Bilateral circumscribed haemangioma of the choroid not associated with systemic vascular syndrome

EDITOR—Circumscribed choroidal haemangioma (CCH) is considered congenital, vascular, relatively rare hamartoma which typically occurs as a localised, monolateral lesion in patients without other vascular malformation. This tumour generally is discovered in adulthood and it is located in the macular area. CCH may be ophthalmoscopically confused with amelanotic melanoma, metastatic tumour, choroidal osteoma, disciform scar, serous detachment, and central serous chorioretinopathy, but may be differentially diagnosed with fluorescein angiography (FA), indocyanine green angiography (ICGA), ultrasoundography, and periodic observation. The bilateral CCH localisation represents an extremely uncommon condition which, in literature, has been only reported in association with Sturge-Weber syndrome or Klippel-Trenaunay-Weber syndrome. To the best of our knowledge, this is the first documented case of bilateral CCHs in the absence of any other evidence of systemic vascular abnormalities.

CASE REPORT
A 81 year old white man was referred to our institution in June 1999 to undergo conservative therapy because of malignant choroidal melanoma of the left eye. He reported a 6 month history of bilateral, progressive reduction of the central vision, greater in his left eye. His best corrected visual acuity was 20/30 in the right eye and 20/40 in the left. Biomicroscopy of the anterior segment did not reveal any notable alterations with the exception of a bilateral nuclear cataract, more evident in the left eye. Intraocular pressure was 18 mm Hg in both eyes. Ophthalmoscopic examination of the left temporal posterior pole showed a lesion, about five optic disc diameters in size and red-orange in colour (Fig 1B), while, in the right macular area, an irregular appearance of the retinal surface was detected (Fig 1A). Bilateral B-scan echography confirmed the presence of a dome-shaped solid lesion, with regular profile and without choroidal cup, in the left eye, revealing a small solid lesion also in the right posterior choroid. Standardised A-scan ultrasonography documented that the maximum thickness of these solid lesions was 1.56 mm in the right eye (Fig 2A) and 3.32 mm in the left (Fig 3A). In the left eye the high and regular internal reflectivity of the lesion was consistent with the presence of a benign tumour, reliably of an angiomatous type. FA did not detail any significant abnormality in the right posterior pole (Fig 2B), showing an irregular fluorescence of the orange-coloured lesion previously described in the left eye (Fig 3B). ICGA confirmed the diagnosis of CCH of the left eye (Fig 3C, D) and documented an early hyperfluorescence, followed by a relative decrease in fluorescence (“washout”), corresponding to the echoraphic findings observed in the right macula (Fig 2C, D). The patient underwent chest x ray, abdominal and chest computed tomographies, total body scintigraphy, liver ultrasonography, blood, and urine analyses.

These investigations did not show any abnormality, reliably excluding the possible metastatic origin of the bilateral choroidal lesions. In the course of a 15 month follow up period, we periodically reassessed this patient, and did not diagnose any ocular or systemic modification.

COMMENT
Atypical CCH can cause differential diagnostic problems by its appearance at the time of presentation. Moreover, bilateral choroidal localisation of tumoral lesions raises the question about their primary or metastatic onset. At our department we observed approximately one haemangioma of the choroid for every 15 malignant melanomas, referred to us yearly for conservative treatment. In spite of this relatively high frequency of haemangioma, this represents the first case in whom we diagnosed a bilateral circumscribed vascular hamartoma, which was not associated with any systemic syndrome. During the mid-term follow up (15 months) there were neither ocular nor systemic significant modifications. The echographic and ICGA features of these choroidal lesions, together with the lack of neoplastic or vascular abnormality in another part of the body, lead us to confirm the first documented diagnosis of bilateral CCHs. Last but not least, our findings demonstrate that FA and echography are not always capable of documenting the specific characteristics of small CCH; thus, when this kind of lesion is
suspected, ICGA represents the most important non-invasive tool for the diagnosis to differentiate amelanotic choroidal melanoma, choroidal metastasis, and choroidal haemangioma.1,2

**Figure 3** (Left eye). (A) Standardised A-scan ultrasonography at 1660 m/s demonstrates the temporal paramacular solid lesion of the choroid, with its high and regular internal reflectivity, consistent with the presence of an angiomatous benign lesion. The maximum thickness of this circumscribed choroidal haemangioma is 3.32 mm. (B) Late phase fluorescein angiogram shows an ill defined hyperfluorescent and hypofluorescent area in correspondence with choroidal haemangioma (arrows). (C) Early indocyanine green photograph reveals a rapid and complete fill up of the lesion. The haemangioma has a “mulberry appearance” at the stage of maximal fluorescence. (D) Late indocyanine green angiogram shows clearing of the dye from the tumour, associated with diffusion into the choroidal and subretinal space (“toashout” phenomenon).

**Trabecular Ciliary Body Detachment, Choroidal Effusion, and Hypotony caused by severe anterior lens capsule contraction following cataract surgery**

**Editor,—**Continuous curvilinear capsulotomy (CCC) first described by Gimbel and Neumann1 has become the procedure of choice for cataract extraction by phacoemulsification. Untoward effects of capsulorhexis have not been frequently noted. Davidson first described capsular contraction syndrome as an exaggerated reduction in anterior capsular diameter, and possibly intracapsular lens (IOL) placement.

Tractional ciliary body detachment and associated hypotony is an uncommon complication of severe anterior lens capsule contraction. Only three such cases have been reported in the literature.1,2 We report a case of transectional ciliary body detachment caused by a severe anterior lens capsule fibrosis, in which Nd:YAG laser anterior capsulotomy was effective in relieving the traction caused by the capsular contraction. We illustrate the value of ultrasound biomicroscopy (UBM) in the diagnosis and management of such conditions.

**CASE REPORT**

A 72 year old woman with primary open angle glaucoma and previous bilateral trabeculectomies (performed twice in the left eye) was followed up in our clinic since December 1999 for an ischaemic central vein occlusion in her right eye. She had a dense cataract in her left eye, which prevented the use of the fundus. The biometry of the left eye showed an axial length of 22.60 mm. Preoperatively intraocular pressures were 15 mm Hg in both eyes. She underwent an uncomplicated phacoemulsification through a superior temporal clear corneal wound. A capsulorhexis of about 5 mm was fashioned. A foldable three piece silicone IOL with poly(methylmethacrylate) (PMMA) haptics (Allergan SH40 NB) was implanted “in the bag.” The lens had an optic diameter of 10.0 mm and a haptic diameter of 13.0 mm. In the immediate postoperative period she was noted to have a well centred IOL “in the bag” and fundus showed an inferior hemiretinal vein occlusion involving the macula in the left eye. At this time she had a visual acuity of counting fingers at 2 metres in her right eye and 6/60 in her left eye.

Two and a half months following her cataract surgery she was referred by an optician with deterioration of vision in her left eye. Visual acuity was counting fingers at 2 metres in both eyes. Slit lamp biomicroscopy of the left eye showed a quiet anterior chamber. Severe contraction of the CCC opening with eccentric displacement of the CCC orifice was noted and the IOL was displaced superiorly (Fig 1, above). Gonioscopy showed an open iridocorneal angle. There was no evidence of any iris changes or changes at the pupillary border, consistent with pseudoexfoliation in either eyes. Goldmann applanation tonometry revealed an intraocular pressure of 5 mm Hg in the left eye and 14 mm Hg in the right. Posterior segment examination revealed severe anterior lens capsule fibrosis. A neodymium: YAG (Nd:YAG) laser anterior capsulotomy was performed. Four relaxing radial anterior capsulotomy cuts were made at 2, 5, 8, and 10 o’clock. The Nd:YAG capsulotomy comprised 50 shots with a power of 1.4 mJ each. During the procedure the anterior capsule was noted to be thick. Immediate widening of the CCC orifice was noted following this procedure (Fig 2, above). The IOL also returned to a well centred position.

Topical prednisolone acetate 1% (Predforte, Allergan, Westport, Ireland) four times a day was prescribed to the left eye. Three days after the anterior capsulotomy, the visual acuity remained at counting fingers at 2 metres in both eyes. The left eye showed a quite deep anterior chamber, well centred IOL and fundus showed resolution of the choroidal effusion, which was confirmed by B-scan ultrasonography. UBM examination showed reattachment of the ciliary body (Fig 2, below) and applanation tonometry showed an intraocular pressure of 14 mm Hg.

COMMENT
Capsulorhexis has become the preferred method of anterior capsulotomy, and untoward effects have not often been noted. Nevertheless, distinct complications of continuous tear capsulotomy are now recognised. This includes capsular bag hyperdistension, shrinkage of the anterior capsule opening with visual loss, and/or IOL decentration and lens shrinkage of the anterior capsule opening with significant capsule shrinkage. Some authors believe that the initial diameter of the CCC is an important factor in its pathogenesis. It is postulated that the more epithelium that is left the greater the potential for capsule contraction. The IOL optic composition may influence the development of anterior capsule fibrosis. Davidson suggested that one piece PMMA IOL with a large optic would help counterbalance the centripetal forces of capsular fibrosis. Werner et al[4] in their histopathological study comparing different IOL styles found that the rate of anterior capsule contraction was relatively high with plate-haptic silicone lenses. The lowest rate was noted with the three piece acrylic optic PMMA haptic IOLs. In their histopathological grading of anterior capsule contraction with IOL materials and designs, silicone optic-PMMA haptic IOL as used in this case was rated third after plate haptic silicone lenses with large holes and small holes.

Anterior capsular shrinkage shifts the relative position of the lens equator, moving it to a more anterior location. This centripetal movement induces an inward pulling force on the zonular apparatus. Depending on the strength of the zonular apparatus, a ciliary body detachment might result. We feel that the smaller capsulorhexis size and the use of silicone IOL predisposed our patient to develop severe anterior lens capsule contraction. Severe anterior lens capsule contraction can exert continuous traction on the ciliary body resulting in a ciliary body detachment. In this case Nd:YAG radial anterior capsulotomy was helpful in relieving the phimosis and thereby removing the tractional force on the ciliary body.

The authors have no proprietary interest in any of the products described in this paper.

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Angle closure in fellow eye with prophylactic plicarcine treatment

Editor,—Prophylactic plicarcine is often used in patients presenting with unilateral primary acute angle closure until definitive treatment with a peripheral iridotomy can be performed.1 We present two cases of unilateral primary acute angle closure glaucoma treated with prophylactic plicarcine that subsequently developed angle closure in the fellow eye within 24 hours of admission.

CASE REPORTS

Case 1
An 81 year old woman was referred from the orthopaedics department with increasing pain and redness in the right eye. Visual acuities were hand movements on the right and 6/24 improving to 6/9 with pinhole on the left. The right cornea was oedematous with intraocular pressures (IOP) of 56 mm Hg in the right and 17 mm Hg in the left. The iridocorneal angle was closed on the right eye, and narrow on gonioscopy (grade 1 inferiorly and closed superiorly) on the left, with bilateral moderate nuckleoscleral cataracts.

She was treated with intravenous Diamox 500 mg, topical levobunolol, 2% pilarcine, and dexamethasone 0.1%. Review 1 hour later showed decreased oedema with IOP of right eye 15 mm Hg and left eye 15 mm Hg. Prophylactic 2% pilarcine four times daily was started in the fellow eye and she was admitted to hospital. On review 8 hours after admission her IOP was 16 mm Hg in the right eye and 46 mm Hg in the left. The left cornea had minimal oedema and closed iridocorneal angle on gonioscopy.

