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, unilateral 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, discoform scar, serous detachment, and central serous chorioretinopathy, but may be differentially diagnosed with fluorescein angiography (FA), indocyanine green angiography (ICGA), ultrasonography, and periodic observation. The bilateral CCH localisation represents an extremely uncommon condition which, in literature, has been only reported in association with Sturge-Weber syndrome or Klippel-Trenaunay-Weber syndrome. To the best of our knowledge, this is the first documented case of bilateral CCHs in the absence of any other evidence of 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. Intraocular pressure was 18 mm Hg in both eyes. Ophthalmoscopic examination of the left temporal posterior pole showed a lesion, about five optic disc diameters in size and red-orange in colour (Fig 1B), while, in the right macular area, an irregular appearance of the retinal surface was detected (Fig 1A). Bilateral B-scan echography confirmed the presence of a dome-shaped solid lesion, with regular profile and without choroidal cup, in the left eye, revealing a small solid lesion also in the right posterior choroid. Standardised A-scan ultrasonography documented that the maximum thickness of these solid lesions was 1.56 mm in the right eye (Fig 2A) and 3.32 mm in the left (Fig 3A). In the left eye the high and regular internal reflectivity of the lesion was consistent with the presence of a benign tumour, reliably of an angiomatous type. FA did not detail any significant abnormality in the right posterior pole (Fig 2B), showing an irregular fluorescence of the orange-coloured lesion previously described in the left eye (Fig 3B). ICGA confirmed the diagnosis of CCH of the left eye (Fig 3C, D) and documented an early hyperfluorescence, followed by a relative decrease in fluorescence (‘washout’), corresponding to the 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 echographic and ICGA 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 to differentiate amelanotic choroidal melanoma, choroidal metastasis, and choroidal haemangioma.

**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 fornix incision. A capsulorhexis of about 5 mm was fashioned. A foldable three-piece silicone IOL with poly(methylmethacrylate) (PMMA) haptics (Allergan SH40 NB) was implanted “in the bag”. The lens had an optic diameter of 1.0 mm and a haptic diameter of 1.30 mm. In the immediate postoperative period she was noted to have a well centred IOL “in the bag” and fundus showed an inferior hemiretinal vein occlusion involving the macula in the left eye. At this time she had a visual acuity of counting fingers at 2 metres in her right eye and 6/60 in her left eye.

Two and a half months following her cataract surgery she was referred by an optician with deterioration of vision in her left eye. Visual acuity was counting fingers at 2 metres in both eyes. Slit lamp biomicroscopy of the left eye showed a thick lens anterior capsule. Severe contraction of the CCC opening with eccentric displacement of the CCC orifice was noted and the IOL was displaced superiorly (Fig 1). Ultrasound biomicroscopy 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, below).

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, below). The IOL also returned to a well centred position.

![Image](http://example.com/image.png)
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 aplation tomometry 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 pseudoxfoliation, 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.

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

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

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Angle closure in fellow eye with prophylactic pilocarpine treatment

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

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

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

Case 2
A 46 year old hypermetropic woman (right eye +2.75DS −0.5 × 45, left eye +4.50DS) with no significant ocular history presented to casualty with intermittent visual disturbance followed by pain, redness, and decreased vision in the left eye. Visual acuity on presentation was right eye 6/9 and left eye 6/24. The left cornea was hazy with a shallow anterior chamber with IOP of 62 mm Hg. The right iridocorneal angle was narrow but open with pigmented grade 1 angle on gonioscopy. She was admitted and treated with topical apraclonidine, levobunolol, dexamethasone, and intravenous Diamox 500 mg. Pilocarpine 4% every 15 minutes for 1 hour was used in the left eye and a single dose of 4% pilocarpine was instilled in the right eye.

