Management of Sudden, Painless Vision Loss

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Abstract

Sudden, painless loss of vision is quite alarming to patients and may signify a true ocular emergency that requires swift and accurate diagnosis and management by eye care providers in cooperation with multiple sub-specialties.
Case Report

Case History
Patient CDG, a 62-year-old African American male, presented to the Optometry Department of the Baltimore Veterans Affairs Medical Center (VAMC) on August 10, 2011, with a chief complaint of having no vision in his left eye, which was noted upon awakening four days ago. His vision has remained “black” since. The patient denied any pain before or since the event. The patient also reported his vision had been going black for periods of 2-3 hours in the left eye since his trabeculectomy in 2007, with full recovery after the attacks. The patient stated his episodes of blacking out of vision had never persisted as long as his current attack. The patient denied any numbness or tingling in his hands/legs, difficulty speaking or walking, as well as symptoms of headache, fatigue, ocular pain, jaw ache, or scalp tenderness.

Ocular History
The patient has received eye care with Optometry and Ophthalmology services at the Baltimore VAMC since 2000. At initial presentation in 2000, he was being followed as a glaucoma suspect secondary to pigment dispersion syndrome OU. The patient’s central corneal thickness was documented as thinner than average (OD 480 microns, OS 470 microns), and gonioscopy revealed open angles with Sampaolesi’s line OU. In October 2001, treatment was initiated for pigmentary glaucoma OU. In 2003, the patient had cataract surgery with placement of a posterior chamber intraocular lens (PC IOL) in the left eye outside of the VA system. A PC IOL was placed in the right eye in February 2006 following a pars plana vitrectomy (PPV)/air fluid exchange (AFX)/lensectomy/endolaser treatment for a macula-on rhegmatogenous retinal detachment, after a laser/pneumatic retinopexy failed to correct the retinal detachment completely. PPV/A XF/endolaser was performed again in April 2006 after the patient presented with a macula-off retinal detachment in the right eye, which resulted in best corrected visual acuity of Hand Motion (HM) in the right eye. In 2007, a trabeculectomy with mitomycin C (MMC) was performed in the left eye at the Wilmer Eye Institute. The patient was lost to follow up from September 2007 to April 2009. The bleb became non-functional in May 2009 as evidenced by another IOP spike. Currently intraocular pressure is controlled by Brimonidine 0.2% BID OS and Cosopt BID OS. In 2009, the patient presented with another retinal detachment in the right eye due to Proliferative Vitreoretinopathy (PVR). He refused further surgical intervention and was left with light perception only in that eye. In October 2005, the left eye was treated with prophylactic peripheral laser 360 degrees after multiple atrophic retinal holes were discovered. In June 2006, the patient presented with a macula-off retinal detachment in the left eye, which was treated with PPV/AFX/silicone oil (SO)/endolaser with resulting best corrected visual acuity of 20/40.

Medical History
The patient’s medical history is significant for Type 2 Diabetes Mellitus, coronary artery disease, cerebral vascular accident (CVA) without ocular involvement in 2001, hyperlipidemia,
hypertension with good control since January 2010, enlarged prostate, and erectile dysfunction. The patient is also a cigarette smoker (1-3/day) with occasional marijuana use.

**Current Medications**

- Acetaminophen 300mg/Codeine 30mg
- Diclofenac BID OS for chronic iritis
- Cosopt BID OS
- Brimonidine BID OS
- Albuterol HFA oral inhaler
- Allopurinol 100mg
- Atenolol 25mg
- Dipyridamole 200mg/Aspirin 25mg BID
- Hydrochlorothiazide 25mg
- Lisinopril 40mg
- Niacin 500mg
- Simvastatin 80mg
- Terazosin 2mg
- Vardenafil HCl 20mg
- Aspirin 325mg
- Etodolac 400mg TID/PRN

