LETTERS TO THE EDITOR

Haemodynamic changes in ocular Behçet’s disease

EDITOR,—Behçet’s disease (BD) is an immune system-related obstructive vasculitis characterized by recurrent inflammation that affects multiple systems. Posterior uveitis and retinal vasculitis are the common features of ocular involvement. Colour Doppler imaging (CDI) is a non-invasive ultrasonographic method which permits simultaneous grey scale imaging of structure and colour coded imaging of blood velocity. CDI has successfully demonstrated changes in orbital haemodynamics associated with a variety of pathological conditions. In the present study the haemodynamic changes in ophthalmic (OA), central retinal (CRA), and posterior ciliary arteries (PCA) of the patients with BD were investigated using CDI.

CASE REPORT
The study group consisted of 26 patients with BD (five females, 21 males), aged between 22 and 58 (mean 32.19 (SD 8.27)). Diagnosis was made according to the criteria recommended by the International Study Group for Behçet’s disease. Control group consisted of 20 males, six females, aged between 21 and 58 (31.49 (8.60)), who had no ocular or systemic pathology other than presbyopia.

After performing full ophthalmological examination, transocular CDI was performed in all the patients and controls with a colour Doppler sonographic unit (General Electric Sonochrome 625L, Les Moulineaux, France) with a 7.5 MHz linear transducer. CDI was performed to both eyes of Behçet’s patients and left eyes of the controls. Activity status of the patients was evaluated according to the ocular findings. During CDI, eyes of Behçet’s patients who had cells in anterior chamber, cells in the vitreous, macular oedema, papillary oedema, retinal vasculitis, and exudation in pars plana or retina were encountered as inactive. The left eye of a single patient who had no ocular or systemic pathology other than presbyopia. Diagnosis of the inflammatory process for small arteries in BD.

Findings are yet to be determined.

TABLE 1  Comparison of flow velocities and indexes (SD) in OA, CRA, and PCA in left eyes of Behçet’s patients irrespective of activity status and controls

<table>
<thead>
<tr>
<th></th>
<th>Left eye (n=26)</th>
<th>Control eye (n=26)</th>
<th>p Value</th>
</tr>
</thead>
<tbody>
<tr>
<td>OA</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>PSV</td>
<td>33.458 (9.46)</td>
<td>34.400 (7.94)</td>
<td>0.8101</td>
</tr>
<tr>
<td>EDV</td>
<td>9.125 (3.44)</td>
<td>8.680 (2.57)</td>
<td>0.6437</td>
</tr>
<tr>
<td>AFV</td>
<td>14.208 (4.35)</td>
<td>15.400 (4.49)</td>
<td>0.350</td>
</tr>
<tr>
<td>RI</td>
<td>0.727 (0.067)</td>
<td>0.743 (0.06)</td>
<td>0.3726</td>
</tr>
<tr>
<td>PI</td>
<td>1.782 (0.495)</td>
<td>1.716 (0.395)</td>
<td>0.7039</td>
</tr>
<tr>
<td>CRA</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>PSV</td>
<td>7.600 (2.26)</td>
<td>8.000 (1.658)</td>
<td>0.4250</td>
</tr>
<tr>
<td>EDV</td>
<td>2.320 (1.069)</td>
<td>2.800 (0.913)</td>
<td>0.0242</td>
</tr>
<tr>
<td>AFV</td>
<td>3.880 (1.269)</td>
<td>4.600 (1.354)</td>
<td>0.0531</td>
</tr>
<tr>
<td>RI</td>
<td>0.704 (0.108)</td>
<td>0.646 (0.087)</td>
<td>0.0394</td>
</tr>
<tr>
<td>PI</td>
<td>1.420 (0.523)</td>
<td>1.177 (0.374)</td>
<td>0.0575</td>
</tr>
<tr>
<td>PCA</td>
<td></td>
<td></td>
<td></td>
</tr>
<tr>
<td>PSV</td>
<td>8.625 (2.46)</td>
<td>11.880 (1.74)</td>
<td>0.000</td>
</tr>
<tr>
<td>EDV</td>
<td>2.458 (1.14)</td>
<td>4.20 (1.90)</td>
<td>0.000</td>
</tr>
<tr>
<td>AFV</td>
<td>4.304 (1.43)</td>
<td>6.920 (1.56)</td>
<td>0.000</td>
</tr>
<tr>
<td>RI</td>
<td>0.715 (0.124)</td>
<td>0.644 (0.092)</td>
<td>0.0209</td>
</tr>
<tr>
<td>PI</td>
<td>1.527 (0.364)</td>
<td>1.1980 (0.432)</td>
<td>0.0324</td>
</tr>
</tbody>
</table>

