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 chorioretinopathy, 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 vascular systemic 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 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. Intracocular pressure was 18 mm Hg in both eyes. Ophthalmoscopic examination of the left temporal posterior pole showed a lesion, about five optic disc diameters in size and red-orange in colour (Fig 1B), while, in the right macular area, an irregular appearance of the retinal surface was detected (Fig 1A). Bilateral B-scan echography confirmed the presence of a dome-shaped solid lesion, with regular profile and without choroidal cup, in the left eye, revealing a small solid lesion also in the right posterior choroid. Standardised A-scan ultrasonography documented that the maximum thickness of these solid lesions was 1.56 mm in the right eye (Fig 2A) and 3.32 mm in the left (Fig 3A). In the left eye the high and regular internal reflectivity of the lesion was consistent with the presence of a benign tumour, reliably of an angiomatous type. FA did not detail any significant abnormality in the right posterior pole (Fig 2B), showing an irregular fluorescence of the orange-coloured lesion previously described in the left eye (Fig 3B). ICGA confirmed the diagnosis of CCH of the left eye (Fig 3C, D) and documented an early hyperfluorescence, followed by a relative decrease in fluorescence (“washout”), corresponding to the 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. 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 ICGA6,10 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.126

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 hyperfluorescent and hypofluorescent area in correspondence with choroidal haemangioma (arrows). (C) Early indocyanine green photographic 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 differentiation amelanotic choroidal melanoma, and choroidal haemangioma.126

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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 view of the fundus. The biometry of the left eye showed an axial length of 22.60 mm. Preoperatively intraocular pressures were 15 mm Hg in both eyes. She underwent an uncomplicated phacoemulsification through a superotemporal limbal wound. A capsulorrhexis of about 5 mm was fashioned. A foldable three piece silicone IOL with poly(methylmethacrylate) (PMMA) haptics (Allergan SH4-3 Bur) 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 ophthalmologist 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 deep and 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). Gonioscopy showed an open iridocorneal angle. There was no evidence of any iris changes or changes at the pupillary border, consistent with pseudoexfoliation in either eyes. Goldmann applanation tonometry revealed an intraocular pressure of 5 mm Hg in the left eye and 14 mm Hg in the right. Posterior segment evaluation of the left eye showed diffuse choroidal effusion. This was confirmed by B-scan ultrasonography, which showed total choroidal detachment. Ultrasound biomicroscopy (UBM, 50 MHz probe, Humphrey) showed a ciliary body detachment with central rotation of the ciliary body, as the underlying cause of the hypotony (Fig 1). Repeated Goldmann applanation tonometry revealed an intraocular pressure of 5 mm Hg in the left eye and 14 mm Hg in the right. Posterior segment evaluation of the left eye showed diffuse choroidal effusion. This was confirmed by B-scan ultrasonography, which showed total choroidal detachment. Ultrasound biomicroscopy (UBM, 50 MHz probe, Humphrey) showed a ciliary body detachment with central rotation of the ciliary body, as the underlying cause of the hypotony (Fig 1).

A neodymium: YAG (Nd:YAG) laser anterior capsulotomy was performed. Four relaxing radial anterior capsulotomy cuts were made at 2, 5, 8, and 10 o’clock. The Nd:YAG capsulotomy comprised 50 shots with a power of 1.4 mJ each. During the procedure the anterior capsule was noted to be thick. Immediate widening of the CCC orifice was noted following this procedure (Fig 2, above). The IOL also returned to a well centred position.

Tractical ciliary body detachment, choroidal effusion, and hypotony caused by severe anterior lens capsule contraction following cataract surgery

EDITOR,—Continuous curvilinear capsulotomy (CCC) first described by Gimbel and Neubham1 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.2 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. Tractical 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.2,3 We report a case of tractical ciliary body detachment caused by a severe anterior lens capsulotomy fibrosis, in which Nd:YAG laser anterior capsulotomy was effective in relieving the traction caused by the capsular contraction. We illustrate the value of ultrasound biomicroscopy (UBM) in the diagnosis and management of such conditions.

