LETTERS TO
THE EDITOR

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 choroiditis, but may be differentially diagnosed with fluorescein angiography (FA), indocyanine green angiography (ICGA), ultrasonography, and periodic observation.1 The bilateral CCH localisation represents an extremely uncommon condition which, in literature, has been only reported in association with Sturge-Weber syndrome2–6 or Klippel-Trenaunay-Weber syndrome.7 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 1A), 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 angiomatosus 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 echographic 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.1 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 echographic1 and ICGA1–10 features of these choroidal lesions, together with the lack of neoplasm 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 of choroidal metastasis, and choroidal haemangioma.

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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 fundus examination. 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 superonasal limbal wound. A capsulorhexis of about 5 mm was fashioned. A foldable three piece silicone IOL with poly(methylmethacrylate) (PMMA) haptics (Allergan SH4 NB) was implanted “in the bag.” The lens had an optic diameter of 7.0 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 irido-corneal 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 an equatorial capsular diameter, and possibly associated hypotony is an uncommon complication of severe anterior lens capsular contraction.

Tractional 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 Gimble and Neuhann 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 capsulotomy and capsular bag diameter after cataract surgery. This specific clinical entity of “capsular contraction syndrome” is usually associated with a reduction in the capsular opening, malposition of the opening, reduction in the equatorial capsular diameter, and possibly intraocular lens (IOL) displacement.

Tractional ciliary body detachment and associated hypotony is an uncommon complication of severe anterior lens capsular contraction. Only three such cases have been reported in the literature.1 2 We report a case of tractional 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 ultrasonic biomicroscopy (UBM) in the diagnosis and management of such conditions.

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 angiomatic benign lesion. The maximum thickness of this circumscribed choroidal haemangioma is 3.32 mm. (B) Late phase fluorescein angiogram shows an ill defined hypofluorescent 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 (“washout” phenomenon).

In general, shrinkage of the anterior capsule according to Davidson is produced by an imbalance between the centrifugal and centripetal forces on the capsular bag. Although the pathogenic mechanism responsible for excessive capsule fibrosis and contracture are not well understood, several histopathological studies have identified the cell types associated with pseudophakic fibrosis.  Frezotti et al. attributed constriction of the anterior capsule opening to fibrogenic transformation of the subcapsular and equatorial lens epithelial cells (LECs). Nishi and Nishi suggested that this fibrosis might be induced by interleukin 1 or 6 and other cytokines synthesised by residual LECs, which in turn affect the epithelial cells in an autocrine manner.

The following three main factors may account for anterior capsule contraction: (1) IOL material, (2) IOL design, and (3) CCC opening. The sphincter effect of an intact capsulorhexis seems to be important in creating 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. 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 counteracting force 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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Figure 1 (Above) Anterior segment photograph showing severe anterior capsular contraction with superior displacement of the IOL (arrow). Note the partly visible tridectomy (arrowhead). (Below) Ultrasound biomicroscopy showing ciliary body detachment with central rotation of the ciliary body (arrow).

Figure 2 (Above) Anterior segment photograph showing the radial capsulotomies and the widened anterior capsule. (Below) Post laser, ultrasound biomicroscopy showing reattached ciliary body.
Angle closure in fellow eye with prophylactic pilocarpine treatment

EDITOR,—Prophylactic pilocarpine is often used in patients presenting with unilateral primary acute angle closure until definitive treatment with either peripheral iridotomy can be performed.1

We present two cases of unilateral primary acute angle closure glaucoma treated with prophylactic pilocarpine 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 ophthalmologists in 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 nuclear cataracts.

She was treated with intravenous Diamox 500 mg, topical levobunolol, 2% pilocarpine, and dexamethasone 0.1%. Review 1 hour later showed decreased oedema with IOP of right eye 45 mm Hg and left eye 15 mm Hg. Prophylactic 2% pilocarpine 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 with subsequent resolution of the attack.

Case 2

A 46 year old hypermetropic woman (right eye +2.75DS−0.5 × 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% pilocarpine 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 goniprism, 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 pilocarpine until formal iridotomy can occur remains controversial. In a survey of members of the American Glaucoma Society pilocarpine 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 Pilocarpine 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 pilocarpine by a dose-dependent shallowening of the anterior chamber, potentially precipitating angle closure in compromised eyes.4,5

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

Early prophylactic peripheral iridotomy without pilocarpine treatment may be the treatment of choice.

