Imaging Modalities in the Diagnosis of Transient Central Retinal Artery Occlusion

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KEY WORDS: transient central retinal artery occlusion, cotton wool spots, optical coherence tomography, fluorescein angiography, indocyanine green angiography

IMAJ 2012; 14: 329–330

Central retinal artery occlusion is an ophthalmic emergency, causing acute painless loss of vision. Typical clinical findings include retinal pallor, appearance of a foveal "cherry-red spot," and in some cases narrow and segmented arteries [1]. Hayreh and Zimmerman [1] divided CRAO into four distinct subtypes: non-arteritic, non-arteritic with cilioretinal sparing, arteritic, and transient. The diagnosis of transient CRAO is challenging as it lacks the aforementioned classical findings. It occurs when the central retinal artery is not completely occluded and retinal perfusion pressure is reduced, but not enough to cause complete retinal infarction as seen in complete CRAO. The peripapillary and periarterial regions remain perfused, and beyond them the retina becomes ischemic. At the border between the perfused and ischemic retina the axoplasmic transport is interrupted, which results in the appearance of cotton wool spots [2]. Oji and McLeod [3] described seven patients with transient CRAO who presented with acute mild decrease of vision, a ring-like pattern of CWS around the optic disk and along the arcades, and perifoveal ischemia. Several causes of transient CRAO and retinal hypoperfusion have been suggested, such as transient emboli that dislodge or dissolve, nocturnal arterial hypotension, or transient vasospasm induced by serotonin released from atherosclerotic plaques [4].

A literature review revealed only a few studies that acknowledge this diagnosis; most studies consider all subtypes of CRAO as a single clinical entity. We present a case of transient CRAO and demonstrate the imaging modalities that were used to confirm this unique diagnosis.

CRAO = central retinal artery occlusion
CWS = cotton wool spots

PATIENT DESCRIPTION

A 58 year old man presented to our clinic with a 3 day history of acute painless decreased vision in his right eye. The patient was otherwise healthy and did not use any medications. Ocular history included only presbyopia.

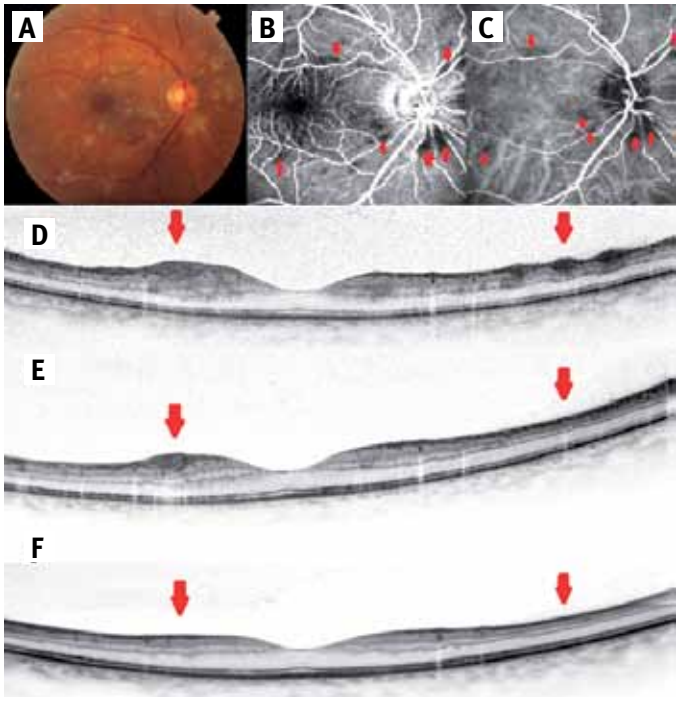
Best corrected visual acuity at presentation was 20/70 in the right eye and 20/20 in the left eye. There was no afferent pupillary defect or ocular movement limitation, anterior segments were both normal, and applanation tonometry was normal. Dilated fundus examination of the right eye revealed multiple fluffy whitish lesions around the optic disk and along the arcades, with perifoveal ischemia [Figure A]. The left eye was normal. HR-OCT (high resolution optical coherence tomography) (Spectralis, Heidelberg, Germany) of the retinal lesions demonstrated focal thickening and hyper-reflectivity of the nerve fiber layer, compatible with CWS [Figure D]. The macula appeared normal. Fluorescein angiography demonstrated focal areas of hypofluorescence which matched the size and location of the CWS [Figure B]. There was no leakage from the blood vessels or the optic disk. Importantly, a marked delay of 24 seconds in the arterial filling time was documented. Indocyanine green angiography demonstrated areas of hypofluorescence that matched the size and location of the CWS [Figure C]. No disturbance in choroidal filling or other chorioretinal pathology was detected. Automated visual field testing demonstrated decreased sensitivity of the right eye (mean deviation -4.7 dB OD compared with -0.7 dB OS), with no scotoma.

