CASE REPORT

Multimodal imaging of central retinal artery occlusion with retained cilioretinal perfusion

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SUMMARY

A man aged 59 years old presented with sudden, painless, monocular visual loss due to central retinal artery occlusion. Central vision was retained and peripheral vision lost due to retained cilioretinal perfusion. Increased inner retinal thickening and reflectivity followed by subsequent reduction was documented by sequential imaging. This is the first report of such events monitored with spectral-domain optical coherence tomography where central visual acuity was maintained.

BACKGROUND

Central retinal artery occlusion (CRAO) is often considered a potential ophthalmic emergency; analogous to cerebral ischaemic stroke. Patients typically present with sudden, painless, extensive monocular visual loss. Clinical signs include relative afferent pupillary defect, severe retinal oedema and often with a 'cherry red spot' at the macula. Visual loss is often permanent. When CRAO occurs in the presence of a patent cilioretinal artery visual function may be somewhat better preserved. The prevalence of a cilioretinal artery varies between 25% and 50% of the population.¹ Early restoration of retinal perfusion is critical but rarely possible in clinical practice as ischaemic retinal damage quickly occurs. The retinal survival time is not known although damage is thought to be irreversible after about 4–6.5 hours based on animal experiments and clinical studies of thrombolysis.¹ Critically clinical staff need to be aware of CRAO's natural history and the evidence base for any interventional management. Ritter *et al*² reported a case series of patients with branch retinal artery occlusion (BRAO) imaged with spectral-domain optical

coherence tomography (SD-OCT) technology. These patients subsequently developed inner retinal thinning in the area of BRAO.² To the best of our knowledge the changes to retinal layers imaged by SD-OCT following CRAO in the presence of preserved cilioretinal perfusion and good visual acuity have not been reported to date.

This case is of clinical and academic interest given recent reports of modern imaging modalities showing novel pathophysiological changes within the retina following BRAO. Furthermore, our somewhat unique case (in terms of serial SD-OCT imaging of CRAO with a patent cilioretinal circulation) adds to the current knowledge base within this emerging field of clinical academic endeavour.

CASE PRESENTATION

A man aged 59 years old presented with sudden, painless reduction of left vision of 48 hours duration. He had hypercholesterolaemia for which a statin had been prescribed. He was a long-term smoker. The visual acuity was 6/12 Snellen in left eye. Funduscopy revealed atherosclerotic arteriolar narrowing and cotton wool spots with ischaemic grey pallor in an annular pattern of opacification in the mid-peripheral retina surrounding an oblique area of central macular sparing of normal colour (figure 1). The working clinical diagnosis on presentation was of CRAO with a perfused cilioretinal circulation.

INVESTIGATIONS

SD-OCT imaging 1-week after initial symptoms showed extensive inner retinal thickening and increased reflectivity surrounding an oblique central papillomacular peninsula of retina of normal thickness and reflectivity (figure 2).

Figure 1 (Left panel) Colour fundus image; whitening of retina from central retina artery occlusion with central, oblique wedge of normal retinal colour retained due to a cilioretinal artery. (Right panel) Fluorescein angiogram; dye visible in cilioretinal artery and laminar flow in superior temporal vein.

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Figure 2 SD-OCT en face C-scan 1-week after initial symptoms. Extensive annular inner retinal thickening surrounding an oblique central papillomacular peninsula of retina of normal thickness is seen. B-scan scout lines shown at point of interest 102 vertical and 330 horizontal.

Fundus fluorescein angiography (FFA) 4 weeks later revealed extensive retinal non-perfusion surrounding the peninsula of perfused central retina with rapid filling of the capillaries within the territory supplied by a cilioretinal artery (figure 1).

Full blood count, erythrocyte sedimentation rate, C reactive protein, electrolytes and random glucose were within normal ranges for our laboratory. Serum cholesterol was raised 7.4 mmol/L (normal range is 3–5.4 mmol/L). Blood pressure was not raised. Carotid artery Doppler ultrasound scan showed <50% stenosis of each vessel. Small vessel ischaemic change was seen on CT and MRI neuroimaging. MR carotid angiogram showed <70% carotid stenosis.

