Epithelial downgrowth following clear cornea phacoemulsification in a buphthalmic eye

Epithelial downgrowth is a rare complication which can occur following anterior segment surgery, and penetrating ocular injury. A prevalence as high as 16–29% among eyes enucleated after cataract surgery has been reported in the older literature, but advancements in the microsurgical techniques have decreased that rate to less than 0.2%. To our knowledge only two cases of epithelial downgrowth following clear corneal phacoemulsification have been reported.1,2 We present the clinical and histopathological features, and the successful surgical management, of a patient with extensive intraocular involvement of epithelial downgrowth with follow-up clear corneal phacoemulsification.

Case report
A 26 year old man was followed up for bilateral congenital glaucoma. He had undergone several bilateral trabeculectomies in the past. Preoperatively his intraocular pressures (IOP) were well controlled with no medication. He underwent phacoemulsification with implantation of a polymethylmethacrylate (PMMA) lens through a 5 mm superior clear corneal wound in the right eye. The corneal wound was closed with Nylon sutures, as the eye wall was thin secondary to buphthalmos and the temporal end of the wound was irregular extending to the limbus. At the end of the surgery the wound appeared secured with a deep anterior chamber. Two weeks postoperatively he presented with wound leak from the corneal section. A conjunctival hood was fashioned over the corneal wound in an attempt to prevent wound leak. Four weeks later he developed an epithelial downgrowth in the form of a retrocorneal membrane with involvement of the iris from the 7 o’clock to 2 o’clock position (fig 1, top). He underwent a corneosclero-iridocyclectomy with removal of the IOL and posterior capsule combined with anterior vitrectomy. No ad- junctive cryotherapy was used. A horseshoe-shaped corneoscleral graft was sutured in place. Histopathological examination confirmed extensive downgrowth of non-keratinised stratified squamous epithelium involving the cornea, the angle, anterior and posterior surface of the iris, the ciliary body, and extending on to the posterior capsule (fig 1, bottom and fig 2, top). At 17 months follow up he had a vision of 6/12 with aphakic correction and there was no evidence of recurrence of the downgrowth (fig 2, bottom).

Comment
Epithelial downgrowth is characterised by progressive advancement of an epithelial membrane onto the intraocular surfaces and it can lead to intractable and painful glaucoma. It can occur in three forms: (1) “pearl tumours” of the iris, (2) epithelial cyst, (3) growth of epithelium as a sheet on to the anterior chamber structures. This last form has been shown to have the worst prognosis.3 Although there is no single predisposing factor, wound fistula, damage to the corneal endothelium, and corneal stromal vascularisation can have a role in the pathogenesis of the epithelial invasion.4 Various methods have been tried in the treatment of epithelial downgrowth.5 It has been shown that patients treated surgically underwent fewer recurrences than patients managed conservatively. However, surgical management of epithelial downgrowth is difficult, particularly when the anterior chamber contains sheets of epithelium. Knauf et al6 reported surgical success following surgical excision of a localised cystic downgrowth. Extensive disease involvement is associated with a poor outcome.6 We feel that early detection and surgical intervention can result in improved outcome even in cases where the downgrowth has extensive intraocular involvement as demonstrated in this case. Experimental evidence suggests that the presence of endothelium inhibits epithelial downgrowth through direct intercellular interactions.7 There are previous reports documenting decreased endothelial cell density in eyes with congenital glaucoma.8 It is possible that the decreased endothelial cell count combined with persistent wound leak would have contributed to the extensive downgrowth of the epithelium in our patient. We hope this case report highlights this rare complication of small incision cataract surgery.

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References
Posterior uveal effusion syndrome after trabeculectomy in an eye with ocular venous congestion

Schepens and Brockhurst,1 in 1963, used the term uveal effusion syndrome to describe spontaneous bilateral serous detachments of the choroid and ciliary body with exudative retinal detachment, primarily occurring in middle and late adult females. Many cases have been reported since then, especially in nanophthalmic eyes.1,2

We report an interesting case of a young, healthy female patient with mild ocular venous congestion who developed posterior serous retinal detachment and uveal effusions after trabeculectomy for open angle glaucoma.

