Acute unilateral visual disturbance

The diagnosis in this case was a bit surprising, especially since the patient was relatively young—and healthy.

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A PREVIOUSLY HEALTHY 37-YEAR-OLD RUNNER presented to his primary care physician with acute-onset floaters and scotoma in his left eye, which he first noticed less than 24 hours earlier. He denied eye pain, diplopia, headache, fever, chills, slurred speech, weakness, or other focal neurologic deficits. His vital signs were normal.

Despite the acute visual disturbances, visual acuity was 20/20 in both eyes with corrective lenses; pupils were equal, round, and reactive to light and accommodation; and extraocular movements were intact. On a dilated funduscopic exam, the physician discovered edema of the optic cup, tortuous vasculature, and microhemorrhages in the left eye (**FIGURE**).

- WHAT IS YOUR DIAGNOSIS?
- HOW WOULD YOU TREAT THIS PATIENT?

FIGURE

Dilated funduscopic exam at presentation

IMAGES COURTESY OF NEHMAN ANDRY, MD

Funduscopic exam shows the unaffected right eye (**A**) and the affected left eye (**B**) with edema of the optic cup, tortuous vasculature, and microhemorrhages.

Diagnosis: Central retinal vein occlusion

The patient was given a diagnosis of central retinal vein occlusion (CRVO). In this condition, a blockage causes the central retinal vein to leak blood and excess fluid into the retina. This fluid can collect in the macula, leading to visual disturbance.

Retinal vein occlusion is the second most common retinal vascular disease in the United States and is one of the most common causes of vision loss in the elderly.[1] Advancing age (≥ 70 years), increasing mean arterial blood pressure, and retinal atherosclerotic signs (focal narrowing, arteriovenous nicking, and opacification) are significant predictors of retinal vein occlusion.[2] Other risk factors include diabetes, hyperlipidemia, cardiovascular disease, smoking, obesity, hypercoagulable state, and glaucoma.[3-7] However, retinal vein occlusion may also occur in younger, healthier patients who lack the aforementioned risk factors. In such cases, thrombophilic risk factors should be considered.[8]

CRVO is classified as either ischemic or nonischemic (perfused) based on retinal angiography. More than 80% of CRVO cases are nonischemic,[9] of which the majority has visual acuity better than 20/400, mild or no pupillary defect, and mild, unilateral visual changes.[10] Nonischemic CRVO can progress to ischemic CRVO, which can result in permanent vision loss. Visual outcome is good in nonischemic CRVO and poor in ischemic CRVO.[11] Early detection of poor prognostic features, such as macular edema and neovascularization, is essential for minimizing the risk for permanent damage.[12]

Dilated funduscopic exam of a patient with CRVO may reveal widespread retinal hemorrhages, markedly dilated and tortuous retinal vessels, cotton wool spots, optic disc or macular edema, and/or vitreous hemorrhages.[10]

Cases of central retinal vein occlusion have been linked to dehydration as well, with acute vision changes occurring after strenuous exercise.

Differential includes varied conditions that can affect vision

CRVO may manifest similarly to the following:

Proliferative diabetic retinopathy can manifest with retinal edema or vitreous and retinal hemorrhages, which also are seen in CRVO.[13] Macular edema, retinal hemorrhage, and neovascularization on the optic disc or retinal surface also may be seen on funduscopy in proliferative diabetic retinopathy.[14] However, proliferative diabetic retinopathy is often bilateral and gradual in onset in patients with longstanding, uncontrolled diabetes.

Hyperviscosity retinopathy, which is commonly caused by plasma cell and erythrocyte disorders, also manifests similarly to CRVO. Two noticeable differences include its bilateral presentation and Roth spots, neither of which are commonly seen in CRVO. In addition to visual abnormalities, mucosal bleeding and neurologic abnormalities complete the classic triad of hyperviscosity.[15]

Ocular ischemic syndrome is often confused with diabetic retinopathies and CRVO on funduscopy. However, patients with this condition may have narrowed retinal arteries, perifoveal telangiectasias, and periorbital pain—findings rarely seen in CRVO.[16] Because ocular ischemic syndrome is a manifestation of severe carotid artery atherosclerosis, constitutional symptoms also may be present.

