A Case of Persistent Ophthalmic Artery Occlusion after Migraine in the Setting of Cerebral Vascular Anomalies

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Background

Ophthalmic Artery Occlusion: A sight threatening condition where blood flow to the retinal tissue is interrupted via full or partial occlusion of the ophthalmic artery

- Anatomy:
  - In the majority of the population, the central retinal artery is responsible for the blood supply of the inner retina while a minority also have a collateral cilioretinal artery supplying the macula.
  - Posterior ciliary artery supplies the choroid
  - Both are supplied by the ophthalmic artery which is in turn a branch of the internal carotid artery
- Etiology:
  - Embolism
  - Thrombosis from atherosclerotic disease, carotid artery disease, hypercoagulable states, diabetes, collagen vascular disease, infectious disease, and rarely, migraines
- Signs and Symptoms:
  - Vision Loss
  - Relative afferent pupillary defect
  - Narrowing and boxcarring of retinal arterioles due to segmentation of blood flow
  - Global paling of the retina rather than the characteristic cherry red spot of the more common central retinal artery occlusion
- Treatment:
  - Investigate underlying etiology and minimize risk for secondary ischemic events

Retinal Migraines: Transient and in rare cases permanent monocular vision loss not better explained by amaurosis fugax in association with a migraine.

- Etiology:
  - Vasospasm caused by substance P and nitric oxide release leading to extravasation of plasma protein and irritation of local structures innervated by trigeminal nerve
- Diagnosis:
  - Aura with reversible monocular positive/negative symptoms
  - And two of the following: gradual spread of aura over 5+ minutes, aura lasting 5-60 minutes or headache during or within 60 minutes of aura
- Risk factors:
  - female, 2nd-3rd decade of life, alcohol, tobacco, oral contraceptives, nasal decongestants, selective serotonin reuptake inhibitor (SSRI), hypertension
- Treatment:
  - Migraine preventative strategies (calcium channel blockers, antiepileptics, tricyclic antidepressants)
  - Aspirin and risk modification

Case

37-year-old African American female presents with sudden vision loss in the right eye associated with migraine.

Past Medical History: Migraine with aura, seasonal allergies, tobacco use

Past Ocular History: Numerous episodes of transient vision loss associated with migraines (undiagnosed)

Family History: Blindness, diabetes, heart disease, hypertension, colon and breast carcinoma

Medications:

- Topiramate 50mg QD (for migraine)
- Fluticasone furoate 100mcg QD, cetirizine 10mg QD, hydroxyzine hydrochloride 25mg PRN (for seasonal allergies)
- Prednisone 60mg QD taper (prescribed by discharging hospital 4 days prior after being treated for presumptive giant cell arteritis

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Case and Figures

Presentation

- Visual acuity: no light perception (right eye), 20/25+2 (left eye)
- IOP: 10mmHg (right eye), 12mmHg (left eye)
- OCT macula: central foveal subfield thickness 82µm right eye with diffuse intraretinal edema
- Ocular exam: +RAPD right eye, ocular pain on movement (ocular pain improved with prednisone)
- Carotid doppler: within normal limits
- MRI/MRA:
  - Sluggish vascular flow in right ophthalmic artery
  - Congenital aplasia of bilateral posterior communicating arteries (PCoA)
  - Early transition to A/P branch of right middle cerebral artery (MCA) at sphenoidal (M1) segment with decreased flow
- B Scan of right eye: attached retina, proximal hyperlucency of optic nerve
- Labs: negative for syphilis, lyme, tuberculosis, HIV, RA, SLE, COVID-19, dyslipidemia. CBC, ESR/CRP levels within normal limits. Positive for reduced levels of antithrombin III
- **Diagnosis: Ophthalmic artery occlusion**

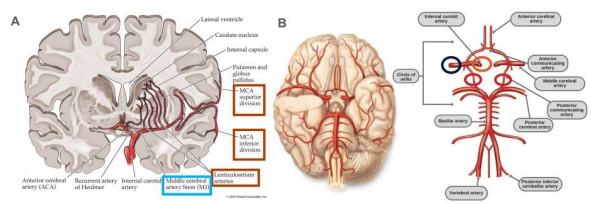

Figure 1: (A) Branches of the middle cerebral artery (adapted from Rhoton AL et al.) Our patient had an early branching of the MCA at the M1 segment (highlighted in blue) with resulting decreased flow. (B) Circle of Willis overview (adapted from Ashish). Our patient had bilateral absent posterior communicating arteries (green). Middle cerebral artery had decreased flow due to early branching on the right (blue). Ophthalmic artery (branch of internal carotid artery) had decreased flow on the right (orange).

Figure 2: Fundus photo OD. Pale optic nerve, diffuse NFL edema, attenuated retinal vessels with boxcarring. 2+ vitreous cell. 6mm optic disc diameter.

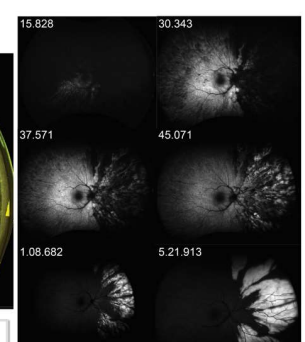

Figure 3: IV fluorescence angiography OD from 15.828 seconds to 5.21913 minutes. Global absence of retinal perfusion with minimal patchy choroidal filling.

Discussion

Vascular variations: The significance of variations of the PCoA and MCA are generally neurosurgical. Clinical significance is controversial with investigation showing an increased risk of ischemic stroke in the small vessels of the thalamus with PCoA aplasia and increased risk of aneurysm with early MCA branching. Even though the PCoA and MCA do not have direct connections to the ophthalmic artery, they are both part of the circle of Willis which does share a connection and creates vital collateral circulation.

Optic neuritis symptoms: With pain on eye movement controlled with prednisone and a proximal optic nerve hyperlucency found on ultrasound, multiple sclerosis or some other inflammatory process was suspected. Multiple sclerosis is an autoimmune process shown to be associated with impaired retrobulbar hemodynamics. These hemodynamic changes are thought to come about secondary to optic neuritis and retinal vasculitis causing decreased flow in the central retinal arteries and posterior ciliary arteries, or via endothelin-1, a vasoconstrictor that could increase blood flow resistance. Further workup however, ruled out multiple sclerosis.

This patient likely suffered from a constellation of risk factors for vasospasm and hypercoagulability that led to permanent vision loss in the right eye:

- History of retinal migraines secondary to vasospasm
  - Risk factors: tobacco use, age (37), gender (female), topiramate (SSRI mechanisms), nasal decongestants, and positive history
- Inflammatory process potentially indicated by the proximal hyperlucency
- Antithrombin mutation: associated with hypercoagulability
- COVID-19: Even though the patient was negative, post-COVID-19 hypercoagulable state associated with central retinal artery occlusion has been documented
- Congenital vascular variants
  - Bilateral PCoA aplasia: increase ischemic stroke risk and decrease collateral circulatory options
  - Early branching of the MCA with decreased blood flow potentially leading to stasis and clot formation
  - Family history of blindness could be related to these variants

Overall this is the first case of persistent ophthalmic artery occlusion in association with various hypercoagulable and vasospastic states including retinal migraines and congenital vascular anomalies.

This case report highlights the importance of considering an anatomic origin of monocular vision loss during workup especially with a family history of blindness.

References

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Full list of references available on request.

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