A Nd:YAG laser peripheral iridotomy was performed in the left eye that night that with subsequent resolution of the attack.

Case 2
A 46 year old hypermetropic woman (right eye +2.75DS −0.5 x 160 left eye +4.5DS) with no significant ocular history presented to casualty with intermittent visual disturbance, followed by pain, redness, and decreased vision in the left eye. Visual acuity on presentation was right eye 6/9 and left eye 6/24. The left cornea was hazy with a shallow anterior chamber and IOP of 62 mm Hg. The right iridocorneal angle was narrow but open with pigmented grade 1 angle on gonioscopy. She was admitted and treated with topical apraclonidine, levobunolol, dexamethasone, and intravenous Diamox 500 mg. Pilocarpine 4% every 15 minutes for 1 hour was used in the left eye and a single dose of 4% pilarcine was instilled in the right eye.

On review 2 hours after admission IOP was 45 mm Hg in the right eye and 26 mm Hg in the left. The right cornea remained clear, the anterior chamber appeared shallow, and repeat gonioscopy showed a closed iridocorneal angle on the right. The angle was opened by compression with a Zeiss gonipram and she underwent a Nd:YAG laser peripheral iridotomy initially in the right eye and subsequently in the left eye the following day.

COMMENT

The management of the fellow eye in acute glaucoma is controversial. Although Nd:YAG peripheral iridotomy has established itself as the treatment of choice,2 the use of prophylactic pilarcine until formal iridotomy can occur remains controversial. In a survey of members of the American Glaucoma Society pilarcine was used as the treatment of the fellow eye when iridotomy was deferred by more than half the respondents, whereas close observation was the choice of a third.3 Pilarcine results in miosis thereby pulling the peripheral iris from the anterior chamber angle, relieving pupillary block and increasing aqueous outflow facility. Of more concern is the possibility of a paradoxical effect of pilarcine by a dose dependent shallowing of the anterior chamber, potentially precipitating angle closure in compromised eyes.4 5

The above cases highlight concerns on the use of prophylactic pilarcine (especially in higher concentrations) to the fellow eye. In these cases, prophylactic treatment with pilarcine did not prevent and probably contributed to angle closure.

Early prophylactic peripheral iridotomy with pilarcine treatment may be the treatment of choice.

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Keratolysis in a patient with pemphigus vulgaris

Editor,—Pemphigus vulgaris is an auto-immune, blistering disease of the skin and mucous membranes.1 The characteristic ocular finding is conjunctivitis, and corneal involvement is rare.2 We present a case with pemphigus vulgaris with severe keratolysis that required a corneal transplantation.

CASE REPORT

A 41 year old man had suffered from pemphigus vulgaris for 2 years, and prednisolone 40 mg/day and cyclosporine 300 mg/day had been prescribed. He was admitted to the Hamamatsu University Hospital on 15 March 1999 with an acute exacerbation of the symptoms because of non-compliance with corticosteroid therapy. He returned on 17 March 1999 because of increased discharge and visual loss in both eyes. His visual acuity was 20/20 right eye and 20/20 left eye, and his intraocular pressures was 24 mm Hg right eye and 20 mm Hg left eye. No remarkable findings were observed in both visual fields and optic discs. Slit lamp examination showed mild erosions of his eyelid and cornea. The treatment with prednisolone 40 mg/day and cyclosporine 300 mg/day was continued.

He returned on 9 April 1999 because of acute deterioration of vision in both eyes. His visual acuity was light perception in the right eye and counting fingers in the left. The right conjunctiva showed marked oedema and the anterior chamber was flat. Slit lamp examination showed that the lower two thirds of the right cornea had eroded leaving only Descemet’s membrane and endothelium (Fig 1). The left conjunctiva showed mild oedema and slit lamp examination demonstrated anterior stromal opacities in the lower half of the cornea and bulla-like central corneal epithelial changes. Olofoxin ointment was prescribed for both eyes.

The opacity of the right corneal stroma gradually increased, and scar-like tissue formed in the area of the erosion. Slit lamp examination showed that Descemet’s membrane was touching the iris and lens. The corneal bullous degeneration in the left eye formed an erosion. At the same time, corneal erosion and conjunctival oedema resolved, faint stromal opacities were observed in the region of the corneal lesion in the left eye.

Because of the overall improvement of the cornea of the right eye, an 11 mm right penetrating keratoplasty was performed on 22 September 1999. The dissected lens was extracted and anterior vitrectomy was also performed. His visual acuity on 11 November 1999 was improved to 8/200 with +9.0 − 2.5 × 65° right eye, and 20/25 with −3.0 D −3.75 × 180° D left eye.

COMMENT

Corneal involvement is a rare complication in patients with pemphigus vulgaris. Severe corneal involvement has never been reported except in the case of a 56 year old man with severe ocular involvement including conjunctivitis, corneal ulceration, and perforation despite immunosuppressive therapy.3 Although a causative organism was not isolated, the authors suggested that the complications were due to an infectious agent.

Two mechanisms have been suggested to cause the corneal erosion—bacteria or other pathogenic organisms that infect the cornea because of the epithelial defect and tear film disorder brought on by the corticosteroid and immunosuppressive therapy. Although the culture obtained from right ocular discharge before starting ofloxacin ointment showed a negative result, we could not deny the bacterial infection. We did not perform a bacterial or viral culture or polymerase chain reaction examinations using a corneal sample.

The second mechanism is an autoimmune mechanism against one of the intercellular adhesion molecule—for example, desmoglein (Dsg). The patient was diagnosed as pemphigus vulgaris by histological examination, direct immunofluorescent staining of the skin

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Isolated episcleral plasmacytoma mimicking episcleritis in a patient with benign monoclonal gammopathy

COMMENT
Our case is unique in several respects. The isolated extramacular plasmacytoma of our patient mimicked an episcleritis with mild pain and inflammatory reaction. As it turned out to be resistant to anti-inflammatory therapy a biopsy was performed which finally allowed for the correct diagnosis. Thus solitary plasmacytoma has to be included in the spectrum of ocular masquerade syndrome.1

Another interesting aspect is that our patient developed a monoclonal gammopathy, apparently not related to the isolated episceral plasmacytoma. The latter showed a kappa light chain restriction, whereas in the serum the level of IgA lambda was increased. As a thorough general examination did not reveal any signs of systemic disease or isolated plasmacytoma elsewhere, the monoclonal component was attributed to a monoclonal gammopathy of unknown significance (MGUS) which is considered as a benign or premalignant disorder.

Lymphocytes and plasma cells of the MALT, especially the GALT, are characterised by integrin $\alpha_4\beta_1$, instead of integrin $\alpha_4\beta_2$ which is displayed by plasma cells homing to the bone marrow.4 According to this extramacular plasmacytoma tend to occur more often in the MALT or GALT than in other locations except for the solitary plasmacytoma of the bone. Ninety per cent of the isolated plasmacytomas grow in the head and neck area, especially in the upper respiratory tract, but they are surprisingly rare in the gastrointestinal tract,6 though 80% of all immunoglobulin producing cells of the body are located here.7 The atypical location of the plasmacytoma presented here may be mediated through a specific repertoire of adhesion molecules.8

REFERENCES
molecules. Since antibodies for detection of the above mentioned homing receptors in paraffin sections are not available until now, we were not able to find out whether specific integrins or a total loss of them was responsible for the peculiar location of the tumour in our patient who was only speculated that the isolated plasmacytoma in our case arose from a monoclonal proliferation of plasma cells in an originally inflammatory infiltrate. Whether tissue specific immunoregulatory mechanisms involving accessory cells are also implicated in the localised episcleral tumour growth remain to be elucidated.

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Crystalluria with sulphadiazine

EDITOR,—Toxoplasmosis is the commonest cause of crystalluria with sulphadiazine, and pyrimethamine. Folic acid rescue is added to prevent bone marrow suppression. Steroids are frequently used in combination with antimicrobials to combat threatening inflammatory foci of infection.

We report a case of acute ureteric obstruction in a young female with her first presentation of recurrent ocular toxoplasmosis. We would like to draw the attention of ophthalmologists the risk of crystalluria in patients being treated with sulphadiazine.

CASE REPORT

A 22 year old, otherwise fit woman presented with floaters in the left eye. She had had poor vision since childhood when she had been diagnosed as “amblyopic” and undergone strabismus surgery for esotropia. A pigmented and atrophic scar was present at the left macula, and involving the fovea. At the infero-nasal edge of the scar was a raised creamy area of activity with overlying vitritis. A diagnosis of recurrent toxoplasmosis was made.

Despite the poor visual prognosis of this eye, the symptomatic nature of this lesion and the intensity of the inflammatory response prompted treatment. Pyrimethamine (75 mg immediately then 25 mg twice daily) and sulphadiazine (1 g four times daily) were started immediately then 25 mg twice daily) and sulphadiazine (1 g four times daily) were started followed by administration of intravenous fluids. Ureteroscopy demonstrated crystalluria, and urinary acidification to above a pH of 7.5. This usually achieves prompt dissolution of even large calculi.4 It is not always necessary to stop sulphadiazine.

We refer to a case of acute ureteric obstruction in a young female with her first presentation of recurrent ocular toxoplasmosis. We would like to bring to the attention of ophthalmologists to the risk of crystalluria in patients being treated with sulphadiazine.

COMMENT

Within 24 hours of starting treatment the patient felt unwell, with nausea, anorexia, and oligo-dipsia. She developed pink discoloration of the urine in which she noted sediment, and intense loin pain. Hospitalisation followed. Urinalysis demonstrated a pH of 5.0, urinary blood and protein. An intravenous urogram showed an obstructed right kidney—ureteral retrograde uroscopy demonstrated crystalluria, and insertion of a temporary ureteric stent at this time, with administration of intravenous fluids, effected symptomatic relief. Sulphadiazine was suspended.

We refer to a case of acute ureteric obstruction in a young female with her first presentation of recurrent ocular toxoplasmosis. We would like to bring to the attention of ophthalmologists to the risk of crystalluria in patients being treated with sulphadiazine.

COMMENT

The majority of reports of sulphadiazine crystals or pseudomembrane toxoplasmosis encephalitis.4 These patients usually have various factors predisposing them to the development of crystalluria such as poor fluid intake, fever, diarrhoea, hypalbuminuria, and acidification of the urine. The associated polypharmacy of many AIDS patients may contribute to crystal or stone formation through the latter mechanism, or because of crystallisation of other drugs such as acciocil, triamterene, primate done, or other sulphonamides.

Historically sulphadiazine crystalluria has been reported in non-AIDS patients and may cause renal impairment in 1–4% of HIV negative patients.6 To our knowledge, however, this complication has not been reported in the ophthalmic literature. Ophthalmologists we surveyed were not aware of this potential complication, nor is it documented in the British National Formulary.

Although it occurred quickly in our patient, the complication usually occurs after a median of 10 days in HIV negative subjects at a cumulative sulphadiazine dose of 40 g.6 Microscopy of freshly voided urine commonly shows characteristic “sheaves of wheat” crystalluria and haematuria. Ultrasonography can reveal echogenic foci in the renal parenchyma as well as in the collecting systems, and hydro nephrosis.x Ray examination has a low diagnostic sensitivity.

Treatment Management can be conservative with prompt analgesia, intravenous fluids, plus or minus diuretics, and alkalisation of urine with sodium bicarbonate to above a pH of 7.5. This usually achieves prompt dissolution of even large calculi.6 It is not always necessary to stop sulphadiazine.