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 hyperfluorescent and hypofluorescent area in correspondence with choroidal haemangioma (arrows). (C) Early indocyanine green photographic 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 differentiation amelanotic choroidal melanoma, and choroidal haemangioma.126

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

COMMENT
Capsulorhexis has become the preferred method of anterior capsulotomy, and untoward effects have not often been noted. Nevertheless, distinct complications of continuous tear capsulotomy are now recognised. This includes capsular bag hyperdistension, shrinkage of the anterior capsule opening with visual loss, and/or IOL decentration and lens epithelial hyperproliferation on the posterior lens capsule.

In 1993 Davidson first described the capsule contraction syndrome as a complication of continuous curvilinear capsulorhexis. This syndrome is characterised by an exaggerated reduction in the equatorial diameter of the capsular bag, fibrosis of the anterior capsule, and shrinkage of its opening. It has been associated with various eye diseases including pseudoexfoliation, pars planitis, low grade vitritis, high myopia, retinitis pigmentosa, and myotonic dystrophy. It has also been seen in elderly patients. Commonly observed expressions of these diseases are weakened zonules or a chronic inflammation.

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. Frezzotti 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 capsule 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 cataract might result. We feel that the smaller capsulorhexis size and the use of silicone IOL predisposed our patient to develop severe anterior lens capsule contraction. Severe anterior lens capsule contraction can exert continuous traction on the ciliary body resulting in a ciliary body detachment. In this case Nd:YAG radial anterior capsulotomy was helpful in relieving the phimosis and thereby removing the tractional force on the ciliary body.

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

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Angle closure in fellow eye with prophylactic pilocarpine treatment

EDITOR,—Prophylactic pilocarpine treatment is often used in patients presenting with unilateral primary acute angle closure until definitive treatment with laser 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 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 50 mm Hg in the right and 34 mm Hg in the left. The iridocorneal angle was closed on the right eye, and narrow on gonioscopy (grade 2 inferiorly and closed superiorly) on the left, with bilateral moderate nucloescleral 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 38 mm Hg in the right, and 17 mm Hg in the left. Pilocarpine 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 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 after subuent resolution of the attack.

Case 2
A 46 year old hypermetropic woman (right eye +2.75DS−0.50DS) was admitted to hospital. On review 8 hours after admission her IOP was 16 mm Hg in the right eye and 24 mm Hg in the left eye.

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. After successful 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 +9.0−0.25×65° right eye, and 20/25 with −3.0−3.75×180° left eye.

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.1 We present a case with pemphigus vulgaris with severe keratolysis that required a corneal transplantation.

CASE REPORT

A 41 year old man had suffered from pemphigus vulgaris for 2 years, and prednisolone 40 mg/day and cyclosporine 300 mg/day had been prescribed. He was admitted to the Hamamatsu University Hospital on 15 March 1999 with an acute exacerbation of the symptoms because of non-compliance with the 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 cyclosporine 300 mg/day was continued.

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 the 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 shallowing 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 higher concentrations) 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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showing intracellular deposition of immunoglobulin O and high titres 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.

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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 O (right).

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

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. He had been diagnosed as episcleritis. There was no evidence of rheumatic disease; ANA and ANCA were negative. Neither dexamethasone eye drops 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.

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 syndrome.

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 α7β1, instead of integrin αβ2 which is displayed by plasma cells homing to the bone marrow. 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. The atypical location of the plasmacytoma presented here may be mediated through a specific repertoire of adhesion receptors.

Figure 1 Conjunctival and episcleral injection and intraocular invasion has been reported yet.
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 special integrins or a total loss of them was responsible for the peculiar location of the tumour in our patient. It may only be speculated that the isolated plasmacytoma in our case arose from a monoclonal proliferation of plasma cells in an originally inflammatory infiltrate. Whether tissue specific immunoregulatory mechanisms involving accessory cells are also implicated in the localised episcleral tumour growth remain to be elucidated.

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We report a case of acute urteric 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 sulphasalazine.

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 sulphasalazine (1 g four times daily) were started with folic acid (5 mg twice weekly). Topical dexamethasone, cyclopentolate, and oral prednisolone (60 mg reducing course) were added later.

Within 24 hours of starting treatment the patient felt well, with nausea, anorexia, and oligidia. 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 juncture. Retrograde ureteroscopy demonstrated crystalluria, and insertion of a temporary ureteric stent at this time, with administration of intravenous fluids, effected symptomatic relief. Sulphasalazine was suspended.

