Spontaneous venous pulsations should be monitored during glaucoma therapy

It is well established that lowering intraocular pressure slows or halts progression of glaucoma. None the less, changes in intraocular pressure also affect the pressure gradient across the lamina cribrosa. Normal intraocular pressures combined with low intraocular pressures produce the same pressure differential across the laminae as elevated intraocular pressures in conjunction with normal intracranial pressures.1–3 Failing to factor in both intraocular and intracranial pressures may create an inappropriate distinction in the pathogenesis of glaucomas currently labelled as normal tension from those where elevated pathogenesis of glaucomas currently labelled create an inappropriate distinction in the

References


Acanthamoeba keratitis in Ghana

Suppurative keratitis due to Acanthamoeba spp is most commonly associated with poor contact lens hygiene. However, recently there have been reports of keratitis caused by Acanthamoeba spp in the tropics in non-contact lens wearers.1 We report one such case of Acanthamoeba keratitis in Ghana, west Africa.

Case report

A 25 year old male driver (from Accra) reported to a hospital in Bawku, northern Ghana. He presented with symptoms of chronic corneal ulceration (duration of symptoms 31 days) and visual acuity in his affected eye was reduced to perception of hand movements.

On examination there was a central corneal ulcer (greatest diameter 8 mm) involving more than 50% of the corneal epithelium and more than two thirds infiltrate. The lesion was white, dry in texture, with a well defined margin and raised slough; flare, AC cells, and a posterior corneal abscess were present. Satellite lesions, immune ring, fibrin, keratic precipitates, and hypopyon were absent.

No organisms were isolated from culture on blood agar or Sabouraud agar. Non-nutrient agar was not available at this rural hospital. Corneal ulcer material stained with Gram stain and lactophenol cotton blue mountant revealed Acanthamoeba spp as a potential causative agent of chronic keratitis. In patients presenting at rural hospitals in the tropics. In addition, the usefulness of simple stains such as lactophenol cotton blue and Gram stain to visualise Acanthamoeba spp is demonstrated as previously described in the literature.1

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Acanthamoeba keratitis in Ghana

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Need for measurement of porphyrins in teardrops in patients with congenital erythropoietic porphyria

Congenital erythropoietic porphyria (CEP: MIM #263700) is an extremely rare disorder inherited as an autosomal recessive trait. The cause of this disease is the deficient activity of uroporphyrinogen III synthase (UROS: EC 4.2.1.75). Since a cloning of UROS gene (UROS: Genebank NM0000735), efforts have been made to clarify underlying mutations that cause CEP. To date, more than 20 mutations of UROS have been described. Identification of UROS mutations at the molecular level is important for genetic counselling and prenatal diagnosis of affected families.

Clinically, CEP is characterised by severe cutaneous photosensitivity, chronic haemolysis, and massive porphyrinuria resulting from the accumulation in the bone marrow, peripheral blood, and other organs of large amounts of predominantly type I porphyrins, which are not substrates for haem synthesis. Red urine may be observed from infancy, and the teeth become stained red. Haemolytic anaemia, an additional complication, may be helped by splenectomy. Besides such classic manifestations, ocular involvement, including scleral changes, has been reported in patients with CEP. Furthermore, we recently reported the evidence of the accumulation of porphyrins in teardrops in a Japanese patient with CEP who showed scleral changes. To confirm whether the accumulation of porphyrins is a common feature and is a direct cause of ocular involvement, we analysed and confirmed the presence of porphyrins in teardrops in an additional three Japanese patients with CEP.

Case reports

We analysed three Japanese patients with CEP, all of whom were diagnosed by their typical clinical manifestations and by the elevation of porphyrins (uroporphyrin I and coproporphyrin I) in urine (Table 1). For case 2, we performed sequence analysis of UROS and identified a T to C transition of nucleotide 634, which predicted a serine to proline and identified a T to C transition of nucleotide 2. We performed sequence analysis of Acanthamoeba cysts in corneal scrapings by lactophenol cotton blue staining. Arch Ophthalmol 1990; 108:168.

Table 1 Tear drop analysis of porphyrins in Japanese patients with CEP. Case 1 has been reported previously.

<table>
<thead>
<tr>
<th>Case 1</th>
<th>Case 2</th>
<th>Case 3</th>
<th>Case 4</th>
<th>Control</th>
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Figure 1 Scleral involvement shown in case 2.

Table 2 shows some of the Tear drop analysis of porphyrins in Japanese patients with CEP. Case 1 has been reported previously.