The following laboratory tests were normal: complete blood count, kidney and liver function tests, lipid profile, blood clotting test, erythrocyte sedimentation rate, C-reactive protein, collagenogram, and serology testing for human immunodeficiency virus and syphilis. Homocysteine levels were normal and there was no hypercoagulability. Echocardiography and carotid ultrasound Doppler were normal. Ultrasound Doppler of the orbital vessels revealed no vascular abnormalities in the ophthalmic, posterior choroidal and central retinal arteries. To demonstrate any vascular abnormalities that may have been undetected by the previous modalities, magnetic resonance angiography of the head and neck was performed, and was found normal.

A diagnosis of transient CRAO was made, and the patient was treated with aspirin. He was followed closely for 6 weeks, during which BCVA improved to 20/20, the CWS completely resolved, and only mild foveal retinal pigment epithelial changes remained. HR-OCT documented the gradual disappearance

BCVA = best corrected visual acuity

[A] Presentation of the right eye with multiple cotton wool spots around the optic disk and along the arcades. **[B]** Fluorescein angiography at end-transit time, demonstrating areas of hypofluorescence (arrows) that match the size and location of the cotton wool spots seen in [A]. **[C]** ICG demonstrating areas of hypofluorescence (arrows) that match the size and location of the cotton wool spots. Choroidal filling is normal. **[D–F]** Spectralis HR-OCT scans demonstrating the gradual absorption of two cotton wool spots (arrows) and restoration of normal retinal configuration, **[D]** at presentation, **[E]** after 2 weeks, and **[F]** after 6 weeks



of the cotton wool spots and restoration of normal retinal configuration [Figure D-F]. Repeated visual field testing after 6 weeks was normal.

COMMENT

We report a case of multiple CWS secondary to transient CRAO. To our knowledge, this is the first case in which multiple imaging modalities were used to confirm the suspected diagnosis. This is the first report on the use of ultrasound Doppler of orbital vessels and MRA to ascertain patency of the central retinal artery in transient CRAO.

HR-OCT is a valuable tool for identifying retinal abnormalities, and a review of the literature revealed a few studies, which used this modality, on the appearance of CWS. Only two studies have shown that HR-OCT is efficient in demonstrating CWS in both acute and late stages [5]. HR-OCT has also been shown to detect retinal hyper-reflectivity even after CWS resolution, consistent with residual retinal damage [5]. Our data are consistent with these studies.

Fluorescein angiography and ICG aided in establishing the diagnosis of transient CRAO. The arterial filling delay is highly suggestive of CRAO (both complete and transient). ICG ruled out ocular ischemic syndrome and other chorioretinal pathology. Unique to our case is the demonstration of CWS by ICG, which were seen as focal areas of hypofluorescence.

After CWS resolution, foveal retinal pigment epithelial changes were noted, which may result from an initial perifoveal ischemia [1,3]. It has been suggested that foveal retinal thickening may prevent reperfusion of the fovea, which may result in foveal ischemia. This mechanism has been dubbed "no-reflow phenomenon," and may explain residual ischemic changes or even a central scotoma [4]. As occurred in most reports of patients with transient CRAO, our patient's BCVA improved and his visual field returned to normal.

In summary, our case emphasizes the role of different imaging techniques in establishing the rare diagnosis of transient CRAO. HR-OCT is very helpful in identifying and monitoring CWS. Fluorescein angiography and ICG aid in ruling out other potential causes for a similar clinical presentation. Ultrasound Doppler of the orbital vessels may be a significant adjunct to these tests, as it may directly demonstrate occlusion or patency of the arteries in question. MRA may be useful in finding the cause of CRAO, but since it is relatively expensive and less available we suggest it be reserved for cases in which all other modalities fail to reveal any significant findings and where the clinical picture supports its use.

Acknowledgments

The authors thank Mrs. Galit Yair-Pur for her photographic and technical support.

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MRA = magnetic resonance angiography

ICG = indocyanine green (angiography)