COMMENT
The majority of reports of sulphasalazine crystalluria occur in patients with AIDS under treatment for toxoplasmosis encephalitis.6 These patients are exposed to various factors predisposing them to the development of crystalluria such as poor fluid intake, fever, diarrhoea, hypoalbuminuria, and acidification of the urine. The associated polypharmacy of many AIDS patients may contribute to crystalluria or stone formation through the latter mechanism, or because of crystallisation of other drugs such as aciclovir, trimeterene, prilimdone, or other sulphonamides.

Historically, sulphasalazine crystalluria has been reported in non-AIDS patients7 and may cause renal impairment in 1–4% of HIV negative patients.8 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 sulphasalazine dose of 40 g.6 Microscopy of freshly voided urine commonly shows characteristic "sheets of wheat" crystalluria and haematuria. Ultrasonography can reveal echogenic foci in the renal parenchyma as well as in the collecting systems, and hydropnephrosis.1 x Ray examination has a low diagnostic sensitivity.

Management can be conservative with prompt analgesia, intravenous fluids, plus or minus diuretics, and alkalisation of urine with sodium bicarbonate to above a pH of 7.5. This usually achieves prompt dissolution of even large calculus.9 It is not always necessary to stop sulphasalazine.

Editor.—Congenital erythropoietic porphyria (CEP: MIM No 263700) is an extremely rare disorder inherited as an autosomal recessive trait, which is characterised by an 80–98% reduction in the activity of uroporphyrinogen III synthase (UROS: EC 4.2.1.75). Clinically, CEP is characterised by severe cutaneous photosensitivity, chronic haemolysis, and massive porphyrinuria resulting from the accumulation in the bone marrow, peripheral blood, and other organs of large amounts of the non-physiological and pathogenic porphyrin isomers, uroporphyrin I and coproporphyrin 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 hypertrophy between palpebral fissures in both eyes. A 3 × 4 mm area of scleral necrosis was observed at the limbus in the right eye (Fig 1). Hypertrophy of the temporal limbus and pigmentation of eyelids were also observed, but lid closure was normal. Corneal changes were not observed. Visual activity was right eye: 20/50, left eye: 20/20.

In order to cover the region of scleral necrosis, 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


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

1 2 3 4 5 6 7 8 9
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 each mutation, and confirmed that each of them had no residual activity. We can expect that both mutations in this case are “disease causative.”

Scleral changes at the body surface lesions in CEP are mainly caused by the accumulation of porphyrins. Here we proved the accumulation of porphyrins in tear drops with a single case of CEP. Additional cases are needed to confirm the presence of porphyrins in eye involvement. Since our finding demonstrates the likelihood that accumulated porphyrins in tear drops directly exerted a toxic effect in scleral lesions, the protection of sunlight by ultraviolet cut glasses is strongly recommended for prevention against the initiation and progression of scleral lesions in the patients with CEP.

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

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

CASE REPORT

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

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

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

COMMENT

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

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

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

In this case, other conditions more commonly associated with positive ANCA titres were excluded on clinical grounds and follow-
ing investigations. Magnetic resonance imaging is sensitive for central vasculitis, and excluded CNS involvement. The optimum treatment of ANCA associated vasculitis is generally considered to consist of a combination of corticosteroids and immunosuppressive agents. Azathioprine, 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-

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

Bilateral facial nerve palsy is rarely seen in vasculitic conditions. Isolated reports of bilateral facial nerve paralysis associated with Sjögren’s syndrome and polyarteritis nodosum exist. Rheumatoid arthritis is a common condition, and life threatening complications, although rare, are well recognised. Initial presentation may be to the ophthalmologist and awareness of such situations, will improve the prognosis for these patients.

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 recurrent 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. This clinical pattern in patients with granuloma-
tous cheilitis, facial palsy, and fissured tongue called lingua plicata was added to the classic features by Rosenthal. This clinical pattern in patients with granulomatous cheilitis, facial palsy, and fissured tongue was first called Melkersson-Rosenthal syn-
drome (MRS) by Lüschner in 1949. We report on a patient with the typical clini-
cal signs who had chronic bilateral conjunctival 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 equally affected.

