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Retinal Artery Occlusions

Retinal artery occlusions are a rare but serious ocular finding. In addition to causing permanent vision loss or deficits, retinal artery occlusions may portend a serious underlying medical issue and are associated with a significant risk for subsequent stroke and cardiovascular disease. As such, retinal artery occlusions provide an opportunity for timely diagnosis by an eyecare specialist to be truly life-saving.

In this review, we present three cases that highlight the variable causes of retinal artery occlusions and the need for prompt systemic workup in patients with this diagnosis.

Case 1:

A 44-year-old man with no known past medical history presented with one week of vision changes in the left eye. He reported the sudden onset of a "shade" blocking the bottom half of his vision in the left eye. There was no associated eye pain, flashes, floaters or headache, so he did not present for evaluation initially because he thought his symptoms would resolve. On examination, his vision was 20/20 in the right eye and 20/25 in the left. Intraocular pressure was normal in each eye. There was a subtle afferent pupillary defect in the left eye and a dense scotoma of the inferior visual field was noted on confrontational testing in the left eye.

Fundus examination was normal in the right eye, but in the left eye demonstrated diffuse whitening of the superior hemi-retina as well as an intravascular thrombus consistent with a retinal artery occlusion (Figure 1).

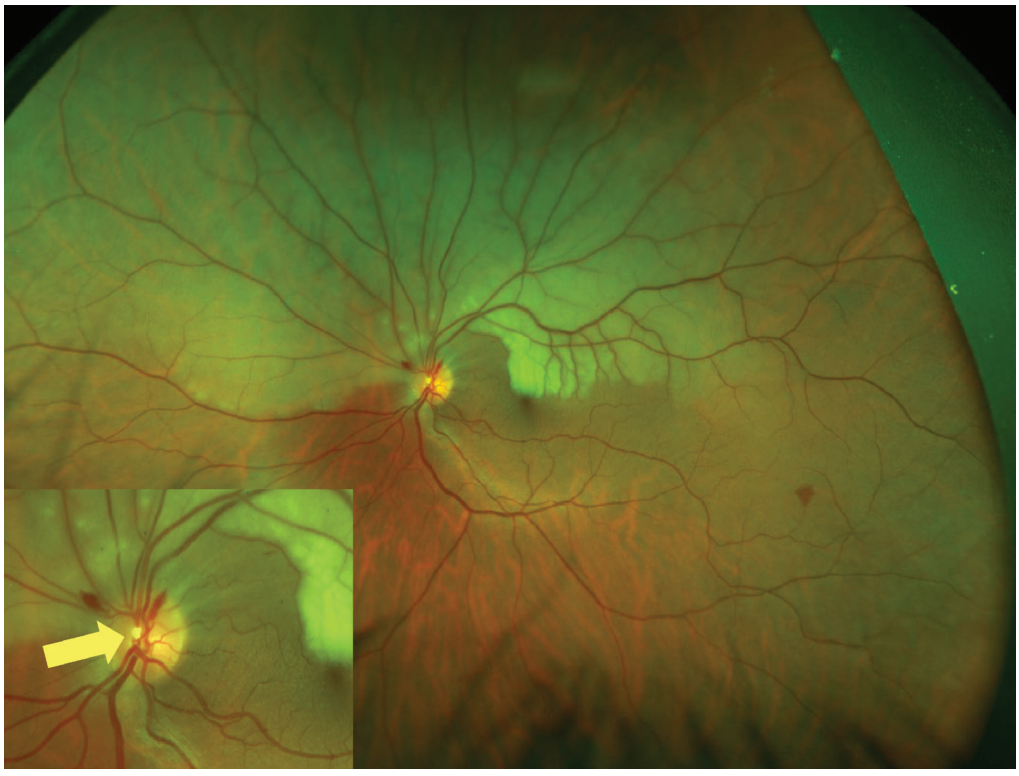

Figure 1

The patient was sent directly to the Emergency Department for prompt evaluation and workup. A transthoracic echocardiogram was performed which revealed severe aortic valve calcification and aortic stenosis due to a congenital bicuspid valve. The patient was started on systemic anticoagulation by cardiology and underwent an aortic valve replacement shortly thereafter. The AVR surgery was done to mitigate risk of subsequent ischemic events and thus the retinal diagnosis led identifying and fixing a more serious problem.