Comment

Sclerotic changes at the body surface lesions in CEP are mainly caused by the accumulation of porphyrins. Here, we demonstrated the first evidence that the accumulation of porphyrins in teardrops is a common feature in patients with CEP. No accumulation was observed in normal control, whereas the elevation of porphyrins in teardrops was observed in all patients with CEP. Furthermore, in case 4, who manifested relatively mild phenotype of ocular involvement, also showed mild accumulation of porphyrins, suggesting that the accumulation of porphyrins, especially uroporphyrin I and coproporphyrin I, is a common feature and is a direct cause of ocular involvement, and that its severity depends on the level of porphyrins accumulated in tear capsule. Therefore, it is important to measure tear drops in patients with CEP. We strongly recommend the use of ultra violet spectacles to protect against sunlight and to prevent the initiation and progression of scleral lesions, although some patients do not have eye involvement.

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Uveitis initiating an autoimmune reaction resulting in Goodpasture’s syndrome in a Chinese man

Goodpasture’s syndrome is an autoimmune disease caused by IgG directed against the alpha-3 chain of type IV collagen found in basement membrane. It causes pulmonary haemorrhage and renal failure. The antibody has been demonstrated in the basement membranes in the eye. However, ophthalmological complications in Goodpasture’s syndrome are rare. As with other autoimmune diseases, Goodpasture’s syndrome patients have a strong susceptibility based on a genetic background. Much evidence supports the concept that cross reactivity with exogenous epitopes or insult to the basement membrane can initiate the process of autoimmunity resulting in Goodpasture’s syndrome. We present a case of a Chinese man in whom we believe uveitis precipitated the autoimmune reaction causing Goodpasture’s syndrome.

Case report

A 77 year old Chinese man from Hong Kong presented to the eye casualty department of the Eye Institute of the Central Hospital, Tokorozawa, Japan. He had a 20 year history of Goodpasture’s syndrome, which was diagnosed in 1981. He had a history of haemoptysis and proteinuria, and was being treated with prednisolone 40 mg/day and azathioprine 100 mg/day. He was admitted to hospital because of progressive haemoptysis and renal failure. On examination, he had a patent pupils with no relative afferent pupillary defects. There were no lens opacities, and no corneal changes. Fundus examination revealed no changes. There were no retinal vasculature abnormalities. Visual acuity was 20/20 in each eye.

Further examination revealed a positive indirect ophthalmoscope. There was an active choroiditis with sectoral hypopigmentation. There was no evidence of choroidal neovascularisation. There were no subretinal fluid or serous retinal detachments. There was no evidence of retinal vasculitis. There was no evidence of retinal neovascularisation. The patient had no evidence of anterior uveitis. He had no evidence of posterior uveitis. There were no evidence of retinal vasculitis. There was no evidence of retinal neovascularisation. The patient had no evidence of anterior uveitis.

The patient was treated with oral prednisolone 60 mg/day and azathioprine 100 mg/day. The patient’s haemoptysis resolved within 2 hours. His renal function improved. Further examination revealed no evidence of choroiditis. There were no evidence of choroidal neovascularisation. There was no evidence of retinal vasculitis. There was no evidence of retinal neovascularisation. The patient had no evidence of anterior uveitis. He had no evidence of posterior uveitis. There were no evidence of retinal vasculitis. There was no evidence of retinal neovascularisation. The patient had no evidence of anterior uveitis.

The patient was discharged with oral prednisolone 10 mg/day and azathioprine 100 mg/day. He was reviewed in 4 weeks. The patient had no evidence of choroiditis. There were no evidence of choroidal neovascularisation. There was no evidence of retinal vasculitis. There was no evidence of retinal neovascularisation. The patient had no evidence of anterior uveitis. He had no evidence of posterior uveitis. There were no evidence of retinal vasculitis. There was no evidence of retinal neovascularisation. The patient had no evidence of anterior uveitis.
with a 1 month history of a black patch in the centre of his visual field in the left eye. Before this he noticed floaters and a “black cloud” in the left eye. He did not complain of pain or photophobia.

Three months earlier he had an episode of acute anterior uveitis, which resolved with dexamethasone and cyclopentolate drops. His vision at that episode was 6/24 in the right eye and 6/12 in the left eye. Six months earlier he had developed septicaemia secondary to a urinary tract infection treated with a course of intravenous flucloxacillin and fusidic acid and he made a good recovery.

He came to the United Kingdom 30 years ago and returned to Hong Kong for holidays every few years. There was no family history of note.

