

Atypical Presentation of Central Retinal Artery Occlusion

Dear Editor,

Central retinal artery occlusion (CRAO) typically presents with a retinal “cherry red spot”. In CRAO, infarction of the retinal nerve fibre layer renders the retina opaque. The fovea, where this layer is absent, retains its normal orange-red colour due to perfusion from the underlying choroidal vessels, resulting in the appearance of the “cherry red spot”. Retinal survival time has been shown to range from 0 to 105 minutes.¹ If the embolus gets dislodged within this time, retinal ischaemia can occur but to a milder and limited severity.

We describe 2 cases of limited CRAO highlighting the need for awareness of atypical presentations where the “cherry red spot” is absent.

Case Reports

A 74-year-old Indian woman presented with sudden acute, painless loss of vision in her left eye upon awakening. She had well-controlled diabetes and hypertension. Visual acuity at presentation was 6/7.5 on the right and 6/60 on the affected left eye. No relative afferent pupillary defect (RAPD) was detected and confrontation visual field testing, which was normal in the right eye, revealed a central scotoma in the left. Subtle retinal oedema which was initially passed off as normal was noted on dilated fundal examination (Fig. 1a). A diagnosis of CRAO was made. Her blood pressure was 150/88 mmHg. There was no atrial fibrillation, carotid bruit or cardiac murmurs. Retinal nerve fibre layer thickness measured by the stratus ocular coherence tomogram was normal and fundus fluorescein angiography (FFA) showed delayed arm-retinal time of 43 seconds in the left eye. The patient declined anterior chamber paracentesis and hospital admission. Left ocular massage, carbogen inhalation, anti-glaucoma drops and aspirin were commenced. Ultrasound of the carotids and an echocardiogram were normal.

There was significant improvement in vision of the affected left eye the following day from 6/60 to 6/9 but colour vision with Ishihara plates was abnormal (responses were slower and only 9 of 15 plates were read correctly). The retinal oedema, however, had subsided. Six months later, her vision remained stable at 6/9.

The second case was a 52-year-old hypertensive Chinese female, who presented with an acute painless “darkening” of vision in her left eye. Visual acuity at presentation was 6/7.5 on the right and counting fingers on the affected left eye. No RAPD was detected. Colour vision was normal at presentation. Confrontation visual field testing, normal on the right, revealed a central scotoma on the left. Dilated fundus examination was unremarkable. In particular, no retinal oedema was seen. Her blood pressure was 145/90 mmHg and her pulse was regular. An urgent FFA showed no compromise in vascular perfusion. A provisional diagnosis of amaurosis fugax, limited CRAO was given.

Her left eye vision improved to 6/12 the following day although she still reported a central scotoma. A mild left RAPD was now detected, colour vision was abnormal (only 1 of 15 Ishihara plates identified), dilated fundus examination revealed a mild “cherry red spot” in the affected left eye (Fig. 1b), and Humphrey’s visual field testing demonstrated a left inferior paracentral scotoma. A diagnosis of limited CRAO was made. Ultrasound carotids were normal and transthoracic echocardiography reported mild mitral and tricuspid valve regurgitation but no cardiac thrombus. In view of her young age, she was screened for autoimmune vasculitis and hypercoagulable states. Full blood count, electrolytes, fasting lipids and glucose, erythrocyte sedimentation rate, syphilis screen, lupus screen, anticardiolipin antibodies, homocysteine levels and protein S and C were normal. Lipid panel tests revealed a slightly high low density lipoprotein level and a transoesophageal

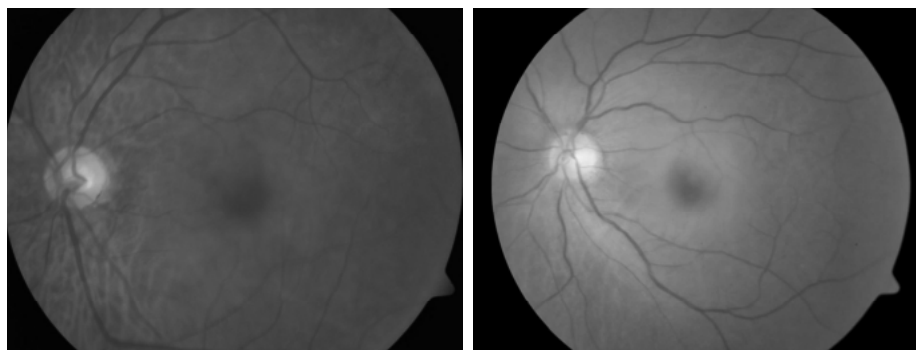


Fig. 1a. Fundal photograph of left eye of Patient 1 with mild macular oedema on Day 2 of presentation.