Case 2:

An 80-year-old woman with a history of systemic hypertension presented for evaluation of a sudden-onset painless "shade" coming down over the top half of her vision in the right eye. Visual acuity was 20/20 in each eye but there was a superonasal visual field defect on confrontational testing in the right eye. Fundus examination demonstrated a Hollenhorst plaque near the inferior optic disc margin and retinal whitening along the inferotemporal arcade. Fluorescein angiography was obtained which demonstrated markedly delayed and partially obstructed flow within the inferotemporal arteriole, consistent with branch retinal artery occlusion (Figure 2). Of note, fundus autofluorescence images highlighted calcific plaques at the disc margin in both eyes, including the asymptomatic left eye (Figure 3A and 3B).

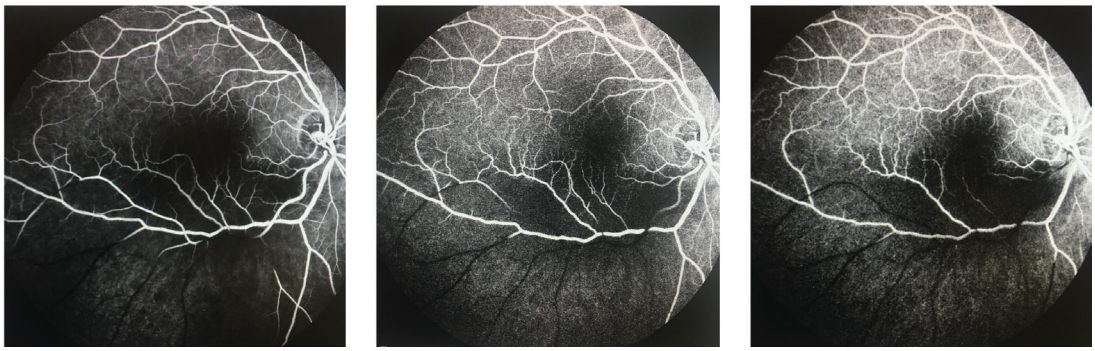

Figure 2

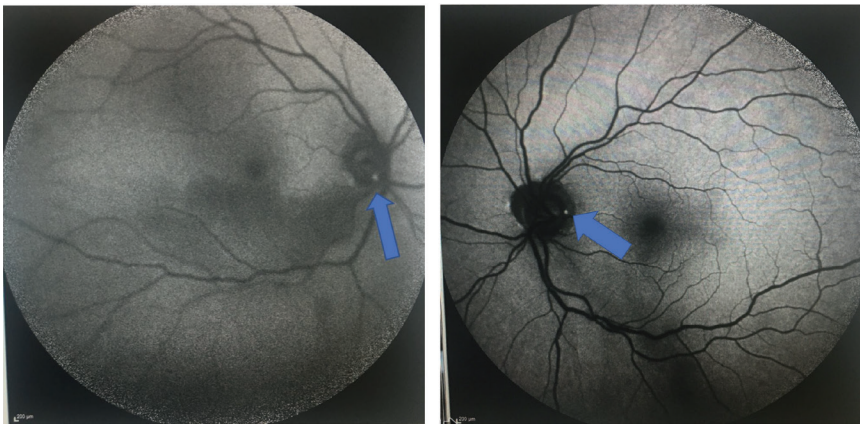

Figure 3A and 3B

The patient was sent to the hospital for prompt evaluation. EKG was remarkable for atrial fibrillation and hyperlipidemia was noted on laboratory testing. The patient was started on systemic anticoagulation and a statin and was discharged from the hospital with a Holter monitor and further follow-up with cardiology.

Case 3:

A 61-year-old man with no known past medical history presented with five days of vision loss in the left eye. He described the sudden, painless onset of a black area in the left eye that had gotten larger and denser since onset. He could no longer make out shapes or figures in the left eye.

On examination, visual acuity was 20/20 in the right eye and hand motions in the left eye. There was an afferent pupillary defect in the left eye. Anterior examination was otherwise normal for age. Dilated fundus exam in the right eye demonstrated arteriolar attenuation and A-V nicking. In the left eye, there was marked retinal whitening along the arcades and in the macula as well as a "cherry-red spot" consistent with a central retinal artery occlusion (Figure 4).