DIFFERENTIAL DIAGNOSIS

Differential diagnosis of sudden loss of visual acuity in one eye includes:

- ▶ CRAO and BRAO
- ▶ Central and branch retinal vein occlusion
- ▶ Non-arteritic and arteritic anterior ischaemic optic neuropathy
- ▶ Posterior ischaemic optic neuropathy
- ▶ Vitreous haemorrhage
- ▶ Retinal detachment

The diagnosis was made after thorough history taking and clinical examination. The retinal findings described above along with a normal vitreous and optic nerve appearance in conjunction with, subsequent retinal imaging including OCT scanning and FFA showed findings consistent with CRAO with cilioretinal sparing.

TREATMENT

Treatment with digital ocular massage, intraocular pressure-lowering therapy and carbon dioxide inhalation was initiated with the intention to dislodge an embolus or thrombus more peripherally. Secondary prevention of cardiovascular risk factors was started along with urgent referral to stroke services. Smoking cessation was advised. A prescription for 75 mg of aspirin was given.

OUTCOME AND FOLLOW-UP

Sequential SD-OCT scans taken at serial follow-up visits demonstrate progressive inner retinal thinning in the mid-peripheral retina over a 14-month follow-up period. Scout lines set at identical points on the affected macula allow accurate comparison of

retinal thickness over the course of follow-up (figure 3). The central macular thickness remained normal due to 'macular sparing'. The structural changes of mid peripheral inner retinal thinning on OCT imaging correlate with the functional mid-peripheral visual field loss with central visual field sparing. Sequential SD-OCT linked B-scans at scout lines 102 vertical and 330 horizontal are presented with retinal thickness measurements. Three years after presentation, the left eye acuity was 6/6 Snellen with severe left visual field constriction or 'tunnel vision' on perimetry. Left optic atrophy was observed. Repeat FFA imaging 14 months after presentation using confocal scanning laser ophthalmoscopy (Spectralis, Heidelberg Engineering, Germany) with video angiography showed sustained central cilioretinal perfusion with retrograde flow. SD-OCT shows a persistent zone of inner retinal layer thinning surrounding a central zone of normal retinal thickness. This corresponds to a retained island of central visual field surrounded by severe peripheral scotoma (figure 4).

DISCUSSION

The central retinal artery (CRA) is a branch of the ophthalmic artery, which originates from the internal carotid artery. The CRA supplies blood to quadrants of the inner retina via its superior and inferior nasal and temporal branches.¹ The outer retina is supplied by the choriocapillaris which is perfused by the ciliary arteries. Both circulations must be present to maintain normal retinal function.

The SD-OCT imaging showed thinning of inner retinal layers in our patient following initial thickening and increased reflectivity correlates with the recent case series by Ritter *et al.*² Cornut *et al.*³ reported similar events. Our patient showed initial inner retinal swelling in the mid-peripheral retinal areas not

Figure 3 Images depicting inner retinal layer thinning over 3 years. Explanation: scout lines (not shown) were placed at a 'point of interest'; mapped as position at 102 vertical and 330 horizontal for each linked B-scan optical coherence tomography (OCT) image in the macular cube OCT volume scan. (A) At presentation (retinal thickness at the 'point of interest' 332 mm); (B) 1 month later (thickness 223 mm) at same point and (C) 3 years later (retinal thickness 199 mm) at same point. Sequential and linked spectral-domain OCT images of left inferior macula are shown.

Figure 4 Humphrey visual field test, left eye, 4 weeks after presentation, (A) 120 point supra threshold test and (B) central 24–2 threshold test.

perfused by a cilioretinal artery and subsequently progressive inner retinal thinning in this annular zone (figure 3). As far as we know this is the first report of the acute and chronic changes in vivo with SD-OCT imaging of the inner retinal profile in CRAO in the presence of cilioretinal artery perfusion of the fovea. It has been observed that selective thinning of inner retinal nerve fibre layers on SD-OCT may be useful to diagnose longstanding BRAOs from other conditions—such as patients with glaucoma who present later.⁴

