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Combined central retinal artery and medial posterior ciliary artery occlusion: Localizing the lesion

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ABSTRACT

Purpose: To report a rare case of a combined central retinal artery (CRA) and medial posterior ciliary artery (MPCA) occlusion due to an atherosclerotic lesion in the common trunk supplying both arteries.

Observations: A 75-year-old man presented with acute vision loss associated with elevated intraocular pressure in the right eye. Multi-modal imaging revealed a combined retinal and choroidal infarction in the distribution of the CRA and MPCA, localizing the lesion to the common trunk of the ophthalmic artery supplying both the CRA and MPCA. Neurovascular imaging provided supportive evidence for the diagnosis.

Conclusions and importance: A simultaneous retinal and choroidal vascular occlusion is an uncommon presentation. Familiarity with the anatomy of the ophthalmic arteries and its branches facilitates localizing the lesion.

Disclosures

None.

Summary statement

We demonstrate a case of a combined central retinal artery and medial posterior ciliary artery occlusion localized by ophthalmic imaging and computed tomography angiography of the orbits to a common trunk that supplies both vessels.

1. Introduction

A combined central retinal artery (CRA) and medial posterior ciliary artery (MPCA) occlusion is a rare phenomenon with few cases documented in the literature. Patients present with profound vision loss and a relative afferent pupillary defect in the affected eye.[1–3] While embolism secondary to atherosclerotic disease is the most common cause of retinal artery occlusion, systemic vasculitides including giant cell arteritis must be considered.[4] Diagnosis of posterior ciliary circulation compromise is usually inferred from delayed choroidal filling and evidence of Amalric choroidal infarcts on fluorescein angiography (FA).[5] Choroidal infarcts have also been associated with various etiologies including sickle-cell hemoglobinopathy, cocaine use, and panretinal photocoagulation (PRP).[6–8] Though exceedingly rare, iatrogenic injury resulting in posterior ciliary artery occlusion during spinal surgery and middle meningeal artery embolization has also been reported.[9,10]

An understanding of the anatomy of the ophthalmic artery and its branches explains the unusual occurrence of a combined central retinal artery and medial posterior ciliary occlusion. In a seminal cadaveric study published in 1962, Hayreh noted that when the ophthalmic artery crosses over the optic nerve as it travels toward the globe, the first branch is most often a common trunk supplying both the CRA and MPCA.[11] The MPCA supplies the nasal choroid. The lateral posterior ciliary artery (LPCA), supplying the temporal choroid, arises from the ophthalmic artery separately.[12] An embolic or thrombotic event at this common trunk would therefore cause occlusion of both the CRA and MPCA.

We present a rare case of profound vision loss in which the eye exam and retinal imaging led to the diagnosis of a combined CRA and MPCA occlusion, with computed tomography angiography (CTA) of the orbits lending support to the hypothesis that an occlusion of the common trunk supplying the CRA and MPCA is the causative mechanism. To our knowledge, this is the first report in the literature to describe this rare phenomenon with multimodal imaging and supportive neuroimaging, offering *in vivo* confirmation of the findings from seminal cadaveric

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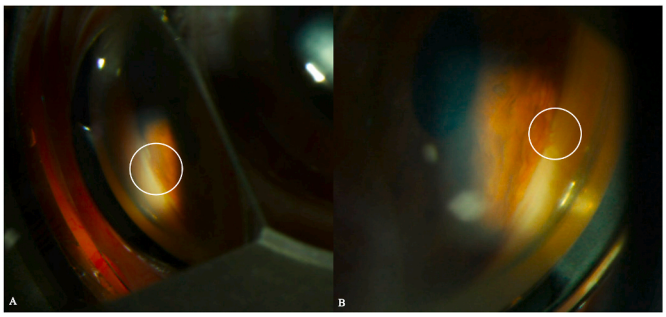

Fig. 1. Slit lamp gonioscopy photograph of the right eye shows **(A)** peripheral anterior synechiae and **(B)** lacy neovascularization of the angle.

studies on ophthalmic artery branching conducted by Hayreh more than 50 years prior.

2. Case report

A 75-year-old Caucasian man was referred to the Retina clinic with a history of 2 months of sudden profound vision loss in the right eye. Ocular history was significant for pseudophakia in the right eye and ocular hypertension in both eyes, for which he was using timolol daily in both eyes. Medical history was notable for essential hypertension.