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

COMMENT
The management of the fellow eye in acute glaucoma is controversial. Although Nd:YAG peripheral iridotomy has established itself as the treatment of choice,1 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.1 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 autochthonous shallowing of the anterior chamber, potentially precipitating angle closure in compromised eyes.2 3

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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Keratolysis in a patient with pemphigus vulgaris

EDITOR—Pemphigus vulgaris is an auto-immune, blistering disease of the skin and mucous membranes.1 The characteristic ocular finding is conjunctivitis, and corneal involvement is rare.1 2 3 We present a case with pemphigus vulgaris with severe keratolysis that required a corneal transplantation.

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

COMMENT
Corneal involvement is a rare complication in patients with pemphigus vulgaris. Severe corneal involvement has never been reported except in the case of a 56 year old man with severe ocular involvement including conjunctivitis, corneal ulceration, and perforation despite immunosuppressive therapy.1 Although a causative organism was not isolated, the authors suggested that the complications were due to an infectious agent.

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

The second mechanism is an autoimmune mechanism against one of the intercellular adhesion molecule—for example, desmoglein (Dsg). The patient was diagnosed as pemphigus vulgaris by histological examination, direct immunofluorescent staining of the skin.
showing intracellular deposition of immunoglobulin G and high titres of circulating anti-Dsg 1 antibodies (Fig 2). Because the cornea usually does not have Dsg 3, 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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Isolated episcleral plasmacytoma mimicking episcleritis in a patient with benign monoclonal gammopathy

EDITOR,—We present the unique case of a patient with an isolated plasmacytoma of the episclera mimicking a painful episcleritis. Plasmacytomas usually grow in the bone marrow probably because of their special homing receptors—for example, αβ, integrin. Solitary plasmacytic tumours outside the bone marrow are rare. They mostly involve the oropharynx and the upper respiratory tract, but have also been encountered in the lids, the orbit, and the palpebral conjunctiva. Only one case of a solitary epibulbar plasmacytoma with intraocular invasion has been reported yet.

CASE REPORT
A 61 year old patient presented with an "inflammatory" episcleral nodule within the lower temporal quadrant and mild pain in his left eye (Fig 1), which had already lasted 5 months and had been diagnosed as episcleritis. There was no evidence of rheumatic disease; ANA and ANCA were negative. Neither dexamethasone eyedrops nor oral fluocortolone (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.

Surprisingly, the histopathological examination revealed a monomorphous infiltrate of plasma cells with characteristic eccentric nuclei and basophilic cytoplasm (Fig 2). The cells stained positively with the plasma cell marker V$38$ and showed a kappa light chain restriction. The proliferation rate was increased with an MIB1 positivity of 5%. Immunohistochemistry for CD20, IgA, IgD, and IgG was negative.

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

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

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

Lymphocytes and plasma cells of the MALT, especially the GALT, are characterised by integrin αβ, from integrin αβ, 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

Figure 1 Conjunctival and episcleral injection and flat subconjunctival tumour mass at the temporal part of the left eye.

Figure 2 Monomorphous infiltrate of plasma cells with characteristic eccentric nuclei and basophilic cytoplasm (haematoxylin and eosin, ×400).

Figure 2 Histological examination by haematoxylin and eosin staining of lesional skin disclosed intraepidermal clefts which contained several acantholytic cells (left). The direct immunofluorescent staining of the skin showed intercellular deposition of immunoglobulin G (right).
We thank Mrs Renate Buchen for technical assistance, Dr Sarah Coupland for providing the VS38 immunostaining, and Dr Flemming Stehbach for reading the manuscript.

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Crystalluria with sulphadiazine

EDITOR.—Toxoplasmosis is the commonest cause of uveitis in people. Ocular toxoplasmosis may occur as part of the primary acquired infection or through reactiva- tion of encysted organisms at the edge of an atrophic scar. Current indications for treatment include sight threatening lesions at or adjacent to macula or papillomacular bundle and disc or marked vitritis.

Treatment is commonly with a combination of the synergistic antagonists of folate metabo- lism, sulphadiazine, and pyrimethamine. Folic acid rescue is added to prevent bone mar- row suppression. Steroids are frequently used in combination with antimicrobials in sight threatening inflammatory foci of infection.