Ocular involvement caused by the accumulation of porphyrins in a patient with congenital erythropoietic porphyria

EDITOR.—Congenital erythropoietic porphyria (CEP, MIM No 263700) is an extremely rare disorder inherited as an autosomal recessive trait, which is characterised by an 80–98% reduction in the activity of uroporphyrinogen III synthase (UROS; EC 4.2.1.75).1 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 the non-physiological and pathogenic porphyrin isomers, uroporphyrin I and coproporphyrin I.2 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 manifestations, we reported a scleral change in the patient with CEP,3 who had a remarkable increase of porphyrins in tear drops. Our case report strongly suggests that the accumulation of porphyrins in tear drops may directly cause the scleral changes in the patients with CEP.

CASE REPORT

A 24 year old man presented typical manifestations of CEP such as skin ulcer and scarring. He was diagnosed with CEP in childhood, because of the elevation of porphyrins in urine. At the time of visit, slit lamp examination of bulbar conjunctiva revealed irregular hypertrophy between palpebral fissures in both eyes. A 3 × 4 mm area of scleral necrosis was observed at the limbus in the right eye (Fig 1). Hypertrophy of the temporal limbus and pigmentation of eyelids were also observed, but lid closure was normal. Corneal changes were not observed. Visual activity was right eye: 20/50, left eye: 20/20.

In order to cover the region of scleral necrosis, an amniotic membrane grafting was performed, but postoperative wound healing was slow and the graft failed to be attached. Histological finding with a tissue taken during this operation showed an inflammatory infiltration of neutrophils and plasma cells in connective tissue under conjunctival layer (data not shown).

To confirm whether this scleral necrosis is caused by the direct effect of the accumulation

of porphyrins in tear drops, analysis of tear drop porphyrins was performed after obtaining informed consent. In normal control, no porphyrin isomers were observed, whereas in this patient, remarkable elevations of type I porphyrins and protoporphyrin were observed (Fig 2).

Furthermore, sequence analysis of UROS was performed and an A to G transition of nucleotide 184 that predicted a threonine to alanine substitution at residue 62 (T62A), and a C to T transition of nucleotide 745 that predicted a glutamine to premature stop codon (Q249X). These mutations have been previously reported by Xu et al.

COMMENT
This patient was confirmed to have compound heterozygous mutations, T62A/Q249X. These mutations had been described by Xu et al in a Japanese patient with CEP. They performed in vivo expression study for blood porphyrins and protoporphyrin were observed (data not shown), whereas in a normal sample, no porphyrin isomers were observed (Fig 2). Here we proved the accumulation of porphyrins in tear drops with a single case of CEP. Additional cases are needed to confirm the presence of porphyrins in tear drops although they are asymptomatic for eye involvement. Since our finding demonstrates the likelihood that accumulated porphyrins in tear drops directly exerted a toxic effect in scleral lesions, the protection of sunlight by ultraviolet cut glasses is strongly recommended for prevention against the patients’ own IgG, and is an inherited complication in congenital erythropoietic porphyria.

Bilateral facial nerve palsy associated with p-ANCA positive vasculitis in a patient with rheumatoid arthritis

EDITOR.—Rheumatoid arthritis is a chronic, generalised, symmetrical inflammatory polyarthritis. Extra-articular associations may involve the eyes, heart, lung, skin, and more rarely, the central and peripheral nervous system. We describe a case of bilateral facial paresis associated with a p-ANCA positive vasculitis in a patient with rheumatoid arthritis.

CASE REPORT
A 67 year old woman presented with 2 days of left sided facial weakness. She was known to suffer from rheumatoid arthritis, and displayed the characteristic hand and finger deformities of this condition. Additional features of vitiligo, hypothyroidism, and splenomegaly were present. Her medication consisted of methotrexate 5 mg weekly, thyroxine 100 µg once daily, and folic acid 5 mg once daily. Examination revealed isolated left sided lower motor neuron facial palsy, and a left Bell’s palsy was diagnosed. One week later, she returned with right sided facial weakness. No improvement on the left side had occurred and bilateral lower lid paralytic ectropion was evident. A provisional diagnosis of rheumatoid associated mononeuritis multiplex was made, and a rheumatological consultation was obtained. Haematological investigations revealed a positive rheumatoid factor (RF) and p-ANCA, and a raised plasma viscosity of 1.80. Other autoimmune studies including ANA, anti-Ro and La antibodies, and c-ANCA were negative, and renal function was normal. Chest radiography and magnetic resonance imaging of the brain were unremarkable.

Three pulses of intravenous methylprednisolone 500 mg were given over 3 days, with commencement of oral prednisolone 1 mg/kg. Despite intensive topical lubrication, developing exposure keratopathy necessitated the surgical correction of the bilateral paralytic ectropion. The oral prednisolone was rapidly tapered down to 5 mg/day, and then discontinued after 3 months. p-ANCA levels subsequently became undetectable.

Full orbicularis function gradually recovered, but only partial recovery of the lower facial muscles occurred. Renal function remained normal throughout and there was no significant exacerbation of the polyarthritis.

COMMENT
Facial nerve weakness may be the result of a number of underlying disorders including vasculitis. The development of bilateral signs in rapid succession, in association with rheumatoid arthritis, highlighted a potential vasculitic process in this case. Other causes of bilateral weakness such as pontine disease—for example, demyelination, or primary muscular disorders—for example, myasthenia gravis, and post-infective polyneuropathy were excluded on clinical grounds and after investigation.

Rheumatoid factor consists of IgM antibodies against the patients’ own IgG, and is an important diagnostic feature in rheumatoid arthritis. However, RF may also be seen in...
polyarteritis nodosa, scleroderma, Wegener’s granulomatosis, systemic lupus erythematosus, and sarcoidosis. No clinical or other investiga
tive features of these conditions were demonstrated in the case described here, and the patient displayed typical erosive joint features of rheumatoid arthritis. RF may lead to immune complex (IC) mediated vascular injury due to IC formation and deposition in the joints and vessels causing endothelial damage, perivascular cellular infiltration, and thrombosis formation.

Another mechanism of a vasculitic process is through leucocyte mediated cytotoxicity caused by ANCA. ANCA may promote neutrophil activation and endothelial injury, targeting the neutrophil granule enzymes protease 3 (c-ANCA) and myeloperoxidase (p-ANCA). ANCA are useful diagnostic serological markers in a number of vasculitic conditions such as Wegener's granulomatosis, microscopic polyangiitis, and Churg-Strauss syndrome. They may be found less commonly in rheumatoid arthritis, systemic lupus erythematosi,
inflammatory bowel disease, and autoimmune hepatitis diseases.

In one study, the incidence of p-ANCA in patients with rheumatoid arthritis was 21%, and was strongly associated with nephropathy, more severe disease, and increased inflammation.

In this case, other conditions more commonly associated with positive ANCA titres were excluded on clinical grounds and follow-up investigations. Magnetic resonance imaging is sensitive for cerebral vasculitis, and excluded CNS involvement.

The optimum treatment of ANCA associated vasculitis is generally considered to consist of a combination of corticosteroids and immunosuppressive agents. Additional immunosuppression was not required as widespread evidence of disease activity was absent. Gradual improvement of the facial paresis occurred and vigorous treatment of the exposure keratopathy prevented visual loss in this case.

Bilateral facial nerve palsy is rarely seen in vasculitic conditions. Isolated reports of bilateral facial nerve paralysis associated with Sjogren’s syndrome and polyarteritis nodosa exist.

Rheumatoid arthritis is a common condition, and life threatening complications, although rare, are well recognised. Initial presentation may be to the ophthalmologist and awareness of such situations, will improve the prognosis for these patients.

**COMMENT**

Symptomatic or asymptomatic clinical courses are frequently observed in MRS. As the symptoms rarely appear simultaneously MRS often can be diagnosed only by longitudinal follow up series. Males and females are equally affected. Symptoms usually manifest during adolescence and have rarely been seen in childhood or individuals older than 50 years. The pathogenesis of MRS still remains obscure. Several predisposing factors have been considered such as heredity, infection, allergy, or derangement of cranial autonomic vasomotor innervation. Cranial nerve dysfunction (that is, trigeminal nerve), para sympathetic (flush, pain), and ocular involvement have been associated with MRS. Summarised ocular involvement includes granulomatous blepharitis, exophthalmus with lagophthalmus, and burning sensations, which may be related to exposure keratitis. In rare cases, palpies of the medial rectus muscle, papilloedema, and retrobulbar neuritis have been described.

Conjunctival involvement has not been reported as yet. By conjunctival biopsy taken from our patient we have shown that conjunctival lesions may be present in MRS.
Choroidal detachment following extracapsular cataract extraction in a patient treated with latanoprost

EDITOR,—Adverse reactions associated with the topical administration of the synthetic prostaglandin F₂α analogue latanoprost have been described.¹ We would like to report a case of choroidal detachment following extracapsular cataract extraction in a patient treated with topical latanoprost.

CASE REPORT
A 78 year old man initially presented with primary open angle glaucoma in 1981. This was well controlled on timolol and ophthalmic follow up was uneventful except for the development of left age related maculopathy in 1995 reducing the vision to 6/9. In November 1999 the intraocular pressure (IOP) became uncontrolled and a left sided cataract noted. Latanoprost was substituted with subsequent control of the IOP.

He underwent an uneventful left extracapsular cataract extraction by a traditional, non-phacoemulsification technique at another facility in January 2000 (the operating surgeon did not perform phacoemulsification on any cataract patient). Postoperative drops were betamethason, chloramphenicol, and latanoprost. Immediately postoperatively he experienced nocturnal eye pain and subsequent photophobia. He also noticed a shadow in his left vision. Two weeks postoperatively he still had persistent eye pain and the IOP was recorded as 25 mm Hg. Acetzolamide (orally) and Timolol LA (MSD) were added to the above medications. Three days later examination revealed a visual acuity of 6/24 and IOP 16 mm Hg. Fundoscopy showed the presence of a large temporal choroidal effusion.

An opinion was requested and we first saw the patient 3 days later. Visual acuity was 6/60 at best, and examination revealed corneal folds, a marked anterior uveitis with 3+ cells, and a 360 degree choroidal detachment most marked temporally. The IOP measured 10 mm Hg. The latanoprost, chloramphenicol, and acetazolamide were stopped, the Timolol LA continued and dexamethasone 0.1% 2 hourly and cyclopentolate 1% twice daily commenced. Three days later the choroidal detachment had resolved completely and there were no signs of uveitis. The IOP was 22 mm Hg and the visual acuity had improved to 6/12 at best.

COMMENT
The development of choroidal detachment in a patient with primary open angle glaucoma following cataract extraction has been described.² However, this patient had previously had a trabeculectomy, undergone phacoemulsification, and had severe hypotony postoperatively. In another report choroidal effusion and hypotony were noted in a patient who 8 months before commencing latanoprost had undergone a combined cataract extraction and trabeculectomy. It is suggested that, in our case, the choroidal detachment was present from a short time following surgery in view of the subjective shadow in the patient’s vision. It would appear that the detachment developed and persisted in the presence of an elevated IOP. Withdrawal of the latanoprost led to complete resolution of the choroidal detachment but the IOP remained elevated. Uveal effusion has been noted following phacoemulsification without concurrent use of latanoprost. However, in this study all effusions were small and correlated with the presence of hypotony following surgery.³ Latanoprost would appear to lower IOP by increasing uveal outflow and it has been suggested that the increased outflow facility while on latanoprost may contribute to hypotony and the development of choroidal effusions. Although our patient may have had an episode of hypotony immediately following his surgery, IOP measurements did not suggest this. The possibility of latanoprost initiating or potentiating choroidal detachment in the absence of hypotony following cataract extraction should be considered. This hypothesis is supported by the presence of significant uveitis in this case some time following the surgery.