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Keratolysis in a patient with pemphigus vulgaris

EDITOR,—Pemphigus vulgaris is an autoimmune 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 ciclosporine 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 pressure 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 ciclosporine 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. Ofloxacin 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. A dislocated lens, 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 dislocated 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° 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...
showing intracellular deposition of immunoglobulin G and high titer of circulating anti-Dsg 3 and anti-Dsg 1 antibodies (Fig 2). Because the cornea usually does not have Dsg 3 but Dsg 2, an autoimmune mechanism cannot be considered. However, prolonged epithelial defect by limbal damage may have resulted in the corneal erosion because of the expression of Dsg 3 by the epithelium of the corneal limbus. Although no infection was observed in both corneas, the association with an infectious mechanism may be involved in the pathogenesis of corneal erosion in our case.

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

EDITOR—We present the unique case of a patient with an isolated plasmacytoma of the episclera mimicking a painful episcleritis. Plasmacytomas usually grow in the bone marrow are rare. They mostly involve the oropharynx and the upper respiratory tract, but have also been encountered in the lids, the orbit, and the palpebral conjunctiva.1 Only one case of a solitary epibulbar plasmacytoma with intraocular invasion has been reported yet.2

CASE REPORT

A 61 year old patient presented with an “inflammatory” episcleral nodule within the lower temporal quadrant and mild pain in his left eye (Fig 1), which had already lasted 5 months and had been diagnosed as episcleritis. There was no evidence of rheumatic disease; ANA and ANCA were negative. Neither dexamethasone eyedrops nor oral fluorocortolone (60 mg) were helpful, thus an excisional biopsy was performed. The tumour seemed to be attached only to Tenon’s capsule and could easily be removed.

Figure 2 Monomorphous infiltrate of plasma cells stained positively with the plasma cell marker CD20, IgA, IgD, and IgG (right). The proliferation rate was increased with an MIB1 positivity of 5%. Immunohistochemistry for CD20, IgA, IgD, and IgG was negative.

Three months later an IgA lambda monoclonal gammonophathy with an IgA level of 5.6 g/l (normal 0.7–4.0 g/l) was found, but neither bone marrow biopsy nor bone scan showed any abnormalities. A local recurrence of the episcleral tumour with infiltration of the lateral rectus muscle 6 months after the initial diagnosis, was irradiated with 46 Gy over 2 months. The tumour resolved completely and did not recur. The IgA level of the serum ranged between 5.2 and 6.7 g/l over a period of almost 3 years.

COMMENT

Our case is unique in several respects. The isolated extramedullary 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 syndromes.

Another interesting aspect is that our patient developed a monoclonal gammopathy, apparently not related to the isolated episcleral 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 αβ2, instead of integrin αβ1 which is displayed by plasma cells homing to the bone marrow.1 According to this extramedullary 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, though 80% of all immunoglobuline 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 receptors that are specific for certain locations.8

Figure 1 Conjunctival and episcleral injection occurred in both eyes (left). The direct immunofluorescent staining of the skin showed intercellular deposition of immunoglobulin G (right).

Figure 2 Histological examination by haematoxylin and eosin staining of lesional skin disclosed intraepidermal clefts which contained several acantholytic cells (left). The direct immunofluorescent staining of the skin showed intercellular deposition of immunoglobulin G (right).
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. We can only speculate 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 epithelial tumour growth remain to be elucidated.

We thank Mrs Renate Buchen for technical assistance, Dr Sarah Coulband for providing the VS38 immunostaining, and Dr Fleming Staubach for reading the manuscript.

We report 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 the risk of crystalluria in patients being treated with sulphanilamide.

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 inferonasal 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 sulphanilamide (1 g four times daily) were added after 48 hours of starting treatment the patient felt unwell, with nausea, anorexia, and oligidipsia. 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 suggested an obstruction at the right vesico-ureteric junction. Radiographic retrograde ureteroscopy demonstrated crystalluria, and insertion of a temporary ureteric stent at this time, with administration of intravenous fluids, effected symptomatic relief. Sulphadiazine was suspended.

Within 24 hours of starting treatment the patient felt well, with urine, anemia, and oligidipsia. 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 suggested an obstruction at the right vesico-ureteric junction. Radiographic retrograde ureteroscopy demonstrated crystalluria, and insertion of a temporary ureteric stent at this time, with administration of intravenous fluids, effected symptomatic relief. Sulphadiazine was suspended.

