LETTERS TO
THE EDITOR

Involutional type of entropion in a child with cutis laxa

Editor,—The diffuse elastic tissue disease called cutis laxa (CL) is a serious, even lethal systemic illness, involving not only the skin but connective tissues throughout the body. The skin hangs in loose folds, producing the appearance of premature ageing. Internal manifestations such as emphysema, ectasia of the aorta, and multiple hernias are usually present.

We report a child with cutis laxa, who presented with an unusual ophthalmic manifestation of the disease.

CASE REPORT

Our patient, who is now a 4 year old boy and the third child to a normal first degree cousin couple, was noted to have redundant skin and a hoarse cry at the age of 3 months. Skin biopsy was consistent with cutis laxa (elastin stain showed focal thickening of the elastic fibres with tapered ends). His 7 month old sister was also diagnosed as having cutis laxa at 3 months of age. Her ophthalmic examination revealed no abnormalities. Otherwise, the family history was negative for such skin problems.

Recently, he presented to our clinic with a 2 month history of a red right eye. Examination revealed an entropion of the right lower lid (Fig 1A). The lid position corrected temporarily upon manual traction but re-inverted shortly after. The lids were hyperextensible but inelastic. Manual inversion of the lower lid resulted in significant fat prolapse into the fornix (Fig 1B). Slit lamp examination revealed moderate inferior corneal staining and injection of the medial and inferior bulbar conjunctiva. Fundus examination was normal. There was significant skin laxity over the eyelids, cheeks, neck, and trunk.

Surgical correction was carried out using a lateral tarsal strip in addition to two full thickness lid sutures. A small piece of resected eyelid tissue was sent for pathological examination.

Histopathologically, the eyelid specimen showed significant granular degeneration of the elastic fibres (Fig 2). There was also a decreased number of elastic fibres, especially in the superficial dermis.

Postoperatively, the lower lid position was normal at 12 months follow up.

COMMENT

Cutis laxa was first described by Alibert in 1832. The rare syndrome of cutis laxa is a heterogeneous group of disorders characterised by inappropriate laxity of the skin which appears loosely folded to form a cuticular layer larger than the body it envelopes. This leads to the production of a typical grotesque facies and the appearance of premature ageing. The skin is hyperextensible but inelastic. These skin changes are frequently associated with systemic abnormalities, particularly of the lungs and heart. There may be ophthalmic manifestations.

Cutis laxa may be inherited in an autosomal dominant or recessive manner. The clinical features and prognosis differ considerably in the two forms. In the autosomal dominant variety, complications are mild, and the patients have a normal life span. Conversely, in the autosomal recessive type, there is a high incidence of illness and death in childhood from pulmonary and cardiac involvement. Furthermore, autosomal recessive forms of CL can be divided into two types: CL with emphysema and CL with retarded development. The first disorder usually leads to death within the first years of life from cardiopulmonary complications. The second disorder is not associated with pulmonary disease, but there are many systemic defects, among which gross delay in motor development is the most important.

Extensive analysis of the skin and other organs of patients with CL has demonstrated defective elastic fibres throughout the body. This defect consists of a reduction in the amount and size of the elastic fibres and granular degeneration and fragmentation of the fibres with disruption of their normal arrangements; hence the term "generalised elastolysis".

Goltz and coworkers suggested an imbalance between the circulating pancreatic elastase and its inhibitor (pancreatic elastase inhibiting substance, EIS), with a diminution of the latter in patients with CL. Recently, frame shift mutations in exon 30 of the elastin gene were identified in three affected individuals.

The reported ophthalmic manifestations of CL include entropion, blepharochalasis, epicanthic folds, hypertelorism, bilateral macular colobomas, fine retinal pigmentary changes, and bilateral orbital fat prolapse. The case reported here presents a new manifestation of the disease—namely, lower lid entropion, with all the criteria of the involutional type. Several anatomical abnormalities have been identified as causative factors in involutional entropion, including (a) horizontal lid laxity, (b) dehiscence or attenuation of lower lid retractors, (c) overriding of the preseptal over the pretarsal orbicularis muscle, and (d) enophthalmos, the role of which has been recently proved to be insignificant.