To our knowledge there have been no studies examining the incidence and severity of uveitis following cataract surgery where latanoprost has been continued. This case emphasises the possibility that systemic reactions can occur in patients undergoing surgery while continuing to use antiglaucoma medications which may potentiate the inflammatory response. Such patients may require more frequent review and should be warned to attend urgently if unexpected symptoms occur in the early postoperative period.

Surgeons who perform cataract surgery on eyes in which the breakdown of the blood-aqueous barrier is expected to be greater than that produced by routine phacoemulsification surgery should consider substituting another IOP lowering agent for latanoprost in the immediate preoperative and postoperative period.

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2 Wu G. Severe hypotony following cataract extraction in a patient on latanoprost. Eye 2000; 14:915–16.

TIT AND CNV

EDITOR,—We thank Ergun and Stur for their interest in our paper and agree with their comments that it is not possible to directly compare a pilot study with a randomised controlled study. We also pointed out that the angiographic follow up data were not complete, as only primary closure was obtained the patients were followed up clinically.

The issue of the laser spot size in transpupillary thermotherapy (TTT) is confusing; however, it is known that more irradiance (W/cm²) is needed for smaller laser spots because heat conduction from choroidal blood flow cools smaller spots more efficiently than larger spots.¹ This physiological phenomenon was established in experiments,¹ theoretical,¹ and clinical¹ studies. Furthermore, it is true that overlapping zones occur when multiple spots are used for very large treatment areas. None the less, these zones experience the same temperature rise as every other treated area and no clinical abnormalities have been noted in the small overlapping zones. Although TTT is mainly used for occult membranes our results indicate that it may have a place in classic
membranes and in this study stabilisation of vision was obtained in the majority of these patients and in a minority an improved vision was noted.

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NOTICES

Affordable eye care
The latest issue of Community Eye Health (37) discusses affordable eye care. For further information please contact Community Eye Health, International Centre for Eye Health, Institute of Ophthalmology, 11–43 Bath Street, London EC1V 9EL. (Tel: (+44) (0) 20-7608 6908; fax: (+44) (0) 7250 3207; email: eyeresource@ucl.ac.uk) Annual subscription £25. Free to workers in developing countries.

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

41st St Andrew’s Day Festival Symposium on Therapeutics
The 41st St Andrew’s Day Festival Symposium on Therapeutics will be held on 6–7 December 2001 at the Royal College of Physicians of Edinburgh. Further details: Ms Eileen Strawn, Symposium Co-ordinator (tel: 0131 225 7324; fax: 0131-220 4393; email: e.strawn@rcpe.ac.uk; website: www.rcpe.ac.uk).

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

International Society for Behçet’s Disease
The International Society for Behçet’s Disease was inaugurated at the 9th International Congress on Behçet’s Disease. Professor Shigeaki Ohno represents the ophthalmology division (Department of Ophthalmology and Visual Sciences, Hokkaido University Graduate School of Medicine, Sapporo, Japan: tel: +81-11-716-1161 (ext 5944); fax: +81-11-736-0952; email: solno@med.hokudai.ac.jp). The 10th International Congress on Behçet’s Disease will be held in Berlin 27–29 June 2002. Further details: Professor Ch Zouboulis (email: zoubbere@zedat.fu-berlin.de).
Conjunctival changes associated with yellow nail syndrome

The yellow nail syndrome (YNS) is a rare clinical entity characterised by slow growing yellow discoloured nails in association with peripheral lymphoedema, pulmonary manifestation (bronchiectasis, pleural effusions), and chronic sinusitis. About 100 cases of YNS have been described in the literature but it has never been associated with ocular manifestations. We report one patient with YNS in whom chronic chemosis and conjunctival degenerative lesion was observed.

Case report

A 61 year old man was referred with a 2 month history of ocular irritation in the left eye. His past medical history was significant for yellowish slow growing nails, chronic maxillary sinusitis, and bronchiectasis for 5 years diagnosed as YNS. On examination, nails of both hands and feet showed yellow discoloration and thickening (Fig 1). He also had oedema over the lower limbs and subcutaneous bronchial infection for 2 months. On ocular examination, corrected visual acuity was 20/20 in both eyes. Slit lamp examination of the left eye revealed an area of conjunctival injection and thickening in the palpebral fissure adjacent to nasal limbus. A mild nasal chemosis and superficial corneal micropannus were also observed (Fig 2 top). Tear secretion was normal. Examination of the anterior and posterior segment in both eyes was unremarkable. Treatment with topical corticosteroid (dexamethasone 0.1%, four times per day) and artificial tears was begun. Two weeks after presentation the patient reported an incomplete resolution of the irritation on the left eye and conjunctival lesion persisted. Computed tomographic scan of the brain and the orbits was normal. A conjunctival biopsy specimen showed multilayered epithelium, nuclear pleomorphism with malpighian differentiation. The underlying connective tissue was normal (Fig 2 bottom). Ocular symptoms improved rapidly and postoperative antibiotic and steroid eye drops were discontinued after 2 weeks. There was no recurrence of the conjunctival lesion but a mild chemosis persist in the left eye and appeared in the right eye.

Comment

YNS was first described by Samman and White in 1964. The syndrome has been defined as the complete triad of slow growing yellow nails, lymphoedema, and pleural effusions but today it is accepted that the presence of two of the three symptoms is sufficient to establish the diagnosis, even though the abnormalities may appear separately with intervals of several years.

The aetiology of YNS is obscure, while pathogenesis seems to involve impaired lymphatic drainage. Ocular involvement has never been described in the YNS. However, conjunctival chemosis observed in our patient could be related to YNS which may involve the pleura, the lungs, but also other serosal or mucosal membranes such as the conjunctiva. Moreover, ocular symptoms and corneal micropannus appeared at the same time of a rise of pulmonary symptoms. An inflammatory component, which could alter blood flow and capillary permeability, has not been excluded and, interestingly, topical dexamethasone used to treat chemosis is efficient.

There is very little information on histopathological findings observed in the YNS. Nail biopsies demonstrated features of maturation disarray in the nail bed epithelium associated with dense, fibrous tissue replacing subungual stroma. The changes that we observed in the conjunctival epithelium are similar to that in the nail bed in YNS.

Since it appears that there is a pathogenic association between the YNS and ocular findings, this association is probably not coincidental and could be the first report of ocular manifestation of YNS.

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References


Biliteral anterior uveitis as a presenting manifestation of sarcoidosis and syphilis

We report an unusual case of bilateral acute anterior uveitis in an asymptomatic patient in which ophthalmic examination and laboratory tests showed the diagnosis of syphilis and sarcoidosis.

Case report

A previously healthy 34 year old Hispanic woman with a 2 week history of blurred vision was referred for evaluation. The patient had no systemic complaints. On examination her best corrected visual acuity was 20/30 in...
The diagnosis of syphilitic uveitis requires a high index of suspicion and the patient's clinical picture should be taken into consideration. The polymerase chain reaction (PCR) assay has been used to detect *Treponema pallidum* in CSF and serum but has not been used in routine diagnosis. In this case, syphilis was presumed to be the cause of the ocular inflammation because there was an associated inflammatory pleocytosis in CSF and this structure is embryologically related to the aqueous humour and, additionally, there was a marked improvement in the anterior segment inflammation with the syphilitic treatment. Moreover, the association of both diseases in this patient could be coincidental; however, it might be possible that the anergy induced by sarcoidosis could help the development of syphilis. We suggest a routine investigation in all cases of unexplained ocular inflammation including chest x-ray and syphilitic serology to screen for sarcoidosis and syphilis, owing to their great mimicry. Other laboratory tests should be performed following a tailored approach.

### References


**Tangent screens are still useful in the assessment of vigabatrin induced visual field defects**

Vigabatrin induced constriction of peripheral visual fields was first reported in 1997. The potential mechanisms of vigabatrin induced peripheral field constriction are many, but in our opinion, not enough attention has been...
The visual field, whether constricted or not should be conical in shape and expand geometrically with increasing distances. Patients with functional visual field constriction can often be detected by the fact that on repeated testing of the visual field at an increased distance from the tangent screen they will not report this change in field diameter in an attempt to be consistent with their first field (tunnel visual field). This is not physiologically possible and is clear evidence of functional visual impairment.

Vigabatrin may well induce visual field constriction as a result of retinal toxicity but until studies are reported using tests of patients on vigabatrin at two viewing distances then this issue will remain open to debate.

**References**


**Successful treatment of squamous cell carcinoma of the lower eyelid with intralesional cidofovir**

Cidofovir (HPMPC), an acyclic nucleoside phosphonate analogue, is a promising drug that acts against a wide number of DNA viruses. In 1997, the US Federal Food and Drug Administration approved cidofovir (for intravenous use only) for the treatment of cytomegalovirus retinitis in patients with AIDS. Over the last few years, cidofovir in a 1–3% gel or cream vehicle has been found to be effective against unmanageable viral cutaneous lesions induced by herpes, pox, and papilloma families. Recent studies have explored intralesional administration of cidofovir for the treatment of HPV related tumours, such as cervical epithelial neoplasia, oesophageal carcinomas, and HSV-8 induced Kaposi’s sarcoma. We report a squamous cell carcinoma (SCC) of the eyelid which was successfully treated with intralesional cidofovir.
SCC was confirmed by the histological examination of a punch biopsy. As the patient refused conventional surgery, after obtaining written consent the lesion was treated with a dose of cidofovir 0.1 ml (7.5 mg of active principle). The drug was injected both intralesionally and perilesionally with a fine needle (26 gauge). Care was taken to avoid intravascular inoculation. Erythema and ulceration were evident after 3 days, then the lesion became progressively smaller and flatter until it disappeared within the month. A skin punch biopsy was performed after 12 months on the previous lesion area, but revealed no presence of neoplastic cells. No systemic side effects were noted and the cosmetic result was excellent (Fig 2). The patient is free from recurrences after a 24 month follow up.

Comment
Surgical excision is the treatment of choice for SCC. Alternatively, liquid nitrogen, electrocautery, radiotherapy, or laser photocoagulation may be used. Decisions regarding treatment depend on the age of the patient, the location, extension, and severity of the neoplasm. For cases in which surgery or alternative cytodestructive techniques are not practicable, local treatment with 5-fluorouracil, nitrogen mustard, bleomycin, mitomycin C, photodynamic therapy, or imiquimod may be considered as an alternative therapeutic option. Recent studies have shown that cidofovir exerts tumoricidal activity towards HPV-related cervical intraepithelial neoplasia, oesophageal and respiratory papillomatous tumours,1,2 or HSV-8 related Kaposi’s sarcoma.3 To our knowledge, the regression of SCC after the intralesional injection of cidofovir has not been previously reported.