COMMENT
The majority of reports of sulphanilamide crystalluria occur in patients with AIDS under treatment for toxoplasmosis encephalitis. These patients show various factors predisposing them to the development of crystalluria such as poor fluid intake, fever, diarrhoea, hypalbuminaemia, and acidification of the urine. The associated polypharmacy of many AIDS patients (particularly those who use crystal-morphine or stone formation through the latter mechanism, or because of crystallisation of other drugs such as aciclovir, triamterene, prine- done, or other sulphonamides. Historically, sulphanilamide crystalluria has been reported in non-AIDS patients and may cause renal impairment in 1–4% of HIV negative patients. 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 sulphanilamide dose of 40 g.

Microscopy of freshly voided urine commonly shows characteristic “sheaves of wheat” crystalluria and haematuria. Ultrasonography can reveal echogenic foci in the renal paren- chyma as well as in the collecting systems, and hydroprophias. x Ray examination has a low diagnostic sensitivity.

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

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

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 reces- sive trait, which is characterised by an 80–98% reduction in the activity of uroporphyrinogen III synthase (UROS; EC 4.2.1.75). Clin- ically, CEP is characterised by severe cutane- ous photosensitivity, chronic haemolysis, and massive porphyrinuria resulting from the accumulation in the bone marrow, peripheral blood, and other organs of the porphyrins in the non-physiological and pathogenic porphyrin isomers, uroporphyrin I and coprotoporphyrin I. 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, 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-shaped, atrophic area of conjunctiva. The conjunctival vessels were dilated. A 4 mm area of scleral necrosis was observed at the limbus in the right eye (Fig. 1). Hypertrophy of the temporal limbis and pigmentation of eyelids were also observed, but lid closure was normal. Corneal changes were not observed. Visual acuity was right eye: 20/50, left eye: 20/20.

In order to cover the region of scleral necrosis, 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

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Figure 1  Four direction images of right eye. Irregular hypertrophy at the temporal limbus and scleral necrosis at the limbus were observed.

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 (210:183–5).

Furthermore, sequence analysis of UROS was performed and an A to G transition of nucleotide 184 that predicted a glutamine to premature stop codon (Q249X). These mutations had been previously reported by Xu et al.6

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 Q249X. These mutations had been described by Xu et al in a Japanese patient with CEP.


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

COMMENT
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 paresis, 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 mononeuropathy multiplex was made, and a rheumatology 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 facial weakness such as pontine disease—for example, demyelination, or primary muscular disorders—for example, myasthenia gravis, and post-infective polyneuropathy were excluded, but only partial recovery of the lower facial muscles occurred. Renal function remained normal throughout and there was no significant exacerbation of the polyarthritis.

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polyarteritis nodosa, scleroderma, Wegener’s granulomatosis, systemic lupus erythematosus, and sarcoidosis. No clinical or other investiga-
tive features of these conditions were demo-
strated in the case described here, and the patient displayed typical erosive joint features
of rheumatoid arthritis. RF may lead to immune complex (IC) mediated vasculitis due to
IC formation and deposition in the joints
and vessels causing endothelial damage, perivascular cellular infiltration, and throm-
bus formation.

Another mechanism of a vasculitic process is through leucocyte mediated cytotoxicity
caused by ANCA. ANCA may promote neutrophil activation and endothelial injury,
by targeting the neutrophil granule enzymes protease 3 (c-ANCA) and myeloperoxidase
(p-ANCA). ANCA are useful diagnostic sero-
logical markers in a number of vasculitic con-
ditions such as Wegener’s granulomatosis,
microscopic polyangiitis, and Churg-Strauss
syndrome. They may be found less commonly in
rheumatoid arthritis, systemic lupus ery-
thematosus, inflammatory bowel disease, and
autoimmune haematopoietic diseases. In
one study, the incidence of p-ANCA in patients
with rheumatoid arthritis was 21%, and was
strongly associated with nephropathy, mental severe disease, and increased
inflammation.

In this case, other conditions more com-
onnally associated with positive ANCA titres
were excluded on clinical grounds and follow-
ing investigation. Magnetic resonance imaging is
sensitive for cerebral vasculitis,1 and ex-
cluded CNS involvement.

