LETTERS TO THE EDITOR

Haemodynamic changes in ocular Behçet's disease

EDITOR,—Behçet's disease (BD) is an immune system related obstructive vasculitis characterised by recurrent inflammation that affects multiple systems. Posterior uveitis and retinal vasculitis are the common features of ocular involvement.¹⁻³ 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.⁴⁻⁷ 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 by 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 International Study Group for Behçet's disease.⁸ 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 evaluation, 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 active, and the ones without any activity but with permanent damage were encountered as inactive. Peak systolic velocity (PSV), end diastolic velocity (EDV), and average flow velocities (AFV) were calculated by using flow spectrums. Resistivity (RI) and pulsatility indexes (PI) were calculated by using flow

Table 2 Comparison of flow velocities and indexes (SD) in OA, CRA, and PCA in right eyes of Behçet's patients irrespective of activity status and controls

	Right eye (n=26)	Control eye (n=26)	p Value
OA			
PSV	33.560 (9.99)	34.400 (7.937)	0.869
EDV	9.5200 (3.980)	8.680 (2.765)	0.5011
AFV	14.600 (4.933)	15.400 (4.491)	0.6129
RI	0.7200 (0.079)	0.7428 (0.06)	0.3311
PI	1.8132 (0.471)	1.7160 (0.395)	0.4469
CRA			
PSV	7.2308 (1.796)	8.000 (1.658)	0.0959
EDV	2.2400 (1.165)	2.8000 (0.913)	0.0221
AFV	3.7692 (1.107)	4.6000 (1.354)	0.0190
RI	0.7281 (0.139)	0.6464 (0.087)	0.0546
PI	1.4758 (0.489)	1.1772 (0.374)	0.0182
PCA			
PSV	8.6923 (2.363)	11.8800 (1.740)	0.0000
EDV	2.8846 (1.177)	4.2000 (1.190)	0.000
AFV	4.5769 (1.554)	6.9200 (1.552)	0.0000
RI	0.6835 (0.111)	0.6444 (0.092)	0.2302
PI	1.4108 (0.672)	1.1980 (0.432)	0.1507

velocities. Statistical analysis was performed in three stages by using Mann-Whitney U test for independent groups and Wilcoxon signed rank sum test for paired data. p Values were taken from Mann-Whitney U test.

During CDI 21 eyes were active and 30 eyes were inactive. The left eye of a single patient was phthisical and it was encountered as inactive. Results were analysed in three stages. In the first stage, the flow velocities and indexes in right and left eyes of the patients were compared regarding the activity status during CDI. No statistically significant difference was detected in the flow velocities and indexes of three arteries in any of the groups. In stage two, only flow velocities and indexes of OA, CRA, and PCA in left eyes of the patients were compared with the controls. There was a statistically significant reduction in PSV, EDV, and AFV of PCA in both inactive and active eyes compared with controls. However, the increase in RI and PI of PCA in patients compared with controls was short of statistical significance. In the third stage flow velocities and indexes of three arteries in all left eyes and right eyes of the patients were compared with control eyes irrespective of the activity status (Tables 1 and 2). In all patients there was a statistically significant reduction in EDV and AFV, and a significant increase in RI and PI of

CRA than controls. Also the reduction of flow velocities of PCA in patients was statistically significant. The increase in RI and PI of PCA in left eyes was significant; however, in right eyes it was fell short of significance.

COMMENT

In the present study, no significant difference in between 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 and CRA 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. It could be 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.⁹ 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.¹⁰ 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.

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

	Left eye (n=26)	Control eye (n=26)	p Value
OA			
PSV	33.458 (9.46)	34.400 (7.94)	0.8101
EDV	9.125 (3.44)	8.680 (2.77)	0.6437
AFV	14.208 (4.35)	15.400 (4.49)	0.350
RI	0.727 (0.067)	0.743 (0.06)	0.3726
PI	1.782 (0.495)	1.716 (0.395)	0.7039
CRA			
PSV	7.600 (2.26)	8.000 (1.658)	0.4250
EDV	2.320 (1.069)	2.800 (0.913)	0.0242
AFV	3.880 (1.269)	4.600 (1.354)	0.0531
RI	0.704 (0.108)	0.646 (0.087)	0.0394
PI	1.420 (0.523)	1.177 (0.374)	0.0575
PCA			
PSV	8.625 (2.46)	11.880 (1.74)	0.000
EDV	2.458 (1.14)	4.20 (1.190)	0.000
AFV	4.304 (1.43)	6.920 (1.56)	0.000
RI	0.715 (0.124)	0.644 (0.092)	0.0209
PI	1.527 (0.564)	1.1980 (0.432)	0.0324

The authors wish to thank Professor Refik Burgut for advice on the statistical methods and assistance with the data analysis.