OCT scan of the left eye showed significant thickening and intracellular edema of the inner retinal layers, but relative preservation of the outer retina, consistent with a central retinal artery occlusion disrupting blood flow to the inner retina.
The patient's blood pressure was checked in the office and found to be 210/138 mmHg. He was sent directly to the emergency department for management of hypertensive urgency. His blood pressure improved with medications and he was admitted to the hospital for titration of blood pressure medications and workup.

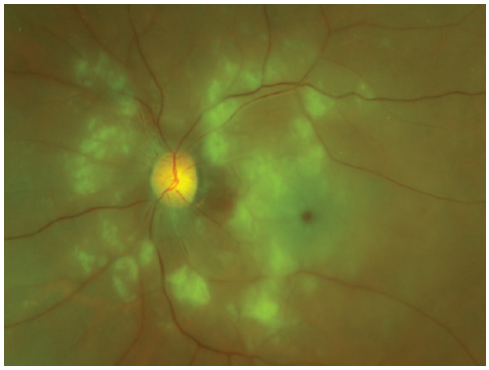

Figure 4

Discussion:

Retinal artery occlusion (RAO) including central retinal artery occlusion and branch retinal artery occlusion are serious and urgent diagnoses. Prompt referral and expedited workup can be truly life-saving in many cases of retinal artery occlusion and may reveal serious conditions such as carotid atherosclerosis, atrial fibrillation, cardiac valve disease and others.[1]

Correctly diagnosing RAO may be difficult depending on the patient's presentation. Retinal whitening takes several hours to become pronounced and may resolve after several days or weeks. Retinal whitening may not be present in patients presenting immediately after symptom onset or in cases of delayed presentation. Emboli are often difficult to appreciate ophthalmoscopically and may break up with time. In a patient with concerning symptoms of sudden painless vision loss, in-office imaging is helpful to establish the correct diagnosis of RAO. Fundus photography provides static, editable images and may help identify emboli that are difficult to appreciate funduscopically (Figure 1).

Fundus auto-fluorescence is especially helpful for identifying small emboli because calcium- containing emboli are brightly hyper auto-fluorescent and are readily apparent on FAF images (Figure 3 A/B).

OCT imaging is also very helpful in the identification of acute and chronic RAO. In acute artery occlusion, OCT demonstrates marked inner retinal edema with loss of the cell layer distinctions. Owing to the dual retinal circulation, the inner retina is supplied by the retinal vessels, and thus becomes ischemic in RAO, whereas the outer retinal layers are supplied by the choroid and remain relatively preserved in RAO.[2] Weeks to months after RAO, the edema resolves, and the ischemic inner retina atrophies, resulting in marked retinal thinning and loss of the inner retinal layers, again with relative preservation of the outer retinal layers – this finding is often most readily apparent on the macular thickness scan which reveals diffuse thinning in CRAO or sectoral thinning that respects the horizontal and is confined to a vascular territory in the case of BRAO.

Finally, fluorescein angiography can be used to assess ocular perfusion and will demonstrate significantly delayed arterial filling times in RAO (Figure 3). Unfortunately, there are no evidence-based ocular therapies for retinal artery occlusion, so the main emphasis of care is to identify systemic risk factors or embolic sources and enable appropriate systemic therapy.[1,3,4] Eye care providers may measure the patient's blood pressure, administer a chewable aspirin and consider IOP-lowering therapies for acute presentations. Heroics such as intravenous tPA ("clot-busters") or YAG laser to the thrombus are not recommended owing to the high risk of complications and lack of proven benefit.[1] In the eye care setting, the main focus remains correct diagnosis and timely referral to enable secondary prevention of further embolic events.[1,3,4]

Acute, symptomatic retinal artery occlusion is considered a stroke equivalent.[1,5] Patients presenting within a week of RAO symptom onset are at extremely elevated risk of stroke and cardiovascular complications and should be sent to the nearest stroke center or capable ED for urgent evaluation.[6,5] At minimum, workup should include an electrocardiogram to rule out cardiac arrhythmia, echocardiogram to look for cardiac valve calcification or structural heart disease, carotid imaging to evaluate for atherosclerosis and a CT or MRI brain to rule-out concurrent stroke (Table 1). In patients younger than 50, laboratory testing should include causes of hypercoagulability and vasculitis such as antiphospholipid antibodies, Factor V Leiden and malignancy. In patients older than 50 years of age, it is prudent to include giant cell arteritis in the differential and to include ESR, CRP and platelets in lab testing and consider starting steroids and arranging for temporal artery biopsy if there is any clinical suspicion of GCA.