He looked well on examination and he was afebrile. Systemic examination was unremarkable apart from an itchy rash on both his legs and his blood pressure was 104/60.

His visual acuity was 6/9, N6 in the right eye and CF, N36 in the left eye. His right eye was white and quiet with evidence of old keratic precipitates (KPs) only. His left eye had mutton fat KPs with ++ cells in the anterior chamber. The right fundus had a healthy disc and retinal vessels, but there were tiny white lesions at the level of the retinal pigment epithelium (RPE) which could represent drusen. In the left eye he had ++ cells in the vitreous, and similar white lesions in the fundus. There were no haemorrhages or cotton wool spots. In the left eye showing multiple dots of hyperfluorescence under the neuroretina at the level of the RPE (Fig 1) showed multiple dots of hyperfluorescence. These dots look more like active disease rather than drusen. There is no leakage from the disc, vessels, or macula.

Investigations were unhelpful so he was treated symptomatically with dexamethasone 0.1% and cyclopentolate drops and an orbital floor injection of 40 mg of Kenalog (triamcinolone) in 1 ml.

The following week he was reviewed in clinic. His left eye had improved to 6/12, N8. However, he complained of generally feeling unwell with dizziness and nausea. There was no temperature or vomiting.

A blood test revealed that he was in acute renal failure with sodium of 125, potassium 6.9, urea 56.9, and creatinine 1681. ESR was 120, FBC 8.0, WCC 10.2, and platelets of 610. Liver function tests were normal apart from albumin of 21. He underwent haemodialysis and a renal biopsy showed Goodpasture’s disease (Fig 2). His glomerular basement membrane antibodies (ELISA confirmed by western blot) were also positive with a level of 77% (0–15 reference range), as were his PEG immune complexes, ANA, ANCA, and anti-DNA were all negative. He was given high dose steroids and had a course of plasma exchange (three 3 litre volume exchanges with FFP/PPF replacement).

His eyes continued to improve during the following month and the inflammation settled. At last review his visual acuity was 6/9 right eye and 6/12 left eye. Unfortunately, he developed haemoptysis and overwhelming septicemia and died 2 months later. A post mortem was declined.

**Comment**

In 1919 Ernest Goodpasture described a patient with haemoptysis, anaemia, and proteinuria and a post mortem showing pulmonary alveolar haemorrhage and proliferative glomerulonephritis. Goodpasture’s disease, is a rare autoimmune organ specific disease in which IgG antibodies are directed against the NCI domain of the alpha-3 chain of type IV collagen, leading to pulmonary haemorrhage, glomerulonephritis, and renal failure. Goodpasture’s syndrome, however, describes Goodpasture’s disease with either haemoptysis or crescentic glomerulonephritis, or both. Clearly, this patient had the complete syndrome. Thirty per cent of patients are ANCA positive and lung or kidney biopsy shows linear immunofluorescent staining for IgG along the basement membrane. The death rate has dramatically improved from 75% to 25% with the advent of plasmapheresis combined with immunosuppressants. However, it is rare to escape irreversible dialysis dependency unless the disease is detected and anti-GBM antibody levels are reduced before oliguria has ensued.

Immunofluorescent and immunohistochemistry studies show that the deposit of the anti-basement membrane antibodies can also occur in the eye in the basement membrane of the choroidal vessels, ciliary body, lens capsule, and Bruch’s membrane. The pathogenic significance of this is still unknown. Furthermore, there have not been any studies confirming the actual presence of type IV collagen or of the alpha-3 epitope in the eye, although it is widely assumed that they are both present.

Documented ophthalmological abnormalities in Goodpasture’s include retinal haemorrhages and exudates and serous retinal detachment. It is possible that these signs may reflect hypertension or ANCA positivity rather than direct damage from antiglomerular basement membrane antibodies possibly because the small percentage of alpha-3 chain or its NCI domain in the eye. As the disease is so florid in the kidneys and the lung, one would expect a disease of similar severity in the eye if the anti-basement membrane antibodies were pathogenic in the eye.