MERİH SOYLU
ISMAL ISIGÜZEL
NIHAL DEMİRCAN
MÜSLİME YALAZ

Department of Ophthalmology,
Çukurova University Medical School

KAIRQUELY S AIKIMBAEV

Department of Radiology,
Çukurova University Medical School

Correspondence to: Merih Söylü, MD, Department of Ophthalmology, Çukurova University Medical School, Balçalı, Adana, Turkey.

Accepted for publication 11 March 1998

- 1 Namba K. Ocular complication and prognosis in Behçet's disease. *Asia-Pacific J Ophthalmol* 1991;3:12-4.
- 2 Atmaca LS. Fundus changes associated with Behçet's disease. *Graefes Arch Clin Exp Ophthalmol* 1989;27:340-4.
- 3 Nussenblatt RB, Palestine AG, eds. Behçet's disease and other vasculitides. In: *Uveitis: fundamentals and clinical practice*. Chicago: Year Book Medical Publishers, 1989:212-32.
- 4 Keyser BJ, Flaharty PM, Sergott RC, et al. Color Doppler imaging arterial blood flow in central retinal vein occlusion. *Ophthalmology* 1994;101:1357-61.
- 5 Regillo CD, Sergott RC, Ho AC, et al. Hemodynamic alterations in the acute retinal necrosis syndrome. *Ophthalmology* 1993;100:1171-6.
- 6 Williamson TH, Baxter GM. Central retinal vein occlusion, an investigation by color Doppler imaging. Blood velocity characteristics and prediction of iris neovascularization. *Ophthalmology* 1994;101:1362-72.
- 7 Ho AC, Lieb WE, Flaharty PM, et al. Color Doppler imaging of the ocular ischemic syndrome. *Ophthalmology* 1992;99:1453-62.
- 8 The International Study Group for Behçet Disease. Evaluation of diagnostic criteria in Behçet's disease towards internationally agreed criteria. *Br J Rheumatol* 1992;31:299-308.
- 9 Özdemir H, Atilla H, Atilla S, et al. Diagnosis of ocular involvement in Behçet's disease: Value of spectral and color Doppler sonography. *Am J Roentg* 1995;164:1223-7.
- 10 Güven D, Özdemir H, Yilmazbas P, et al. Haemodynamic changes in Behçet's disease. *Retina-Vitreus* 1995;3:177-81.

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 ocular 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 and 0.2 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 dilatations, microaneurysms, and arteriovenous shunts. The arm-retina time was prolonged with wide peripheral avascular

Figure 1 Colour Doppler imaging of the ophthalmic artery in case 1. Note the marked decrease in the systolic peak of the antegrade ophthalmic artery blood flow.

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

Case	Age/sex	Eye	Vmax	Vmin (cm/s)	Vmean	PI
1	34/M	R	8	3	5	1.00
		L	11	6	9	0.55
2	75/F	R	19	4	12	1.25
		L	17	4	10	1.30
Control*	58.6		34.4	9.0	18.0	1.46
	+10.0		+6.7	+2.3	+3.9	+0.26

*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.

areas. Two years later, left iris rubeosis and episcleral vessel dilatation developed. IOP was 20 mm Hg. CDI showed a marked decrease in both the left ophthalmic artery antegrade blood flow and the systolic peak of the arterial waveform (Fig 1). Gosling and King's pulsatility index (PI) values² were calculated at 1.0 in the right eye and 0.55 in the left (Table 1).