Case report

A 32 year old white female was referred to the glaucoma service at Doheny Eye Institute for high intraocular pressure in the left eye. Visual acuity was 20/20 right eye and 20/25 left eye. Manifest refraction was +2.75+1.50 x13 right eye and +0.50+2.00 x160 left. An afferent pupillary defect was noted in the left eye. Applanation intraocular pressures were 17 mm Hg right eye and 36 mm Hg left.

Slit lamp examination of both eyes revealed dilated and tortuous conjunctival and episcleral vessels, although more obvious in the left eye than the right eye. The anterior chambers were deep and quiet. The cornea and lens were clear.

Gonioscopic evaluation of both eyes showed open, grade IV Shaffer angles with no peripheral anterior synchiae. Blood was noted in the canal of both eyes. Cup to disc ratios were 0.80 right eye and 0.95 left eye. The remainder of the posterior segment was normal.

Past ocular history was remarkable for strabismus surgery on both eyes at the age of 2.

Because of clinical evidence of mild venous outflow congestion in both eyes, further tests were done to evaluate for a possible carotid-cavernous fistula. Magnetic resonance angiography revealed no evidence of a carotid-cavernous fistula. Ultrasonography revealed no evidence of any orbital mass. Orbital soft tissues were normal bilaterally. Superior ophthalmic veins were not dilated. Choroid and retinal layers were normal. However, bilateral diffuse moderate thickening of sclera was noted without a T sign in either eye. Axial lengths were 21.5 mm in the right eye 22.5 mm in the left.

No history of collagen vascular disease was elicited. Complete blood count (CBC) and erythrocyte sedimentation rate (ESR) were normal; antinuclear antibody (ANA) and rheumatoid (Rh) factors were negative.

Because the patient continued to have high uncontrolled intraocular pressure on maximum medical therapy, trabeculotomy of the left eye was performed. Intraoperatively, after the scleral fistula was created, a sudden lowering of the anterior chamber pressure and anterior displacement of the lens-iris diaphragm occurred. An inferotemporal sclerotomy was used to drain 0.5 ml of clear serous suprachoroidal fluid, thereby relieving the posterior pressure.

On the first postoperative day, visual acuity was hand movements. A posterior retinal detachment was noted in the macular area, extending to the equator on the temporal side. The subretinal fluid was loculated in the posterior pole. Posterior retinal folds were noted, but no retinal holes were found in the macula or the periphery. Applanation intraocular pressure was 30 mm Hg.

Ultrasonography revealed posteriorly thickened choroid, along with the diffusely thickened sclera. (Preoperatively, ultrasonographically, sclera was thickened bilaterally but choroid was normal.) A diagnosis of uveal effusion syndrome was made.

The shallow serous macular detachment seemed unchanged 3 days postoperatively. However, on postoperative day 4, the macular detachment resolved, but the choroid still looked boggy and macular choroidal folds were seen. The visual acuity improved from hand movements to 20/200 within a week, and to 20/60 over the next 3/2 months.

Comment

Schepens and Brockhurst reported a series of 17 patients with uveal effusion syndrome in 1963,5 predominately noted in middle aged females. Flat, annular choroidal detachments were seen in five (30%) of their patients. Rarely, glaucoma occurred secondary to anterior displacement of the ciliary body. Thickened sclera (about 2 mm) was found in 47% of their cases.

Since then, numerous cases have been reported.6,13 Gass described the features of idiopathic serous detachments of the choroid, ciliary body, and retina (uveal effusion syndrome) in nine patients. He noted dilated episcleral vessels, iris, ciliary processes, posteriorsuprachoroidal pigment epithelium (RPE) changes, and characteristic ultrasonographic and angiographic findings. None of these patients had previous trauma, surgery, ocular or inflammatory disease, or systemic disease.