This man had a granulomatous panuveitis with choroidal infiltrates. These ophthalmic features have not previously been described in Goodpasture’s syndrome. The serum from his initial admission with uveitis in February, 2 months after his episode of septicemia, was normal apart from raised ESR and CRP levels and his antiglomerular basement membrane antibodies were 9%, which is within normal limits. Four weeks later in March, the titre of antibodies had risen to a grossly abnormal level of 77%. The temporal relation of developing antiglomerular basement membrane antibodies and disease means it is likely that these complexes occurred in response to his septicemia and produced a reactive uveitis. As type IV collagen is also found in the choroidalplexus, the uveitis may have been exposed type IV collagen to the immune system causing antibody production which results in cross-reactivity with the glomerular basement membrane in the kidney to produce Goodpasture’s syndrome. As with other autoimmune diseases, Goodpasture’s syndrome patients have a strong susceptibility based on a genetic background. Much evidence supports the concept that insult to the basement membrane or cross-reactivity with exogenous
epitopes can initiate the process of autoimmunity resulting in Goodpasture’s syndrome. Interestingly, uveitis and Goodpasture’s share some similar characteristics. The immunological profiles both involve the cytokines IL-6, IL-12, and TGFβ, and both are associated with HLA B7 (indicating a particularly poor prognosis in Goodpasture’s) and HLA DR 15. This case is extremely rare. Other events associated with the induction of anti-GBM disease in humans have included lung damage by infections such as influenza and toxin exposure with tobacco and hydrocarbons. Renal injury by ischaemia and membranous nephropathy can also induce disease. Other causes include lymphoma and adenoma. To our knowledge this is the first reported case where uveitis was the likely precipitating factor for the induction of Goodpasture’s disease. Moreover, ophthalmologists should be aware of all blood test results and check the results of ordered tests. They should follow up the patient with repeated history and examination and repeat tests if the diagnosis is uncertain. Unsuspected fatal diseases as we have seen in this case, may occasionally present to ophthalmologists.

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Fusarium solani keratitis following LASIK for myopia

Fungal keratitis is a rare complication of photorefractive keratectomy (PRK),1 LASIK,2 and LASIK enhancement.3 We describe a patient who developed keratitis in one eye after bilateral LASIK procedures. The keratitis progressed despite intensive topical and systemic antibacterial and antifungal treatment. Initial cultures were negative, but a diagnosis of fungal keratitis was suggested by confocal microscopy. Fusarium solani was isolated following penetrating keratoplasty.

Case report

A 45 year old woman noted blur and discomfort of the right eye 3 days after bilateral LASIK procedures for myopia. The visual acuity was 6/9–3 unaided and there was a 1.5 mm diameter infiltrate beneath the flap (Fig 1A). The left eye was unaffected with an unaided visual acuity of 6/6. Cultures were not taken but her topical antibiotic was changed from chloramphenicol to ofloxacin, and dexamethasone 0.1% four times daily was continued. After a further 4 days the vision had reduced to 6/36 and there was ulceration through the flap over the infiltrate; treatment was changed to hourly gentamicin 1.5%, cefuroxime 5%, and econazole 1%, and dexamethasone was continued to reduce the risk of further flap melt. A culture from the ulcerated corneal surface over the infiltrate was negative. There was continued deterioration and treatment was therefore stopped for 24 hours before the flap was lifted and cultures taken for routine bacteriology, acid fast bacilli (Mycobacterium), and fungi. No organism was identified. Because of the uncertain diagnosis a loading oral dose of fluconazole 200 mg was given followed by 100 mg daily, and a 7 day course of intensive topical vancomycin (5%) and amikacin (2.5%) was started with oral clarithromycin 300 mg twice daily. Confocal microscopy of the edge of the lesion demonstrated filamentary structures in the deep stroma (Fig 1B), but a subsequent tissue biopsy of the flap and the deeper stroma was again negative. Despite intensive treatment with topical amphotericin (0.15%) and natamycin (5%), and courses of oral itraconazole (400 mg daily) and voriconazole (400 mg daily) there was continued deterioration and the cornea perforated (Fig 1C). On the 32nd day following LASIK a 9 mm penetrating corneal graft was performed and the anterior chamber was irrigated with amphotericin (5 µg in 0.1 ml). Dexamethasone 0.1% four times daily was continued after surgery to treat severe intraocular inflammation. Histology of the excised corneal tissue showed filamentary fungal elements anterior to Descemet’s membrane but no evidence of hyphae extending to the margin of the excised tissue. Fusarium solani was subsequently grown from the corneal button and the isolate was reported sensitive in vitro to amphotericin (1.0 µg/ml), partially sensitive to clotrimazole (8 µg/ml), econazole (4 µg/ml), and miconazole (8 µg/ml), and resistant to itraconazole (>16 µg/ml) and fluconazole (>16 µg/ml). Despite continued topical and oral antifungal treatment, two further penetrating grafts were necessary to remove recurrences at the graft interface. Following the last graft topical steriod was substituted with topical cyclosporin A 2% four times daily for 4 weeks, after which fluorometholone 0.1% four times daily was started to control inflammation and prevent vascularisation. All topical antifungal treatment was stopped after 3 months. The final vision at 8 months was reduced to 6/60 as a result of irregular corneal astigmatism, cataract, and graft oedema.