Case 2

A 75 year old woman was referred for evaluation. She reported left visual disturbance, was diagnosed as having TA based on systolic blood pressure difference between the arms (right 88/46 mm Hg, left 122/86 mm Hg), a neck bruit, left common carotid artery occlusion, and left subclavian artery stenosis on carotid angiography, and positive HLA assays. The best corrected Landolt visual acuities were 1.0 right eye and 0.02 left. IOP was 6 mm Hg in both eyes. Left eye funduscopy showed inferior optic disc paleness and inferotemporal artery narrowing with a cholesterol plaque. Arm-retina circulation time was delayed. Two years later, left iris neovascularisation developed. IOP was 10 mm Hg. CDI showed reduction in the left ophthalmic artery antegrade blood flow (Table 1). Gosling and King's PI values were 1.25 right eye and 1.3 left eye.

COMMENT

CDI has only recently been introduced as a useful adjunct to clinical ophthalmological examination.¹ Although most useful in the evaluation of vascular disorders, CDI is also of value in the diagnosis of ocular ischaemic syndromes by allowing direct measurement of the waveform of orbital arteries, thus enabling direct assessment of the orbital vascular

supply.³⁻⁷ 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.³⁻⁷ 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. 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 retrobulbar vessel flow abnormalities, particularly in the ophthalmic artery, and in the evaluation of ocular manifestations in TA.

MICHAEL FRANCIS T ESCANO
MASANORI INO-UE
ATSUSHI AZUMI
YUKO YAMADA
MISAO YAMAMOTO

Department of Ophthalmology, Kobe University School of Medicine, Kobe, Japan

Correspondence to: Masanori Ino-ue, MD, Department of Ophthalmology, Kobe University School of Medicine, 7-5-1 Kusunoki-cho, Chuo-ku, Kobe 650, Japan.

Accepted for publication 2 April 1998

- 1 Belden CJ, Abbitt PL, Beadles KA. Color Doppler US of the orbit. *Radiographics* 1995;15:589-608.
- 2 Gosling RG, King DH. Arterial assessment by Doppler-shift ultrasound. *Proc R Soc Med* 1974; 67:447-9.
- 3 Ho AC, Lieb WE, Flaharty PM, *et al.* Color Doppler imaging of the ocular ischemic syndrome. *Ophthalmology* 1992;99:1453-62.
- 4 Hu HH, Sheng WY, Yen MY, *et al.* Color Doppler imaging of orbital arteries for detection of carotid occlusive disease. *Stroke* 1993;24: 1196-203.
- 5 Riordan-Eva P, Restori M, Hamilton AMP, *et al.* Orbital ultrasound in the ocular ischemic syndrome. *Eye* 1994;8:93-6.
- 6 Kerty E, Horven I. Ocular hemodynamic changes in patients with high grade carotid occlusive disease and development of chronic ocular ischemia. *Acta Ophthalmol Scand* 1995; 73:66-71.
- 7 Mawn LA, Hedges TR III, Rand WR, *et al.* Orbital color Doppler imaging in carotid occlusive disease. *Arch Ophthalmol* 1997;115:492-6.

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 neck, facial erythema, dyspnoea, cough, hoarseness, dysphagia, and mental status changes.¹ It may, however, present to an ophthalmologist through features such as conjunctival venous engorgement, chemosis, non-pulsatile proptosis, glaucoma, retinal venous changes, and papilloedema.^{2,3} It is an important diagnosis as there is usually associated serious pathology, frequently malignancy.¹ We present a case of SVCO presenting with minor ocular complaints and findings.

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.

Figure 1 View of patient's head and upper trunk on presentation showing plethora and swelling of the face and neck.

Figure 2 Posteroanterior chest x ray demonstrating the right paratracheal mass, and a stent (arrow) inserted after angioplasty into the superior vena cava.

He had no cardiovascular or respiratory complaints and his weight was stable; he did however smoke 15 cigarettes per day.

On examination he had a plethoric complexion, large neck, and thickening of his facial and upper limb skin; his periorbital skin was also slightly swollen (Fig 1). There was anterior blepharitis, but otherwise ocular examination was unremarkable; in particular there was no proptosis or conjunctival injection, the intraocular pressures were normal and funduscopy showed no vessel or disc changes. The nasolacrimal ducts were patent on syringing. In view of the upper body signs, a chest x ray was performed; this demonstrated a right paratracheal mass. A clinical diagnosis of superior vena cava obstruction was made.