Although vision is unlikely to improve after RAO, it is nonetheless necessary to monitor these patients closely to ensure that no retinal or iris neovascularization develop. If neovascularization does develop, this complication can be treated with Anti-VEGF injections followed by panretinal photocoagulation to the ischemic retina. Patients with poor vision in one eye after RAO should be advised to follow monocular precautions to help prevent vision loss in the fellow eye.

References:

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6. *Park SJ, Choi N-K, Yang BR, et al. Risk and Risk Periods for Stroke and Acute Myocardial Infarction in Patients with Central Retinal Artery Occlusion. Ophthalmology 2015;122:2336- 2343.e2.*

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At the Forefront of Clinical Research NJRetina currently conducts clinical trials at key locations. Our clinical research coordinators who conduct the trials will be happy to discuss the inclusion/ exclusion criteria or any other aspect of these studies with you or your patients. If you have any questions, please feel free to contact:

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Enrolling Studies:

Dry AMD

Teaneck & Edison (Closed)

A Genetic Screening and Registry Study to Evaluate Long-term Clinical Outcomes and Disease Progression in Subjects with Non-Central Geographic Atrophy (GA) Who Are Carriers of High-Risk Genetic Complement Variants Associated with Dry Age-related Macular Degeneration (AMD) A Prospective Natural History Study to Evaluate Clinical Characteristics and Disease Progression in Subjects with Non-Central Geographic Atrophy (GA) Who Are Carriers of High-Risk Genetic Variants of Complement Factor H (CFH)

Teaneck (Closed)

Phase II, Randomized, Double-Masked, Placebo-Controlled Clinical Study to Evaluate the Safety, Efficacy, and Pharmacokinetics of Subcutaneous Injections of Elamipretide in Subjects with Age-Related Macular Degeneration with Geographic Atrophy (SPIAM)

Teaneck & Toms River

A Phase II, Multi-Center, Randomized, Single-Masked, Sham Injection Controlled Study of the Safety, Tolerability, and Evidence of Activity of Intravitreal Injection of R7171009 in Patients with Geographic Atrophy Secondary to Age-Related Macular Degeneration (Gallego)

Vauxhall

A Study of Disease Progression in Genetically Defined Patients with Geographic Atrophy Secondary to Age-Related Macular Degeneration (Gyroscope)

Wet AMD

Edison and Teaneck (Closed)

A Randomized, Single-Masked, Active-Controlled Phase 2 Study of the Safety, Tolerability, and Efficacy of Repeated Doses of Hight-Dose Aflibercept in Patients with Neovascular Age-Related Macular Degeneration

Soon to Enroll Studies:

Dry AMD:

- A Phase 2 Multicenter, Randomized, Double-Masked, Sham-Controlled Study of the Safety and Efficacy of Intravitreal Injections of NGM621 in Subjects with Geographic Atrophy (GA) Secondary to Age-Related Macular Degeneration (AMD) (NGM study) – Teaneck

Wet AMD:

- A Randomized, Double-Masked, Active-Controlled Phase 2/3 Study of the Efficacy and Safety of High - Dose Aflibercept in Patients with Neovascular Age-Related Macular Degeneration (Pulsar) – Teaneck and Edison

Diabetic Retinopathy:

- A Phase 2, Randomized, Dose-escalation, Observation-controlled Study to Evaluate the Efficacy, Safety, and Tolerability of RGX-314 Gene Therapy Delivered via One or Two Suprachoroidal Space (SCS) Injections in Participants with Diabetic Retinopathy (DR) Without Center Involved-Diabetic Macular Edema (CI-DME) (ALTITUDE) – Teaneck