The insignificant difference in the flow velocities of OA between the patients and the controls might be due to the predilection of the inflammatory process for small arteries in BD.

In conclusion, CDI is a useful technique in evaluating orbital haemodynamic changes in BD non-invasively, and the quantitative findings could help to determine the ultimate degree of ocular damage and visual outcome; however, the clinical significance of our findings are yet to be determined.

COMMENT
In the present study, no significant difference in the flow velocities of active and inactive eyes of Behçet’s patients was detected. However, there was a significant reduction in EDV and AFV of PCA in eyes of Behçet’s patients regardless of the activation, than the controls. This shows that haemodynamic changes occur in BD which are not always correlated with clinical presentation. CDI was postulated that CRA involvement plays an important role in the pathogenesis of BD, as it is shown that inner retinal layers are the main region affected in BD. However, the results of our study showed that choroidal circulation is also affected by vasculitis although it is rarely detected clinically. In a previous study carried out in Behçet’s patients, PSV and EDV of PCA of the patients with ocular involvement was found to be significantly reduced compared with those without ocular involvement and the controls. A preliminary report on CDI in BD revealed that PSV of CRA and CRV were significantly lower in patients with ocular involvement irrespective of the activation compared with the cases without ocular involvement. This might be the result of occlusive vasculitis seen in retinal vessels in BD. However, no difference was detected in flow velocities in PCA between the patients and controls, which is not in accordance with our findings. The insignificant difference in the flow velocities of OA between the patients and the controls might be due to the predilection of the inflammatory process for small arteries in BD.

In conclusion, CDI is a useful technique in evaluating orbital haemodynamic changes in BD non-invasively, and the quantitative findings could help to determine the ultimate degree of ocular damage and visual outcome; however, the clinical significance of our findings are yet to be determined.
Colour Doppler imaging in Takayasu’s arteritis

EDITOR,—Takayasu’s arteritis (TA) is a systemic necrotising vasculitis of unknown aetiology whose characteristic ocular manifestations relate to ischaemia. Orbital Colour Doppler imaging (CDI) is useful to evaluate haemodynamics of the orbital circulation and its abnormalities.1 We report the ophthalmic artery blood flow in two patients with TA using CDI.

CASE REPORT

Case 1

A 34 year old man, previously diagnosed with TA was referred for ophthalmic evaluation. He had absence of radial artery pulses, unrecordable upper extremity blood pressure measurements, lower extremity hypertension, arteriographic evidence of total left brachiocephalic artery occlusion, right brachiocephalic and bilateral common carotid artery stenoses, increased erythrocyte sedimentation rate (11 mm in the first hour), and positive C reactive protein. The best corrected Landolt visual acuity was light perception in the right eye. In the left, intraocular pressures (IOP) were 9 mm Hg in the right eye and 12 mm Hg in the left. Left funduscopy revealed venous dilations, microaneurysms, and arteriovenous shunts. The arm-retina time was prolonged with wide peripheral avascular areas. Two years later, left iris rubeosis and episcleral vessel dilatation developed. IOP was 20 mm Hg. CDI showed a marked decrease in the waveform of orbital arteries, thus enabling direct assessment of the orbital vascular supply.2 The peak systolic flow velocity of the ophthalmic artery in TA was markedly reduced as in patients with carotid atheromas. In ocular ischaemic syndrome secondary to carotid atheroma, the PI is reported to be higher than normal. This is thought to be due to the increase in the peripheral vascular resistance secondary to the arteriosclerotic process involving the orbital vessels.3 In contrast, the PI in TA was lower (case 1) or almost normal (case 2) compared with control eyes. Since only large vessels are affected in TA, the peripheral vascular resistance would not be as increased as in carotid atheroma. Moreover, the low PI values in case 1 reflect a decreased peripheral vascular resistance which may be due to the formation of arteriovenous anastomoses in the retina and to the formation of collateral blood flow between the internal and external carotid arteries manifesting as dilated episcleral vessels.4 In case 2, the lack of arteriovenous anastomosis formation could have resulted in the almost normal PI value despite the decrease in total peripheral ocular vascular resistance.