The patient was referred to a cardiothoracic surgeon. The diagnosis was confirmed by angiography, and angioplasty with insertion of a stent was carried out (Fig 2). Anterior mediastinotomy allowed biopsy of the mass, histology reporting a low grade carcinoma, probably of pulmonary or thymic origin. This was treated with a course of palliative radiotherapy. Following these measures his symptoms and signs resolved.

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.

ALAN W D FITT
S RAUZ
G R KIRKBY

Birmingham and Midland Eye Centre, City Hospital
NHS Trust, Dudley Road, Birmingham B18 7QU

Correspondence to: Mr Fitt.

Accepted for publication 15 April 1998

- 1 Escalante CP. Causes and management of superior vena cava syndrome. *Oncology* 1993;7:61-8.
- 2 Alfano JE, Alfano PA. Vena caval obstruction syndrome. *Am J Ophthalmol* 1956;42:685-96.
- 3 Bedrossian HE. Increased intraocular pressure secondary to the mediastinal syndrome. *Arch Ophthalmol* 1952;47:64.

Paediatric varicella choroiditis

EDITOR.—Unifocal choroiditis occurs in children and adults with primary (chickenpox) and secondary varicella zoster virus (VZV)

infection.¹ 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.² 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

Figure 1 A choroidal lesion inferonasal to the macula, with overlying serous detachment of the retina.

Figure 2 A late fluorescein angiogram (8 minutes post-injection) showing collection of subretinal fluid involving the macula.

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

Chorioretinal 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.³ Kelly and Rosenthal also describe multifocal choroiditis in an otherwise healthy adult with primary VZV infection, where oral aciclovir resulted in regression of lesions.⁴

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 exanthema.

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 exanthema, with a rapid and complete resolution of her symptoms with return of excellent central acuity. Only a small residual pigmentary disturbance of the retinal pigment epithelium persisted. Significant visual changes may occur as a result of this common childhood illness, which may benefit from ophthalmic examination and treatment.

NADER MOINFAR
DAVID G WAGNER
GEORGIA A CHROUSOS
HOWARD P CUPPLES
MARSHALL KEYS

Department of Ophthalmology, Center for Sight,
Georgetown University Medical Center, PHC-7, 3800
Reservoir Road, NW, Washington, DC 20007, USA

Correspondence to: Dr Moinfar.

Accepted for publication 24 March 1998

- 1 Deegan W, Duker J. Unifocal choroiditis in primary varicella zoster. *Arch Ophthalmol* 1994;112:735-6.
- 2 Committee on Infectious Diseases, American Academy of Pediatrics. The use of oral acyclovir in otherwise healthy children with varicella. *Pediatrics* 1993;91:674-6.
- 3 Barondes MJ, Tellez F, Siegel A. Acute retinal necrosis after chickenpox in a healthy adult. *Ann Ophthalmol* 1992;24:335-6.
- 4 Kelly SP, Rosenthal AR. Chickenpox chorioretinitis. *Br J Ophthalmol* 1990;74:698-9.

Spontaneous hyphaema in hereditary haemorrhagic telangiectasia

EDITOR,—Hereditary haemorrhagic telangiectasia (HHT) is characterised by multiple dilations 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%).¹ Intraocular involvement is however extremely rare.

We report a patient with HHT with spontaneous bilateral hyphaemas

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 pupil. There was no history of any trauma. He had suffered from intermittent epistaxis, haematuria, gastrointestinal bleeds, and post-coital penile bleeding associated with telangiectasia on his face, external ear, chest, and glans penis. Past medical history included HHT, iron deficiency anaemia, and hypertension. Current medication included captopril, omeprazole, and ferrous sulphate. He recently had blood transfusion for severe iron deficiency anaemia.

On ophthalmic examination, visual acuity was 6/12 right eye and 6/18 left eye. Slit lamp examination showed hyphaema in the left eye and microhyphaema in the right. Mulberry-like iris vascular malformations were present at 12 o'clock position of both pupillary margins (Fig 1). Intraocular pressures were normal. He had moderate cataract and fundi were normal.