The mechanism of cidofovir as an anti-neoplastic agent is unknown. The involution of the neoplastic tissue could be due to the inhibition of rapidly proliferating cells through a decrease in DNA thymidine incorporation, the activation of tumour suppressor genes, the induction of apoptosis, and the inhibition of angiogenesis.4-6

Systemic administration of cidofovir is burdened with serious, dose related side effects. Kidney toxicity is the most common but less frequently uveitis, macular oedema, neuropenia, thrombocytopenia, nausea, fever, hair loss, and muscle pain have also been observed.7 When administered topically or intralesionally cidofovir has not, to date, shown systemic toxicity.

Surgical excision remains the best possible treatment for SCC, as it is usually curative and permits the histopathological evaluation of margins. Herein, this successful outcome with intralesional cidofovir in this case might be worth considering.

References

Multiple idiocrinal cysts in patients with mucopolysaccharidoses

The mucopolysaccharidoses (MPSs) are rare hereditary diseases. They are classified into six hereditary diseases. They are classified into six parts: MPS type 1,2,3,4,5, and 6. MPS 1 is also known as Hurler syndrome, MPS 2 as Hunter syndrome, MPS 3 as Sanfilippo syndrome, MPS 4a as Morquio syndrome, MPS 4b as Maroteaux-Lamy syndrome, and MPS 6 as Hurler-Scheie syndrome. MPSs are characterized by accumulation of glycosaminoglycans, which give rise to the progressive clinical features with involvement of multisystems. Ophthalmic complications, such as corneal stromal opacity, pigmented retinal degeneration, optic atrophy, and glaucoma, are common in patients with MPSs.

Cysts in various organs have been reported in patients with MPSs—for example, multiple demingent cysts, multifocal large cysts in the white matter and arachnoid of the brain, and bone cysts.1,2 In the eye, membrane bound vacuoles in the non-pigmented epithelium of the ciliary processes have been observed by electron microscopy.7 However, iridocrinal cysts have never been reported in patients with MPSs.

We present two cases of multiple iridociliary cysts in two patients with MPSs, one with Scheie syndrome and the other with Maroteaux-Lamy syndrome.

Case reports
Case 1
A 18 year old woman, who was diagnosed with Scheie syndrome (MPS type IS) by enzyme assay. The activity of α-L-iduronidase in peripheral blood lymphocytes was not detectable. She had bilateral corneal stromal opacities, shallow anterior chambers, and high intraocular pressures. On 6 April 1998 her corrected visual acuity was 20/30 in both eyes. Her right intraocular pressure was 24 mm Hg and the left was 20 mm Hg with topical medication. Ultrasound biomicroscopy revealed multiple round cystic lesions with uniformly low echo density similar to anterior chamber fluid in all quadrants of the posterior iris, iridociliary sulcus, and pars plicata of both eyes (Fig 1).

Case 2
A 23 year old woman, who was diagnosed with Maroteaux-Lamy syndrome (MPS type VI). The activity of arylsulfatase B in the peripheral blood lymphocytes was significantly low. At the age of 13 years, she underwent penetrating keratoplasty on her right eye because of corneal stromal opacity. At the age of 23 years, she underwent deep lamellar keratoplasty on her left eye. On 10 September 1997, slit lamp examination disclosed a clear graft and the shallow anterior chamber in both eyes. The correct visual acuity in her right eye was 20/30 and left was 20/40. Her right intraocular pressure was 12 mm Hg and left was 18 mm Hg without medication. Ultrasound biomicroscopy revealed multiple round echo lesions in the posterior iris and ciliary body similar to case 1 in both eyes (Fig 2).

We examined an additional two patients with Scheie syndrome; however, no iridocrinal cysts were found in either patient.

Comment
We have demonstrated the presence of multiple round cystic lesions. From this echoographic finding, we interpret these lesions as cystic lesions in patients with MPSs, since there is a wide gap in the incidence of ciliary body cysts on the posterior ciliary body because of the difficulty in detecting them by conventional methods. Marigo et al.1 retrospectively reported that cystic lesions were identified in 108 eyes of 86 out of 4632 patients by ultrasound biomicroscopy and the incidence of the multiple cysts occupying more than 180° was 13.3%. Kunishimo et al. studied the ciliary body in 232 eyes of 116 healthy people by ultrasound biomicroscopy. They reported that ciliary body cysts were detected in 54.3%, and all the cysts were located at the iridocrinal sulcus or pars plicata. Cysts in our patients were located at the posterior iris as...
MPS has been reported. Multiple iridociliary cysts in a patient without the other hand, angle closure that is caused by rigidity of the cornea in the MPSs patients. On work by the glycosaminoglycan, or a false glaucomas or ocular hypertension. It has been cysts was performed in our cases. Because no evidence of the progression of the organs in MPSs patients support the notion reports.

All of our patients were diagnosed with glaucomas or ocular hypertension. It has been suggested that the high intraocular pressure was due to a blockage of the trabecular meshwork by the glycosaminoglycan, or a false high ocular pressure because of the higher rigidity of the cornea in the MPSs patients. On the other hand, angle closure that is caused by multiple iridociliary cysts in a patient without MPS has been reported.1 So we suggest that angle closure by the cysts may be another cause for the high intraocular pressure in some MPSs cases.

In summary, some of the patients with MPSs with shallow anterior chamber demonstrated the presence of multiple iridociliary cysts and ultrasound biomicroscopy is very useful tool for finding the cysts.

Figure 1 Cross sectional appearance of the iris by ultrasound biomicroscopy in case 1. Multiple iridociliary cysts are seen in the posterior iris of both eyes: (A) 1.2 mm diameter cyst in the right eye; (B) 0.8 mm diameter cyst in the left eye.

Figure 2 Cross sectional view of the iris by ultrasound biomicroscopy in case 2. Multiple iridociliary cysts are seen in the posterior iris of both eyes: (A) 0.8 mm, 0.4 mm, and 0.8 × 1.3 mm diameter cysts in the right eye; (B) 1.1 mm and 1.0 mm diameter cysts in the left eye.

A case of encephalocranio-cutaneous lipomatosis

Encephalocraniocutaneous lipomatosis (ECCL) is a rare neurocutaneous syndrome characterised by cranial and facial asymmetry, cutaneous lesions, central nervous system abnormalities, and ocular abnormalities. A case is described of a young man who presented with limbal dermoids, subcutaneous lipomas, and scalp alopecia. Further investigation revealed cranial and facial asymmetry, intracranial lipomas, and calcification and an arachnoid cyst, supporting a diagnosis of ECCL. This patient also had the additional ocular abnormality of bilateral optic disc colobomas, an association with ECCL not previously reported in the literature.

Case report

A 23 year old Asian man first presented to the eye clinic aged 16 with a left conjunctival lesion. On examination he had bilateral conjunctival dermoliopmas, preauricular lipomas, and bilateral optic disc colobomas. Surgery was not advised at this time and no follow up was arranged, although photographs were taken (Fig 1). Aged 20, he presented to the eye clinic again. Before excision biopsy of the left conjunctival dermopma, a computed tomograph (CT) scan of the head was requested to delineate the posterior extent of the lesion. This revealed some asymmetry of the skull vault, intracranial calcification within the right cerebellar hemisphere, and a possible cystic lesion in the left parietal area (Fig 2 (left)). Subsequent examination by a neurologist was entirely normal with no stigmata of the phacomatoses. Histology of the conjunctival lesion confirmed a complex corneoscleral choristoma comprising collagenous tissue with fat and a focus of cartilage.

Three years later the patient was seen by a dermatologist complaining of a lesion on his left eyebrow, which was clinically a lipoma. Skull and facial x rays revealed asymmetry of the skull vault and facial bones including the zygomatic arches. Further investigation in the form of an magnetic resonance imaging (MRI) scan of the brain showed lipomas within the subcutaneous fat of the scalp and also intracranial lipomas. In addition, there was an arachnoid cyst anterior and inferior to the left temporal lobe (Fig 2 (right)). There was no connection between the soft tissue tumour on the left eyebrow and the intracranial cavity. Subsequent histology of the excised lesion revealed a lipoma.

Review of the patient’s childhood medical records revealed that since birth he had had large patches of scalp alopecia and aged 7 he was noted to have a large suprapubic fat pad. A final diagnosis of ECCL was made based on the findings of limbal dermoids, subcutaneous lipomas, scalp alopecia, cranial and facial asymmetry, intracranial lipomas and calcification, and an arachnoid cyst.

Comment

ECCL is a rare neurocutaneous syndrome of unknown aetiology, first described by Haberland and Perou in 1970. All cases described in the literature have been sporadic and there does not appear to be any geographic, racial,

References


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or sex predilection. The syndrome is characterised by cranial and facial asymmetry, cutaneous lesions, central nervous system abnormalities, ocular abnormalities, and occasionally visceral lipomas. The abnormalities tend to be unilateral, although bilateral involvement has been described.4

Cutaneous lesions consist mainly of subcutaneous scalp lipomas with overlying alopecia but lipomas involving the limbs and paravertebral areas have also been reported.4-6. Popular skin lesions are also common and histologically have been found to be lipomas, fibrolipomas, and angiofibromas.4 Central nervous system abnormalities are numerous and include cerebral lipomas, cerebral calcifications, ventricular dilatation, cerebral atrophy, arachnoid cysts, seizures, spasticity, and mental retardation.4 The most common ocular lesions in ECCL are epibulbar choristomas and small skin nodules around the eyelids, which histologically represent connective tissue naevas.4 Other reported ocular abnormalities include a subcutaneous choristoma of the eyelid,4 a small tag of tissue in the anterior chamber,4 a persistent posterior hyaloid system,4 a dysplastic iris,4 papilloedema,4 and epicanthus inversus plus hypertelorism.4

The patient in this case report demonstrates the main features of ECCL; cranial and facial asymmetry, subcutaneous lipomas and scalp alopecia, intracranial calcification and lipomas, an arachnoid cyst, and limbal dermoids. There was also the additional finding of optic disc cobolomas, an association with ECCL not previously reported in the literature. Other common findings in ECCL are seizures and mental retardation,4 but neither were apparent in this case and may explain the comparatively late presentation of this patient; most patients present in early childhood.4-11. The abnormalities in ECCL are usually unilateral but bilateral involvement does occur4-6 and was evident in this patient. Although epibulbar choristomas and limbal dermoids can occur sporadically in isolation or in a Mendelian inherited pattern4-6 there may be systemic associations such Goldenhar’s syndrome, the linear naevoid sebaceous syndrome, or ECCL and the ophthalmologist should consider these diagnoses when a patient presents with an epibulbar choristoma or limbal dermoid.

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References

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Bilateral non-specific orbital inflammation (orbital “pseudotumour”), posterior scleritis, and anterior uveitis associated with hypothyroidism in a child

Posterior scleritis and non-specific orbital inflammation (NSOI), also known as orbital “pseudotumour,” are rarely seen in children. Paediatric posterior scleritis and NSOI seldom have an underlying systemic association and, to our knowledge, hypothyroidism has not been reported as an association, although thyroid abnormalities are recognised in adults.

We present a case of a child with bilateral anterior NSOI, posterior scleritis, and anterior uveitis who was also found to be hypothyroid.

Case report

A previously fit and well 13 year old girl presented with a 3 week history of bilateral red eyes and painless puffy left upper and lower lids for 1 week. She had had a headache for 10 days which had failed to respond to oral antibiotics. Systemic enquiry revealed a sore throat for 10 days but no other symptoms, in particular no thyroid related symptoms. Her visual acuity was 6/9 right, and 6/18 unaided, improving to 6/12 with pinhole in the left eye. She read all the Ishihara plates with the right eye, but missed three out of 17 plates with the left. No relative afferent pupillary defect (RAPD) was present. She had mild left periorbital swelling with 1+ of cells while the left eye had 2+ of cells in the anterior chamber. There was no evidence of posterior segment involvement, with normal vitreous and fundi.