The optimum treatment of ANCA associ-
ated vasculitis is generally considered to consist of a combination of corticosteroids and
immunosuppressive agents. Azathioprine,
trebrox, cyclosporin, azathioprine, or cyclo-
phosphamide may be used although the most
effective treatment protocols are yet to be
determined. Evidence of renal or CNS
involvement should prompt aggressive therapy because of potentially life threatening
complications. In this case, therapy consisted of
pulsed intravenous methylprednisolone in the
initial phase, followed by oral pred-


nisolone. Additional immunosuppression was
not required as widespread evidence of disease
activity was absent. Gradual improvement of
the facial paresis occurred and vigorous treat-
ment of the exposure keratopathy prevented
visual loss in this case.

Bilateral facial nerve palsy is rarely seen in
vascular conditions. Isolated reports of bilat-
eral facial nerve paralysis associated with
Sjögren’s syndrome2 and polyarteritis nodosa3
exist.

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

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Bilateral conjunctival lesions in
Melkersson-Rosenthal syndrome

EDITOR,—The Melkersson syndrome is a rare
granulomatous disease of unknown origin. The
typical clinical picture consists of recur-
rent facial oedema associated with peripheral
facial palsy and was first described by
Melkersson in 1928. Three years later a
fissured tongue called lingua plicata was
added to the classic features by Rosenthal.

The classical clinical picture includes
granulomatous cheilitis, facial palsy, and
fissured tongue was first called Melkersson-Rosenthal syn-
drome (MRS) by Lüscher in 1949.1

We report on a patient with the typical clini-
cal signs who had chronic bilateral conjunc-
tival lesions. To our knowledge an association of
conjunctival lesions with MRS has not been
described previously.

CASE REPORT

A 64 year old man presented with a fissured
tongue, recurrent painless facial oedema,
 especially of the eyelids, and facial flush for 6
years. A review of the other systemic diseases
was unremarkable. He required blepharo-
plasty for the facial malformation.

The histopathological findings of the skin
biopsy confirmed the clinical suspicion of
MRS by the typical granulomatous infiltration
(Fig 1A).

Furthermore, he complained bilateral con-
 junctival swelling had been present for 6
months. Visual acuity was 20/30. Slit lamp
examination revealed a bilateral fleshy mass
extending from the upper fornices to the lim-
bus and conjunctival fornices (Fig 1B).

Motility of the eyeball was normal and an
exophthalmus has not been present. In the
magnetic resonance image (MRI) of the orbit
a bilateral enlargement of lacrimal glands
and a swelling of the lateral rectus and superior
rectus muscle of the right eye were observed.

Staging examinations for lymphoma or other
malignancies were uneventful. Since differen-
tial diagnosis included a bilateral orbital
lymphoma a conjunctival biopsy was per-
formed. The histopathological examination of
the conjunctival specimen by light microscopy
revealed a subepithelial process. An infiltrate
of small lymphocytes without any differentia-
tion and with septate orientation was found

Figure 1 (A) Histopathological examination of
skin biopsy with epitheloid infiltration
(haematoxylin and eosin, original magnification ×400); (B) Biomicroscopic view of right eye shows a
fleshy mass in the superior fornix; (C) Histopathological examination of conjunctival
mucosa shows an infiltration of small lymphocytic
cells with septate orientation (haematoxylin and
eosin, original magnification ×200).

(From 1C). By immunohistochemical examina-
tion no monoclonal pattern was determined,
therefore, excluding a lymphoma.

Consequently, the patient was treated with
clobazam 100 mg daily. No progression in
clinical findings was seen afterwards while he
was on a maintenance dosage of the drug.

COMMENT

Disymptomatic or monosymptomatic clinical
courses are frequently observed in MRS. As
the symptoms rarely appear simultaneously
MRS often can be diagnosed only by longitudi-
nal follow up series.1 Males and females are
equally affected.2 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.1 Cranial nerve dys-
function (that is, trigeminal nerve), para-
sympathetic (flush, pain), and ocular involve-
ment have been associated with MRS. Summa-
rised ocular involvement includes granulomatous blepharitis, exophthalmus with
lagophthalmus, and burning sensations,
which may be related to exposure keratitis. In
rare cases, palsies of the medial rectus muscle,
papillodema, and retrobulbar neuritis have
described.1