Posterior suprachoroidal fluid was noted in half of the patients. Ultrasonography revealed slightly smaller than normal globes, even in the presence of myopic refractive error in one patient. The two patients who had gonioscopy had partial or complete filling of Schlemm’s canal with blood. The author attributed the above clinical findings to partial obstruction of the vortex veins. Fluorescein abnormalities included slow perfusion of the choroid and prolonged choroidal fluorescence, suggesting choroidal vascular congestion.1

Uveal effusion syndrome is occasionally associated with a normal or narrow eye (nanophthalmos). This association was first reported by Brockhurst in 1974 in five nanophthalmic patients who underwent glaucoma surgery for either acute or chronic angle closure glaucoma. The thickened sclera seen in this anomaly may lead to partial obstruction of the vortex veins, causing the choroidal effusions.7

In our case, although the patient had slightly shortened globes (axial length of 21.5 mm right eye, 22.5 mm left), they were not nanophthalmic. The thickened sclera in each eye was consistent with slightly small hyperopic eyes. The corneal diameters were normal. The anterior chambers were deep.

The dilated episcleral vessels and blood in Schlemm’s canal suggested compromised venous outflow. Moderately thickened sclera, noted on ultrasonography, probably caused impaired venous drainage by mechanically compressing the vortex veins. Such an eye would be predisposed to uveal effusions. An abrupt lowering of intraocular pressure during sclerostomy may have triggered the formation of choroidal effusions. The periph- eral serous choroidals were drained intraoperatively, leaving behind loculated, suprachoroidal fluid, which resulted in an unusual serous macular detachment. In a normal eye, this small amount of suprachoroidal effusion and subretinal fluid is readily absorbed. In our patient, however, we presume that the thickened choroid impaired the retinal pigment epithelial pumping mechanism. Although the serous retinal detachment resolved within a week, the choroidal folds remained visible for months.

Elective surgical intervention should be avoided in patients with compromised venous outflow. Potentially blinding complications can occur intraoperatively or postoperatively in these eyes. Patients with ocular venous congestion should be advised of the potential complications of uveal effusion syndrome with any intraocular surgery.

Recognition of the classic signs and symptoms of uveal effusion syndrome is very important. The natural course is variable. The effusions may resolve spontaneously. Medical treatment with steroids and anti-metabolites is usually not helpful. Decompression of vortex veins has had limited success. Scleral windows to improve venous outflow of suprachoroidal fluid have been successful.2

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Intravitreal triamcinolone acetonide for ischaemic macular oedema caused by branch retinal vein occlusion

No proved treatment exists for ischaemic macular oedema associated with branch retinal vein occlusion (BRVO) despite the potential for significant visual loss in affected eyes.

We report a patient with an ischaemic BRVO with associated macular oedema treated with intravitreal triamcinolone acetonide.

Case report

A 38 year old man was referred to our department with a 6 week history of acute visual loss in his right eye (RE). On examination, visual acuity was 6/60 RE and 6/6 LE. A right relative afferent pupillary defect was present. Intraocular pressures were 9 and 10 mm Hg, respectively. Anterior segment examination showed no evidence of neovascularisation. There was no vitreous inflammation. Dilated fundus examination revealed a right inferotemporal BRVO with macular oedema. Collarette disc vessels were present. Systemic and haematological examinations, including full blood count, erythrocyte sedimentation rate, C-reactive protein, clotting screen, serum angiotensin converting enzyme, serum cholesterol and triglycerides, clotting screen, chest x ray, and ECG, were negative for systemic vascular or vasculitic disease.