Comment

Fungal keratitis is rare in the United Kingdom but accounts for 17–37% of microbial keratitis isolates in warmer countries. To date all reported cases of fungal keratitis after LASIK have been due to filamentous organisms.4 The diagnosis may be difficult to confirm as cultures and biopsies of the anterior stroma can be negative because of the tendency of filamentary fungi to proliferate in the posterior corneal stroma. A delay in identification of the causative agent often contributes to the poor outcome. Of the four previous reported cases of presumed flap interface infection, excisional keratoplasty was required in three,5,6 and perforation and vascularisation of the corneal occurred in one.7 The only case with a medical cure had a surface infection of a persistent epithelial defect.8 The origin of this infection is uncertain. The patient had visited Florida 2 months previously, where Fusarium sp is the most common isolate from cases of fungal keratitis.9 On the day of treatment the ambient temperature in London was 80°F, but subsequent air culture plates from the treatment area and from the air conditioning system were negative. Because of a reluctance to disturb the LASIK flap there is a temptation to treat infiltrates empirically with broad spectrum antibiotics and steroid,9 rather than lifting the flap to culture the lesion directly. Unfortunately, in this case, two cultures were negative, and the diagnosis of a filamentous fungal infection was suggested by confocal microscopy. A further negative biopsy then delayed a decision for surgical intervention. Corticosteroid may
increase the virulence of pathogenic fungi and
it is normally contraindicated in fungal keratitis, although it has a role in reducing
inflammation after antifungal treatment has been started.⁷ The role of cyclosporin A as an
alternative to steroid after surgery for fungal keratitis requires further evaluation.⁸

Acknowledgement
Mr John Dart provided helpful advice with the man-
agement of this case.

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Uveal metastasis 43 years after resection of bronchogenic carcinoid

Carcinoids are a heterogeneous group of
tumours believed to arise from enteroendo-
crine cells scattered primarily throughout the
gastrointestinal tract, but these tumours are
also found at other sites such as the lungs. Although the term carcinoid underscores the
benign nature of the tumour with an indolent course, its potential for metastasis is widely
recognised.⁹ The most frequent sites for
metastasis include the lymph nodes, liver, and
bone.¹ Metastasis to the eye and orbit is rare. Gastrointestinal carcinoids tend to metasta-
sise to the orbit while bronchial carcinoids have the propensity to metastasise to the
uvea.¹² The interval between diagnosis of the
primary tumour and the recognition of ocular
or orbital metastasis varies. The longest reported interval is 24 years for intraocular
metastasis, and 15 years for orbital metastasis.¹³ We report on a patient with metastatic
ca102carcinoid to the uvea occurring 43
years after excision of the primary bronchial
carcinoid and confirmed by histological and
immunohistochemical tests, and we propose a
mechanism for such delayed metastasis.

Case report

A 63 year old white woman was found to have a
superiorly located choroidal mass in the
right eye and an inferonasal elevated mass in
the left eye on routine examination. Medical
history is significant for the diagnosis in 1958
of bronchial adenoma, presently known as
bronchial carcinoid. The patient underwent
 lung surgery, details of which are unavailable.
She denies any history of smoking. The bron-
chial carcinoid recurred in 1965 and a right
lower lobectomy was performed. The hist-
opathological diagnosis was bronchial ad-
ena, cylindroid type, with metastasis to the
peribronchial lymph node of the basilar
segments.

On initial examination, visual acuity was 20/25 in each eye. Fundus examination of the
right eye revealed a lightly pigmented dome-
shaped golden brown mass at the 12 o’clock
position involving the pars plana ciliaris and
peripheral choroid. Echography revealed a
solid ciliary body and peripheral choroidal
mass lesion located superiorly with an eleva-
tion of 4 mm and a base diameter of 8 mm.
Internal reflectivity was medium to high, with
a moderately irregular internal structure. The
left eye revealed an irregularly demarcated non-
pigmented orange-redish choroidal lesion
with overlying subretinal fluid and calcification
(Fig 1). Echography of this lesion showed an
elevation of 2.2 mm and a base diameter of
7 mm with medium internal reflectivity.