**Pertinent Clinical Findings**

The patient presented with light perception without projection in the right eye and light perception with projection in the left eye. Extra-ocular muscles were smooth and full; confrontation visual fields were not possible to assess due to vision. The pupils appeared to be equal and round, though an afferent pupillary defect was present in the right eye, this had been previously noted. Slit lamp examination revealed clear lids and lashes. The conjunctiva of the right eye exhibited nasal pinguecula. The corneas of both eyes presented with arcus senilis 360 degrees and krukenberg spindle. The cornea of the right eye had an inferior stromal scar as well as a temporal pterygium. In the left eye, pingueculae were present both nasally and temporally, and a superior/temporal bleb with microcystic changes was present. The anterior chamber of the right eye had trace cells, 50% of which were pigmented. A bubble of silicone oil was present superiorly. The left eye had grade 1+ cells, 50% of which were pigmented. No neovascularization of the iris was present in either eye. Intraocular pressures by applanation tonometry were 11 mmHg in the right eye and 17 mmHg in the left eye. PC IOLs were present in both eyes, clear and centered. A dilated fundus exam revealed 0.90 cup to disc ratio with diffuse pallor in the right eye, and 0.90 cup to disc ratio with collateral vessels, which were first noted in April 2009. No optic disc swelling was present in either eye. The posterior pole of the right eye revealed a macula-off retinal detachment with many areas of fibrovascular proliferation. The left eye exhibited mild traction superior/temporal to the macula due to fibrosis with a chorioretinal scar. Laser scars were seen in the periphery of both eyes 360 degrees.
Pertinent Physical Findings
Blood Pressure May 2011-August 2011: between 125/83 to 140/84
Body Temperature May 2001-August 2011: between 97.2-98.2 degrees
The patient was accompanied by a guide dog which he has had since 2007. The patient also attends meetings with the Visual Impairment Services (VIST) through the VA to assist with orientation and mobility training, as well as emotional support.

The differential diagnoses that one should consider in a case of sudden, painless vision loss include:

**Non-Arteritic Posterior Ischemic Optic Neuropathy (NAPION)**
Typically unilateral, sudden, painless vision loss ranging from mild to count fingers or worse and central visual field defect. Generally occurs in patients between 40 and 60 years of age. Ophthalmic exam shows presence of relative afferent papillary defect in the involved eye, but is otherwise normal with no fundus or optic disc abnormalities on ophthalmoscopy or fluorescein fundus angiography. The fellow eye, unlike NAAION, will not be anomalous. Later, optic atrophy will set in. Diagnosis of exclusion.¹

**Arteritic Anterior Ischemic Optic Neuropathy (AAION)**
Also known as Giant Cell Arteritis (GCA). Unilateral sudden, painless vision loss at the level of count fingers or worse, but may become bilateral quickly. Generally occurs in patients over 50 years of age, but more commonly over 70 years of age. Patient may experience headache, jaw claudication, scalp tenderness over the superficial temporal arteries, proximal muscle and joint aches, anorexia, weight loss or fever before or during the episode. Clinical signs include afferent papillary defect, and a chalky white, swollen optic disc. After resolution of edema, optic atrophy and cupping occur. On laboratory analysis, erythrocyte sedimentation rate (ESR), C-reactive protein (CRP) and platelet count may be increased. There is generally presence of a visual field defect, either altitudinal or central. The clinician may be able to palpate a tender, non-pulsatile temporal artery.²,³,⁴,⁵