At review 4 months later, right visual acuity had reduced to counting fingers. Fluorescein angiography showed significant macular ischaemia and non-perfusion with a broken foveal capillary ring (fig 1A). There was diffuse leakage and additional masking of normal foveal contour 2 weeks after first intravitreal triamcinolone injection demonstrating marked macular oedema with intraretinal cysts. (B) Late phase fluorescein angiogram showing macular oedema.

Vein Occlusion Study group showed that argon laser grid photocoagulation can improve visual acuity in eyes with BRVO with macular oedema reducing vision to 20/40 or worse.6

However, laser treatment is inappropriate in eyes with foveal capillary non-perfusion. Eyes with BRVO and a broken foveal capillary ring have a poorer prognosis for visual acuity.7 Although one study reported a favourable visual outcome in 91% of eyes with BRVO and macular oedema associated with incomplete macular perfusion,8 the majority of cases with a good visual outcome were presented with visual acuities of 20/100 better. Poor presenting visual acuity is correlated to poor visual prognosis.4

In this case, a patient with a chronic BRVO and ischaemic macular oedema had complete resolution of the macular oedema 2 weeks following intravitreal injection of triamcinolone. The resolution of macular oedema was associated with a significant improvement in visual acuity from counting fingers to 6/24. This final visual acuity was presumably limited because of residual ischaemic macular damage.

Although the mode of action by which triamcinolone induces resolution of macular oedema remains poorly understood, in vitro studies and clinical observations indicate that triamcinolone has the capacity to reduce the permeability of the outer blood-retinal barrier.3

Intravitreal triamcinolone has been shown to be safe and effective when used for the treatment of cystoid macular oedema caused by uveitis,9 diabetic maculopathy,10 central retinal vein occlusion,11 and post-cataract surgery.12 IOP elevation may occur in up to 90% of eyes after triamcinolone injection.13 Other potential risk includes cataract development, retinal detachment, and endophthalmitis. Despite good initial anatomic and visual response to intravitreal triamcinolone when used in these conditions, macular oedema has been reported to recur following treatment, often necessitating repeated injections. Further follow up is required to determine if macular oedema recurs following treatment in branch retinal vein occlusions.

This case suggests that the use of intravitreal triamcinolone acetonide for the treatment of macular oedema due to BRVO warrants further study.

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360 degree giant retinal tear as a result of presumed non-accidental injury

Giant retinal tears are defined as retinal tears extending 90 degrees or more around the circumference of the fundus. They most commonly occur spontaneously but are associated with direct trauma in 20–25% of cases. We report the case of a 360 degree giant retinal tear occurring in a baby girl as a result of presumed non-accidental injury (NAI).

Case report

A 4 month old girl presented with iris is periorbital oedema with subconjunctival haemorrhage. Other manifestations reported include retinal detachment and retinoschisis. The collection of signs seen in this case, including retinal detachment, anterior lens opacity, bruising of the contralateral eye, MRI and electrophysiological findings are highly suggestive of NAI. The electrophysiological findings were suggestive of multiple contusions to the head and it is our belief that the giant retinal tear and subsequent retinal detachment occurred as a result of a direct blow to the eye rather than violent shaking. This is, to our knowledge, the first reported case of a 360 degree retinal tear associated with NAI. Practitioners should be alert to the possibility of NAI when faced with a giant retinal tear in a young child.

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A new look at ophthalmodynamometry

It was with great interest that I read the article by Jonas 1. It is indeed noteworthy that efforts are being undertaken for revising ophthalmodynamometry. This is especially pertinent as this method is, despite the many modern techniques published within the past 20 years, the only way to gain insights on the position of the central retinal vessels. A direct measuring device has demonstrated the usefulness of this examination.2 Nevertheless, Jonas's recent article raises some questions that must be clarified in light of additional articles that will soon be published by the same author as well as for those who are no longer familiar with ophthalmodynamometry.