The pathogenesis of MRS still remains obscure. Several predisposing factors have been considered such as heredity, infection, allergy, or derangement of cranial autonomic vasomotor innervation. Cranial nerve dys-
function (that is, trigeminal nerve), para-
sympathetic (flush, pain), and ocular involve-
ment have been associated with MRS. Summarised ocular involvement includes granulomatous blepharitis, exophthalmus with lagophthalmus, and burning sensations, which may be related to exposure keratitis. In rare cases, palies of the medial rectus muscle, papilloedema, and retrobulbar neuritis have been described.

Conjunctival involvement has not been reported as yet. By conjunctival biopsy taken from our patient we have shown that conjunc-
tival lesions may be present in MRS.
A conservative satisfactory therapy has not been established so far. The results of various modes of symptomatic treatment, including systemic or topical glucocorticosteroids, are questionable.7,8 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.9 Finally, a surgical excision of the mass has been suggested for granulomatous chelitis or blepharitis, in order to improve the motility of the eyeball 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. Hygica 1928;90:77, 111.


Choroidal detachment following extracapsular cataract extraction in a patient treated with latanoprost

EDITOR.—Adverse reactions associated with the topical administration of the synthetic prostaglandin F2 analog latanoprost have been described.7 We would like to report a case of choroidal detachment following extracapsular cataract extraction in a patient treated with topical latanoprost.

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

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

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

COMMENT
The development of choroidal detachment in a patient with primary open angle glaucoma following cataract extraction has been de-scribed.9 However, this patient had previously had a trabeculoplasty, undergone phacoemul-sification, and had severe hypotony postopera-tively. 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 trabeculoplasty. It is evident 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 detach-ment but the IOP remained elevated. Uveal effusion has been noted following phacoemul-sification without concurrent use of latanoprost. However, in this study all effusions were small and correlated with the presence of hypotony following surgery.1

Latanoprost would appear to lower IOP by increasing uveoscleral outflow and it has been suggested that the increased outflow facility while on latanoprost may contribute to hypo-tony 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 detach-ment in the absence of hypotony following cataract extraction should therefore be con-considered. This hypothesis is supported by the presence of significant uveitis in this case some time following the surgery.

To our knowledge there have been no stud-ies 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 inflam-matory response. Such patients may require more frequent review and should be warned to attend urgently if unexpected symptoms occur in the early postoperative period.

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

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


MAILBOX

TIT and CNV

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

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

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NOTICES

Affordable eye care
The latest issue of Community Eye Health (37) discusses affordable eye care. For further information please contact Community Eye Health, International Centre for Eye Health, Institute of Ophthalmology, 11–43 Bath Street, London EC1V 9EL. (Tel: (+44) (0) 20-7608 6900/6910/6923; 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 Co-ordinator (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, CCL 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: opthalm@icmsaust.com.au; website: www.opthalmology.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: sohno@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).
Bilateral facial nerve palsy associated with p-ANCA positive vasculitis in a patient with rheumatoid arthritis

OWEN G STEWART, IAN G SIMMONS, MIKE F R MARTIN and ANDREW J MORRELL

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Conjunctival changes associated with yellow nail syndrome

The yellow nail syndrome (YNS) is a rare clinical entity characterised by slow growing yellow discolouration of nails, chronic peripheral lymphoedema, pulmonary manifestations (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 maxillary sinusitis, and bronchiectasis for 5 years diagnosed as YNS. On examination, nails of both hands and feet showed yellow discoloration, increased curvature, and thickening of the nail plates in fingers and posterior segment in both eyes was unremarkable. Treatment with topical corticosteroid (dexamethasone 0.1%, four times per day) and artificial tears was begun. Two weeks after presentation the patient reported an incomplete resolution of the irritation on the left eye and conjunctival lesion persisted. Computed tomographic scan of the brain and the orbits was normal. A conjunctival biopsy specimen showed multilayered epithelium, nuclear pleomorphism with malpighian differentiation. The underlying connective tissue was normal (Fig 2 bottom). Ocular symptoms improved rapidly and postoperative antibiotic and steroid eye drops were discontinued after 2 weeks. There was no recurrence of the conjunctival lesion but a mild chemosis persisted 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 been excluded and, interestingly, topical dexamethasone used to treat chemosis is efficient.