Both lid lamellae and the canthal tendons contain elastic fibres. Histopathologically, the eyelid specimen showed significant granular degeneration in addition to a decreased number of the elastic fibres, thus accounting for the horizontal lid laxity present in our patient. In addition, the lower lid retractors also contain elastic fibres. The shallowness of the lower fornix and the fat herniation into it upon manual inferior lid traction confirm the laxity of the lid retractors and the orbital septum secondary to the disorder. These factors, horizontal lid and retractor laxity, allowed for the overriding of preseptal over pretarsal orbicularis and the inward rotation of the lid margin in a fashion similar to that which occurs with involutional entropion in elderly people.

In summary, cutis laxa is a systemic disease that relates to the presence of abnormal elastic fibres throughout the body. Involutional entropion in a 4 year old child is an unusual finding that was associated with marked laxity of the eyelid tissues present in cutis laxa. Step unidentified.

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Figure 1 (A) Entropion of the right lower lid. (B) A shallow lower fornix with significant fat herniation upon manual inferior lid traction.

Figure 2 Eyelid tissue stained for elastic fibres showing marked granular degeneration of the elastic fibres. Aldehyde-fuchsin, x400.
Unilateral arcus lipoides corneae with contralateral Sturge–Weber syndrome

EDITOR,—Arcus lipoides corneae usually occurs bilaterally and symmetrically.1 Pronounced unilateral arcus lipoides corneae occurs in atrophic eyes, less marked with relative ocular hypotony1 or contralateral carotid artery stenosis.2,3 We report on a patient with unilateral arcus lipoides in the normal eye sparing the other one with Sturge–Weber syndrome.

CASE REPORT
A 33 year old patient showed a left sided secondary juvenile open angle glaucoma due to Sturge–Weber syndrome. The glaucoma diagnosed at the age of 10 months with a maximum intraocular pressure (IOP) of 40 mm Hg had been treated twice by diathermy of the ciliary body, and once by trabeculotomy during the first 6 years of life. Since then, the IOP has ranged between 16 and 20 mm Hg in the left eye, constantly being 4–6 mm Hg lower in the right eye.

Visual acuity was 20/15 right eye and 20/30 left eye, intraocular pressure 16 mm Hg right eye and 20 mm Hg left eye. The right eye disclosed a definite yellow-whitish arcus lipoides extending through the entire corneal thickness, separated from the limbus by a clear zone (Fig 2A). The arcus was more pronounced and inferiorly than in the medial and lateral quadrant. Apart from the arcus lipoides, the right eye did not reveal any abnormalities. No arcus lipoides could be detected in the left eye (Fig 2B). The eye showed increased episcleral vascularisation. The optic disc presented with moderate glaucomatous disc change (optic disc size 3.0 mm, cup/disc ratio 0.78).

COMMENT
Arcus lipoides corneae is known to be usually associated with hyperlipoproteinaemia, especially type IIA and IIB, but may occur without predisposing factors.1 Histologically, it mainly consists of extracellular deposits of cholesterol, phospholipids, and triglycerides in stroma, Bowman and Descemet membrane.1 Generally, it is developed bilaterally and symmetrically.1 Single cases of unilateral arcus lipoides corneae were reported in eyes with absolute or relative ocular hypotony,4,5 or in patients with contralateral carotid artery stenosis.6,7

Our patient presented with a narrow, but clearly recognisable arcus lipoides which occasionally can occur in younger people, even in the absence of marked hyperlipoproteinaemia.1 In earlier reports,1 it was suggested that vascular congestion and increased vascular permeability of limbal blood vessels might possibly be involved in the development of unilateral arcus lipoides. Analogously to cases of absolute ocular hypotension, one could assume that in our patient, the relative hypotension in the right eye with consecutively relatively higher blood supply may have facilitated the lipid deposition in the cornea, whereas the development of arcus was prevented by the higher IOP in the left eye.

In our opinion, this case implies that unilateral arcus lipoides corneae can occur in morphologically normal eyes with relatively ocular hypotony.

Sickle form macular whitening in a child after viewing a solar eclipse

EDITOR,—Individual cases of retinopathies after unprotected exposure to sunlight are frequently reported in patients with psychiatric disorders,8 after religious practices,9 or related to drug abuse.10 Even though knowledge of the harmful effects of sun gazing dates back to ancient times11 there are still many reports on epidemics of patients with solar retinopathy after viewing solar eclipses.2 This case report demonstrates that, despite the availability of cheap and safe protection,12 cases of eclipse retinopathy are still observed and so strong preventive efforts should be made for future eclipses, especially for groups at risk.