An important anatomical variant in the fundus is the presence of a cilioretinal artery. If present, a cilioretinal artery may supply the papillomacular bundle. In such patients, this alternative blood supply may allow central vision to be maintained despite CRA occlusion. However, this matter is variable and may inter alia depend on the size and perfusion area of any such cilioretinal artery and duration of occlusion and penumbra factors.^{5 6} Cilioretinal arteries are said to be present in 32% of the eyes, and bilaterally in 15%.⁷ When present such perfusion contributes to some portion of the macular area in 19% of the patients and can on occasion supply the posterior pole.^{6 8}

Emboli from carotid atheroma and valvular cardiac disease are clinically important sources of ocular emboli. In all, 74% of such emboli are composed of cholesterol, while 10.5% are calcific material and 15.5% are fibrin. Thrombus can also occur.⁹

CRAO can be considered as part of the stroke continuum and worthy of urgent consideration as these patients have been shown to be at risk of imminent stroke.¹⁰ Helenius *et al*¹¹ reported use of MRI cerebral imaging to investigate patients with transient and permanent retinal arterial ischaemia showed such matters have poor prognosis similar to cerebral ischaemia. However, unlike cerebral cortex, ischaemic penumbral retina can persist indefinitely, and McLeod¹² considers that reperfusion of this tissue may explain the recovery of the ERG b-wave after arterial unclamping in experimental animals following 4 hours of CRAO.

In terms of treatment, thrombolysis has been investigated in the treatment for non-arteritic CRAO. This is analogous to the treatment in acute ischaemic stroke or coronary artery occlusion. A prospective, randomised, multicentre clinical trial, the European Assessment Group for Lysis in the Eye (EAGLE) study,¹³ found no significant improvement between the thrombolysis and standard therapy groups (60.0% vs 57.1%) in patients presenting within 20 hours of symptom onset. Furthermore adverse events were more frequent in the thrombolysis arm of the EAGLE study. Development of intracerebral haemorrhage was the main safety concern.¹³ The use of intra-arterial tissue-type plasminogen activator (tPA) administered within 24 hours of symptoms has been investigated by a large multicentre study

and reported negative results but did observe a three letter gain in visual acuity in the subgroup of patients treated within 6 hours of symptom onset.¹⁴ This agrees with CRA clamping studies in monkeys by Hayreh,¹⁵ which suggest the time to irreversible retinal ischaemia is under 4 hours. Current research using experimental arterial occlusions suggests that emergency fibrinolytic intervention is inappropriate after 2 hours post CRAO as hypoxic areas of the retina, termed the ischaemic penumbra, are not at risk of ischaemic apoptosis as they remain marginally oxygenated by the choroid or by residual circulation.¹⁶

Prompt assessment and treatment of patients with CRAO is required to optimise general cerebrovascular outcomes.⁵ In addition to limited benefit from recent thrombolysis interventional studies, previous case series studies found anterior chamber paracentesis disappointing.¹⁷ Thus it is pragmatic to opine that such interventions after more than a day of visual loss from CRAO is not appropriate. In terms of prognosis outcome is dependent on variability of retinal vascular anatomy. Watchful waiting of such patients and attention to vascular risk factors is indicated. Use of novel SD-OCT imaging and multimodal confocal scanning laser ophthalmoscopy may provide new insights into the natural history of such patients and aid with better categorisation of CRAO subtypes.

Learning points

- ▶ Central retinal artery occlusion (CRAO) presents with sudden painless profound monocular visual loss.
- ▶ Central vision may be retained in some patients with CRAO in the presence of cilioretinal artery perfusion.
- ▶ SD-OCT imaging is of merit in studying inner retinal morphology in CRAO and may aid in the identification of a perfused cilioretinal circulation.
- ▶ There was correlation between the inner retinal layer swelling on acute presentation and later inner retinal layer loss imaged in vivo with angiography and perimetry mirroring the structural/functional relationships.
- ▶ These findings can be used to aid diagnosis during acute phase and are of prognostic significance and may lead to a better understanding of natural history in CRAO.

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Contributors As first author AW has written the case report after managing and diagnosing the patient in question. SPK has also managed and treated the patient and has edited and revised the submitted manuscript.

Competing interests None declared.

Patient consent Obtained.

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