She was commenced with 2 hourly dexamethasone eye drops to both eyes and on review 3 days later her vision improved to 6/6 in both eyes. A week later, she returned with pain and increasing peribular swelling, left eye greater than right. Her visual acuity was reduced to 6/12 (right eye) and 6/60 improving to 6/18 with a pinhole (left eye). A mild left RAPD was present. Significant non-tender, mildly erythematous peribular swelling was present in the left eye. She had 2 mm left relative aprotinin, with generalised restriction of extraocular movements. There was mild bilateral anterior uveitis. The left disc was more swollen than the right and retinal striae were present in both eyes (Fig 1). There was no evidence of vitreous or choriorretinal inflammation. An orbital computed tomograph (CT) scan suggested a diagnosis of bilateral anterior NSOI without enlargement of the extraocular muscles (Fig 2). A-mode ultrasonography showed bilateral diffuse thickening of the posterior coats of the eyes (maximum left thickness of 4.4 mm) and fluid in Tenon’s capsule consistent with scleritis.

Full blood count, urea, and electrolytes, liver function tests, serum angiotensin converting enzyme, serum calcium, autoantibodies (including ANA, ANCA, and rheumatoid factor), C reactive protein (CRP), and chest x ray were normal. She had negative serology for Borrelia, HTLV-1, and HTLV-2. Her erythrocyte sedimentation rate (ESR) was 32 mm in the first hour and a mildly raised Ig M was found. She was biochemically hypothyroid (raised TSH of 25.5 mU/L (0.4-4.0) and T4 of 10.6 pmol/L (9-20)) with positive anti-thyroid M antibodies. Thyrotoxin releasing hormone stimulation test confirmed primary hypothyroidism.

Treatment was commenced with prednisolone 50 mg which was reduced gradually over 3 months with resolution of her symptoms and signs. She had no relapse at 1 year follow up. She was also treated with thyroxine for her hypothyroidism.

Comment

The distinction between posterior scleritis and diffuse anterior NSOI is not always clear. As in this case, they commonly have evidence of inflammatory changes of the posterior coats of the eye and periorcular tissues.*** In a series of 29 paediatric NSOI cases, there was an association with peripheral blood eosinophilia, raised ESR, and positive ANA. Normal thyroid function tests (TFTs) were present in all of the nine children tested.9 Other reports of paediatric NSOI do not state whether TFTs were performed.9 Similarly, children with posterior scleritis tend not to have any clinical or laboratory evidence of associated systemic disease, but TFTs are not specifically mentioned.8 This is in contrast with our patient who was found to be biochemically hypothyroid. There is some evidence of thyroid autoimmunity in adult patients with NSOI, as shown by Atabay et al who found antibodies to eye muscle membrane antigens and thyroid microsomal antigen in patients with NSOI. Each of their cases was clinically and biochemically euthyroid.8

NSOI and scleritis in children may be associated with iritis, unlike the adult form of this disorder.8 Bloom et al report that children with NSOI and anterior uveitis tend to have a worse prognosis and increased recurrence.4 Our patient responded well to steroid treatment with no relapse at 1 year.

References


Unilateral enlargement of the blind spot: a diagnostic dilemma

Unilateral blind spot enlargement occurs as an isolated entity (acute idiopathic blind spot enlargement) or in association with other conditions such as multiple evanescent white dot syndrome, multifocal choroiditis with panuveitis, or punctate inner choridopathy. It remains controversial whether blind spot enlargement in these conditions serves to unify them as a diagnostic group. The patient presented here had clinical features suggesting “diagnostic overlap” with some of these conditions, suggesting that diagnostic “lumping” of these diseases may have more logic than “splitting” them.

Case report

A 30 year old female patient presented to us in September 2000 with a blind spot close to the centre of vision in her left eye. She was uncertain as to how long it had been present, having noticed it only when the other eye was temporarily covered by chance. She was fit and well, with no recent viral illness or previous eye problems. She had noted difficulty with myopic correction (−3.00 dioptre sphere right, −3.25 dioptre sphere left) was recorded at 6/6 right and left, and discrete foci of chorioretinal scarring were noted above and nasal to the optic disc in the left eye (Fig 1). There was no evidence of vitreous inflammatory activity in...
either eye. Humphrey C24-2 testing revealed an enlarged blind spot on the left (Fig 2), while on the right it was normal. Fluorescein angiography demonstrated window and masking defects consistent with chorioretinal scarring, and late leakage at the optic disc margin.

No treatment was given, and the blind spot has gradually decreased over 12 months.

Comment

Fletcher et al were the first to describe a syndrome of acute idiopathic blind spot enlargement (AIBSE) without optic disc oedema in a series of seven patients. This phenomenon has since become well recognised both as an isolated finding, and in association with various forms of chorioretinitis including multiple evanescent white dot syndrome (MEWDS), multifocal choroiditis with panuveitis (MCP), acute macular neuroretinitis (AMN), diffuse subretinal fibrosis (DSF), and punctate inner choroidopathy (PIC). Together, these diagnoses span a wide spectrum of clinical disease expression, and it remains controversial whether blind spot enlargement serves to unify the group or whether other clinical features are sufficiently distinctive for them to be regarded as separate disorders. This patient is young, myopic and female, and presented with unilateral blind spot enlargement and chorioretinal scarring in the absence of acute symptoms such as photopias. These features make it difficult to assign a specific diagnosis, as discussed below.

AIBSE and MEWDS share many common features. Both tend to present acutely with visual loss and photopias in young myopic females, and follow a prodromal viral illness. They occasionally recur and can both be bilateral. The visual prognosis is good, with early disappearance of the white spots and later resolution of blind spot enlargement in most, though not all, cases. The principal distinguishing feature of MEWDS is the presence of white spots at the level of the outer retina or retinal pigment epithelium (RPE), and the variable presence of vitreous cells, retinal vascular sheathing, and optic disc swelling. Since the white spots can be fleeting and hard to see, it has led some to believe that AIBSE is really a subset of MEWDS patients first seen after resolution of the white spots. However, this has been strongly refuted by Hoyt and Imes, who argued that, in contrast with MEWDS, the peripapillary visual loss in AIBSE is absolute in density and has steep edged margins. The presence of chorioretinal scarring in our patient would not be in keeping with either of these conditions.

In multifocal choroiditis with panuveitis (MCP), patients again present acutely with visual loss, scotomata, and/or photopias, anterior and/or posterior uveitis is present and foci of chorioretinitis become apparent, most commonly in the peripapillary region. Inflammation leads to a variable degree of chorioretinal scarring, which can gradually enlarge and develop a subretinal component. Blind spot enlargement can occur, and does not always correlate with disc swelling or peripapillary chorioretinal scarring. MCP tends to be recurrent, with asymmetric bilaterality. Subfoveal choroidal neovascularisation (CNV) is the commonest cause of permanent vision loss, with resolution of blind spot enlargement occurring in most patients. Presumed ocular histoplasmosis syndrome (POHS) can cause similar chorioretinal scarring, but is not usually associated with uveitis or blind spot enlargement, and has no female preponderance. So-called “pseudo POHS” has been linked with MEWDS, AMN, and AIBSE, though the absence of acute symptoms or vitreous inflammatory activity in our patient is at variance with most previous reports of patients with MCP or POHS.

Punctate inner choroidopathy (PIC) is similar to MCP in many ways, including the presence of an enlarged blind spot in some cases. It is rarer than MCP and no cells or other signs of inflammation are seen in the vitreous or anterior chamber. Our patient would perhaps be closest to PIC in clinical findings, though again the lack of acute symptoms or bilaterality would be atypical.

AMN is less well associated with blind spot enlargement. It occurs predominantly in young adult females, and presents with rapid onset of dense paracentral scotomata. Reddish brown retinal lesions corresponding to the dense scotomata become evident, and are best seen with red free light. Uveitis is not present, and the scotomata diminish over months or years. Diffuse subretinal fibrosis (DSF) is very rare and regarded by some as a variant of MCP. In addition to many of the clinical features of MCP already discussed, this condition is distinguished by widespread and progressive subretinal fibrosis not preceded by CNV.

The blind spot enlargement in AIBSE, MEWDS, MCP, PIC, AMN, and DSF, coupled with their tendency to present in young adult females, has led to a proposal that they be
grouped under the term “acute zonal occult outer retinopathy” (AZOOR). Added to the clinical similarities already described, Jacobsen et al demonstrated electroretinographic (ERG) abnormalities in a group of 24 AZOOR patients, though in some cases there were only subtle intereye differences detected. It was concluded that ERG findings help to unify this diagnostic group, as well as indicating that the primary pathophysiology lies at the level of the photoreceptor outer segment. This view was not supported by Jampol and Wiredu, who argued that the above entities did not fit neatly into any of the diagnoses discussed above, and the principal clinical features of blind spot enlargement with choriotinal scarring in the absence of acute symptoms or evidence of vitritis suggest that there is a degree of diagnostic overlap in her case. To the extent that a single case report can inform this debate, it does indicate that some patients may not fit neatly into diagnostic groups, strengthening the case for those who would “lump” these diagnoses rather than “split” them. Perhaps there are other cases which remain unreported because of this diagnostic uncertainty.

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References

Acquired ocular toxoplasmosis in pregnancy

We describe the management of a case of acquired ocular toxoplasmosis that occurred in the first trimester of pregnancy.

Case report

A 27 year old apparently healthy Pakistani woman, at 9 weeks’ gestation, presented to the Birmingham and Midland Eye Centre with a 1 week history of blurred vision in the left eye. She had no past ocular problems. Her first uncomplicated pregnancy was 2 years previously. On examination, the best corrected visual acuities were 6/6 in each eye. The anterior segments were normal and the intraocular pressures were 16 mm Hg right eye, 14 mm Hg left eye. Funduscopy showed a diffuse, elevated white lesion in the left retina half a disc diameter in size located one disc diameter superior to the fovea (Fig 1). There was an area of associated perivascular sheathing and minimal vitreous activity. An active left retinochoroiditis with associated retinal vasculitis was diagnosed. The right fundus was normal.

Sero logical testing revealed antibodies to Toxoplasma gondii at a titre of 1/4096 (2000 IU/ml) using the dye test and a toxoplasma IgM enzyme linked immunosorbent assay (ELISA) on the patient’s peripheral blood was positive. Subsequent serological tests at the PHS Toxoplasma Reference Unit, Swansea, confirmed these results and IgG avidity testing demonstrated that the infection was acute (Table 1). The polymerase chain reaction (PCR) on peripheral blood for T gondii was negative. At 3 weeks after presentation (12 weeks’ gestation), the patient’s visual acuities were unchanged. There was no occlusion of a small retinal arteriole crossing the lesion. After consultation with the obstetrician the patient decided to proceed with the pregnancy. She was commenced on oral spiramycin 1 g three times daily.

Eight weeks after presentation (17 weeks’ gestation) the vision remained the same and the focus of retinochoroiditis was beginning to scar. The patient underwent an amniocentesis for PCR against T gondii. This was negative suggesting the fetus was unaffected. Spiramycin was continued until the end of pregnancy. Sequential fetal ultrasounds were normal.