Conjunctival involvement has not been
reported as yet. By conjunctival biopsy taken
from our patient we have shown that conjunc-
tival lesions may be present in MRS.
A satisfactory conservative therapy has not been established so far. The results of various modes of symptomatic treatment, including systemic or topical glucocorticosteroids, are questionable. They may reduce the patients’ complaints, at least temporarily. Another treatment consists of clofazimine, which is an oral phenazine. This drug had been useful in other conditions with granulomatous inflammation. Finally, a surgical excision of the masses had been suggested for granulomatous chelitis or blepharitis, in order to improve the motility of the eyelid when exophthalmus occurred. Conjunctival biopsy with histopathological and immunohistochemical examination may be helpful to differentiate it from other lymphoid lesions.

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1 Melkersson E. Case of recurrent facial paralysis with angioneurotic edema. Hygiea 1928;90:717–21.


7 Sussman GL, Yang WH, Steinberg S. Miescher’s cheilitis or blepharitis, in order to improve the motility of the eyelid when exophthalmus occurred. Conjunctival biopsy with histopathological and immunohistochemical examination may be helpful to differentiate it from other lymphoid lesions.

COMMENT

The development of choroidal detachment in a patient with primary open angle glaucoma following cataract extraction has been described. However, this patient had previously had a trabecelecotomy, undergone phacoemulsification and had severe hypotony postoperatively. In another report, choroidal effusion and hypotony were noted in a patient who 8 weeks before commencing latanoprost had undergone a combined cataract extraction and trabeculectomy. It is possible 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. 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 must therefore be considered. This hypothesis is supported by the presence of significant uveitis in this case some time following 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 idiosyncratic 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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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).

22nd Annual Meeting of the Glaucoma Society (UK & Eire)
The 22nd Annual Meeting of the Glaucoma Society (UK & Eire) will take place on 22 November 2001 at the Central Conference Centre, 90 Central Street, London EC1V 8AQ.

The Allergan Guest Lecture will be delivered by Professor Jost Jonas of the University of Erlangen, Germany on the subject of the optic disc.

Further details: Mrs Janet Flowers, Administrator, 29 Quarry Hill, Grays, Essex, RM17 5BT (tel/fax: 01375 383172; email: glaucomasocukeire@talk21.com; website: www.iga.org.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 Coordinator (tel: 0131 225 7324; fax: 0131-220 4393; email: e.strawn@rcpe.ac.uk; website: www.rcpe.ac.uk).

4th International Conference on the Adjuvant Therapy of Malignant Melanoma
The 4th International Conference on the adjuvant therapy of malignant melanoma will be held at The Royal College of Physicians, London on 15–16 March 2002. Further details: Conference Secretariat, CCI Ltd, 2 Palmerston Court, Palmerston Way, London SW8 4AJ, UK (tel: + 44 (0) 20 7720 0600; fax: + 44 (0) 20 7720 7177; email: melanoma@confcomm.co.uk; website: www.confcomm.co.uk/Melanoma).

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: ophthalm@icmsaustralia.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: solho@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 chronic nasal 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 subacute 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 thickening of the nail plates 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 where 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.

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


Bilateral 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 syphils 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
treated with 12 million units of aqueous penicillin G intravenously daily for 14 days with resolution of the anterior segment inflammation. A chest computed tomograph (CT) confirmed the findings of the chest x-ray (Fig 2B). A transbronchial lung biopsy showed non-caseating granulomas (Fig 2C), in which no micro-organisms could be found with Ziehl-Nielsen, PAS, Grocott’s silver, or Wharthin-Starry stains and the diagnosis of sarcoidosis, stage 2 disease, was made. Pulmonary function tests were normal. No treatment was indicated for sarcoidosis because the patient was asymptomatic. The radiological lesions remained unchanged on the control performed after 6 months.

Comment
During the past decade, there has been a significant resurgence of syphilis, especially among black and Hispanic patients, and an increased number of patients with ocular syphilis has been reported. Syphilitic uveitis has no specific pattern of ocular involvement and, currently, it has been accepted that practically all patients with uveitis should be tested for syphilis. Sarcoïdosis is another condition that can imitate any form of ocular inflammation. We are unaware of any previous reports of the association of sarcoidosis and syphilis in an asymptomatic patient with uveitis, and could find no reference to it in a computerised search using Medline.

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.