PI measurements by CDI could be useful in the detection of retroorbital vessel flow abnormalities, particularly in the ophthalmic circulation, and in the evaluation of ocular manifestations in TA.

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Table 1 Ophthalmic artery flow velocity and pulsatility index (PI) in patients with Takayasu’s arteritis and a control group

<table>
<thead>
<tr>
<th>Case</th>
<th>Age/sex</th>
<th>Eye</th>
<th>Vmax</th>
<th>Vmin(cm/s)</th>
<th>Vmean</th>
<th>PI</th>
</tr>
</thead>
<tbody>
<tr>
<td>1</td>
<td>34/M</td>
<td>R</td>
<td>8</td>
<td>3</td>
<td>5</td>
<td>1.00</td>
</tr>
<tr>
<td>2</td>
<td>75/F</td>
<td>L</td>
<td>11</td>
<td>6</td>
<td>9</td>
<td>0.55</td>
</tr>
<tr>
<td></td>
<td></td>
<td>L</td>
<td>17</td>
<td>4</td>
<td>10</td>
<td>1.30</td>
</tr>
<tr>
<td>Control*</td>
<td>58.6</td>
<td>+1.0</td>
<td>34.4</td>
<td>9.0</td>
<td>18.0</td>
<td>1.46</td>
</tr>
</tbody>
</table>

*The control group comprises 12 eyes of 12 patients with no ocular or carotid artery disease. The flow velocity and PI values were averaged accordingly.
Obstruction of the superior vena cava

EDITOR,—Obstruction of the superior vena cava (SVCO) is a condition which causes a variety of symptoms and signs in the upper body, such as oedema of the face, arms and trunk on presentation showing plethora and swelling with inability to button his shirt. His symptoms were more marked on swallowing. Only on direct questioning did he admit to facial and neck swelling with inability to button his shirt collar. His symptoms were more marked on waking. He had previously been in good health, with no past medical or ocular history.

CASE REPORT

A 68 year old white man was referred with a 3 month history of "chronic conjunctivitis" not responding to topical medications; at his consultation he complained of persistent watering of both eyes with lid swelling. Only on direct questioning did he admit to facial and neck swelling with inability to button his shirt collar. His symptoms were more marked on waking. He had previously been in good health, with no past medical or ocular history.

COMMENT

Our patient is interesting considering the paucity and seemingly trivial nature of his presenting complaints, the potentially distracting presence of blepharitis, and also the relative lack of ocular findings associated with SVCO. Many patients present with epiphora and puffy eyes. It is as well to remember that these symptoms may be due to serious pathology.

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Paediatric varicella choroiditis

EDITOR,—Unifocal choroiditis occurs in children and adults with primary (chickenpox) and secondary varicella zoster virus (VZV) infection. Current recommendations by the American Academy of Pediatrics do not include the routine use of oral aciclovir for uncomplicated varicella in otherwise healthy children; individual cases, however, may justify a "modest clinical benefit" from oral aciclovir therapy, provided it can be initiated within the first 24 hours of illness. We describe here an otherwise healthy child with chickenpox who developed a unilateral, unifocal choroiditis with overlying serous detachment of the macula. A prompt visual response was observed with oral aciclovir, initiated 5 days after the onset of systemic illness. We suggest, therefore, a role for even delayed systemic antiviral therapy of varicella choroiditis in the immunocompetent patient.