CASE REPORT
An 11 year old girl was evaluated after complaining of bilateral central scotoma after observing the subtotal solar eclipse of 11 August 1999 in south eastern Switzerland without protective eyewear. Visual acuity 5 days after the event was 20/25 in both eyes. Amsler grid testing showed bilateral small central scotoma. Anterior segments were unremarkable. Biomicroscopy showed sickle form oedematous areas parafoveal in the deep retinal layers in both eyes, corresponding to the shape of the subtotal solar eclipse (Fig 1).

After 3 months the visual acuity was 25/20, the patient denied any residual scotoma in the Amsler grid testing. The retinal edema had resolved, but there were some subtle pigmen-
tary irregularities to be observed (Fig 2).

COMMENT
Areas of retinal edema outlining a sickle— that is, the ‘brand’ of the sun, have been described after the solar eclipses of 17 June
Figure 1 Sickle form oedematous areas parafocally in the deep retinal layers in both eyes. They were similar in shape to the development of the subtotal solar eclipse calculated for the point of observation of our patient.

Figure 2 Suble pigmentary irregularities in both maculas 3 months after eclipse retinopathy.

1890 and 29 April 1976. If the patient does not gaze at the sun for brief periods but observes it constantly for a longer period of time during a defined phase of the eclipse the subsequent retinal lesion can be sharply defined. From the shape of the “brand” one can determine the time phase of exposure, which in the present case was the maximum of the subtotal solar eclipse (Fig 1).

Approaching the 1999 solar eclipse there was extensive information in the media about appropriate eye protection and special glasses for eclipse viewing were widely available. In addition, there was cloudy weather over most parts of central Europe. Only a few mild cases of eclipse retinopathy have therefore been reported in the eastern part of Switzerland.

Our patient observed the eclipse from the mountains of south eastern Switzerland, where the sky was clear. Although the family was in possession of appropriate protective glasses, the 11 year old girl did not use them. She was in possession of appropriate protective glasses, the 11 year old girl did not use them. Although prevention strategies have proved to be very effective children seem to be the most at-risk group in the population for retinal damage from solar eclipses. We therefore recommend that special efforts be made to ensure eclipse viewing safety in children.

The next total solar eclipse will take place over southern Africa and Madagascar on 21 June 2001.

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Serum antibodies to HSC71 in Vogt-Koyanagi-Harada disease

EDITOR,—Heat shock proteins (HSPs) are highly conserved immunogenic intracellular molecules, and are induced by heat, inflammatory mediators, and physiological stress. The presence of antibodies to HSPs or heat shock cognate protein (HSC) has been reported in several autoimmune diseases. However, despite the prevalence of these antibodies in these autoimmune diseases, their significance is not fully understood.

In this report, we investigated antibodies to HSC71 specific antibody levels in sera from patients with Vogt-Koyanagi-Harada disease (VKH), a systemic disorder that affects various organs that contain melanocytes and is believed to be an autoimmune disease.1

CASE REPORT
Serum samples were obtained from eight patients with VKH, and 54 unaffected volunteers. All samples were obtained from VKH patients during the acute phase of the illness with severe uveitis and were taken before steroid administration. Mean ages were 43 and 40 for VKH and controls, respectively. Informed consent was obtained from all of the patients and volunteers. All sera were stored at −70°C until use.

Cloning and expression of the recombinant HSC71 (HSC71) were performed as previously described.1 Anti-HSC71 antibody levels of sera were assayed by enzyme linked immunosorbent assay. In brief, flat bottomed microtitre plates were coated with 10 µg/ml rHSC71 in phosphate buffered saline (PBS). After incubation overnight, non-specific binding sites were blocked with PBS containing 0.05% Tween 20, 1 mM EDTA, 0.25% BSA, and 0.05% NaN3. Wells were incubated with serum diluted 1:200 for 2 hours at room temperature. The plates were then washed and incubated with alkaline phosphatase conjugated anti-human polyclonal immunoglobulins (Sigma BioSciences, St Louis, MO, USA) diluted 1:40 000 for 2 hours at room temperature. For the colour reaction, the washed wells were incubated with p-nitrophenyl phosphate substrate solution and analysed by measuring the optical density (OD) at 405 nm. Results were analysed using the two tailed Student’s t test. A p value of 0.05 or less was considered significant. Levels of anti-HSC71 antibody were significantly raised in patients with VKH (p=0.00065) compared with healthy volunteers (Fig 1).

