (LDF) was incorrect.

The original formula described by Lauande-Pimentel et al is:

\[ \text{LDF} = -3.131 + (0.994 \times \text{ellipse modulation}) - (0.017 \times \text{the number}) - (0.086 \times \text{average thickness}) + (0.111 \times \text{ellipse average}) \]

The correct suggested cut-off point, defined as abnormal, is <0.43 instead of <-0.547. The use of such formula, with the suggested cut-off point, resulted in an enhancement in the sensitivity (sensitivity = 90.4%, specificity = 82.4%) of the scanning laser polarimeter to detect glaucomatous changes.

An error occurred in the article: Additive effect of unoprostone and latanoprost in patients with elevated intraocular pressure Br J Ophthalmol 2002;86:75–9. The authors should have been listed as Tin Aung, Paul T K Chew, Francis T S Oen, Yiong-Huak Chan, Lennard H Thean, Leonard Yip, Boon-Ang Lim, Jade Soh, and Steve K L Seah.

An error occurred in the article: Fluoxetine oral administration increases intraocular pressure. Br J Ophthalmol 1996;80:678. The authors should have been listed as C Costagliola, I Mastropasqua, I Steduto, N Testa.