On examination, his visual acuity was count fingers in the right eye and 20/30 in the left eye. Intraocular pressure (IOP) was 52 mm Hg in the right eye and 17 mm Hg in the left eye. A right afferent pupillary defect was noted. Gonioscopy of the right eye revealed neovascularization of the angle with peripheral anterior synechiae (Fig. 1). Peripapillary cotton wool spots and subtle nasal triangular zones of retinal pigment epithelium changes, consistent with Amalric choroidal infarction, were observed on fundus examination (Fig. 2A). The optic nerve was noted to have moderate cupping, mild pallor and no disc hemorrhages. Examination of the left eye was unremarkable. FA was notable for marked absence of dye in the nasal choroid in the early frames and marked delay in retinal arterial filling, which began 1 minute 15 seconds after dye injection (Fig. 2B). Late phase hyperfluorescent staining corresponding to the triangular zones of RPE changes was noted, consistent with Amalric choroidal infarcts (Fig. 2C; 2D). Spectral domain optical coherence tomography (SD-OCT) demonstrated diffuse inner retinal thinning in the right macula, consistent with retinal ischemia due to retinal artery occlusion (Fig. 3).

A systemic workup was pursued. Westergren erythrocyte sedimentation rate (ESR) and C-reactive protein were unremarkable, and a normal temporal artery biopsy ruled out giant-cell arteritis. Interferon gamma release assay was negative, ruling out tuberculosis. Syphilis antibody was negative. An aqueous fluid specimen sent for PCR was negative for the Herpesviridae (VZV, HSV, and CMV). Carotid duplex

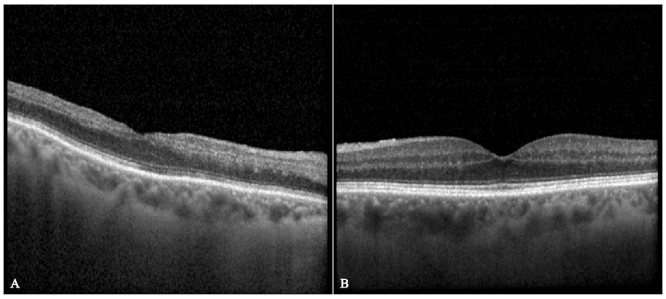

Fig. 3. Spectral domain optical coherence tomography (SD-OCT). **A.** SD-OCT of the right macula shows diffuse inner retinal thinning consistent with an ischemic insult. **B.** Normal SD-OCT of the left macula.

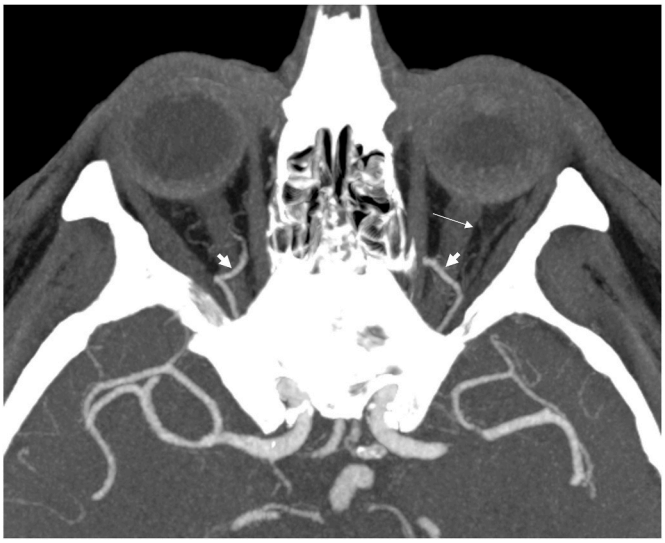

Fig. 4. Computed tomography angiography, focused view centered on orbits. The ophthalmic arteries cross over the optic nerves bilaterally (short arrows). Note enhancing tortuous vessel (long arrow) in the left orbit, not detectable in the right, suggestive of an occlusion of the common trunk supplying the CRA and MPCA in the right.