We report a case of acute ureteric obstruc- tion in a young female with her first presenta- tion of recurrent ocular toxoplasmosis. We would like to bring to the attention of ophthalmologists the risk of crystalluria in patients being treated with sulphadiazine.

CASE REPORT

A 22 year old, otherwise fit woman presented with floaters in the left eye. She had had poor vision since childhood when she had been diagnosed as “amblyopic” and undergone strabismus surgery for esotropia. A pigmented and atrophic scar was present at the left macula, and involving the fovea. At the 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 sul- phadiazine (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 oligidipsia. She developed pink discoloration of the urine in which the 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 vesi- coureteric junction. A retrograde ureteroscopy demonstrated crystalluria, and insertion of a temporary ureteric stent at this time, with administration of intravenous fluids, effected symptomatic relief. Sulphadia- zine was suspended.

COMMENT

The majority of reports of sulphadiazine crys- talluria occur in patients with AIDS under treatment for toxoplasmosis encephalitis.1-3 These patients exhibit several factors predisposing them to the development of crystalluria such as poor fluid intake, fever, diarrhea, hypoalbuminemia, and acidification of the urine. The associated polypathy of many AIDS patients may contribute to cystine or stone formation through the latter mech- anism, or because of crystallisation of other drugs such as aciclovir, triamterene, prami- done, or other sulphonamides.

Historically, sulphadiazine crystalluria has been reported in non-AIDS patients4 and may cause renal impairment in 1–4% of HIV nega- tive patients.5 To our knowledge, however, this complication has not been reported in the ophthalmic ophthalmologists we surveyed were not aware of this potential complication, nor is it documented in the British National Formulary. Although it occurred quickly in our patient, the complication usually occurs after a median of 10 days in HIV negative subjects at a cumulative sulphadiazine dose of 40 g.6 Microscopy of freshly voided urine com- monly shows characteristic “sheaves of wheat” crystalluria and haematuria. Ultrasonography can reveal echogenic foci in the renal paren- chyma as well as in the collecting systems, and hydropnephrosis.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 calculi.7 It is not always necessary to stop sulpha- diazine.

REFERENCES


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

EDITOR.—Congenital erythropoietic porphy- ria (CEP; MIM No 263700) is an extremely rare disorder inherited as an autosomal reces- sive trait, which is characterised by an 80–98% reduction in the activity of uroporphyrinogen III synthase (UROS; EC 4.2.1.75).1 Clin- ically, CEP is characterised by severe cutane- ous photosensitivity, chronic haemolysis, and massive porphyrinuria resulting from the accumulation in the bone marrow, peripheral blood, and other organs am- monium, of the non-physiological and pathogenic porphyrin isomers, uroporphyrin I and copro- porphyrin I. Red urine may be observed from infancy, and the teeth become stained red. Haemolytic anaemia, an additional complica- tion, may be helped by splenectomy. Besides such manifestations, we reported a scleral change in the patient with CEP,2 who had a remarkable increase of porphyrin 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 manifesta- tions of CEP such as skin ulcer and scaring. He was diagnosed with CEP in childhood, because of the elevation of porphyrins in urine. At the time of visit, slit lamp examina- tion of bulbar conjunctiva revealed irregular meatal excrescences in both eyes. A 3 x 4 mm area of sceral necrosis was observed at the limbus in the right eye (Fig 1). Hypertrophy of the temporal limbus and pigmentation of eyelids were also ob- served, but lid closure was normal. Corneal changes were not observed. Visual activity was in right eye: 20/50, left eye: 20/20.