The hyphaemas resolved spontaneously within 2 weeks with no sequelae. A month later, spontaneous subconjunctival haemorrhage occurred in the left eye. Conjunctival telangiectasia were seen on the palpebral conjunctiva in both eyes. This haemorrhage resolved spontaneously.

COMMENT

HHT is a rare autosomal dominant disorder characterised by abnormal multiple dilations of capillaries and venules which have a tendency to bleed. These occur 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 thrombi, 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.¹ Although retinal lesions seldom cause symptoms, retinal haemorrhages, beading of retinal arteries, branch, and central retinal artery occlusion,² vitreous haemorrhage, secondary to areas of retinal neovascularisation, and inner retinal ischaemic atrophy can occur. Spontaneous hyphaema has been reported in juvenile xanthogranuloma, retrolental fibroplasia, persistent hyaloid, blood dyscrasias, and delayed

contusional injury.³ In patients of all ages, Doggart found spontaneous hyphaema associated with intraocular neoplasia, herpes simplex keratitis, zoster involvement of the iris, retrolental fibroplasia, persistent hyperplastic primary vitreous, and retinoschisis.⁴ Other causes of spontaneous hyphaema include inflamed iris vessels, rubeosis, iris haemangioma,⁵ iris neovascular tufts,⁶ neovascularisation of cataract surgery incisions, both small incision⁷ and extracapsular,⁸ and aneurysmal dilatation from persistent strand of vascularised pupillary membrane.⁹ Iris vascular tufts are uncommon acquired lesions associated with non-insulin dependent diabetes, myotonic dystrophy, retinal vein occlusion, and old age. They are elevated, and confined to the pupillary border. Recurrent hyphaema arising from vascular tufts can be successfully prevented by laser.⁶ Cataract surgery in such patients may be complicated by "in the bag hyphaemata".⁸

Our case demonstrates that iris vascular malformations may occur in HHT and give rise to spontaneous hyphaema. Patients with HHT are frequently not aware of having the disease unless it has been previously diagnosed in their family.¹⁰ Hence it would be prudent to look for evidence of HHT in patients with iris vascular malformations and consider HHT in the differential diagnosis of spontaneous hyphaema.

NOLAN R COTA

C O PECKAR

Warrington Hospital NHS Trust, Lovely Lane,
Warrington WA5 1QG

Correspondence to: Mr Cota.

Accepted for publication 17 March 1998

- 1 Green WR. Retina. In: Spencer WH, ed. *Ophthalmic pathology: an atlas and textbook*. 4th ed. Philadelphia: WB Saunders, 1996:1128.
- 2 Martin Diaz E, Obrer A, Ciscar Vilanova MA, *et al*. A patient with Rendu Osler Weber disease and pulmonary arteriovenous malformations treated satisfactorily by embolization. *Arch Bronchopneumol* 1996;32:307-9.
- 3 Howard GM. Spontaneous hyphema in infancy and childhood. *Arch Ophthalmol* 1962;68:615-20.
- 4 Doggart JH. Spontaneous hyphaema XVI. *Conciliium Ophthalmol* 1950;1:450-5.
- 5 Lam S. Iris cavernous hemangioma in a patient with recurrent hyphema. *Can J Ophthalmol* 1993;28:36-9.
- 6 Coleman SL, Green WR, Patz A. Vascular tufts of pupillary margin of iris. *Am J Ophthalmol* 1977;83:881-3.
- 7 Oduduwa TO, Bolger J. Late hyphema after small incision cataract surgery. *J Cataract Refract Surg* 1994;20:342-3.
- 8 Zigelbaum BM, Hersh PS. Endocapsular hyphema. *J Cataract Refract Surg* 1993;19:413-14.
- 9 Laidlaw DA, Bloom PA. Spontaneous hyphaema arising from a strand of persistent pupillary membrane. *Eur J Ophthalmol*. 1993;3:98-100.
- 10 Hodgson CH, Burchell HB, Good CA, *et al*. Hereditary haemorrhagic telangiectasia and pulmonary arteriovenous fistula. Survey of a large family. *N Engl J Med* 1959;261:625.