Molecular analysis of specificity of anti-HSC71 antibody of VKH patient serum was done by western blot analysis as described previously.1 The antisera from VKH patients

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Several investigators have previously reported retinal and choroidal damage following argon or Nd:YAG laser peripheral iridotomy. This included peripheral retinal and foveal photocoagulation, choroidal and retinal detachment, and cystoid macular oedema and unexplained visual loss. Most of the complications were attributed to radiant laser energy rather than mechanical stress. In our patient, this is a remote possibility in view of the good laser focus on the targeted iris tissue through the Abraham lens. A likely explanation is a rebleeding of fragile NVD secondary to the posterior propagation of shock waves. This mechanism has been contemplated to explain retinal complications after posterior capsulotomy as well as after excimer laser photorefractive keratotomy. Experimentally, a pressure wave of 9–16 bar (130–230 psi) has been measured by Vogel et al. 18 mm from the focal point of a Q switched Nd:YAG laser using a pulse energy of 5 mJ. Changes in intraocular pressure during the procedure or while removing the contact lens is another possible explanation although unlikely, the patient, already experienced with laser procedures, was very cooperative. The iridotomy was technically easy and performed with particular care since this was her only eye.

The Nd:YAG laser is used in the management of premacular haemorrhages. In our patient, performing the PI with the same laser was the cause of a single. Regardless of the mechanism(s) involved, the procedure should be performed with special care particularly in predisposed patients such as those with proliferative retinopathy and/or with a recent bleed. Using the least amount of energy is an obvious precaution. Pretreatment of the anterior segment with the argon laser in a photocoagulative mode to stretch or thin the iris bed might also be helpful in lowering the Nd:YAG laser energy required.

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OBJECTIVE—Various vascular and haematological disorders can cause a subhyaloid premacular haemorrhage leading to decrease in vision. So far, no treatment for subhyaloid haemorrhage has not been recognised as a complication of peripheral laser iridotomy (PI). The following is a case report of such an occurrence with consideration of possible mechanisms.

CASE REPORT
A 58 year old diabetic woman complained of sudden reduction in vision immediately after a prophylactic laser peripheral iridotomy to her left eye. Fundus examination revealed a dense premacular haemorrhage. The iridotomy was performed through an Abraham lens using a Coherent 7970 laser. Ten shots were needed with energy of 3.5–5 mJ.

The patient had proliferative diabetic retinopathy for which she underwent extensive peripheral retinal photocoagulation with residual neovascularisation at the disc (NVD). Several weeks before her current event, she presented with a spontaneous premacular haemorrhage following a Valsalva manoeuvre. She refused any surgical or laser intervention and the haemorrhage had completely resolved at the time of the PI.

COMMENT
The Nd:YAG laser creates a PI by photodisruption of tissue. Three mechanisms are involved in this process: (1) very high irradiances strip electrons from atoms creating a gaseous state of electrons and ions called "plasma"; (2) the plasma expands rapidly outward creating shock and acoustic waves that mechanically disrupt adjacent tissue; and (3) latent stress in the tissues causes additional disruption when the laser makes an incision. The peak pressure in the shock wave associated with plasma formation may exceed 1000 atm. A peripheral laser iridotomy could be helpful in lowering the Nd:YAG laser energy required.
Bilateral endogenous bacterial endophthalmitis associated with pyogenic hepatic abscesses

EDITOR—Endogenous or metastatic bacterial endophthalmitis is rare, with a prevalence of approximately 2–8% of all cases of endophthalmitis. 1 Endogenous bacterial endophthalmitis is associated with chronic diseases such as diabetes mellitus and renal failure, invasive medical procedures, and non-ocular surgery, injecting drug abuse, or prolonged placements of central venous lines. 2 Gram positive bacteria are the most common causative organisms of endogenous bacterial endophthalmitis. 3

A small number of cases of endogenous bacterial endophthalmitis due to Klebsiella pneumoniae, a gram negative organism, have previously been reported, with the majority of the cases originating in Taiwan. 4–6 K pneumoniae endophthalmitis is associated with diabetes mellitus and hepatic abscesses, can be bilateral, and is also associated with a poor visual outcome. 4–6 We report the case of a Taiwanese seaman who developed bilateral endogenous bacterial endophthalmitis after presenting with a pyogenic hepatic abscess.