CASE REPORT

An 11 year old girl presented 5 days after the onset of chickenpox, and 1 day after she developed blurring of vision in the left eye. She denied any pain with eye movements or photophobia.

On examination, the patient had diffuse, non-acute varicelliform lesions of the skin with signs of early crusting. Visual acuity was 20/20 in the right eye, and 20/100 in the left. Pupils were equal in size and reactivity, without afferent defects. The anterior segment examination was unremarkable. Fundus examination of the left eye revealed an irregular, 0.75 disc diameter subretinal lesion with an overlying serous detachment involving the macula (Fig 1). There was no evidence of vitritis. Fluorescein angiography showed early blockage, with late hyperfluorescence and pooling of dye in the subretinal space (Fig 2). Fundus examination of the fellow eye was normal.

The patient was started on oral aciclovir 500 mg four times daily for 1 week, based on September 17, 2023 by guest. Protected by copyright.
upon her size and weight. Examination of the involved eye 8 days later revealed visual acuity of 20/40, and regression of her choroidal lesion and sensory detachment. Subtle changes to the retinal pigment epithelium persisted.

COMMENT
Chiorioretinal lesions seen in conjunction with primary and secondary systemic VZV infections are relatively rare, with most cases limited to immunocompromised hosts. Barondes et al have, however, described acute retinal necrosis (ARN) in a healthy man 2 weeks after diffuse varicella eruption, where aciclovir and corticosteroids were associated with a favourable outcome. Kelly and Rosenthal also describe multifocal choroiditis in an otherwise healthy adult with primary VZV infection, where oral aciclovir resulted in regression of lesions.

The patient described here is unusual in that the choroiditis was unilateral, unifocal, and involved the macula. Furthermore, there was a significant response to oral aciclovir despite being administered several days after the acute onset of her exantheme.

We therefore suggest that ophthalmologists consider systemic antiviral therapy in immunocompetent children with chickenpox and choroiditis. As this patient's maculopathy was clinically evident during the convalescent phase of her vesicular lesions, this consideration should still be made independent of the timing of visual symptoms. Furthermore, there was a the acute onset of her exantheme, with a rapid and complete resolution of her symptoms with return of excellent central vision. This occurs in the skin, mucous membranes, and other viscera. Common presentations are epistaxis, dyspnoea on exertion, and gastrointestinal bleeding due to haemorrhage from, or shunting through, abnormal vessels in the nasal mucosa, lung, and the gastrointestinal tract. The most serious consequence of HHT is the potential for paradoxical embolisation and stroke. This occurs when venous thrombus, originating in the leg or pelvis, pass through arteriovenous malformations in the lung and lodge in the brain.

Common ophthalmic features are conjunctival and eyelid telangiectasia. Intraocular involvement is extremely rare. In a study of 20 patients with HHT conjunctival telangiectasia was seen in seven patients and retinal vascular malformation in two. Although retinal lesions seldom cause symptoms, retinal haemorrhages, beading of retinal arteries, branch, and central retinal artery occlusion, vitreous haemorrhage, secondary to areas of retinal neovascularisation, and inner retinal ischaemic atrophy can occur. Spontaneous haemorrhages have been reported in juvenile xanthogranuloma, retrolental fibroplasia, persistent hyaloid, blood dyscrasias, and delayed retinal necrosis (ARN) in a healthy man 2

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Conjunctival infestation of a child with Theromyzon tessulatum

EDITOR—In its normal life cycle, Theromyzon tessulatum (duck leech) enters the nasal cavities of waterfowl to suck blood, staying in its host for hours up to days. Infestation of the human eye is very rare and may be classified as a zoonosis. To our knowledge only one case is described in the literature.

CASE REPORT
A 4 year old girl bathed in a flooded gravel pit located in the western part of Freiburg (southwest Germany) for approximately 15 minutes; occasionally she had put her head

Figure 1 Iris photograph of the right eye showing mulberry-like iris vascular malformation at the 12 o’clock position. The left iris showed a similar appearance.