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.¹

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.

Figure 1 The leech was found to be adherent to the temporal bulbar conjunctiva of the girl's right eye.

under the surface of the water. Within 15 minutes after the girl had left the water, her father found a black coloured, oval shaped animal of about 10 mm length and of soft consistency at the temporal part of her right eye. His attempts to remove the animal were unsuccessful. In the eye hospital of Freiburg University the animal was found to be adherent to the temporal bulbar conjunctiva of the girl's right eye (Fig 1) and provisionally identified as a leech. After amethocaine (Tetracaine) eye drops were instilled into the eye, the animal could be removed. It fell down on to the right cheek, from where it was pulled off. Minor subconjunctival bleeding, but no conjunctivitis, occurred at the affected part of the conjunctiva. Dexpanthenol eye drops were instilled repeatedly. On the following day, no conjunctival injection was seen. The presumptive leech was later identified by a parasitologist (AK) as the duck leech *Theromyzon tessulatum*, a species which is common in the central and western part of Europe (Fig 2).

COMMENT

In wild mallard ducks (*Anas platyrhynchos*) and wild coots (*Fulica atra*), which were shot by hunters in the state of Baden-Württemberg (southwest Germany), up to three specimens of *Th. tessulatum* have been found in their nasal cavities.² A report from Britain deals with the infestation of both young and adult waterfowl, which showed signs of conjunctivitis associated with a watery ocular discharge. On post-mortem examination, leeches were found in the conjunctival sacs. Sometimes there was haemorrhage from the external nares caused by parasites in the nasal cavities and sinuses, which on at least one occasion were present in sufficient numbers to cause asphyxia.³

Infestation of the eyes of four domestic geese was also reported in the former East Germany, the leeches numbered from seven to 19 specimens per host. In these cases, *Th. tessulatum* stuck to all parts of the conjunctiva, especially on the membrana nictitans causing haemorrhagic inflammation of the mucous membranes, oedema, and watery ocular discharge.⁴ Christiansen observed minor bleeding of the pale conjunctiva and mucous purulent exudate, as well as a diffuse corneal

Figure 2 *Theromyzon tessulatum* from the conjunctiva of the 4 year old girl (anterior part) after fixation with formalin (9:1).

opacity with disseminated whitish dots in a domestic goose in Denmark, whose eyes, nasal and buccal cavities were infested with *Th. tessulatum*.⁵

In our case, as well as in the report of Pawlowski,¹ the conjunctiva showed minor bleeding, which resulted from the bite of the leech. Conjunctivitis was not observed in the human cases, which may be due to the short time of attachment by each leech.

C AUW-HAEDRICH
Universitäts-Augenklinik, D-79106 Freiburg, Germany

A KEIM
Institut für Gewässer und Fischerei,
Bruchsal-Heidelberg, Germany

M KIST
Institut für medizinische Mikrobiologie und Hygiene,
Freiburg, Germany

Correspondence to: Dr Auw-Haedrich.
Accepted for publication 17 March 1998

- 1 Pawlowski LK. *Theromyzon tessulatum* (O F Müller)—Egel im menschlichen Auge. *Zool Pol* 1938;3:181–3.
- 2 Keim A. Zur Wirtsspezifität von Blutegeln. Untersuchungen zum Parasitismus des Entenegels *Theromyzon tessulatum* (O F Müller 1774) und des Medizinischen Blutegels *Hirudo medicinalis* Linnaeus, 1758. Dissertation, Universität Hohenheim, 1979.
- 3 Keymer JF. Infestation of waterfowl with leeches. *Vet Rec* 1969;85:632–3.
- 4 Gräfiner G, Baumann H. Blutegelbefall beim Wassergeflügel. *Angew Parasitol* 1975;15:121–4.
- 5 Christiansen M. *Proctocleipsis tessellata* (O F Müller), der Entenegel, als Ursache von Krankheit, ua Konjunktivitis, bei Gänsen und Enten. *Zeitschr Inf Krkh Hyg Haustiere* 1939;55:75–89.