The metastatic examination included mag-
netic resonance imaging of the brain, com-
puted axial tomography of the chest, abdo-
men and pelvis, positron emission
tomography scan, and mammography. It did
not show any evidence of metastasis. Clinical
diagnosis in the left eye waschoroidal
haemangiomaversustauveus. Differential di-
gnooses in the right eye included ciliary body-
 peripheral choroidal melanoma, retinal pig-
ment epithelium (RPE) adenoma, adenocarcinoma, and metastatic lesion. The
patient underwent an iridocyclectomy with
periocular resection of the melanoma.

Table I summarizes the clinical findings in this
case.

Results

We report on a patient with metastatic case of
carcinoid tumour showing infiltrating cells with trabecular, tubular, cord-like or
rosette-like pattern (haematoxylin and eosin; original magnification 300) (Fig 2A).

Figure 1 Fundus photograph of the left eye with a non-pigmented orange-reddish lesion.
We thank Dr James D Sanchez, medical oncologist, for metastatic uveal tumour is generally poor; metastasise as much as four decades after case suggests that carcinoid tumours can bronchial carcinoid led to the clinical impres-
chus as its site of origin.
tumour's histological trabecular pattern and exhibited no staining to TTF-1. Likewise, the carcinoids are compatible with long term negative activity to Ki-67 and non-reactivity to p53 may possibly explain the indolent course of this tumour and its metastasis many years after the resection of the original tumour. Ki-67 is a DNA binding nuclear protein that is expressed in proliferating cells but not in quiescent cells. It is regarded as a fractional measure of tissue and tumour growth.4 P53, a tumour suppres-
sor gene, is the most commonly mutated gene in tumours; its functional inactivation can bring about abnormal cell proliferation. Occa-
sonal staining of the patient's tumour to Ki-67 and the negative staining of the tumour to p53, as well as its histologically well circumscribed appearance, demonstrate low proliferative activity and low aggressiveness. These biological factors may have played a part in the delayed metastasis of the patient's tumour.

**Comment**

Notwithstanding their metastatic potential, carcinoids are compatible with long term survival5 (as attested by our patient's case). To our knowledge, the 43 year interval between diagnosis of the patient's primary tumour and recognition of uveal metastasis is the longest interval reported to date. Low immunorac-
tivity to Ki-67 and non-reactivity to p53 may possibly explain the indolent course of this tumour and its metastasis many years after the resection of the original tumour. Ki-67 is a DNA binding nuclear protein that is expressed in proliferating cells but not in quiescent cells. It is regarded as a fractional measure of tissue and tumour growth.4 P53, a tumour suppres-
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**References**


**Recurrent corneal ulcerations with perforation in keratosis follicularis (Darier-White disease)**

Darier-White disease, also known as keratosis follicularis, is a relatively common genodermatosis(specifically keratosis follicularis (Darier-White disease) and 100,000) with autosomal dominant inheritance and late age of onset. Clinical features consist of disseminated warty papules and plaques with an affinity to the seb-oarhoeic areas of the body. The disease may be exacerbated by sun exposure or sweating and never remits. Ocular involvement in keratosis follicularis is relatively uncommon. Most of the affected patients have dry eye syndrome with and without Sjögren's syndrome. Possible corneal manifestations are peripheral arcus lipoides-like opacifications, asymptomatic nebulor dot-like opacities of the peripheral corneal epithelium, and central epithelial surface irregularities.6,7 Involvement of the eyelids and association with retinitis pigmentosa, subnormal dark adaption, and post-cataract extraction staphy-
lococcal endophthalmitis may also occur.8

We describe the first case of recurrent severe bilateral corneal ulcerations with perforation in a female patient with keratosis fol-
licularis.

**Case report**

A 74 year old woman was referred to the university eye hospital with complaints of redness and reduced visual acuity in the left eye. She had no significant ocular history. She and her two daughters have suffered from keratosis follicularis (Darier-White disease) for many years.

On first examination (February 2001) best corrected visual acuity was 0.8 in the right eye and 0.3 in the left eye. Both eyes showed pathological Schirmer's test tear.

Slit lamp examination showed blepharitis marginalis on both eyes, thickened, knobly lid margins, and a rarefication of the eye-
lashes. In the left eye there was a descemeto-
toe without corneal perforation or intra-
ocular inflammation (Fig 1A).

Microbiological examination of the conjunctiva indicated Staphylococcus aureus, Staphy-
lococcus epidermidis, and Streptococcus viridans. The left eye was treated with ofloxacin and cyclosporin A eye drops. Additionally, both eyes were treated with artificial tears, and the clinical picture slowly improved over the next 9 days.