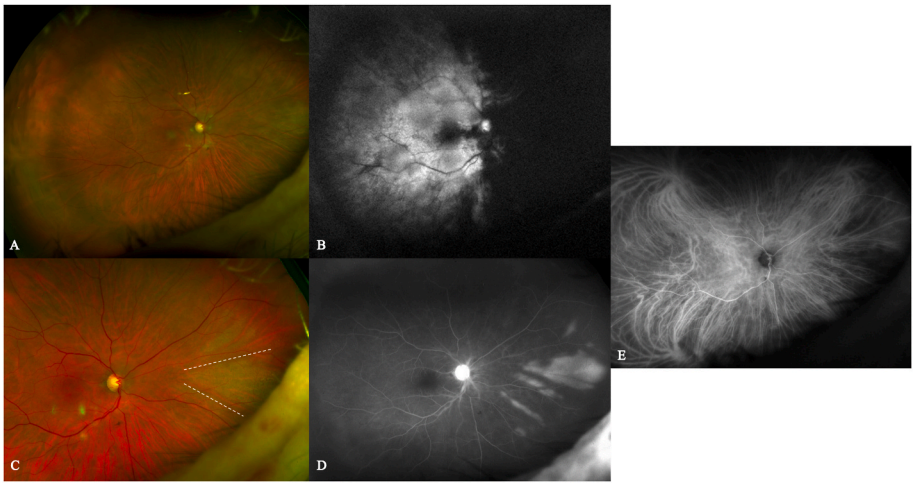

Fig. 2. A. Ultra-widefield fundus image showing peripapillary cotton wool spots and mild pallor of the optic nerve in the right eye. No disc hemorrhage or optic disc edema is observed. **B.** Fluorescein angiography (FA) shows absent central retinal artery (CRA) flow and non-perfusion of the nasal choroid indicating occlusions in both the CRA and the medial posterior ciliary artery. **C.** An Amalric choroidal infarct (demarcated by white dashed lines) is noted in the nasal periphery. **D.** Late phase FA is notable for disc leakage, diffuse vascular leakage and hyperfluorescent staining of the Amalric choroidal infarct. **E.** Late phase indocyanine angiography (ICGA) is notable for hypofluorescence in the area of the Amalric choroidal infarct.

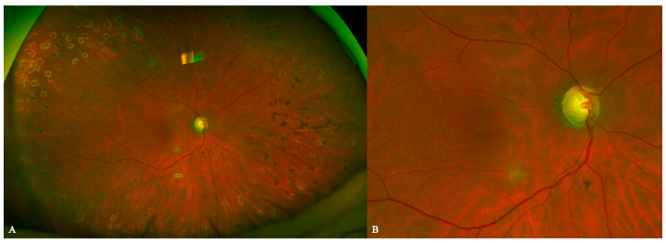

Fig. 5. A. At the most recent follow up, ultra-widefield fundus image of the right eye shows resolution of the peripapillary cotton wool spots seen on initial imaging and panretinal photocoagulation chorioretinal scars. **B.** The optic nerve appears mildly pale with moderate cupping and no disc hemorrhages. The macula is flat.

ultrasonography and transthoracic echocardiography were unremarkable. An extensive autoimmune workup, including consultation with rheumatology, was also unremarkable. Computed tomography angiography (CTA) of the head, neck and orbits showed that the ophthalmic artery crosses over the optic nerve as it heads toward the globe, with absent enhancement in a branch of the right ophthalmic artery, likely corresponding to an occlusion of the common trunk supplying the CRA and MPCA (Fig. 4). Atherosclerotic changes affecting the aorta and carotid vessels were noted on CT angiography. Chronic small vessel ischemic disease of the brain was also noted on CT of the head.

The patient was subsequently diagnosed with a combined CRA and MPCA occlusion due to an occlusion of the common trunk from the ophthalmic artery supplying the CRA and MPCA, likely due to arteriosclerosis or atherosclerosis. He was also diagnosed with neovascular glaucoma, likely a consequence of the retinal ischemia. In conjunction with oral and topical IOP-lowering medications, the patient was treated with panretinal photocoagulation and anti-vascular endothelial growth factor (VEGF) injections (bevacizumab). He was also prescribed aspirin by his primary care physician.

At his most recent follow up, one year following initial presentation, the patient's right eye visual acuity was stable at count fingers and his IOP was 18 mm Hg, maintained on dorzolamide-timolol. The retina was well treated with panretinal photocoagulation and no neovascularization was noted (Fig. 5).