Spontaneous haemorrhage in hereditary haemorrhagic telangiectasia

EDITOR.—Hereditary haemorrhagic telangiectasia (HHT) is characterised by multiple dilatations of capillaries and venules in the skin, mucous membranes, and viscera that have a tendency to bleed. Although a relatively rare condition, ocular involvement is common in these patients (45-65%). Intracocular involvement is however extremely rare.

We report a patient with HHT with spontaneous bilateral haemorrhages

CASE REPORT
A 69 year old man presented with sudden onset of blurred vision in the left eye associated with a “red streak” under his left exantheme, which may benefit from ophthalmic examination and treatment.

angiectasia (HHT) is characterised by multiple lesions with a favourable outcome.
CASE REPORT

A 59 year old man with no history of ocular disease came to our hospital as an emergency patient for treatment of a corneal burn. The patient was working at his desk when he quickly turned his head and his eye accidentally contacted a hot incandescent bulb. His chief complaint was decreased vision without pain. On the first day, his left corrected visual acuity was decreased to finger counting. By slit lamp biomicroscopic examination, the eye did not show any inflammation or apparent reflex tearing; there was, however, an oval whitish nectrotised area approximately 6 mm in diameter on the superficial layer of the central corneal epithelium (Fig 1). This lesion was stained with fluorescein and rose bengal; the remaining portion of the ocular surface epithelial integrity was intact without fluorescein or rose bengal staining. A previously reported system, which can be used for the screening and the evaluation of the severity of dry eye and is now commercially available in Japan (DR-1, Kowa Co Ltd, Tokyo, Japan), was used for further observing the tear film in this case. The system disclosed that, outside the lesion, the tear film was stable and the precorneal tear film lipid layer moved easily with blinking (Fig 2B) and this pattern corresponded to that in normal healthy eyes. The corrected visual acuity returned to 20/20.

COMMENT

The present case clearly demonstrated that tear film stability was completely lost, when the corneal surface epithelium was damaged.

Figure 1 Oval whitish nectrotised lesion on the central corneal epithelium.

Figure 2 (A) Tear appearance on the first day of visit. Inside the lesion, tear film stability was lost and was easily disrupted showing break up on the damaged epithelium; outside the lesion, tear film stability was maintained and the tear film lipid layer moved easily with blinking (star). (B) Tear appearance returned completely to normal on the 4th day after the first visit. A circular area 2 mm in diameter (×100) was observed at the upper nasal portion of the central cornea (A), and in the central corneal portion (B).

Direct observation of tear film stability on a damaged corneal epithelium

EDITOR.—Wettability of the corneal epithelial surface and properties of tear fluids have been regarded as principal factors in the determination of precorneal tear film stability. However, there is no direct evidence indicating which factor is the key to this stability. The role of wettability of corneal epithelial surface in tear film stability can be elucidated in cases where damage occurs only to the corneal epithelium, not to the tear fluid itself. Therefore, we report an unusual case with sudden burning of the superficial layer of the corneal epithelium which clearly shows the relation between corneal surface epithelial integrity and tear film stability.

CASE REPORT

A 59 year old man with no history of ocular disease came to our hospital as an emergency patient for treatment of a corneal burn. The patient was working at his desk when he quickly turned his head and his eye accidentally contacted a hot incandescent bulb. His chief complaint was decreased vision without pain. On the first day, his left corrected visual acuity was decreased to finger counting.

By slit lamp biomicroscopic examination, the eye did not show any inflammation or apparent reflex tearing; there was, however, an oval whitish nectrotised area approximately 6 mm in diameter on the superficial layer of the central corneal epithelium (Fig 1). This lesion was stained with fluorescein and rose bengal; the remaining portion of the ocular surface epithelial integrity was intact without fluorescein or rose bengal staining.