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

To confirm whether this scleral necrosis is caused by the direct effect of the accumulation of...
dicted a glutamine to premature stop codon

...alanine substitution at residue 62 (T62A), and

been performed and an A to G transition of nucleotide 745 that pre-

...protein IX (PP) were observed (8.48, III (UROI + III), coproporpyrin I (CPI), and

...markable elevation of uroporphyrin I + II in a patient with CEP. Additional cases are

...pseudohypoaldosteronism, and splenomegaly were present. Her medication consisted of methotrexate 5 mg weekly, thyrxxine 100 µg once daily, and folic acid 5 mg once daily. Examination revealed isolated left sided lower motor nervous facial nerve paraplegia, and a left Bell's palsy was diagnosed. One week later, she returned with right sided facial weakness. No improvement on the left side had occurred and bilateral lower lid paralytic ectropion was evident. A provisional diagnosis of rheumatoid associated mononeuritis multiplex was made, and a rheumatological consultation was obtained. Haematological investigations revealed a positive rheumatoid factor (RF) and p-ANCA, and a raised plasma viscosity of 1.80. Other autoimmune studies including ANA, anti-Ro and La antibodies, and c-ANCA were negative, and renal function was normal. Chest radiography and magnetic resonance imaging of the brain were unremarkable.

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

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

COMMENT

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

Rheumatoid factor consists of IgM antibodies against the patients’ own IgG, and is an important diagnostic feature in rheumatoid arthritis. However, RF may also be seen in
polyarteritis nodosa, sclerodermia, Wegener’s granulomatosis, Churg–Strauss syndrome, 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 cyto toxicity caused by ANCA. ANCA may promote neutrophil activation and endothelial injury, by targeting the neutrophil granule enzymes protease 3 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 haemolytic 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 cerebral vasculitis, and excluded CNS involvement.

The optimum treatment of ANCA associated vasculitis is generally considered to consist of a combination of corticosteroids and oral immunosuppressive agents. Azathioprine, cyclosporin, azathioprine, or cyclophosphamide 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 predni-
solone.

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 keratitis 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 nodosa exist.

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

**Letters, Mailbox, Notices**

**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 Melkerson in 1928. Three years later a fissured tongue called lingua plicata was added to the classic features by Rosenthal.

This clinical presentation in patients with granulomato-
sus cheilitis, facial palsy, and fissured tongue was first called Melkerson-Rosenthal syn-
drome (MRS) by Lüscher in 1949.

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

**CASE REPORT**

A 64 year old man presented with a fissured tongue, recurrent painless facial oedema, especially of the eyelids, and facial flush for 6 years. A review of the other systemic diseases was unremarkable. He required blepharoplasty for the lid malformation. The histopathological findings of the skin biopsy confirmed the clinical suspicion of MRS by the typical granulomatous infiltration (Fig 1A).

Furthermore, he complained bilateral conjunctival swelling had been present for 6 months. Visual acuity was 20/30. Slit lamp examination revealed a bilateral fleshy mass extending from the upper fornices to the lim-
bus and conjunctiva (Fig 1B). Motility of the eyeball was normal and an exophthalmus has not been present. In the magnetic resonance image (MRI) of the orbit a bilateral enlargement of lacrimal glands and a swelling of the lateral rectus and superior rectus muscle of the right eye were observed. Staging examinations for lymphoma or other malignancies were uneventful. Since differen-
tial diagnosis included a bilateral orbital lymphoma a conjunctival biopsy was performed. The histopathological examination of the conjunctival specimen by light microscopy revealed a subepithelial process. An infiltrate of small lymphocytes without any differenta-
tion and with seprate orientation was found in the conjunctival tissue.

**COMMENT**

Disymptomatic or monosymptomatic clinical courses are frequently observed in MRS. As the symptoms rarely appear simultaneously MRS often can be diagnosed only by longitudinal follow up series. Males and females are equally affected. Symptoms usually manifest during adolescence and have rarely been seen in childhood or individuals older than 50 years. The pathogenesis of MRS still remains obscure. Several predisposing factors have been considered such as heredity, infection, allergy, or derangement of cranial autonomic vasmotor 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, papillodecema, 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 satisfactory conservative therapy has not been established so far. The results of various modes of symptomatic treatment, including systemic or topical glucocorticosteroids, are questionable. They may reduce the patients’ complaints, at least temporarily. Another treatment of choice is clofazimine, which is an oral phenazine. This drug had been useful in other conditions with granulomatous inflammation. Finally, a surgical excision of the masses 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. Hygionica 1928;90:737–41.