At term the patient gave birth to a healthy baby girl. Polymerase chain reaction (PCR) and culture for T gondii were negative on placental tissue. Nevertheless, the dye test on cord blood was positive at 500 IU/ml. As toxoplasma IgM ELISA, IgG, and IgA ISAGA on cord blood were negative, the positive dye test was probably detecting passively transferred maternal IgG. The mother’s vision remained normal, and only a small, pigmented scar was present (Fig 2).

Comment

In the United Kingdom ocular toxoplasmosis is normally thought to occur through the genital route, although recent evidence suggests that we may be underestimating the amount of acquired disease. 

Overall, about 40% of primary maternal infections lead to congenital infection of children, with a transplacental transmission rate of T gondii reported to be 3% in the first trimester, 22% in the second, and 63% in the third.

In our case the features supporting an acquired aetiology included lack retinal scars from previous infection, and the positive IgM serology and IgG avidity results. Sera taken early in infection (<3 months) usually have avidity levels of less than 30%. Most sera taken later in infection (>6 months) have avidity levels of greater than 40%.

There are conflicting reports on the value of the PCR to detect toxoplasma DNA. 

Spiramycin is effective in reducing the risk of transmission to the fetus and therefore was given throughout pregnancy. This treatment may have contributed to the resultant good outcome for both mother and fetus.

Determining an aetiology of acquired T gondii infection was important in this patient as it allowed the most appropriate management plan to be initiated resulting in an excellent outcome. Acquired ocular toxoplasmosis occurring in pregnancy is rare and we hope this case report will raise the awareness of this unusual presentation.

Table 1 Serum dye test titres, IgM levels, and IgG avidity levels

<table>
<thead>
<tr>
<th>Time (days) after onset of symptoms</th>
<th>Dye test titre</th>
<th>Dye test (IU/ml)</th>
<th>ELISA IgM</th>
<th>IgG avidity (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>1/4096</td>
<td>2000</td>
<td>Positive</td>
<td>20</td>
</tr>
<tr>
<td>20</td>
<td>1/2048</td>
<td>1000</td>
<td>Positive</td>
<td>19</td>
</tr>
<tr>
<td>103</td>
<td>1/2048</td>
<td>1000</td>
<td>Positive</td>
<td>--</td>
</tr>
</tbody>
</table>

Figure 1 Left fundus showing active retinochoroiditis with retinal vasculitis.

Figure 2 Left fundus showing healed area of previous retinochoroiditis.
which she had made a full recovery. The infection (chickenpox) 3 weeks earlier, from an uneventful episode of primary varicella zoster.

Case report

We present the case of an 11 year old myopic girl who developed significant refractive changes due to corneal scarring following varicella disciform stromal keratitis. She had had an uneventful episode of primary varicella zoster infection (chickenpox) 3 weeks earlier, from which she had made a full recovery. The general practitioner diagnosed her as having conjunctivitis and she was treated with topical chloramphenicol. One week later, the redness and pain had settled but her vision remained hazy. As a result she was referred to the eye department. Examination confirmed a reduction of visual acuity to 6/24 (Snellen) in the right eye and 6/6 in the left with spectacle correction. She was wearing a correction for myopia with a prescription of −4.75−0.75 × 110° in the right eye and −6.00−0.50 × 90° in the left eye. There was right superficial disciform stromal scar in the central cornea over the visual axis extending towards the periphery at 6 o’clock. Corneal sensation was intact and equal in both eyes. There was no active inflammation with white conjunctiva and quiet anterior chambers. The intraocular pressures were normal. Fundus examination revealed no abnormality. As there was no active inflammation, she was not given any treatment. On review 2 months later, there was no change in her symptoms or in the clinical findings. One year later, she was seen again in the clinic and has a surprising Snellen visual acuity of 6/9−1 unaided, improving to 6/9+2 with −1.75 × 150° in the right eye and 6/6 with −7.00 DS in the left. There was a reduction in opacity of the right corneal scar with stromal thinning (Fig 1). There was no ocular inflammation. Corneal topography showed a flattening of the right corneal surface secondary to disciform stromal scar. Compare this with the unaffected myopic left eye, which has a steeper corneal surface (bottom).

Figure 1. Anterior segment photograph of the right eye showing mild corneal scarring.

Figure 2. Corneal topography of the affected right eye (top) shows a flattening of the corneal surface secondary to disciform stromal corneal scar. Compare this with the unaffected myopic left eye, which has a steeper corneal surface (bottom).

Spontaneous reduction in myopic correction following varicella disciform stromal keratitis

We present the case of an 11 year old myopic girl who developed significant refractive changes due to corneal scarring following varicella disciform stromal keratitis in her right eye. This has markedly reduced the myopia in her right eye and resulted in significant anisometropia.

Case report

This 11 year old girl presented to her general practitioner with a red and painful right eye with reduced vision. She had had an uneventful episode of primary varicella zoster infection (chickenpox) 3 weeks earlier, from which she had made a full recovery. The...
Comment
This is an unusual case of spontaneous myopia correction following corneal scarring secondary to varicella zoster disciform stromal keratitis. Primary varicella zoster infection (chickenpox) is a diffuse vesicular skin rash mainly affecting children and is usually self-limiting. Common ocular findings are eyelid vesicles or marginal erosions and acute conjunctivitis. Corneal changes are infrequent but can occur during the first week or two after the onset of chickenpox. Disceiform stromal keratitis is an unusual but well recognised complication of primary varicella zoster infection.1 Varicella viral antigen and intra-cellular viral inclusions has been found in the corneal epithelium of affected eyes. Wilhelmus et al reported five cases and reviewed the literature and found that this condition is typically unilateral, has a delayed onset, typically several weeks after the onset of skin rash.2 The complications following disceiform stromal keratitis identified in this review are corneal scarring, neurotrophic keratopathy, iridocyclitis with secondary glaucoma, and iris stromal atrophy. The principal cause of loss of vision is corneal scarring. In our case, the corneal scarring had resulted in the flattening of the corneal surface thus reducing the degree of myopia significantly. This was highly unusual and had contributed to improving the unaided visual acuity in her affected eye. The effect on the cornea was similar to post LASIK (laser assisted in situ keratomileusis) and PRK (photorefractive keratectomy) in that the corneal appearance was similar as was the topography. In addition, the degree of cornea haze was limited and there had been a degree of stability for a period of over 1 year. She was unable to tolerate the full refractive corrections because of the significant anisometropia and aniseikonia. Despite this she was pleased as she can see 6/9 unaided and would rather not wear her rather high myopic spectacle correction, in view of her young age the uncertainty of the long term stability of her refractive state and her lack of visual complaint, contact lens and refractive surgery were not considered as appropriate treatment at present. However, they may have possible roles in her future management.

Topical corticosteroid therapy and antiviral agents have a role in the management of herpetic stromal disease following herpes simplex and herpes zoster infection.3 However, their roles in the treatment of stromal keratitis following primary varicella zoster are controversial and have not been determined.4 In our patient, the keratitis settled despite having neither topical corticosteroid nor antiviral agent.

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References

Presumed ocular candidiasis in drug misusers after intravenous use of oral high dose buprenorphine (Subutex)

Heroin drug misusers are a high risk group for disseminated candidiasis.5 Recently, an oral substitute for heroin with oral methadone or high dose sublingual buprenorphine tablets (Subutex) (HDSB) has proved to be effective in management of opioid addiction.6 We report the first four cases of presumed candida endophthalmitis following intravenous injection of HDSB.

Case reports
Case 1
A 22 year old man, HIV negative, former heroin misuser, was seen complaining of blurry vision in his right eye. He used HDSB intravenously after dissolution in saliva. One week following an injection, he presented with a febrile septicemic syndrome associated with scalp nodules. Funduscopy revealed a moderate vitritis and a white tiny perifoveolar lesion with few white snowballs (Fig 1). The bacterial and fungal cultures from both blood and anterior chamber tap were negative. Treatment was begun with intravenous fluconazole associated with three intravitreal amphotericin B injections (IVT). After 15 days of therapy, he was discharged on oral fluconazole. Ten days later, the endophthalmitis relapsed with development of a second paramacular necrotising lesion. A posterior vitrectomy was performed. The vitreous cultures were negative for Candida albicans. He was treated again with intravenous amphotericin B, along with amphotericin B IVT with success.

Case 2
A 27 year old man, a former heroin misuser, was receiving HDSB substitution therapy.

Case 3
A 25 year old man, with history of heroin misuse, was referred for blurry vision and floaters in the right eye. His ocular symptoms started following an intravenous HDSB injection prepared with rotten lemon juice. Funduscopy revealed a parapapillary white lesion and few white snowballs (Fig 2) Treatment was instituted with intravenous amphotericin B and fluconazole but the patient left the hospital against medical advice 4 days later.

Case 4
A 30 year old man, a former heroin misuser, was referred for decreased vision in the left eye. He had been using intravenous HDSB by dissolving the tablets in preserved lemon juice. Ten days earlier, he had a disseminated pustular rash with folliculitis over the chest, shoulders, and back. On funduscopy, there was a 2 + vitritis and a white chorioretinal lesion below the inferotemporal arcade (Fig 4). He improved after a treatment with intravenous amphotericin B rapidly switched to intravenous fluconazole combined with two amphotericin B IVT.

Comment
Endogenous candida endophthalmitis diagnosis is usually based on the combination of

Figure 1 Case 1. Fundus photographs showing tiny white perifoveolar lesion with few white snowballs associated with a mild vitritis on his right eye.

Figure 2 Case 2. Fundus photographs showing yellowish-white macular lesion, retinal vasculitis, and few white snowballs on his right eye.

Figure 3 Case 3. Fundus photographs showing white lesion near the optic nerve and few white vitreous snowballs on his left eye.

Figure 4 Case 4. Fundus photograph showing white chorioretinal lesion below the inferotemporal arcade associated with a marked vitritis.

Occasionally, he injected a preparation of HDSB diluted with preserved lemon juice. Two weeks following such an injection, he developed a skin abscess in which cultures revealed C albicans, posterior cervical lymphadenopathy, nodules of the scalp, and arthritis of the left wrist. Three weeks later he complained of decreased vision in his right eye. Funduscopy revealed a yellowish-white macular lesion and few white snowballs (Fig 3) Improvement was obtained after 14 days of intravenous fluconazole and amphotericin B IVT.

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Aetiology of microbial keratitis in northern Tanzania

The incidence of corneal blindness caused by microbial keratitis in the developing world is far higher than that in the developed world. Microbial keratitis has become a more prominent cause of corneal blindness in east Africa as the uptake of measles immunisation improves, reduces measles keratitis scarring, and with improved recognition and treatment of vitamin A deficiency, reducing its associated xerophthalmia and subsequent corneal scarring.

Clinical cases

Patients referred to, or presenting for the first time to, KCVMC with clinical signs of microbial keratitis, were prospectively recruited to the study, between May 1997 and April 1998.