A previously reported system, which can be used for the screening and the evaluation of the severity of dry eye and is now commercially available in Japan (DR-1, Kowa Co Ltd, Tokyo, Japan), was used for further observing the tear film in this case. The system disclosed that, outside the lesion, the tear film was stable and the precorneal tear film lipid layer moved easily with blinking. However, the tear film on the lesion was discontinuous and the lesion did not change in appearance with blinking (Fig 2A). A combination of 0.1% fluorometholone, 0.3% oxyloxacin, and preservative-free artificial tears was applied to the eye four times a day. By 4 days after the first visit, the corneal epithelium in the burned region had recovered completely to normal without any resultant epithelial abnormalities. Thus, it is likely that only the superficial layer was involved in the lesion. The appearance of tear lipid layer interference returned to show a greyish colour with striped pattern in conjunction with blinking (Fig 2B) and this pattern, corresponded to that in normal healthy eyes. The corrected visual acuity returned to 20/20.

COMMENT

The present case clearly demonstrated that tear film stability was completely lost, when the corneal surface epithelium was damaged.

Figure 1 Oval whitish nectrotised lesion on the central corneal epithelium.

Figure 2 (A) Tear appearance on the first day of visit. Inside the lesion, tear film stability was lost and was easily disrupted showing break up on the damaged epithelium; outside the lesion, tear film stability was maintained and the tear film lipid layer moved easily with blinking (star). (B) Tear appearance returned completely to normal on the 4th day after the first visit. A circular area 2 mm in diameter (×100) was observed at the upper nasal portion of the central cornea (A), and in the central corneal portion (B).
In contrast, outside the lesion, the tear film was normal in appearance and seemed to have an intimate relation with the corneal epithelium. In this case, as it is reasonable to presume that there was no continuous damage to the tear film itself, this observation suggests that tear film stability may be supported much more by corneal surface epithelial integrity (wettability) than by the properties of tear themselves. Factors which contribute to this mechanism probably include intact expression of glycoalyx and the recently demonstrated MUC 1 (epithelial cell derived mucin). Given these observations, it is reasonable to postulate that various corneal epithelial diseases may be accompanied by some tear film instability due to superficial epithelial abnormalities. Thus, in such cases, therapy should be targeted to treat the loss of tear film interaction with the epithelium.

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Pharmacological reversal of ptosis in a patient with acquired Horner’s syndrome and heterochromia

EDITOR,—We present a patient who possesses two aspects of Horner’s syndrome worth re-emphasising: (1) heterochromia iridis with Horner’s syndrome does not always signify a congenital origin,1 2 and (2) sympathomimetics, such as 2.5% phenylephrine, may be used to elevate the lid when desired as a measure to defer surgery.**

CASE REPORT

A nurse brought her 7½ year old daughter for evaluation of the right pupil, which was noted to be larger than that on the left in the dark. Review of systems and past medical history were unremarkable. An isolated deficiency of the dilator muscle in her left pupil without heterochromia iridis or ptosis was diagnosed. Two years later, mild ptosis and mild heterochromia were present. Because of the progression to a Horner’s syndrome, now also demonstrating left facial anhidrosis and acquired heterochromia, magnetic resonance imaging (MRI) of the neck and chest was performed. A large left apical lung ganglioneuroma was diagnosed. This was surgically excised and the patient had an uncomplicated recovery. In response to concerns regarding her asymmetric pupillary sizes, 2.5% phenylephrine was prescribed for use up to four times a day to help equalise her pupils when going on photographic shoots for modelling purposes.

Three years later, at age 12½, the patient shows marked heterochromia. She uses the 2.5% phenylephrine solution, but describes its true value to be for the near immediate relief of her ptosis after placing one drop in her affected eye, and two drops in the contralateral eye (Fig 1). She explains that contralateral drop placement serves to improve and maintain symmetry in the two lid fissures and that the effect lasts approximately 15 minutes with pupillary dilation lagging 10–15 minutes behind.

COMMENT

Even in an otherwise asymptomatic child, the presence of heterochromia iridis with Horner’s syndrome can be the result of an undetected, progressive condition, and does not establish a definite congenital aetiology. Our patient appears to be a well adjusted and happy preteen, who, given a sense of control over her ptotic lid with topical drops, freely declines surgical correction for her ptosis at the moment. Coloured contact lenses were proposed as an option to mask her acquired heterochromia. The best effect of these lenses was obtained when matched to the colour of her darker iris, although the patient’s initial desire had been to match the colour of the eye affected by Horner’s syndrome.

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