Fig. 1b. Fundal photograph of left eye of Patient 2 on Day 2 of presentation showing a mild “cherry red spot”.

echocardiogram detected a small complex atheromatous sessile plaque in the ascending aorta. She was treated with Zocor and aspirin and followed-up with her cardiologist.

Her visual function remained stable at 6 years of follow-up. She has had 2 further episodes of amaurosis in the left eye but stroke workup and rheumatology reviews remain normal. During these attacks, she was treated with glyceryl trinitrate with good response.

Discussion

Central retinal artery occlusion is well documented and typically presents with a “cherry red spot” on the retina. Visual acuity at presentation is usually very poor, with the majority having counting fingers vision or worse. The most common mechanism of CRAO is embolic obstruction, with the carotid artery being the most common source of endogenous emboli.

In a study on Rhesus monkeys, Hayreh et al² temporarily clamped the central retinal artery of 39 eyes in 39 middle-aged monkeys. The duration of occlusion, ranging from 97 to 300 minutes correlated with retinal nerve fiber layer damage. No apparent damage was noted in the eyes with less than 100 minutes of artery occlusion. Therefore, theoretically, CRAO can be totally reversible if the occlusion time is less than the retinal survival time.

Hayreh and Zimmerman³ also reported 38 patients with a milder, limited form of transient CRAO. The retinal findings in this group varied from normal-looking retina and optic discs to patches of retinal whitening.

We have described 2 unusual cases where the patients did not have a “cherry red spot” at presentation and experienced dramatic improvement in vision the following day. The first case only presented with subtle retinal oedema while the second initially presented with a normal fundus.

CRAO in young patients is rare. It has been reported to be caused by multiple mechanisms including autoimmune disease such as the presence of lupus anticoagulant or anticardiolipin antibodies, hypercoagulable states like

Protein C and S deficiencies or embolic cardiac disease such as atrial myxoma, bacterial endocarditis or mitral valve disease.⁴ Migraine occasionally causes CRAO but this should be a diagnosis of exclusion.⁵ Therefore, detailed investigations are necessary to rule out potentially life-threatening thrombotic or vasculitic conditions in young patients.

Conclusion

CRAO can present differently depending on the duration of retinal ischaemia. Although a pale retina and optic disc is common, a normal fundus or retinal vasculature should not exclude CRAO.

We need to be aware of the presentation of a limited form of CRAO as this has the same aetiology as classic CRAO and requires the same work-up and management despite better visual prognosis.

REFERENCES

1. Hayreh SS, Zimmerman MB, Kimura A, Sanon A. Central retinal artery occlusion. Retinal survival time. *Exp Eye Res* 2004;78:723-36.
2. Hayreh SS, Jonas J. Optic disk and retinal nerve fiber layer damage after transient central retinal artery occlusion: An experimental study in rhesus monkeys. *Am J Ophthalmol* 2000;129:786-95.
3. Hayreh SS, Zimmerman B. Fundus changes in central retinal artery occlusion. *Retina* 2007;27:276-89.
4. Greven CM, Slusher MM, Weaver RG. Retinal arterial occlusions in young adults. *Am J Ophthalmol* 1995;120:776-83.
5. Brown GC, Magargal LE, Shields JA, Goldberg RE, Walsh PN. Retinal arterial obstruction in children and young adults. *Ophthalmology* 1981;88:18-25.

Audrey YC Pang,¹ MB BChir, Su Ann Lim,¹ MBBS, MMed (Ophth), FRCS (Edin)

¹ Department of Ophthalmology, Tan Tock Seng Hospital, Singapore

Address for Correspondence: Dr Audrey Pang, Department of Ophthalmology, Tan Tock Seng Hospital, 11 Jalan Tan Tock Seng, Singapore 308433. Email: audreypang@gmail.com