CASE REPORT

A 40 year old male surgical inpatient was reviewed after he complained of a 3 day history of bilateral painful red eyes and a right hepatic lobe abscess measuring 3.6 cm 7.5 cm. This had been treated by open drainage of the abscess followed by peritoneal lavage and intravenous gentamicin 200 µg and a subconjunctival injection of ceftazidime 750 mg twice daily, oral prednisolone 60 mg daily, and dexamethasone hourly plus atropine 1% twice daily. No organisms were isolated from the pus samples and blood cultures grew K pneumoniae sensitive to these antibiotics and the patient’s general condition had improved by the time the ophthalmic review was requested.

Initial best corrected visual acuity (BCVA) was 3/60 in both eyes, while examination revealed bilateral periorbital erythema and oedema with marked conjunctival chemosis and injection. This was accompanied by bilateral severe anterior uveitis (cells 4 plus, flare 4 plus) and bilateral posterior synechiae formation. Funduscopy demonstrated bilateral severe vitritis (plus 4 cells) with temporally located, white choroidal infiltrates associated with exudation corresponding to subretinal abscesses (Fig 1A). Bilateral vitreous aspirates were performed with an intravitreal injection of gentamicin 200 µg and a subconjunctival injection of cefuroxime 200 mg and gentamicin 80 mg. A second dose of these antibiotics was subconjunctivally administered 24 hours later. No organisms were isolated from the vitreous aspirates. The previous intraocular antibiotic regimen was continued to which intravenous ciprofloxacin 400 mg twice daily and hydrocortisone 100 mg three times daily were added. The anterior uveitis was treated with topical gentamicin, cefazidime, and dexamethasone hourly plus atropine 1% twice daily.

After 16 days the BCVA increased to 6/36 and 6/24 in the right and left eyes. The periorbital erythema and oedema improved, while slit lamp examination demonstrated reduced anterior chamber activity (2 plus cells and 1 plus flare bilaterally). There were no residual posterior synechiae. Fundus examination demonstrated reduction of the vitritis (2 plus cells) and the areas of subretinal abscesses had decreased leaving a surrounding area of RPE atrophy (Fig 1B). Medications included oral ciprofloxacin 750 mg twice daily, oral prednisolone 60 mg daily, topical gentamicin and cefazidime six times a day, dexamethasone 2 hourly, and atropine 1% twice daily.

Just before transfer home to Taiwan, the patient developed a right retinal detachment involving the macula (Fig 2). Two vitrectomy procedures involving retinopexy, encircling, and ultimately silicone oil were subsequently performed in Taiwan. The patient’s left retina also detached and required a trans pars plana vitrectomy with retinopexy, which resulted in successful reattachment of the retina. At 4 month follow up the BCVA is 6/12 in the right and left eyes respectively, the right retina is redetached but the left retina remains flat.

COMMENT

Although rare, most cases of endogenous bacterial endophthalmitis are caused by Gram positive bacteria in patients with existing illness or injecting drug abuse. 7 A number of cases caused by the Gram negative organism K pneumoniae have been reported, mainly from Taiwan. 4–6 K pneumoniae is the leading cause of pyogenic liver abscess in Taiwan and patients with K pneumoniae endophthalmitis secondary to hepatic abscess are more likely to have diabetes mellitus. 8–10 However, like this patient, not all patients have diabetes and the role of unrecognised host or environmental factors leading to this unique association between pyogenic liver abscess and endophthalmitis is unclear. 8–10

Typically the clinical symptoms of K pneumoniae endophthalmitis occur 2–3 days after drainage of a pyogenic liver abscess but before bacterial culture results and antibiotic sensitivities are available. 11–13 Systemic antibiotics are more valuable in endogenous rather than traumatic or postoperative endophthalmitis, probably due to breakdown of the blood-ocular barrier at the site of ocular seeding. 14 The choice of antibiotic reflects bacterial sensitivity results and while an increasing prevalence of K pneumoniae infection previously isolated in Taiwan are resistant only to ampicillin and sulbactam. 15

The benefit of intravitreal antibiotics in endogenous endophthalmitis is unproved but potential benefits outweighed the risks in our patient who had bilateral disease. 15–17 Successive drainage of a subretinal abscess has been documented, but up to three quarters of retinal detachments in all types of endophthalmitis due to virulent organisms remain detached despite surgery. 18 Visual prognosis is poor in K pneumoniae endophthalmitis with 90% of reported eyes having visual outcomes of counting fingers or worse. 11–13 In view of this, a high index of suspicion with prompt diagnosis and aggressive treatment is important, particularly in the 25% of patients who have bilateral disease. 15–17

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Sustained remission of CMV retinitis in HIV-2 disease

EDITOR,—The only established risk factor for the development and progression of cytomegalovirus retinitis (CMVR) in individuals with HIV disease is a low CD4+ T lymphocyte count. The initiation of highly active antiretroviral treatment (HAART) has brought a significant change in the clinical outcome of CMV retinitis with studies reporting longer times to CMV relapse. These findings correlate with increased CD4+ T lymphocyte counts and reduced HIV viral loads.