Follow up was uneventful except for the development of left age related maculopathy in 1995 reducing the vision to 6/9. In November 1999 the intraocular pressure (IOP) became uncontrolled and a left sided cataract noted. Latanoprost was substituted with subsequent control of the IOP.

He underwent an uneventful left extracapsular cataract extraction and a trabeculectomy, undergone phacoemulsification on any other treated area and had severe hypotony postoperatively. The IOP was 22 mm Hg and the visual acuity had improved to 6/12 at best.

COMMENT

The development of choroidal detachment in a patient with primary open angle glaucoma following cataract extraction has been described. However, this patient had previously had a trabeculectomy, undergone phacoemulsification, and had severe hypotony postoperatively. In another report choroidal effusion and hypotony were noted in a patient who 8 months before commencing latanoprost had undergone a combined cataract extraction and trabeculectomy. It was noted that, in our case, the choroidal detachment was present from a short time following surgery in view of the subjective shadow in the patient’s vision. It would appear that the detachment developed and persisted in the presence of an elevated IOP. Withdrawal of the latanoprost led to complete resolution of the choroidal detachment but the IOP remained elevated. Uveal effusion has been noted following phacoemulsification without concurrent use of latanoprost. However, in this study all effusions were small and correlated with the presence of hypotony following surgery.

Latanoprost would appear to lower IOP by increasing uveoscleral outflow and it has been suggested that the increased outflow facility while on latanoprost may contribute to hypotony and the development of choroidal effusions. Although our patient may have had an episode of hypotony immediately following his surgery, IOP measurements did not suggest this. The possibility of latanoprost initiating or potentiating choroidal detachment in the absence of hypotony following cataract surgery must be considered. This hypothesis is supported by the presence of significant uveitis in this case some time following the surgery.

To our knowledge there have been no studies examining the incidence and severity of uveitis following cataract surgery where latanoprost has been continued. This case emphasises the possibility that uveitis can occur in patients undergoing surgery while continuing to use antiglaucoma medications which may potentiate the inflammatory response. Such patients may require more frequent review and should be warned to attend urgently if unexpected symptoms occur in the early postoperative period. Surgeons who perform cataract surgery on eyes in which the breakdown of the blood-aqueous barrier is expected to be greater than that produced by routine phacoemulsification surgery should consider substituting another IOP lowering agent for latanoprost in the immediate preoperative and postoperative period.

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MAILBOX

TTT and CNV

EDITOR.—We thank Ergun and Stur4 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 transpupillary thermotherapy (TTT) is confusing; however, it is known that more irradiance (W/cm²) is needed for smaller laser spots because heat conduction from choroidal blood flow cools smaller spots more efficiently than larger spots. This physiological phenomenon was established in experiments,4 theoretical,5 and clinical6 studies. Furthermore, it is true that overlapping zones occur when multiple spots are used for very large treatment areas. None the less, these zones experience the same temperature rise as every other treated area and no clinical abnormalities have been noted in the small overlapping zones. Although TTT is mainly used for occult membranes our results indicated 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/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, CCI Ltd, 2 Palmerston Court, Palmerston Way, London SW8 4AJ, UK (tel: + 44 (0) 20 7720 0600; fax: + 44 (0) 20 7720 7177; email: melanoma@confcomm.co.uk; website: www.confcomm.co.uk/Melanoma).

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

International Society for Behçet’s Disease
The International Society for Behçet’s Disease was inaugurated at the 9th International Congress on Behçet’s Disease. Professor Shigeaki Ohno represents the ophthalmology division (Department of Ophthalmology and Visual Sciences, Hokkaido University Graduate School of Medicine, Sapporo, Japan: tel: +81-11-716-1116 (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: zobbere@zedat.fu-berlin.de).