Direct observation of tear film stability on a damaged corneal epithelium

EDITOR,—Wettability¹ of the corneal epithelial surface and properties of tear fluids² 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 acciden-

Figure 1 Oval whitish necrotised 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 ($\times 100$) was observed at the upper nasal portion of the central cornea (A), and in the central corneal portion (B).

tally 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 necrotised area approximately 6 mm in diameter on the superficial layer of the central corneal epithelium (Fig 1). This lesion stained with fluorescein and rose bengal; the remaining portion of the ocular surface epithelium was intact without fluorescein or rose bengal staining. A previously reported system,³ 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% ofloxacin, 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.³ 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.

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 glycocalyx⁴ and the recently demonstrated MUC 1⁵ (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.

Supported in part by a research fund from Kyoto Foundation for the Promotion of Medical Science, and the intramural research fund of Kyoto Prefectural University of Medicine.

NORIIHIKO YOKOI
HIROSHI SAWA
SHIGERU KINOSHITA

Department of Ophthalmology, Kyoto Prefectural University of Medicine, Kyoto, Japan

Correspondence to: Dr Norihiko Yokoi, Department of Ophthalmology, Kyoto Prefectural University of Medicine, 465 Kajicho, Hirokoji-agaru, Kawaramachi-dori, Kamigyo-ku, Kyoto 602, Japan. Accepted for publication 2 April 1998

- 1 Holly FJ, Lemp MA. Wettability and wetting of corneal epithelium. *Exp Eye Res* 1971;11:239–50.
- 2 Tiffany JM, Winter N, Bliss G. Tear film stability and tear surface tension. *Curr Eye Res* 1989;8:507–15.
- 3 Yokoi N, Takehisa Y, Kinoshita S. Correlation of tear lipid layer interference patterns with the diagnosis and severity of dry eye. *Am J Ophthalmol* 1996;122:818–24.
- 4 Dilly PN. Contribution of epithelium to the stability of the tear film. *Trans Ophthalmol Soc UK* 1985;104:381–9.
- 5 Inatomi T, Spurr-Michaud S, Tisdale AS, et al. Human corneal and conjunctival epithelia express MUC 1 mucin. *Invest Ophthalmol Vis Sci* 1996;37:1684–92.

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

Figure 1 (A) Left sided ptosis, miosis, and iris hypochromia. (B) Within 10 seconds of 2.5% phenylephrine instillation in both eyes, relief of ptosis on the left and widening of the palpebral fissures on both sides has already occurred. Pupillary dilatation and equalisation follow approximately 15 minutes later.

congenital origin,^{1–3} and (2) sympathomimetics, such as 2.5% phenylephrine, may be used to elevate the lid when desired as a measure to defer surgery.^{4–6}

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 dilatation 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.

CAMERON F PARSA
NICOLAS D L GEORGE
CREIG S HOYT

Pediatric Ophthalmology Unit,
Department of Ophthalmology, School of Medicine,
University of California, San Francisco, USA

NICOLAS D L GEORGE
Department of Ophthalmology,
Leeds General Infirmary, Leeds

Correspondence to: Creig S Hoyt, MD, University of California, San Francisco, Department of Pediatric Ophthalmology, Box 0344, 400 Parnassus Avenue, Room 702-A, San Francisco, CA 94143, USA.

Accepted for publication 17 March 1998

- 1 Miller NR. Walsh & Hoyt's clinical neuro-ophthalmology. 4th ed. Vol 2. Baltimore: William & Wilkins, 1985:500–11.
- 2 Loewenfeld IE. The pupil. Ames and Detroit: Iowa State and Wayne State University Presses, 1993:71–3, 907.
- 3 Diefenhouse MC, Palay DA, Newman NJ, et al. Acquired heterochromia with Horner syndrome in two adults. *Ophthalmology* 1992;99:1815–17.
- 4 Campbell, Jr, WW, Hill TA. A good treatment for Horner's syndrome. *N Engl J Med* 1978;299:835.
- 5 Beard C. Ptosis. 3rd ed. St Louis: CV Mosby, 1981:82–3.
- 6 McGregor ML. Adrenergic agonists. In: Mauger TF, Craig EL, eds. *Havener's ocular pharmacology*. 6th ed. St Louis: Mosby, 1994:74.