We report the case of an HIV-2 positive patient who showed no reactivation of CMV retinitis for 25 months despite a low CD4+ T lymphocyte count and a lack of response to HAART treatment. The role of HIV-2 seropositivity in our case is assessed and other relevant factors discussed.

CASE REPORT

A 47 year old black African male was diagnosed with AIDS (HIV-2 positive) in September 1996. The AIDS defining illness was enteric non-Hodgkin’s lymphoma. He was given saquinavir, stavudine (d4T), and zalcitabine (DDC). Owing to his persistently low CD4+ T lymphocyte count (30 cells x10⁹/l) he was referred for ophthalmic evaluation in August 1997. He was diagnosed with peripheral zone III inactive CMV retinitis in his left eye (Fig 1) and was started on maintenance treatment with oral ganciclovir (3 g per day).

Three weeks later this area of retinitis reactivated. He was then given intravenous ganciclovir 5 mg/kg twice daily for 4 weeks and then restarted on oral ganciclovir. His CD4+ T lymphocytes remained low with a peak reaching 70 cells x10⁹/l in January 1998. In April 1998 antiretroviral treatment was discontinued because he showed no response to therapy.

The patient underwent frequent ophthalmic reviews during which inactive CMV retinitis was recorded on funduscopic and photographic assessment. No new lesions have been recorded in either eye. His current CD4+ T lymphocyte count is 7 cells x10⁹/l and his visual acuity is 6/5 in each eye.

COMMENT

In a group of patients not responding to HAART therapy Mitchell et al. showed a median time to CMVR progression of 18 days (95% CI: 8.91). Walker and Popescu’ showed a median time to CMVR reactivation of 122 days (95% CI: 93-186) in patients receiving oral ganciclovir as maintenance treatment.

This remission period far exceeds the time for CD4+ T lymphocyte counts, further suggesting that the immune response to CMV is not solely related to the CD4+ T lymphocyte count.

We believe that HIV-2 seropositivity in this case is a relevant factor in modifying the natural history of CMVR. HIV-2 is biologically similar to HIV-1 but it has a reduced virulence. Thus, our knowledge comparing ocular lesions between HIV-1 and HIV-2 infected individuals is by Monteiro-Grillo et al. They reported less severe complications in patients infected by HIV-2.

We also report the case of two patients with CMVR whose peripheral zone III retinitis remitted after 30 and 94 days, respectively (95% CI: 93-186) in patients receiving standard therapy.

More studies with HIV-2 infected individuals are required to monitor the clinical course of CMVR in this group of patients and clarify whether HIV-2 virulence is an important factor modifying the immune response to CMV.

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Complete regression of retinal neovascularisation after therapy with interferon alfa in Behcet’s disease

EDITOR,—One of the main problems in ocular Behcet’s disease (BD) is severe posterior uveitis with retinal vessel occlusion and secondary ischaemic changes leading to retinal neovascularisation and to bad prognosis despite immunosuppression.

Previously we have shown the efficacy of interferon alfa (IFNo)-2a in posterior uveitis and especially in retinal vascities with reopens of occluded vessels. Now we demonstrate the antangiogenic effect of IFNo-2a in one BD patient with retinal neovascularisations.

The IFNo-2a treatment has resulted in complete remission of the retinal neovascularisation, without laser coagulation of non-perfusion areas, which would have been the standard therapy.

CASE REPORT

A 27 year old man with recurrent oral aphthosis, pustular skin lesions, epididymitis, arthritus (elbow, sacroiliac joint), and retinal vasculitis of the left eye was diagnosed in October 1998 as having Behcet’s disease (BD), according to the criteria of the international study group. At this time therapy with cyclosporin A (3 mg/kg body weight) was initiated. Until June 1999 he had had no relapses. One month later the visual acuity in the right eye decreased to 20/100, but was stable in the left eye (20/600). Biometrically there was no inflammation of the anterior chamber.