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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
paid to the method and appropriateness of recording this constriction. Current recommendations for patients prescribed vigabatrin are that they are screened at regular intervals by automated perimetry. Automated perimetry cannot, however, differentiate between pathological and functional (non-physiological) constriction of the visual field. Furthermore, automated perimetry, although deceptively simple for the operator to perform, is notoriously laborious and fatiguing for the patient. Although several safeguards are built into automated perimetry, in the form of reliability indices, there are traps for the unwary. This is clearly demonstrated in the recent case report where a 10 year old girl’s visual field constriction apparently reversed on cessation of vigabatrin. Baseline visual fields, performed with automated perimetry, showed a classic artefact cloverleaf-shaped pattern that was not recognised by the authors. Automated threshold perimetry involves checking the visual threshold of the retina at set intervals. To reduce the number of presented stimuli starting points for threshold determinations are made at four quadrants 9 degrees from the horizontal and vertical meridians. Not infrequently, poorly cooperative patients are only attentive during this initial stage resulting in a cloverleaf-shaped field. In this situation the reliability indices are of little help as the suprathreshold false negative reliability indices are based on already fatigued thresholded locations.

**Case report**

A 30 year old woman was referred to the neuro-ophthalmology clinic in October 1998 for confirmation of vigabatrin induced constriction of visual fields. Her seizures had started at the age of 12 and consisted of sudden tonic posturing of the limbs preceded by left sided sensory symptoms. Initially she was having 12 seizures a week but by 1998 she was having 18 seizures a day. She was unable to tolerate phenytoin, valproate, carbamazepine, clonazepam, lamotrigine, gabapentin, or topiramate and in 1990 had been started on vigabatrin. In 1998 she was referred for consideration of epilepsy surgery. At that time she complained of bumping into objects and she was noted on simple confrontation testing to have constricted visual fields. Automated perimetry was recommended and this was subsequently performed (Fig 1). Gross peripheral field constriction was noted but tangent screen examination at 1 and 2 metres revealed this to be non-physiological tubular visual field constriction (Fig 2).

**Comment**

The best way to ascertain whether visual constriction is pathological or not is to test the patient at 1 and 2 metres using a wall mounted tangent screen. 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.

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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.

**Case report**

A 70 year old man presented with a nodular lesion 10 × 14 mm in size on his right lower eyelid, which had appeared 3 months earlier. The patient, who was otherwise in good general condition, reported a history of chronic, intense solar radiation exposure because he had lived in Somalia for many years. In the past 3 years he had been repeatedly treated with liquid nitrogen for multiple actinic keratoses of his forehead and upper eyelids. The clinical diagnosis of cutaneous...
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. When administered topically or intrathecally 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. Here we report a successful outcome with intratatal cidofovir in this case might be worth considering.

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Multiple iridociliary cysts in patients with mucopolysaccharidosis

The mucopolysaccharidoses (MPSs) are rare hereditary diseases. They are classified into six types by the distinct lysosomal accumulations 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 dentigerous cysts, multilocal cysts in the white matter and arachnoid of the brain, and bone cysts.

In the eye, membrane bound vacuoles in the non-pigmented epithelium of the ciliary processes have been observed by electron microscopy. However, iridociliary 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 echoic 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 arylsulphatase 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 corrected visual acuity in her right eye was 20/30 and left was 18 mm Hg without medication. Ultrasound biomicroscopy revealed multiple round echoic 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 iridociliary cysts were found in either patient.

Comment

We have demonstrated the presence of multiple round cystic lesions. From this echographic finding, we interpret these lesions as multiple iridociliary cysts. It is important to note that 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 retrospectively reported that ciliary cysts were identified in 108 eyes of 86 out of 4632 patients by ultrasound biomicroscopy and the incidence of the multiple cysts occupying more than 180º was 13.3%.

Kunath 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 iridociliary sulcus or pars plicata. The cysts in our patients were located at the posterior iris as Figure 2 shows.

Figure 1 Cutaneous squamous cell carcinoma of the lower right eyelid.

Figure 2 The same patient 24 months later.
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 was due to a blockage of the trabecular mesh-


A case of encephalocranio-cutaneous lipomatosis

Encephalocraniocutaneous lipomatosis (ECCL) is a rare neurocutaneous syndrome characterised by cranial and facial asymme-
ture, cutaneous lesions, central nervous system abnormalities, and ocular abnormalities.

