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Clinical challenges

Acute visual loss: just the beginning?

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1. Case report

A 47-year-old man presented with sudden visual loss in the left eye (OS). He had a history of a painful red eye lasting for 10 days prior to his visual loss. He was treated with local antihistamine eye drops and referred for further evaluation when he developed sudden visual loss. His past medical history was noncontributory except for hyperlipidemia.

Visual acuity was 20/20 with normal color vision OD and no light perception with an amaurotic pupil OS. He had limited upgaze OS (Fig. 1). Slit lamp examination was normal. The left optic disk was edematous with a cherry red spot in macular area OS. Venous congestion with dot and blot hemorrhages were seen throughout the retina (Fig. 2). The right eye and physical examination were normal.

What is the differential diagnosis of his visual loss and how would you proceed?

1.1. *Comments by Rod Foroozan, MD*

The visual loss in this patient appears to be monocular on the left. To help exclude visual loss on the right I would suggest automated perimetry. Assuming the field on the right is normal, we are dealing with monocular visual loss on the left side with no light perception and funduscopic findings suggestive of a combined retinal artery and vein occlusion.

The mechanisms for combined retinal vascular occlusions are likely multifactorial.[8] Obstruction of the central retinal vein (CRV) may cause increased venous pressure and subsequent impaired retinal arterial flow. Impaired perfusion of the central retinal artery (CRA) may cause venous stasis and thrombosis.

The broad classes of disorders that can cause this include inflammatory conditions such as orbital cellulitis, idiopathic orbital inflammation, scleritis, vascular conditions including coagulopathies and vasculitis, compressive lesions such as orbital and optic nerve tumors, and infiltrative disorders such as metastasis.[4,16]

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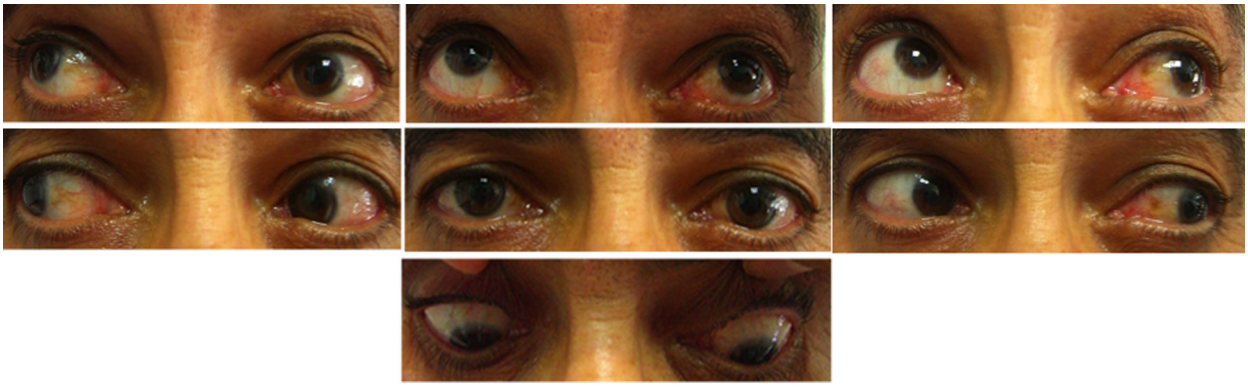

Fig. 1 – Eye movements showing upgaze restriction OS.

Mechanical compression of the vascular structures within the optic nerve has been the cause combined occlusion of the CRA and CRV in some patients. This has been described in patients with optic nerve inflammation and optic nerve tumors.[8]

The history of pain and red eye are not particularly helpful here; however, the limitation in upgaze on the left is suggestive of more than just a retinopathy/optic neuropathy and implies something additional such as an orbital process.

In primary position the left superior sulcus looks slightly full, and assessment for proptosis may be helpful. The supraduction deficit is present in both adduction and abduction, and there is no ptosis or anisocoria suggestive of a third nerve palsy. Forced ductions may help to determine if there is restriction. Neuroimaging to include the orbits becomes more important now that we suspect there is an orbital component to this disorder.

I would suggest magnetic resonance imaging (MRI) of brain and orbits with contrast and fat suppression. I would also perform blood tests for inflammatory/infectious causes such as syphilis, sarcoidosis, and rheumatologic conditions. A lumbar puncture and blood tests for coagulopathies may be helpful, but I would await the results of these other tests first.

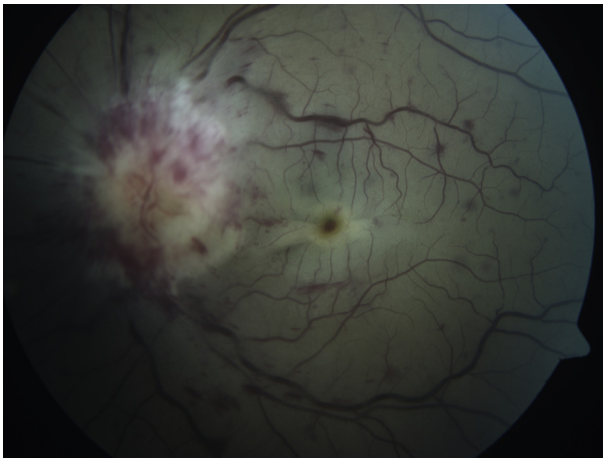

Fig. 2 – Optic disk edema and retinal whitening with cherry-red spot OS. Venous congestion and hemorrhages are seen throughout the retina.

2. Case report, continued

MRI showed diffuse thickening of the left optic nerve and slight enhancement with gadolinium (Fig. 3A, B). No other abnormality was observed in the orbital or cranial structures. CT angiography of cerebral–cervical circulation and echocardiography were normal.

Total white blood cell count was elevated (13,400/μL), dominated by neutrophils, suggesting an inflammatory process. The erythrocyte sedimentation rate (ESR) and C-reactive protein (CRP) were normal. Antinuclear antibody, anti ds-DNA, antineutrophilic cytoplasmic antibody, and extractable nuclear antibody tests were negative for collagenous tissue disorders. Angiotensin-converting enzyme and chest x-ray were normal. The patient was also screened for hypercoagulability. Neither an embolic source nor thrombophilia was detected. Rapid plasma reagin, antibodies to toxoplasma, rubella, cytomegalovirus, *Herpes simplex* virus, and screening for *Brucella* and *Borrelia* were done to exclude an infectious etiology. He refused a lumbar puncture.

Intravenous (IV) methylprednisolone 1 g/day was given for five days, followed by 1 mg/kg oral prednisolone which was tapered over two weeks. Because the *Borrelia burgdorferi* IgM antibody was positive by Western blot, orbital Lyme disease was considered, and he was treated for 21 days with IV ceftriaxone. His eye movements improved to some degree, but there was no recovery of vision.

Does this patient have Lyme disease?

What is the next step?

2.1. *Comments by Dr. Foroozan, continued*

It may be helpful to know the demographics and travel history to see if this patient has been in an area endemic for Lyme disease, also whether he had a history of tick bite or skin rash. Acute phase reactants may be elevated in Lyme disease, but may also be normal, so that the normal ESR and CRP are not particularly helpful here. Lyme disease has been decribed as causing an inflammatory orbitopathy.[15] Patients with orbital