There is very little information on histopathological findings observed in the YNS. Nail biopsies demonstrated features of maturation disarray in the nail bed epithelium associated with dense, fibrous tissue replacing subungual stroma. The changes that we observed in the conjunctival epithelium are similar to that in the nail bed in YNS. Since it appears that there is a pathogenic association between the YNS and ocular findings, this association is probably not coincidental and could be the first report of ocular manifestation of YNS.

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References


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 syphilis and sarcoidosis.

Case report

A previously healthy 34 year old Hispanic woman with a 2 week history of blurred vision was referred for evaluation. The patient had no systemic complaints. On examination her best corrected visual acuity was 20/30 in
pleocytosis and increased protein concentration; examination was abnormal (mononuclear cells). Serological tests were negative. PPD test were performed. A markedly reactive serum FTA-ABS and low RPR titres (1:2) were obtained. A transbronchial lung biopsy showed non-caseating granulomas (Fig 2C), in which no micro-organisms could be found with Zhiel-Nielsen, PAS, Grocott’s silver, or Warthin-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. Sarcoidosis 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 aequous 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 inflammatory 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...
The corresponding tangent fields are those they are screened at regular intervals. Current recommendations for patients prescribed vigabatrin are that they are screened at regular intervals. Automated perimetry of a patient taking vigabatrin. Left, 1990, right, 1998.

Figure 1


Figure 2

The corresponding tangent fields performed at 1 metre with a 1 mm (broken line) white object and 2 metres with a 2 mm object (solid line). The 2 mm isoptre lies within 1 mm isoptre, which is a non-physiological finding. (Top) Right eye, (bottom) left eye.

References

Successful treatment of squamous cell carcinoma of the lower eyelid with intralosomal cidofovir

Cidofovir (HPMPC), an acyclic nucleoside phosphonate analogue, is a promising drug that acts against a wide number of DNA viruses. 1 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. 2 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. 3 Recent studies have explored intralosomal administration of cidofovir for the treatment of HPV related tumours, such as cervical epithelial neoplasia, oesophageal carcinomas, and HSV-8 induced Kaposi’s sarcoma. 4 We report a squamous cell carcinoma (SCC) of the eyelid which was successfully treated with intralosomal 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 (Fig 1). 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 squamous cell carcinoma was confirmed by biopsies. He attempted to live at his home in Somalia despite this diagnosis. Cidofovir (2 mg/mL) was diluted in phosphate-buffered saline to a concentration of 1 mg/mL and successfully infiltrated the tumour.

Successful treatment of squamous cell carcinoma of the lower eyelid with intralosomal cidofovir

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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 (Fig 1). 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 squamous cell carcinoma was confirmed by biopsies. He attempted to live at his home in Somalia despite this diagnosis. Cidofovir (2 mg/mL) was diluted in phosphate-buffered saline to a concentration of 1 mg/mL and successfully infiltrated the tumour.
Kidney toxicity is the most common but less frequently uveitis, macular oedema, neutropenia, thrombocytopenia, nausea, fever, hair loss, and muscle pain have also been observed. When administered topically or intravenously 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. However, these successful outcomes with intralesional cidofovir in this case might be worth considering.

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References

Multiple iridociliary cysts in patients with mucopolysaccharidoses

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 denitrogenous cysts, multifocal large cysts in the white matter and arachnoid of the brain, and bone cysts.1,3 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. Ultrasonic biomicroscopy revealed multiple round cystic lesions with uniformly low echo density similar to anterior chamber fluid in all quadrants of the posterior iris, iridociliary sulcus, and pars plicata of both eyes (Fig 1).

Case 2
A 23 year old woman, who was diagnosed with Maroteaux-Lamy syndrome (MPS type VI). The activity of 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 20/400. Her right intraocular pressure was 12 mm Hg and left was 18 mm Hg without medication. Ultrasonic biomicroscopy revealed multiple round low 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 echo graphic finding, we interpret these lesions as multiple iridociliary cysts. When we observe 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 cystic lesions were identified in 108 eyes of 86 out of 4632 patients by ultrasonic biomicroscopy and the incidence of the multiple cysts occupying more than 180° was 1.3%. Kubo et al. studied ciliary body in 232 eyes of 116 healthy people by ultrasonic 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...
A case of encephalocraniocutaneous lipomatosis

Encephalocraniocutaneous lipomatosis (ECC) is a rare neurocutaneous syndrome characterised by cranial and facial asymmetry, cutaneous lesions, central nervous system abnormalities, and ocular abnormalities.