Four weeks later (March 2001) visual acuity was 0.7 in the left eye. Examination of the left eye revealed corneal staining and thinning at the 6 o'clock position. There were no signs of inflammation. Antibiotic therapy was discontin-
ted.

Seven weeks later (April/May 2001) the patient reported foreign body sensation in the right eye. Slit lamp examination demonstrated 3+ conjunctival injection, 3+ anterior chamber cells, ulceration of the cornea at the peripheral 6 o'clock position, a descemetocele, anterior and posterior synechiae (Fig 1B). The aspect of the left eye was similar to the previous examination 7 weeks earlier.

The patient was again hospitalised and placed on topical ofloxacin and mydriatics. Microbiological examination indicated again Staphylococcus aureus, and additionally Candida albicans and Blastomyces. Therefore, topical antmyotic eyedrops were added.

During follow up the corneal infiltration and intraocular inflammation in the right eye slowly reduced.

Nine weeks after discharge (August 2001) the patient returned to the hospital. She reported burning of the right eye for 5 days. The therapy with ofloxacin had been discontinued 3 weeks previously, only the therapy with artificial tears had been continued.
Visual acuity was 0.02 in the right eye and 0.7 in the left eye. The right eye showed 3+ conjunctival injection, 3+ anterior chamber cells, and ulceration of the cornea. A corneal perforation at the peripheral 6 o’clock position was closed by an incarceration of the iris. The anterior chamber was flattened.

After intensive topical and intravenous antibiotic therapy and insertion of punctum plugs, infection and inflammation slowly resolved. Visual acuity did not improve. The patient was discharged and was again controlled by her ophthalmologist.

Comment

We report a 74 year old woman patient with recurrent bilateral corneal ulceration and unilateral perforation probably associated with keratosis follicularis.

Keratosis follicularis (Darier-White disease) is caused by mutations in the ATP2A2 gene, which encodes the Ca" ATPase 2 isoform. This defect results in disturbed cell adhesion and differentiation of the epithelium.

Histopathological hallmarks of the disease are focal suprabasal clefting due to acantholysis, and subsequent dyskeratotic round epidermal keratinocytes. A historical examination of the eyelids in one patient showed a characteristic epidermal involvement, but no participation of the specific adnexal glands of the lid margin. It was presumed that the knobby, scaling lid margin could provoke symptoms and sequelae similar to seborrhoeic blepharitis. Recurrent localized or widespread cutaneous viral infections and secondary bacterial overgrowth is also common. Bacteria and fungus sometimes cause the keratitis disorders.

In our patient keratosis follicularis probably resulted in dry eye syndrome with reduced corneal protection and microbial colonisation of the skin and ocular surface. Impaired follicular function of the adnexal glands of the cornea epithelium caused the recurrent corneal ulcerations with perforation. Therefore it can be concluded that the increased risk of ocular complications in keratosis follicularis may include severe corneal infection. Beside the long term dermatological medical care an intensive ocular therapy with lubricants and sequelae similar to seborrhoeic blepharitis.
TTT for occult CNV: check the power!

Transpupillary thermotherapy (TTT) was originally introduced for the treatment of small choroidal melanoma. Although the precise mechanism of action for this treatment is unknown yet, the reported data appear to be beneficial compared to the natural course of the disease.

Currently, the following treatment parameters are recommended to treat occult choroidal neovascularisation (CNT) with TTT: one exposure, 60 second exposure time, 800 mW power for the 3 mm spot, 330 mW for the 2 mm spot, and 320 mW for the 1.2 mm spot. We adjusted the parameters applied via the Goldmann fundus lens the power per field is calculated to 247 mW/mm². This usually results in no visible change of the treated area. Thus, if the treated power level would not notice he has to compensate for the lower power by adjusting the instrument's power level.

We have used a diode laser (Iridex Corporation, Mountain View, CA, USA) with the recommended fibretic adapter since 1997 for the treatment of choroidal melanomas and haemangiomas without any apparent problems. Following the report by Reichl and coworkers, we started treating patients with occult CNV according to the recommended treatment parameters described above.

However, we also did not observe satisfactory functional results in the majority of the first consecutive 28 cases treated and in most cases no effect at all could be demonstrated in the fluorescein angiography or OCT.

Lacking any possible explanation other than that the treatment method didn't work, we finally asked a technician to check the laser system immediately and he noticed the fibreoptic had been damaged by its frequent use in treating patients with choroidal tumours. However, this remained unnoticed by the oncologist because in treating choroidal melanomas the end point of an exposure is a visible grey to white lesion. If this is not achieved with the initially chosen power level, the physician usually increases the power in a stepwise manner until this end point is reached.