**Non-Arteritic Anterior Ischemic Optic Neuropathy (NAAION)**
Unilateral sudden, painless vision loss typically less severe than the arteritic form. May become bilateral as well. Generally occurs in patients between 40 and 60 years of age. Symptoms of giant cell arteritis not present and ESR is typically not elevated. Patients also present with afferent pupillary defect and optic disc swelling that is less pale than the arteritic form and may be segmental. Visual field defects are also altitudinal or central with optic atrophy (segmental or diffuse) without increased cupping setting in after edema has resolved. Examination of the fellow eye may show a “disc at risk”, a small, crowded optic disc that is more susceptible to this type of insult.²,³,⁴,⁵
Central Retinal Artery Occlusion (CRAO)
Unilateral sudden, painless vision loss at the level of count fingers to light perception. Prior to presentation, the patient may have experienced incidents of amaurosis fugax. On fundus examination, the retina will appear white in the posterior pole with a cherry-red spot in the center of the macula. The patient will exhibit a marked afferent papillary defect, narrowed retinal arterioles, and boxcarring or segmentation of the blood column in the arterioles. An embolus may be visible on examination. A cilioretinal artery may be present, which spares the foveola. In cases of significant visual loss at the level of light perception of worse, occlusion of the ophthalmic artery should be investigated. In this instance, a cherry-red spot in the macula is not present with global retinal ischemia. Etiology includes embolus, thrombosis or giant cell arteritis, other collagen-vascular disease, hypercoagulation disorders, migraine, Behcet disease, syphilis, sickle cell disease, and trauma.\(^2,3\)

Typical Optic Neuritis
Visual loss typically progresses more slowly over days, which ranges from mild to severe. Usually unilateral presentation in younger patients 18 to 45 years of age. Symptoms include orbital pain, especially on eye movement, decreased color vision and light desaturation. Patients’ symptoms may become worse with exercise or increase in body temperature.\(^2\) Caused by inflammatory demyelination of the optic nerve that is either idiopathic or associated with multiple sclerosis (MS).\(^4,5\)

Atypical/Inflammatory Optic Neuritis
Inflammatory optic neuritis with similar symptoms of typical optic neuritis. Optic disc swelling is more hyperemic and inflamed secondary to uveitis or a systemic inflammatory or infectious disorder, such as tuberculosis, syphilis, sarcoidosis, or cryptococcus.\(^2,4,5\)

Compressive Optic Neuropathy
Visual loss is typically slowly progressive, but may be acute or suddenly noticed. Patients will generally not have symptoms in common with giant cell arteritis. Relative afferent papillary defect will be present and central visual field defect. The optic disc presentation can vary with edema or pallor. The patient may also exhibit proptosis or optociliary shunt vessels. Causes can include optic nerve glioma, optic nerve meningioma, hematoma, thyroid-related orbitopathy, or any intraorbital or suprasellar mass.\(^2,3,4,5\)

Infiltrative Optic Neuropathy
Secondary to sarcoidosis or malignancy, such as lymphoma, leukemia, carcinoma, plasmacytoma, or metastasis from primary malignancy.\(^3,5\)

Central Retinal Vein Occlusion
Painless loss of vision, usually unilateral. Optic disc edema may be present, but this is the only feature that is similar to the other differentials listed above. The presentation diverges from there with diffuse retinal hemorrhages in all four
quadrants, dilated, tortuous retinal veins, cotton-wool spots, and retinal edema. Later, optociliary shunt vessels of the optic disc may develop, as well as neovascularization of the disc, retina, iris or angle. Secondary to atherosclerosis of the central retinal artery causing compression of the central retinal vein. Relative afferent pupillary defect is more commonly present in ischemic CRVO vs. non-ischemic CRVO.³,⁴

In this case, the patient had no pain on eye movements or history of multiple sclerosis. He had multiple risk factors for ischemic optic neuropathy or retinal vascular occlusion. Upon examination, his retina was not opacified and there was no cherry-red spot in the macula. There were no retinal hemorrhages present. His optic discs were not edematous. However, after conferring with a retinal specialist, the working diagnosis was Non-Arteritic Anterior Ischemic Optic Neuropathy OS. Laboratory tests to include complete blood count (CBC) with differential, erythrocyte sedimentation rate (ESR), and C-reactive protein (CRP) were ordered and the patient was instructed to report to have his blood drawn immediately after leaving the office. The patient was scheduled to return for fluorescein angiography. Additionally, a neurology consult was placed for a complete neurologic work-up. Presumably, radiologic studies would be performed such as MRI and MRA to rule out compressive etiologies.

Follow-up #1
The patient returned as scheduled on August 17, 2011, for fluorescein fundus angiography. In the interim, results of the ordered laboratory studies were available, those with flagged values are shown below.