A case is described of a young man who presented with limbal dermoids, subcutan-
eous lipomas, and scalp alopecia. Further investigation revealed cranial and facial asym-
metry, 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 colo-
bomas, an association with ECCL not previ-
ously reported in the literature.

Case report

A 23 year old Asian man first presented to the eye clinic aged 16 with a left conjunc-
tival lesion. On examination he had bilateral conjunctival dermoliopomas, preauricular lip-
mas, 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 exci-
sion biopsy of the left conjunctival dermolioma, 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, intracra-
nial 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 phacoma-
toses. Histology of the conjunctival lesion confirmed a complex corneoscleral choris-
toma 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 intracra-
nial 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, subcutan-
eous lipomas, scalp alopecia, cranial and facial asymmetry, intracranial lipomas and calcifica-
tion, and an arachnoid cyst.

Comment

ECCL is a rare neurocutaneous syndrome of unknown aetiology, first described by Haber-
land and Perou in 1970. All cases described in the literature have been sporadic and there does not appear to be any geographic, racial,
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 puffiness; upper and lower lids for 1 week. She had had a headache for 10 days which had failed to respond to oral antibiotics. Systemic inquiry revealed a sore throat for 10 days but no other symptoms, in particular no thyroid related symptoms. Her visual acuity was 6/6 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 which was not erythematous or tender. There was no proptosis. She had full extraocular movements with no diplopia. The right eye had signs of mild anterior uveitis with 1+ of cells while the left eye had 2+ of cells in the anterior chamber. No evidence of posterior segment involvement, with normal vitreous and fundus.

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 periorbital 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 in the left eye. She had mild periorbital swelling which was not erythematous or tender. There was no proptosis. She had full extraocular movements with no diplopia.

The right eye had signs of mild anterior uveitis 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 fundus.

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 periorbital 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 in the left eye. A mild left RAPD was present. Significant non-tender, mildly erythematous periorbital swelling was present in the left eye. She had 2 mm left relative afferent, 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 choroidal inflammation. An orbital computed tomograph (CT) scan suggested a diagnosis of bilateral anterior NSOI without enlargement of the extraocular muscles (Fig 2). B-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 25.2 mU/l (0.4–4.0) and T4 of 10.6 pmol/l (9–20)) with positive anti-thyroid M antibodies. Thyrotrophin 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 periocular tissues.1,2,4 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.1-3 Other reports of paediatric NSOI do not state whether TFTs were performed.1,4 Similarly, children with posterior scleritis tend not to have any clinical or laboratory evidence of associated systemic disease, but TFTs are not specifically mentioned.1-4 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 al5 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.

NSOI and scleritis in children may be associated with iritis, unlike the adult form of this disorder.1-4 Bloom et al6 report that children with NSOI and anterior uveitis tend to have a worse prognosis and increased recurrence.7 Our patient responded well to steroid treatment with no relapse at 1 year.

Scleritis with uveitis, although rare in children, should be recognised as part of the differential diagnosis of acute paediatric orbital inflammation. Hypothyroidism may be an incidental finding in our case, but the role of thyroid autoimmunity in children with NSOI and scleritis needs to be further investigated.

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 choroidopathy. 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. Her vision acuity 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 chorioidopathy (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 chorioidopathy (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 were sufficiently distinctive to warrant “splitting” rather than “lumping.”

Our patient does not fit neatly into any of the diagnoses discussed above, and the principal clinical features of blind spot enlargement with chorioretinal 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 do 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.

Serological 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 now 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 congenital 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. Serum 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. This may represent the different target DNA strands used in the studies. The negative PCR result on the mother’s serum in our report is therefore not surprising. A negative PCR on amniotic fluid suggested that the fetus was not infected by toxoplasma, which was supported by the normal fetal ultrasounds.

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.

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 with reduced vision. She had had an outbreak of varicella zoster with reduced vision. She had had an outbreak of varicella zoster with reduced vision.

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 significant anisometropia.

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 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 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) 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 active 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 2. Corneal topography of the affected right eye (top) shows a flattening of the 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.
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. Disciform stromal keratitis is an unusual but well recognised complication of primary varicella zoster infection. 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. The complications following disciform 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 unaired 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 cornea 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 of the significant anisometropia and aniseikonia. Despite this she was pleased as she can see 6/9 unaided and was therefore not to wear her rather high myopic spectacle correction, in view of her young age and 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. However, their roles in the treatment of stromal keratitis following primary varicella zoster are controversial and have not been determined. 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. 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. 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 septicaemic 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 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...
clinical setting (febrile-sepsis following an intravenous injection, skin typical lesions) and typical fundus lesions.