Table 1 Details of 44 patients presenting with microbial keratitis at KCVMC

<table>
<thead>
<tr>
<th>Characteristics</th>
<th>Range</th>
<th>Median</th>
</tr>
</thead>
<tbody>
<tr>
<td>Age [years]</td>
<td>8–97</td>
<td>44</td>
</tr>
<tr>
<td>Time from onset of symptoms to presentation (days)</td>
<td>1–90</td>
<td>10</td>
</tr>
<tr>
<td>Number</td>
<td>%</td>
<td></td>
</tr>
<tr>
<td>Sex</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Male</td>
<td>29</td>
<td>65.9</td>
</tr>
<tr>
<td>Female</td>
<td>15</td>
<td>34.1</td>
</tr>
<tr>
<td>Occupation</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Agricultural</td>
<td>22</td>
<td>50</td>
</tr>
<tr>
<td>Student/school</td>
<td>6</td>
<td>13.6</td>
</tr>
<tr>
<td>Driver/conductor</td>
<td>4</td>
<td>9.1</td>
</tr>
<tr>
<td>Maasai*</td>
<td>4</td>
<td>9.1</td>
</tr>
<tr>
<td>Retired</td>
<td>4</td>
<td>9.1</td>
</tr>
<tr>
<td>Soldier/guard</td>
<td>1</td>
<td>2.3</td>
</tr>
<tr>
<td>Miner</td>
<td>1</td>
<td>2.3</td>
</tr>
<tr>
<td>Teacher</td>
<td>1</td>
<td>2.3</td>
</tr>
<tr>
<td>Businessman</td>
<td>1</td>
<td>2.3</td>
</tr>
<tr>
<td>Treatment before presentation at KCVMC of all cases (culture negative cases only)</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Nil</td>
<td>11 (6)</td>
<td>25</td>
</tr>
<tr>
<td>Local remedy</td>
<td>4 (2)</td>
<td>9.1</td>
</tr>
<tr>
<td>Inappropriate treatment</td>
<td>3 (1)</td>
<td>6.8</td>
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<td>“Appropriate”† treatment, inadequate intensity</td>
<td>18 (5)</td>
<td>40.9</td>
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<td>4 (2)</td>
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</tr>
<tr>
<td>Unknown</td>
<td>4 (2)</td>
<td>9.1</td>
</tr>
<tr>
<td>History</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Vegetative trauma</td>
<td>14</td>
<td>32</td>
</tr>
<tr>
<td>All trauma</td>
<td>17</td>
<td>38.6</td>
</tr>
<tr>
<td>Previous corneal scar</td>
<td>14</td>
<td>32</td>
</tr>
<tr>
<td>Lid problems</td>
<td>2</td>
<td>4.6</td>
</tr>
<tr>
<td>Nil</td>
<td>11</td>
<td>25</td>
</tr>
<tr>
<td>Organisms cultured</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Fungi</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Fusarium solani</td>
<td>4</td>
<td>9.1</td>
</tr>
<tr>
<td>Fusarium sp</td>
<td>5</td>
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<tr>
<td>Aspergillus fumigatus</td>
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<td>2.3</td>
</tr>
<tr>
<td>Cladosporium sp + Bacillus sp</td>
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<td>2.3</td>
</tr>
<tr>
<td>Unidentified fungus + Staphylococcus</td>
<td>1</td>
<td>2.3</td>
</tr>
<tr>
<td>All fungi</td>
<td>12</td>
<td>27.3</td>
</tr>
<tr>
<td>Gram positive bacteria</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Staphylococcus epidermidis</td>
<td>3</td>
<td>6.8</td>
</tr>
<tr>
<td>Staphylococcus aureus</td>
<td>2</td>
<td>4.6</td>
</tr>
<tr>
<td>Gram negative bacteria</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Pseudomonas aeruginosa</td>
<td>6</td>
<td>13.6</td>
</tr>
<tr>
<td>Proteus sp</td>
<td>1</td>
<td>2.3</td>
</tr>
<tr>
<td>No organism cultured</td>
<td>20</td>
<td>45.5</td>
</tr>
</tbody>
</table>

* Tribal herdsmen.
† Empirical treatment with broad spectrum topical antibiotic.

References


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Patients with corneal ulceration without infiltration were excluded. These consisted of three patients with herpes simplex keratitis, two patients with Mooren’s ulcer, and one patient with a neurotrophic ulcer. Two patients with secondary infection of a recent penetrating injury were excluded. Two children were unable to undergo slit lamp examination and corneal scraping for microbiology specimens and were also excluded from the study.

Forty-four corneal ulcers were seen. Thirty-eight of the patients had visual acuity in the affected eye of 6/60 or worse, and the mean greatest diameter of the infiltrate on examination was 5.1 mm. Organisms were cultured from 24 of the 44 ulcers (54.6%). Fifteen of these cultures were fungal. Larger diameter ulcers were more likely to be culture positive and have poorer outcomes: mean ulcer diameter was 6.0 mm in culture positive ulcers and 3.7 mm in culture negative ulcers. Fungal growth had been predicted by positive microscopy for fungal elements in nine of the 12 cases that grew fungi (75%), and there was never positive fungal microscopy without fungal growth. All fungal isolates had had Gram positive cocci identified on Gram staining initially but, in contrast, there were three cases where Gram positive cocci were initially seen on Gram staining, but cultures had grown other organisms (two fungi, one Gram negative bacilli). Full details are shown in Table 1.

Comment

In this study fungal keratitis accounted for 50% of culture positive cases of microbial keratitis in northern Tanzania, with the majority of these cases (42%) yielding pure fungal isolates on culture. These figures are similar to those published from west Africa, where 56% of microbial keratitis was caused alone in part by fungi. As in studies from the United States, west Africa, and southern India, the most common genus of fungus isolated was that of the filamentous fungus Fusarium.

There may have been a bias towards fungal ulcers in this study. KCMC is a referral centre, receiving severe ulcers from primary care centres, where topical antibacterial treatments are generally available and topical antifungal treatments are generally not. If the culture negative ulcers were predominantly bacterial ulcers, the relative frequency of fungal keratitis in this study would be artificially high. However culture negative ulcers had a similar range of treatments to culture positive ulcers before presenting to KCMC, as can be seen from Table 1. There is no evidence to suggest the culture negative ulcers were predominantly of (treated) bacterial rather than fungal aetiology.

Study design limitations precluded any estimation of prevalence of microbial keratitis in the region, which would have been useful. The findings relate to northern and parts of central Tanzania; with a similar climate, risk factors and primary care set up, the aetiology of microbial keratitis is likely to be similar in the wider region. The study set out Gram positive isolates had had Gram positive cocci identified on Gram staining initially but, in contrast, there were three cases where Gram positive cocci were initially seen on Gram staining, but cultures had grown other organisms (two fungi, one Gram negative bacilli). Full details are shown in Table 1.

The most important factor in outcome was the initial size of the ulcer on examination. Larger ulcers were more likely to be culture positive and to have a poor outcome, such as a blinding corneal scar or requiring evisceration. Culture results were nevertheless useful in ascertaining antibiotic sensitivities, and in adding antifungal treatment where necessary. Any patient undergoing evisceration had usually been an inpatient for several weeks with a painful blind eye, who had tried multiple treatment regimens, and had often requested evisceration himself. The overall rate of evisceration was 25%. Although there is little published material on outcomes of microbial keratitis in the developing world, this study compares with a series of culture positive fungal ulcers in Madurai, India, where 20% were classified as “severe, with little prospect of recovery”.

Clearly, awareness of proper treatment regimens in northern Tanzania is paramount, particularly at the place of first contact, the village health centre, to prevent this overall picture of severe corneal ulcers with very poor visual prognosis. Treatment regimens should also take into account the high proportion of fungal keratitis. Microscopy looking specifically for fungal elements is a simple, quick, and useful test that could help direct initial treatment, along with clinical appearances suggestive of fungal infection. This might be more economically viable and practical than blanket coverage of all keratitis with an antifungal topical treatment in the first line therapy. Unfortunately consistently effective antifungal topical treatments are not widely available in this part of Africa.

Future studies could examine the efficacy of current treatments for fungal keratitis and how best to improve awareness of appropriate treatment regimens in the region.

Acknowledgements

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We are grateful to the hospital director, the staff, and the patients of KCMC for their collaboration in the above study.

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References


This CD video atlas with accompanying text is the first in a series of three such atlases by these authors and is the first such oculoplastic atlas available. The atlas consists of 99 pages of text supplemented with black and white drawings in six chapters. Two CDs contain all the video sequences and these follow the same chapter layout as the text: surgical anatomy of the eyelid, entropion, ectropion, eyelid retraction, ptosis, and eyelid reconstruction.

There are a total of 17 video procedures typically lasting between 3 and 5 minutes and a 10 minute cadaveric anatomy sequence. The video quality is very acceptable for individual viewing but does not project as successfully. All sequences are well narrated and informative, covering basic and some more advanced oculoplastic procedures. The need for free skin grafting in one of the cases demonstrated is perhaps questionable but this does not detract from the educational value of the atlas. A number of associated procedures are covered in the videos—for example, harvesting skin/cartilage; however, these cannot be instantly located from the menus and an additional separate section including such procedures could perhaps have been usefully included.

The text is clearly written and concise but is not comprehensive and provides limited information relating to patient and procedure selection. A good deal of the text is covered in the narration accompanying the video.

The atlas is aimed at all those who have an interest in oculoplastic surgery but is particularly relevant to the ophthalmic surgeon. It will be a valuable asset for teaching residents and fellows and should make interesting viewing for all those routinely practising oculoplastic surgery.

From a personal point of view, I am always fascinated to watch other surgeons at work, as technical approaches to the same problem/operation are often quite diverse. This is one of the best ways to keep up to date and improve your own surgery.

Overall, this atlas will be a valuable asset to all those in training and a useful tool for those practising oculoplastic surgical procedures. I look forward to the next two atlases in the series and to further editions.

Garry Shuttleworth


**International Centre for Eye Health**

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

**Second Sight**

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

**SPECific Eye ConditionS (SPECS)**

SPECs represents over fifty different organisations related to eye disorders ranging from conditions that are relatively common to very rare syndromes. We also include groups who offer support of a more general nature to visually impaired and blind people. Support groups meet regularly in the Boardroom at Moorfields Eye Hospital to offer support to each other, share experiences and explore new ways of working together. The web site www.eyeconditions.org.uk acts as a portal giving direct access to support groups own sites. The SPECS web page is a valuable resource for professionals and may also be of interest to people with a visual impairment or who are blind. For further details about SPECS contact: Kay Parkinson, SPECS Development Officer (tel: +44 (0)1803 524238; email: ksp@eyeconditions.org.uk; www.eyeconditions.org.uk).

**The British Retinitis Pigmentosa Society**

The British Retinitis Pigmentosa Society (BRPS) was formed in 1975 to bring together people with retinitis pigmentosa and their families. The principle aims of BRPS are to raise funds to support the programme of medical research into an eventual cure for this hereditary disease, and through the BRPS welfare service, help members and their families 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; web site: www.brps.demon.co.uk)

**Ophthalmic Anesthesia Society (OAS) 16th Scientific Meeting**

The 16th Scientific Meeting of the OAS will be held on 4–6 October 2002 in The Westin, Michigan Avenue, 909 North Michigan Avenue, Chicago, USA (reservations +1 800 228 3000). Further details: OAS, 793-A Foothill Blvd, PMB 110, San Luis Obispo, CA 93405, USA (tel: +1 805 771 8300; web site: www.eyeanaesthesia.org).

**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 Eyebanking**

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, paediatric 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 (tel: 01342 410 210 ext 560; fax: 01342 317 181; email: cornea2002@hotmail.com).

**CORRECTION**

An error occurred in the article: Bilateral circumscribed haemangiomata of the choroid not associated with systemic vascular syndrome. *Br J Ophthalmol* 2001;85:1260. The authors should have been listed as P Perri, C Incorvai, C Costagliola, F Parmeggiani, G Lamberti, B Paduano, A Sebastiani.