3. Discussion

Retinal vascular occlusions are seen commonly in the retina clinic, but it is uncommon for occlusions to involve both the retinal and choroidal circulation simultaneously. This case demonstrates the importance of multimodal imaging in evaluating these cases. The FA showed both markedly delayed filling of the nasal choroid, indicating an occlusion of the choroidal circulation, and markedly delayed retinal arterial filling, indicating an occlusion of the retinal circulation. These FA findings confirm a diagnosis of a combined CRA and MPCA occlusion, a rare diagnosis. A cherry red spot, which is typically seen in acute cases of CRA, was not noted as he presented 2 months after vision loss. The branching patterns of the ophthalmic artery, as described by Hayreh, offer a compelling explanation for this unique phenomenon: the existence of a common trunk arising from the ophthalmic artery that supplies both the CRA and MPCA when the ophthalmic artery crosses over the optic nerve as it travels toward the globe, which is likely occluded. An illustration of the ophthalmic artery branching pattern, as described by Hayreh, is provided in Fig. 6. The CTA performed in this case, which shows that the ophthalmic artery indeed crosses over the optic nerve as it travels toward the globe, and absent enhancement in a branch of the ophthalmic artery on the right, relative to the left, provides additional support for an occlusion of the common trunk as the culprit.

As with any diagnostic dilemma, it is important to consider infectious and inflammatory etiologies in the initial evaluation. An extensive workup was unremarkable, leading us to conclude that arteriosclerosis or atherosclerosis, common causes of small vessel disease, likely led to the occlusion in this patient with essential hypertension. While he has unfortunately suffered permanent damage from the retinal and choroidal ischemia, treatment with PRP and intravitreal bevacizumab has resulted in regression of the neovascularization of the angle and stabilized his disease. He was prescribed aspirin by his primary care physician for secondary prevention of atherosclerotic disease.

In summary, we present a rare case of a combined CRA and MPCA occlusion in which multimodal imaging led to the correct diagnosis, supported by CTA of the orbit. This case highlights the importance of familiarity with the anatomy of the ophthalmic artery and its branches to localize the lesion responsible for this atypical presentation.

Patient consent

Written consent to publish this case has not been obtained. This report does not contain any personal identifying information.

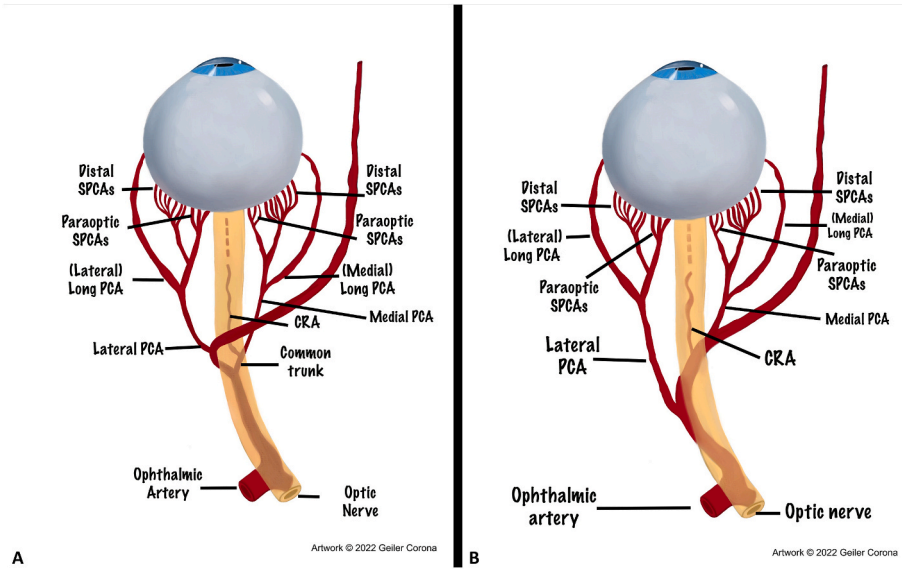

Fig. 6. A. The first branch to come off the ophthalmic artery when it crosses over the optic nerve is a common trunk supplying both the central retinal artery (CRA) and the medial posterior ciliary artery (Medial PCA). An occlusion of the common trunk leads to a combined CRA/MPCA occlusion. B. Conversely, when the ophthalmic artery crosses under the optic nerve, the CRA and medial PCA arise as separate branches of the ophthalmic artery. SPCAs = short posterior ciliary arteries. PCA = posterior ciliary artery. (Artwork © 2021 Geiler Corona. License for use provided to Kareem Moussa.)

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Authorship

All authors attest that they meet the current ICMJE criteria for Authorship.

Declaration of competing interest

All authors in this manuscript have no financial disclosures.

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