A case is described of a young man who presented with limbal dermoids, subcutaneous lipomas, and scalp alopecia. Further investigation revealed cranial and facial asymmetry, intracranial lipomas, and calcification and an arachnoid cyst, supporting a diagnosis of ECC. This patient also had the additional ocular abnormality of bilateral optic disc colobomas, an association with ECC not previously reported in the literature.

Case report

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

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

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

Comment

ECC is a rare neurocutaneous syndrome of unknown aetiology, first described by Haberland and Perou in 1970. All cases described in the literature have been sporadic and there does not appear to be any geographic, racial,
or sex predilection. The syndrome is characterised by cranial and facial asymmetry, cutaneous lesions, central nervous system abnormalities, ocular abnormalities, and occasionally visceral lipomas. The abnormalities tend to be unilateral, although bilateral involvement has been described. Cutaneous lesions consist mainly of subcutaneous scalp lipomas with overlying alopecia but lipomas involving the limbs and paravertebral areas have also been reported. Popular skin lesions are also common and histologically have been found to be lipomas, fibrolipomas, and angiofibromas. Central nervous system abnormalities are numerous and include cerebral lipomas, cerebral calcifications, ventricular dilatation, cerebral atrophy, arachnoid cysts, seizures, spasticity, and mental retardation. The most common ocular lesions in ECCL are epibulbar choristomas and small skin nodules around the eyelids, which histologically represent connective tissue naevi. Other reported ocular abnormalities include a subcutaneous choristoma of the eyelid, a small tag of tissue in the anterior chamber, a persistent posterior hyaloid system, a dysplastic iris, papilloedema, and epicanthus inversus plus hypertelorism.

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

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References
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 of both 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 eye. She had 2 mm left erythematous periorbital swelling was present. Significant non-tender, mildly tender. There was no proptosis. She had full extraocular movements with no diplopia. The left disc was more swollen than the right and retinal striae were present in both eyes (Fig 1). There was no evidence of posterior segment involvement, 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 (left eye). A mild left RAPD was present. Significant non-tender, mildly erythematous peribulbar swelling was present in the left eye. She had 2 mm left relative proptosis, with generalised restriction of 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 (left eye). A mild left RAPD was present. Significant non-tender, mildly erythematous peribulbar swelling was present in the left eye. She had 2 mm left relative proptosis, 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 25.5 mU/l (0.4–4.0) and T4 of 10.6 pmol (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 with no relapse at 1 year.

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.1–3 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.4 Other reports of paediatric NSOI do not state whether TFTs were performed.1,5 Similarly, children with posterior scleritis tend not to have any clinical or laboratory evidence of associated systemic disease, but TFTs are not specifically mentioned.6–8 This is in contrast with our patient who was found to be biochemically hypothyroid. There is some evidence of thyroid autoimmunity in adult patients with NSOI, as shown by Atabay et al7 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.8 Bloom et al reported that children with NSOI and anterior uveitis tend to have a worse prognosis and increased recurrence.9 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 chorioidopathy. 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 acuity with myopic correction (−3.00 sphere 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 or choroidal inflammation.

Figure 1 Fundus photograph of the left eye showing a swollen optic disc, tortuous retinal vessels, and multiple retinal striae.

Figure 2 CT scan showing bilateral soft tissue thickening of the posterior coats of the globe and periorbital tissues. This is predominantly intracanal and adjacent to the left globe. The extraocular muscles appear normal.

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either eye. Humphrey C24-2 testing revealed an enlarged blind spot on the left (Fig 2), while on the right it was normal. Fluorescein angiography demonstrated window and masking defects consistent with chorioretinal scarring, and late leakage at the optic disc margin.

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

Comment

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

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

In multifocal choroiditis with panuveitis (MCP), patients again present acutely with visual loss, scotomata, and/or photopias.
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.

Seroological 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 PHLS 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, IgM, 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).