In our cases, the diagnosis of presumed candida endophthalmitis was based on (1) the characteristic clinical setting, (2) the typical ocular involvement that was characterized by creamy-white chorioretinal lesions with white balls and vitritis, and (3) the response to antifungal therapy. Vitrectomy was performed only in one case. It was not performed in three patients because they presented an isolated chorioretinitis or associated with a mild vitritis and a characteristic clinical presentation.

Patients were treated with intravenous amphotericin B or intravenous fluconazole and intravitreal injections of amphotericin B except in one patient who refused intraocular injections.

In the mid-1980s, in France, an outbreak of candidiasis followed the introduction on the drug market of a new brown heroin. Refusal of the heroin might have been contaminated with C. albicans was demonstrated. Our cases also seem to confirm that the fungi probably come from the lemon juice or the patient himself rather than from the buprenorphine itself. Since March 1995, substitution therapy with HDSB tablets (Subutex) is approved for licence in France. Unfortunately, 8% of the patients enrolled in substitution programmes continued to use the intravenous route. This drug is now widely prescribed in France. Recently, in Europe, this treatment obtained the authorisation for commercialisation in 13 European countries. Our report demonstrates the need to inform general practitioners, pharmacists, and patients of the risks involved with the intravenous use of substitute agents.

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References


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, reducing measles keratitis scarring, and with improved recognition and treatment of vitamin A deficiency, reducing its associated xerophthalmia and subsequent corneal scarring.

It has been shown that in tropical climates, keratitis of fungal aetiology is much more prevalent than in temperate climates. Little information is available about microbial keratitis in east Africa. The aim of this study was to identify the causative organisms of the condition seen in patients presenting to the Kilimanjaro Christian Medical Centre (KCMC) hospital in northern Tanzania, east Africa. KCMC is one of the largest hospitals in Tanzania, situated on the foothills of Mount Kilimanjaro, serving five regions in northern and central Tanzania—Kilimanjaro, Arusha, Singida, Tanga, and Dodoma with a population of approximately eight million people.

Clinical cases

Patients referred to, or presenting for the first time to, KCMC 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 KCMC

<table>
<thead>
<tr>
<th>Characteristics</th>
<th>Range</th>
<th>Median</th>
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<tbody>
<tr>
<td>Age [years]</td>
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</tr>
<tr>
<td>Time from onset of symptoms to presentation [days]</td>
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<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>

*Empirical herdsman.
†Empirical treatment with broad spectrum topical antibiotic.

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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 corneal 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 presentation 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 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 bacillus). 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 or in part by fungi. As in studies from the United States, west Africa, and southern India, the commonest 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 (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 part 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 as part of the microbiological examination, but this became impossible after logistical difficulties.

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 excision. Culture results were nevertheless useful in ascertaining antibiotic sensitivities, and in adding antifungal treatment where necessary. Any patient undergoing excisionation had usually been an inpatient for several weeks with a painful blind eye; had had multiple treatment regimens, and had often requested excisionation himself. The overall rate of excisionation was 25%. Although there is little published material on outcomes of microbial keratitis in the developing world, this figure 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.

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References
for 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 ECIV 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)

SPecific Eye ConditionS (SPECS) is a not for profit organisation which acts as an umbrella organisation for support groups of any conditions or syndrome with an integral eye disorder. SPECS represents over fifty different organisations related to eye disorders ranging from conditions that are relatively common to very rare syndromes. We also include groups who offer support of a more general nature to visually impaired and blind people. Support groups meet regularly in the Boardroom at Moorfields Eye Hospital to offer support to each other, share experiences and explore new ways of working together. The web site www.eyeconditions.org.uk acts as a portal giving 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: k@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 821 334; email: lynda@brps.demon.co.uk; web site: www.brps.demon.co.uk) 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 +1 312 228 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, keratoprostheses, advanced keratoplasty techniques, paediatric cornea, keratorefractive surgery, and intraocular refractive surgery. Spaces are limited and a benefical package rate is available prior to 30 September 2002. Further details: CORNEA 2002 organiser at the Cornea 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).