The device, as it is described, appears to measure the appositional force exerted by the examiner's hand on the contact lens placed upon the eye. One has to clearly distinguish between the appositional force and any pressure within the eye itself. In no way does the former reflect any circulatory parameter of the eye, certainly not a collapse pressure. Moreover, this measurement is strongly dependent on the IOP which, in these experiments, seems not to have been considered. The collapse pressure itself strongly depends on the IOP—that is, the tissue pressure around the central retinal vessels, a matter of debate for many years.3 Since the examination itself lowers the IOP considerably, any repetition of the examination must yield a new value. Therefore, it has been recommended to repeat the examination only after a longer rest period for the eye. 4-6, as cited in this study, the examination has been repeated nine times in succession, the results should show a linear decrease of the distribution curve.

Pulsations of the central retinal vein express the fact that the IOP oscillates at levels near the cerebrospinal fluid pressure, a difference in the order of 1–2 mm Hg.4 We wonder whether the set-up and the method are accurate enough to document such subtle changes. In order to obtain answers to questions concerning the circulation of the central retinal vessels in the diseases mentioned in the article, it would be necessary to convert the force in the corresponding pressure values. We look forward to reading the articles the author cited in the references list and to clarification of the points raised in this letter.

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References

Time to learn from what is known

We read with interest the recent editorial by Melese et al.1 Trachoma is responsible for up to two million cases of blindness worldwide, yet to a large extent it is a forgotten disease which affects the poorest and most medically underserved populations.2

The World Health Organization (WHO), together with the International Agency for Prevention of Blindness, jointly launched Vision 2020—the right to sight” in 1999 which aims to eliminate preventable blindness by the year 2020, including blindness from trachoma. WHO has endorsed “SAFE” (Surgery for trichiasis, Antibiotics to reduce the prevalence of chlamydial infection, and Facial cleanliness and Environmental change to reduce the disease transmission) as the strategy implemented by national programmes to achieve the elimination of blinding trachoma. We have recently undertaken a review of the evidence base of the SAFE strategy and judged it to be strong for the surgery and antibiotics components, but weaker for the other components.3 We therefore concur with the opinion of Melese et al that more research is required, not only to develop a protocol for the rational use of antibiotics, as they suggest, but also to strengthen the evidence relating to the “F” and “E” components.

The SAFE strategy has been implemented by the national control programmes of many endemic countries in Africa, Asia, and Latin America with varying degrees of success. Melese et al make the observation that the experiences gained from research studies are not directly transferable to real life settings; the need, therefore, is for the publication of the experiences of countries that have successfully dealt with trachoma through implementation of the SAFE strategy to serve as a best practice model for other countries. Morocco is a good example. A decade ago trachoma was a public health problem in five provinces of Morocco, but with the efficient implementation of the SAFE strategy the prevalence of active trachoma and trichiasis has declined dramatically so that Morocco is now close to eliminating trachoma as a public health problem.4 Real life examples such as Morocco documented as case studies allow policy makers and programme managers to learn from the mistakes and successes of public health programmes, because, as Melese et al point out, real life is messier and more complicated than studies.

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NOTICES

HIV/AIDS and the eye

The latest issue of Community Eye Health (No 47) discusses the impact of the HIV/AIDS epidemic on prevention of blindness programmes. 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)20 7612 7964; email: Anita.Shah@lshtm.ac.uk; website: www.jech.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 SPECs 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 programmes 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 860 344; 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 93101-9838, USA (tel: +805 963 3303; fax: +805 963 3364; email: hsbrown.md@cox.net or seeintl@seeintl.org; website: www.seeintl.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 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.

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. Further details: ISOPT Secretariat (website: www.kenes.com/isopt).

XVth Meeting of the International Neuro-Ophthalmology Society


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 International Global Congress Organisers and Association Management Services, 17 Rue du Cendrier, PO Box 1726, CH-1211 Geneva 1, Switzerland (tel: +41 22 908 0488; fax: +41 22 732 2850; email: autoim04@kenes.com; website: www.kenes.com/autoim2004).
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