Sir,

Branch retinal artery obstruction with simultaneous central retinal vein occlusion

The occurrence of branch retinal artery occlusion together with central retinal vein occlusion is very rare, especially in patients less than 30 years of age.¹ This combination differs clinically from the previously well-described entities of combined central retinal artery and vein occlusion, and combined central retinal vein occlusion (CRVO) with secondary cilioretinal artery occlusion.^{2,3}

A branch retinal artery occlusion (BRAO) occurring in a patient, however, carries the same clinicopathological significance as a central retinal artery occlusion (CRAO) and should be regarded with equal importance with respect to identifying any underlying aetiological factor.⁴ We present a case of branch retinal artery obstruction with simultaneous central retinal vein occlusion in a young patient.

Case report

A 28-year-old woman presented acutely with a 2 day history of loss of vision in the right eye. Two weeks previously she had experienced a blurring of central vision in the right eye lasting 4 h. This resolved with a return to normal vision. Two days prior to presentation the blurring recurred but this time the vision did not return. There was no pain in the eye and her visual field and colour vision were subjectively unaffected.

She had no significant past medical or obstetric history and was generally in good health. She was not on the oral contraceptive pill or any other medication and is a non-smoker.

On examination the Snellen visual acuities were 6/18 in the right eye and 6/5 in the left. The right eye was quiet and there was no relative afferent pupillary defect. Computerised perimetry revealed an inferior half central scotoma. The colour vision by Ishihara test was 16/16 and the intraocular pressure was normal at 12 mmHg. The vitreous was quiet. The right superotemporal retinal artery was occluded with corresponding sectoral retinal whitening of the area supplied along the horizontal raphe. No emboli were seen (Fig. 1). The superotemporal retinal artery had an abnormal branching pattern as it arose from the disc and the right optic disc appeared to be smaller than the left. The optic disc was swollen and mildly hyperaemic with a few flame-shaped retinal haemorrhages scattered in the fundus. These haemorrhages had white centres. The retinal veins were dilated and tortuous. There was no evidence of retinal vasculitis and no focal fundal lesions were seen. There was no macula oedema. The left eye was normal (Fig. 2).

Fig. 1. Right eye

A fundus fluorescein angiogram was performed the same day. It was interesting to note that the affected artery filled before the other retinal arteries and also well after the choroidal phase of the angiogram (Fig. 3). There was no leakage from the veins and the foveal avascular zone appeared unaffected. Late staining of the disc was observed (Fig. 4).

Clinical examination of all major systems was normal. Pulse was 68 min⁻¹, blood pressure 110/70 mmHg.

The patient subsequently underwent extensive haematological, cardiac and medical assessment to search for an underlying cause. This included urinalysis for red cell casts and microscopy which was normal. The following results were obtained from investigations carried out: haemoglobin, 12.9 g/dl; white cell count, $5.9 \times 10^9/l$; platelets, $348 \times 10^9/l$; haemoglobin

Fig. 2. Left eye.

(a)

(b)

(c)

Fig. 3. (a) Early arterial phase (9.8 s). (b) Mid-arterial phase (11.5). (c) Arteriovenous phase (13.5 s).

electrophoresis, normal; erythrocyte sedimentation rate, 11 mm/h; C-reactive protein, 3.8 mg/l; total cholesterol, 4.8 mmol/l; triglycerides, 1.0 mmol/l; blood glucose, 5.0 mmol/l; liver function tests: bilirubin 10 μ mol/l; ALT

Fig. 4. Late venous phase.

22 IU/l; alkaline phosphatase 67 IU/l; syphilis serology, negative for TPHA; autoantibody screen and antineutrophilic cytoplasmic antibody, negative; antiphospholipid antibodies: lupus anticoagulant 1.04; lupus inhibitor not present; serum angiotensin converting enzyme, 32; free thyroxine, 16.8 pmol/l; protein C, 86.3%; protein S, 124%; activated protein C resistance, 3.36; factor XII, 87%; factor VIII, 50%; prothrombin time, 13 s; thrombin time, 16 s; fibrinogen level, 3.65 g/l. Chest radiograph, carotid Doppler ultrasound, electrocardiogram and echocardiography were normal. All of the above were normal.

Antithrombin III, however, was slightly elevated, at 13.7%, but this is not in keeping with a thrombotic event.

Risk assessment for future risk of myocardial infarction and sudden death was unrevealing since this patient did not possess specific risk factors predictive of these events. These risk factors were: prior myocardial infarction, other heart disease, high cholesterol, current smoker, obesity, bilateral oophorectomy, hypertension, diabetes, oral contraceptive use within the previous month and a history of thromboembolism.

Nine months after initial presentation, there have been no further ophthalmic events of note and the final visual acuity in the right eye is 6/36 with corresponding pallor of the superior half of the optic disc.

Comment

Simultaneous arterial and venous obstructions of the retina are uncommon and occur in three clinical syndromes.¹⁻³

(1) The CRAO/CRVO combination was well described by Richards in 1979.² Incomplete CRAO resembling BRAO has also been described by Iijima and Tsumura,⁵ but in our case pupillary reactions, visual field testing and angiographic features confirmed that our patient had BRAO rather than incomplete CRAO. Combined CRAO/CRVO produces the characteristic haemorrhagic picture of ischaemic CRVO following primary arterial occlusion. It is thought that primary vascular occlusion in

the retinal artery causes stagnation of blood in the venous systems with a secondary haemorrhagic ischaemic picture. Our case did not show any ischaemic features attributable to venous disease and the history of amaurosis is more suggestive of a primary arterial insult.