In summary, we highly recommend to all physicians who are treating occult CNV with subthreshold TTT to check their aiming spot regularly, especially if the system has been in use for a longer time. Additionally, the output power of the entire application system (laser and fibretic optic) should be probably be rechecked at regular intervals depending on the frequency of the instrument's use. This might prevent future reports of the (“false negative”) inefficiency of this possible beneficial treatment method.

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References

Sight Savers International Honoured by American Medical Association Award

Sight Savers International, the UK’s leading charity tackling blindness in the developing world, is the 2002 recipient of the Nathan Davis International Award in Medicine and Public Health, awarded by the American Medical Association. This is the first time ever a non-US organisation has received this prestigious Award.

The panel of judges made a unanimous decision to award the $25,000 prize in recognition of Sight Savers’ support for the provision of vital eye care services in some of the world’s poorest societies around the world over the last 50 years.

Richard Porter, Executive Director of Sight Savers who received the award in Chicago commented: “We are delighted and honoured to receive the Dr Nathan Davis International Award. Our vision is of a world where no-one is needlessly blind and where irreversibly blind people share the same opportunities as everyone. This award recognises the enormous commitment made by many people in poor communities through some of the most simple and cost effective treatments available.

Sight Savers is dedicated to combating avoidable blindness in developing countries. The charity works with partner organisations in poor and under served communities to develop and support healthcare programmes that prevent and cure blindness, and provide services to irreversibly blind people.

Technology for Vision 2000

The latest issue of Community Eye Health (No 42) focuses on technology and training, with an editorial by Catherine Cross, chairperson of the International Agency for the Prevention of Blindness (IAPB). 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 7230 3207; email: eyeresource@ucl.ac.uk).

International Centre for Eye Health

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

Second Sight

Second Sight, a US 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 on the charity website (www.secondsight.org.uk) or by contacting Dr Lucy Mathen (lucymathen@yahoo.com).

Specific Eye Conditions (SPECS)

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

The British Retinitus Pigmentosa Society

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

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).

Cornea 2002—Celebrating 50 Years of Eye Banking
The Cornea 2002 meeting will be held in Le Meridien Hotel, London, Gatwick on 14–15 November 2002. Subjects to be covered will include eye banking, penetrating and lamellar keratoplasty, stem cell restoration, keratoprosthesis, advanced keratoplasty techniques, pediatric cornea, keratorefractive surgery, and intraocular refractive surgery. Spaces are limited and a beneficial package rate is available prior to 30 September 2002. Further details: CORNEA 2002 organiser at the Corneo Plastic Unit, The Queen Victoria Hospital, Holtye Road, East Grinstead, West Sussex, RH19 3DZ, UK (tel: 01342 410 210 ext 560; fax: 01342 317 181; email: Cornea2002@hotmail.com).

Introductory Course in Osteo-odonto-keratoprosthesis (OOKP)
The University of Brighton Postgraduate Medical School is holding an introductory course in osteo-odontokeratoprosthesis (OOKP) 20–21 November 2002 in the New Seminar Room, Sussex House, Brighton & Sussex University Hospitals Trust in Brighton. The course will comprise of a variety of lectures with live surgery, two way audio and video links (Stage 1 and Stage 2 OOKP surgery), and examination of patients. Further details: Mrs Erica Strange, University of Brighton, Postgraduate Medical School, Falmer Campus, Brighton, East Sussex BN1 9PH, UK (tel: +44 (0)1273 644 005; fax: +44 (0)1273 644 002; email: e.strange@brighton.ac.uk).

23rd Annual Conference and Dinner Glaucoma Society (UK & EIRE)
The 23rd Annual Conference and Dinner of the Glaucoma Society will be held on Thursday 21 November 2002, 8.30am to 5.00pm at The Royal College of Surgeons, London. The Annual Dinner is from 6.30pm to 10.00pm at The Royal College of Surgeons, London. Conference charges: £60 members, £80 non-members. Price entitles delegates to refreshments, lunch, abstract book, programme, and annual dinner. (Maximum number of places 250—apply now to secure your place). Further details: Janet Flowers, Administrator, 29 Quarry Hill, Grays, Essex, RM17 3BT, UK (tel/fax: 01375 385172; email: glausoc@ukeire.freeserve.co.uk).
Acanthamoeba keratitis in Ghana

A K Leck, M M Matheson, M Hagan and E Ackuaku

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