Fuduscopy of both eyes revealed vitreous infiltra- tion and a macular oedema with glotic changes and central sanguinisation. In addition, optic disc neovascularisation in the right eye (Fig 1) and optic disc oedema in the left eye were present. Perivascular leakage as a sign of active vasculitis was shown by fluorescein angiography. At the same time oral aphthosis and pustular skin lesions recurred.

Owing to active retinal vasculitis with marked decrease of visual acuity we stopped the cyclosporin A therapy and on the next day started IFNo-2a therapy with 6 million units/day subcutaneously. Six days later the visual acuity rose to 20/40 in the right eye and 20/300 in the left eye. Fuduscopy revealed a decrease of vitreous inflammation, macular oedema, and retinal haemorrhages in both eyes; the optic disc oedema in the left eye and the optic disc neovascularisation in the right eye regressed. Seventeen days after initiating IFNo-2a treatment the ophthalmological exammination disclosed a stable visual acuity in the right eye and an increased visual acuity of 20/200 in the left eye because of irreversible macular defects. The retinal vasculitis had improved in both eyes; additionally, optic disc neovascularisation in the right eye had completely disappeared. Another 5 weeks later there was further improvement with a visual acuity of 20/25 in the right eye (Fig 2) and 20/100 in the left eye.

Figure 1 Fluorescein angiography of right eye before starting IFNo-2a therapy. Optic disc neovascularisation is seen among the superior artery branch. Visual acuity 20/100.

Figure 2 Fluorescein angiography of right eye 7 weeks after beginning IFNo-2a treatment. Optic disc neovascularisation had completely disappeared. Visual acuity 20/25.
An unusual case of asthenopia: “pseudo-accommodative insufficiency” associated with a high AC:A ratio

EDITOR,—Asthenopia is characterised by ocular fatigue, frontal headache, and blurred vision, particularly during periods of sustained close work. It can be caused by accommodative insufficiency, a condition in which the effort required to maintain accommodation for near objects produces troublesome symptoms. This may be associated with a low accommodative convergence: accommodation (AC:A) ratio, which the patient has to overcome using positive fusion reserves. When fusion is insufficient symptoms of asthenopia can occur. In contrast, a high AC:A ratio would not normally be associated with asthenopia, but rather with overconvergence, potentially resulting in a convergence excess type of esotropia. We report two cases in which “pseudo-accommodative insufficiency” was identified as the cause of asthenopia and paradoxically associated with a high AC:A ratio.

CASE REPORTS

Case 1

A 12 year old male patient was referred complaining of difficulty with reading. His visual acuities were 6/6, N4.5 in the right eye, and 6/5, N4.5 in the left eye. Cycloglegic refraction showed no significant refractive error. A cover test revealed a 2 prism dioptre exophoria both for distance and near, and he had a full range of ocular motility. Convergence as measured using the RAF rule was well maintained to 6 cm, but accommodative amplitude for an N5 target was only 9 dioptres. It was noted at this stage that when accommodation failed a right esotropia developed transiently. The negative fusional vergence was 4 prism dioptries base-in for near, and 4 prism dioptries base-in for distance. Positive fusional vergence was 20 prism dioptries base-out for near and 14 prism dioptries base-out for distance. The AC:A ratio, measured using the distance gradient method, was 12:1. He was treated with exercises to build up negative fusional vergence.

COMMENT

We believe that we have identified a cause of asthenopia, which paradoxically, is associated with a high AC:A ratio. Although both patients had signs and symptoms, which initially were suggestive of accommodative insufficiency, the sudden transient esotropia that was observed while testing accommodation, together with the high AC:A ratio, indicates that this diagnosis was incorrect. We speculate that these patients choose to relax their accommodation in order to maintain binocular single vision, but at the expense of clarity of vision for near work. If they continued to accommodate their high AC:A ratio resulted in a greater angle of esophoria, and when their negative fusional vergence reserves were no longer sufficient to compensate then a manifest deviation developed. We have coined the term “pseudo-accommodative insufficiency” to describe this phenomenon.

When assessing patients with asthenopic symptoms it is important to distinguish between true accommodative insufficiency and “pseudo-accommodative insufficiency”. Treatment for the former using convex lenses would be inappropriate for the latter, as it does not address the underlying cause of the problem. Instead, management should be aimed at augmenting negative fusion reserves and negative relative vergence.

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