The work-up

When CRVO is suspected, an extensive laboratory work-up is necessary to determine the underlying etiology, including: blood pressure, electrocardiogram, complete blood count, random glucose level, electrolytes, lipid panel, plasma protein electrophoresis, thyroid function tests, and inflammatory markers.[1]

Additional testing may be required for younger patients who lack vasculopathic risk factors, who have bilateral CRVO, or who have a personal or family history of thrombosis.[1] These patients should be screened for thrombophilia, hypercoagulable disorders, and homocysteinuria.[1]

Cases of CRVO have been linked to dehydration as well, with acute vision changes occurring after strenuous exercise, excessive vomiting, or extended periods of fasting.[17-19]

Treatment may include injections, surgery, or nothing at all

Currently, there are no proven treatments to reopen occluded retinal veins. Thus, management is directed at complications that contribute to vision loss, including macular edema

and neovascularization.[20-21] Intravitreal anti-vascular endothelial growth factor (VEGF) agents are recognized as first-line therapy for macular edema in numerous studies.[22-26] Intravitreal corticosteroids are an alternative treatment for patients with macular edema who do not respond to anti-VEGF therapy; however, monitoring is required as these corticosteroids increase the risk for glaucoma and cataract formation.[27] In patients with CRVO with neovascularization, panretinal laser photocoagulation may be used.[28]

Observation and monitoring for the development of complications, rather than initiation of treatment, is appropriate for patients with CRVO without macular edema or neovascularization, with follow-up intervals and duration dictated by the severity of visual loss and whether the CRVO was ischemic or nonischemic.

Our patient's diagnosis was confirmed by retinal specialists with optic coherence tomography, gonioscopy, and fluorescein angiography. He underwent an extensive laboratory work-up and hypercoagulation studies to determine the etiology. All results returned within normal limits with the exception of a nonspecific pattern found on serum protein electrophoresis that suggested dehydration.

Given his negative hypercoagulation studies, normal laboratory values, and new exercise regimen, dehydration was concluded to be the likely etiology. Since his visual acuity was not affected, observation with bimonthly follow-up for 6 months was the management strategy. He was also encouraged to maintain adequate hydration during exercise. His vision returned to normal 2 weeks after the initial event, and he did not have recurrence during the monitoring period. JFP

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Observation and monitoring for the development of complications is appropriate for patients with central retinal vein occlusion without macular edema.

–2.5 to 1.0; P = .42), favoring the group that continued therapy.

Recommendations from others

After reviewing data from multiple clinical trials, the authors of the 2018 report from the National Kidney Foundation–Kidney Disease Outcomes Quality Initiative (NKF–KDOQI) concluded that the decision to continue or stop RAAS therapy in patients with advanced CKD should be individualized.[4] Criteria that should be considered in the decision-making process include the presence or absence of large acute declines in eGFR (> 20% in the absence of a significant decrease in proteinuria), hypotension, or acute kidney injury with significant risk for worsening.

In 2021, the Renal Association and the Association of British Clinical Diabetologists published updated clinical practice guidelines for the management of hypertension and RAAS blockade in adults with diabetic kidney disease.[5] Collective data indicated that, although outcomes varied based on type of diabetes (1 vs 2) and degree of proteinuria, blockade therapy overall led to improved outcomes; this was hypothesized to be due to the effects of reduced blood pressure. However, discontinuation of RAAS blockade therapy may be warranted when the patient (1) has a potassium level > 5 mmol/L pretreatment or ≥ 6 mmol/L with treatment, (2) demonstrates a decrease in eGFR > 25% or an increase in serum creatinine > 30% upon initiation of blockade, without another cause of renal deterioration, (3) is pregnant, or (4) has an acute illness with fluid depletion (in which case, RAAS therapy can be restarted 24 to 48 hours after recovery).

Editor's takeaway

Evidence supports continuation of RAAS blockade, particularly in patients with significant comorbidities (diabetes and cardiovascular disease). Study data indicate continuation is either beneficial or neutral to further morbidity. The only caveat is that these patients should have their renal function and potassium level continuously monitored. The evidence should provide reassurance to patients and physicians that continuation is the correct course of action. JFP

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