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

<table>
<thead>
<tr>
<th>Time (days) after onset of symptoms</th>
<th>Dye test titre</th>
<th>Dye test (IU/ml)</th>
<th>ELISA IgM</th>
<th>IgG avidity (%)</th>
</tr>
</thead>
<tbody>
<tr>
<td></td>
<td></td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>10</td>
<td>1/4096</td>
<td>2000</td>
<td>Positive</td>
<td>20</td>
</tr>
<tr>
<td>20</td>
<td>1/2048</td>
<td>1000</td>
<td>Positive</td>
<td>19</td>
</tr>
<tr>
<td>103</td>
<td>1/2048</td>
<td>1000</td>
<td>Positive</td>
<td>--</td>
</tr>
</tbody>
</table>

From previous infection, and the positive IgM serology and IgG avidity results. Sera taken early in infection (<3 months) usually have avidity levels of less than 30%. Most sera taken later in infection (>6 months) have avidity levels of greater than 40%.

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

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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 practitioner with a red and painful right eye. This 11 year old girl presented to her general eye department. Examination confirmed a reduction of visual acuity to 6/24 (Snellen) in the right eye and 6/6 in the left with spectacle correction. She was wearing a correction for myopia with a prescription of −4.75−0.75 × 110° in the right eye and −6.00−0.50 × 90° in the left eye. There was right superficial disciform stromal scar in the central cornea over the visual axis extending towards the periphery at 6 o’clock. Corneal sensation was intact and equal in both eyes. There was no active inflammation with white conjunctiva and quiet anterior chambers. The intraocular pressures were normal. Fundus examination revealed no abnormality. As there was no active inflammation, she was not given any treatment. On review 2 months later, there was no change in her symptoms or in the clinical findings. One year later, she was seen again in the clinic and has a surprising Snellen visual acuity of 6/9−1 unaided, improving to 6/9+2 with +1.75 × 150° in the right eye and 6/6 with −7.00 DS in the left. There was no ocular inflammation. Corneal topography showed a flattening of the right corneal surface secondary to disciform stromal corneal 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 corneal scar. Compare this with the unaffected myopic left eye, which has a steeper corneal surface (bottom).

Spontaneous reduction in myopic correction following varicella disciform stromal keratitis

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

Case report

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

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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 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 septicemic syndrome associated with scalp nodules. Funduscopy revealed a moderate vitritis and a white tiny perifoveolar lesion with few white snowballs (Fig 1). The bacterial and fungal cultures from both blood and anterior chamber tap were negative. Treatment was begun with intravenous fluconazole associated with three intravitreal amphotericin B injections (IVT). After 15 days of therapy, he was discharged on oral fluconazole. Ten days later, the endophthalmitis relapsed with development of a second paramacular necrotising lesion. A posterior vitrectomy was performed. The vitreous cultures were negative for Candida albicans. He was treated again with intravenous amphotericin B, along with amphotericin B IVT with success.

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

Case 3
A 25 year old man, with history of heroin misuse, was referred for decreased vision. Both his right 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 choriotreal 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 occasional injection of HDSB diluted with preserved lemon juice. Two weeks following such an injection, he developed a skin abscess in which cultures revealed C albicans, posterior cervical lymphadenopathy, nodules of the scalp, and arthritis of the left wrist. Three weeks later he complained of decreased vision in his right eye. Funduscopy revealed a parapapillary white lesion and few white snowballs (Fig 2) Treatment was instituted with intravenous amphotericin B and fluconazole but the patient left the hospital against medical advice 4 days later.

Case 4
A 30 year old man, a former heroin misuser, was referred for decreased vision in the left eye. He had been using intravenous HDSB by dissolving the tablets in preserved lemon juice. Ten days earlier, he had a disseminated pustular rash with folliculitis over the chest, shoulders, and back. On funduscopy, there was a 2 + vitritis and a white choriotreal 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.
Clinical setting (febrile sepsicaemia following an intraocular injection, skin typical lesions) and typical fundus lesions. Isolation of the fungus from a vitreotomy specimen could provide a definitive diagnosis but this is not routinely performed or required. In our cases, the diagnosis of presumed candida endophthalmitis was based on (1) the characteristic clinical setting, (2) the typical ocular involvement that was characterised by creamy-white choriretinal 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. The hypothesis that the lemon juice used to dissolve 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 HDSSB tablets (Subutex) is approved for licence in France. Unfortunately, 8% of the patients enrolled in the study of candida endophthalmitis in brown heroin addicts: response to early vitrectomy preceded and followed by antifungal therapy. Clin Infect Dis 1998;27:1134–6.