(2) The clinical syndrome of *combined occlusion of the CRVO/cilioretinal artery* was reported by McLeod and Ring,³ who described 10 cases, and also reported by Magargal *et al.*⁶ In these cases, however, the CRVO is the primary insult with the corresponding clinical and investigative findings confirming primary venous disease, followed later by cilioretinal artery occlusion.^{6,7} In these cases it is thought that the cilioretinal artery has a lower perfusion pressure than the other retinal arteries. When the optic disc oedema develops following the CRVO, the resulting rise in intraluminal capillary pressure causes a relative occlusion of the cilioretinal artery.⁸⁻¹⁰

(3) The very rare clinical syndrome of *combined branch retinal artery and central retinal vein obstruction*. There has been only one major series describing this entity. This was reported by Duker *et al.* in 1990.¹ They described 7 patients with this rare combination of retinal vascular disease. Based on their clinical findings they classified BRAO/CRVO into two groups: a group of young patients without underlying systemic risk factors and a second group of elderly patients with vasculopathic risk factors.

In the first group visual outcome was generally good and in each case no underlying aetiological factor was found despite extensive medical, cardiovascular, haematological and ophthalmic investigation. They hypothesised that a primary inflammation of the optic nerve resulting in secondary impairment of retinal vasculature had occurred. Two of the three patients in this group had afferent pupillary defects and dyschromatopsia supporting this theory. These features were not present in our patient, whose pupillary reactions and colour vision were normal. Another mechanism was put forward which proposed a primary vasculitis within the optic nerve head affecting both the branch retinal artery and central retinal vein,¹¹ the oedema at the optic nerve head in this case causing simultaneous extravascular compression. This seems to be a plausible explanation for our case. Supporting this theory would be the history of amaurosis, abnormal branching pattern at the optic nerve head of the occluded artery and the smaller size of the optic disc predisposing to congestion. The earlier filling of the affected artery before the other retinal vessels during the arterial phase of the angiogram would be consistent with a lower perfusion pressure in this vessel and explain why this particular artery was relatively occluded when the optic disc oedema set in.

In the series by Duker *et al.*¹ 2 of the 3 cases in the group of young patients without systemic factors had angiographic evidence of delayed vascular filling. The remaining case had slight asymmetry of arterial filling but no other details with respect to this finding were given in their article. Perhaps this asymmetry of filling is

similar to the filling pattern seen in our case. Against the theory of optic disc vasculitis is the absence of any confirmatory clinical or immunological evidence of retinal vasculitis.

The age group and chronological pattern of vascular events makes this case unique. BRAO, especially in a young patient, carries the same clinical significance as a CRAO.⁴ As such, a detailed search for an underlying cause should be made. This should be regarded as a potentially life-threatening event until proven otherwise and, indeed, the prevalence of factors leading to possible serious thromboembolic phenomena is relatively high.¹² Similar to the series by Duker *et al.*¹ we have not been able to identify an underlying aetiology despite extensive investigation.

It is possible that the pathophysiological mechanisms underlying the vascular events in this case are similar to that of combined CRVO/cilioretinal artery occlusion where there is early filling of the affected artery as opposed to delayed or non-filling found in the series by Duker *et al.*¹ However, as far as we are aware, this has not been previously described with involvement of a branch retinal artery.

References

1. Duker JS, Cohen MS, Brown GC, Sergott RC, McNamara JA. Combined branch retinal artery and central retinal vein occlusion. *Retina* 1990;10:105-12.
2. Richards RP. Simultaneous occlusion of the central retinal artery and vein. *Trans Am Ophthalmol Soc* 1979;77:191-209.
3. McLeod D, Ring CP. Cilioretinal infarction after retinal vein occlusion. *Br J Ophthalmol* 1976;60:419-27.
4. Ros MA, Magargal LE, Uram M. Branch retinal-artery obstruction: a review of 201 eyes. *Ann Ophthalmol* 1989;21:103-7.
5. Iijima H, Tsumura T. Combined occlusion of the central retinal artery and vein. *Jpn J Ophthalmol* 1994;38:202-7.
6. Magargal LE, Gonder JR, Maher V. Central retinal vein obstruction in the young adult. *Trans Pa Acad Ophthalmol Otolaryngol* 1985;37:148-53.
7. Shatz H, Fong AC, McDonald HR, Johnson RN, Joffe L, Wilkinson CP, *et al.* Cilioretinal artery occlusion in young adults with central retinal vein occlusion. *Ophthalmology* 1991;98:594-601.
8. Wrigstad A. Central retinal vein occlusion combined with occlusion of a cilioretinal artery: a case report. *Acta Ophthalmol Scand* 1998;76:503-5.
9. Brazitikos PD, Pournaras CJ, Othenin-Girard P, Borruat FX. Pathogenetic mechanisms in combined cilioretinal artery and retinal vein occlusion: a reappraisal. *Int Ophthalmol* 1993;17:235-42.
10. Noble KG. Central retinal vein occlusion and cilioretinal artery infarction. *Am J Ophthalmol* 1994;118:811-3.
11. Hayreh SS. Optic disc vasculitis. *Br J Ophthalmol* 1972;56:652-70.
12. Greven CM, Slusher MM, Weaver RG. Retinal arterial occlusions in young adults. *Am J Ophthalmol* 1995;120:776-83.

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