**Laboratory Values**
- LSL cholesterol 111 (elevated)
- RBC 4.25 (low)
- HGB 12.1 (low)
- HCT 37.4 (low)
- ESR 23 (slightly elevated by VA lab indices)

Laboratory results ruled out arteritic anterior ischemic optic neuropathy (AAION) due to normal ESR and CRP values. Normal ESR value was calculated by taking the patient’s age (62) and dividing by 2, with the result of 31. Fluorescein angiography showed normal filling times with intraretinal microvascular abnormality (IRMA)/telangiectasia in the superior/temporal arcade of the left eye, but no late stage hyperfluorescence of the optic disc. Given the laboratory and fluorescein angiography results, the retinal specialist concluded that there was no ocular cause for the sudden decrease in vision and an ischemic event posterior to the lamina cribrosa was the likely culprit. Therefore, carotid duplex scan and echocardiogram were ordered with follow-up with Cardiology. The results of these, as well as the originally placed neurology consult, are still pending.

**Diagnosis and Discussion**
Sudden, painless vision loss upon waking is the classic symptom of many ischemic optic neuropathies. The fact that the dilated fundus exam revealed no abnormalities, such as disc edema or pallor in the affected eye, would rule out any anterior optic nerve pathologies.
Furthermore, arteritic causes of this type of vision loss can be ruled out with laboratory results and the absence of classic signs/symptoms. Additionally, no abnormal findings in the fluorescein angiography consistent with anterior optic nerve pathology lend credibility to our working diagnosis of posterior optic nerve pathology. Visual fields were not able to be tested due to the visual acuity in both eyes. The patient also has many risk factors of NAPION, which include hypertension, diabetes, cardiovascular disease, smoking, coronary artery disease, and hyperlipidemia.

The blood supply to the posterior portion of the optic nerve originates from the internal carotid artery (ICA). From the ICA, the ophthalmic artery diverges, from which collateral arteries diverge to form the pial vascular plexus, creating the peripheral centripetal vascular system. Thus, an embolus at any one of these junctions could lead to a variety of ischemic events of the optic disc or retina.

Although many different patterns of visual field loss may be seen in NAPION, the most common are central defects. This is explained by the central portion of the posterior optic nerve being a watershed zone, making it more susceptible to damage from ischemia due to a decrease in systemic blood flow. Conversely, this area is less susceptible to ischemic damage due to localized occlusion from diseases like atherosclerosis. As detailed above, the peripheral centripetal vascular system is composed of numerous collaterals of different sizes and locations, leading to potential variability in visual field defect presentation. This organization of vasculature is possibly a protective factor, which explains why PION is much less common than AION.

Due to pending results, the definitive diagnosis of our patient is unknown. Clinical findings indicate a tentative diagnosis of Non-Arteritic Posterior Ischemic Optic Neuropathy (NAPION). As NAPION is a diagnosis of exclusion, it will not be formally made until results of the neurology work-up, echocardiogram and carotid duplex scan have returned.

**Treatment/Management**
First and foremost, one must rule out arteritic causes to reach the diagnosis of exclusion of NAPION. Treatment for NAPION is controversial. Systemic steroids have been found to improve vision, though 34% of patients with NAPION will have spontaneous improvement in vision with no treatment. After review by the Retinal Specialist co-managing this case, medical treatment was not initiated. Especially in the case of binocular patients, communication with the patient’s Primary Care Physician is critical to ensure all systemic risk factors are controlled, thus decreasing the risk of involvement of the fellow eye or subsequent systemic involvement.

**Conclusion**
This case demonstrates the role of patient history, clinical observation, and consultation with subspecialists in the diagnosis and management of patients with sudden, painless loss of vision.
By nature, patients afflicted with these conditions tend to have multiple comorbidities which make partnering with their Primary Care Physician and other medical specialists key to appropriate intervention. Prompt evaluation of patients at the onset of their symptoms is important to rule out true ocular emergencies.

Bibliography