**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.

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**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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</thead>
<tbody>
<tr>
<td>Age [years]</td>
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</tr>
<tr>
<td>Time from onset of symptoms to presentation (days)</td>
<td>1–90</td>
<td>10</td>
</tr>
<tr>
<td>Number %</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Sex</td>
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<tr>
<td>Male</td>
<td>29</td>
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<td>Female</td>
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<tr>
<td>Student/school</td>
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<td>13.6</td>
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<tr>
<td>Driver/conductor</td>
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<td>9.1</td>
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<tr>
<td>Maasai*</td>
<td>4</td>
<td>9.1</td>
</tr>
<tr>
<td>Retired</td>
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<td>9.1</td>
</tr>
<tr>
<td>Soldier/guard</td>
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<td>2.3</td>
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<tr>
<td>Miner</td>
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<td>Teacher</td>
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<td>2.3</td>
</tr>
<tr>
<td>Businessman</td>
<td>1</td>
<td>2.3</td>
</tr>
<tr>
<td>Treatment before presentation at KCMC of all cases (culture negative cases only)</td>
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<td></td>
</tr>
<tr>
<td>Nil</td>
<td>11 (6)</td>
<td>25</td>
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<tr>
<td>Local remedy</td>
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<tr>
<td>Inappropriate treatment</td>
<td>3 (1)</td>
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<tr>
<td>*“Appropriate”† treatment, inadequate intensity</td>
<td>18 (5)</td>
<td>40.9</td>
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<td>4 (2)</td>
<td>9.1</td>
</tr>
<tr>
<td>Unknown</td>
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<td>9.1</td>
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<tr>
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<tr>
<td>Lid problems</td>
<td>2</td>
<td>4.6</td>
</tr>
<tr>
<td>Nil</td>
<td>11</td>
<td>25</td>
</tr>
<tr>
<td>Organisms cultured</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Fungi</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Fusarium solani</td>
<td>4</td>
<td>9.1</td>
</tr>
<tr>
<td>Fusarium sp</td>
<td>5</td>
<td>11.4</td>
</tr>
<tr>
<td>Aspergillus fumigatus</td>
<td>1</td>
<td>2.3</td>
</tr>
<tr>
<td>Cladosporium sp + Bacillus sp</td>
<td>1</td>
<td>2.3</td>
</tr>
<tr>
<td>Undetermined fungus + Staph epidermidis</td>
<td>1</td>
<td>2.3</td>
</tr>
<tr>
<td>All fungi</td>
<td>12</td>
<td>27.3</td>
</tr>
<tr>
<td>Gram positive bacteria</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Staphylococcus epidermidis</td>
<td>3</td>
<td>6.8</td>
</tr>
<tr>
<td>Staphylococcus aureus</td>
<td>2</td>
<td>4.6</td>
</tr>
<tr>
<td>Gram negative bacteria</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Pseudomonas aeruginosa</td>
<td>6</td>
<td>13.6</td>
</tr>
<tr>
<td>Proteus sp</td>
<td>1</td>
<td>2.3</td>
</tr>
<tr>
<td>No organism cultured</td>
<td>20</td>
<td>45.5</td>
</tr>
</tbody>
</table>

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

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

Comment

In this study fungal keratitis accounted for 50% of culture positive cases of microbial keratitis in northern Tanzania, with the majority of these cases (42%) yielding pure fungal isolates on culture. These figures are similar to those published from west Africa, where 56% of microbial keratitis was caused by fungi.

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

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

Acknowledgements

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References

for Bath Street, London EC1V 9EL, UK (tel: +44 (0)20 7608 6910; fax: +44 (0)20 7250 3207; email: eyeresource@ucl.ac.uk; website: www.jceh.co.uk). Annual subscription (4 issues) UK£25/US$40. 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).

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@ucl.ac.uk).

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.eyesconditions.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@eyesconditions.org.uk; www.eyesconditions.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) and a beneficial package rate is available prior to 30 September 2002. Further details: CORNEA 2002 organiser at the Corneo Plastic Unit, The Queen Victoria Hospital, Holtye Road, East Grinstead, West Sussex, RH19 3DZ (tel: 01342 410 210 ext 560; fax: 01342 317 181; email: Cornea2002@hotmail.com).

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