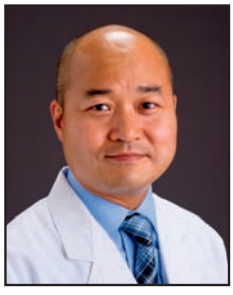
Diagnostic Considerations in Patients Presenting with Transient Vision Loss

by Bokkwan Jun, MD

No standard classification system and no guidelines exist for the diagnosis and management of transient vision loss.



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Abstract

A complete history is essential for determining whether the underlying etiology of transient vision loss is from an ischemic or a nonischemic condition that originates in the eye or elsewhere. Ischemic transient vision loss constitutes a transient ischemic attack and requires a full work-up for ischemic stroke. If carotid artery stenosis is found, early intervention may improve the clinical outcome. This article highlights the diagnostic considerations in the evaluation of patients with transient vision loss.

Introduction

Transient vision loss is clinically challenging for ophthalmologists and debilitating, frustrating and worrisome for patients. Most of the time patients are asymptomatic when they present to an ophthalmology clinic because of episodic transient vision loss, and clinical evaluation during the visit may be unremarkable. Therefore, clinical judgment for further investigation and management is often based on the patient's history alone, underscoring the importance of asking the patient probing questions to elicit a thorough and complete history. How do the symptoms develop? How do they go away? How often does the vision loss occur and how long does it last? Do the symptoms occur with any activity, any particular position or at a specific time of the day? Do other symptoms occur before, after or during the visual disturbance? Does the visual disturbance affect one eye or both eyes? Ask questions to get a sense of whether the visual symptoms are positive or negative visual phenomena. Not only is it important to obtain a thorough history of the features of the episodes of transient vision loss, it is also important to obtain a complete personal, social, family and medication history, which can uncover pertinent information such as the recent start of a new medication, the presence of cardiovascular risk factors, or a history of migraine or motion sickness in the past or in family members.

Obtaining a detailed and extended history is the first step in the diagnostic approach of transient vision loss. Why is an extended history important clinically in ophthalmology? Because patients with ischemic transient vision loss are at risk of irreversible loss of vision. Moreover, transient loss of vision can be followed by an ischemic stroke. An ischemic cause of transient vision loss should not be missed and needs to be managed properly to prevent permanent vision loss or ischemic stroke.

Table 1. Ophthalmic and Nonophthalmic Conditions That May Cause Transient Vision Loss

	Ischemic	Nonischemic
Ophthalmic conditions	Retinal ischemia Temporal arteritis Vasospastic vision loss Carotid dissection Postprandial transient vision loss Light-induced transient vision loss Ocular thrombosis	Tear film abnormalities Corneal endothelial dysfunction Anterior chamber abnormalities Transient elevation of intraocular pressure Vitreous abnormalities Optic nerve anomaly Papilledema Orbital mass
Nonophthalmic conditions	Transient cortical ischemia Cardiac and carotid thromboemboli to the retina Vasculitis Hypercoagulable disorders	Migraine Transient cortical blindness (posterior lobe epilepsy) Uhthoff phenomenon in demyelinating diseases Fluctuating blood glucose

Illustrative Case

A 70-year-old woman with a medical history of hypertension and hyperlipidemia presented to the emergency department because of episodic vision loss in her left eye. She described the episodic vision loss as sudden, painless and associated with a growing white spot that covered the visual field. The episodes lasted for 3 minutes, followed by gradual recovery of vision. The initial neuro-ophthalmologic evaluation showed signs of optic neuropathy in the left eye, with poor visual acuity (light perception), dyschromatopsia (0/4 gross color) and a relative afferent pupillary defect. Interestingly, about 40 minutes after the initial examination, the patient stated that she could see the outline of images, and repeat examination indeed showed slight improvement of visual acuity and color vision, with finger counting at 6 feet and 3/4 gross color.

Magnetic resonance imaging (MRI) of the brain and magnetic resonance angiogram (MRA) of the head and neck were unremarkable for ischemic stroke and carotid artery stenosis. Given the fluctuation of the neuro-ophthalmologic evaluation, vasospastic ischemic optic neuropathy was a concern, and the decision was made to begin calcium channel blocker therapy with amlodipine and increase her aspirin dose.

Two days later, repeat neuro-ophthalmologic examination showed significant improvement of the visual sensory function in the left eye. Visual acuity was 20/15 and color vision was 10/10 on Hardy Rand and Rittler color plate testing. The patient concurred that there was about a 20% reduction of light and red desaturation in the left eye. Humphrey automated visual field testing showed cecocentral scotomas in the left eye, with 25 decibel (dB) fovea threshold and –6.96 dB mean deviation (MD), compared to the right eye, which exhibited a 34 dB fovea threshold and –3.2 dB MD. The patient had several more episodes while she was receiving 5 mg of amlodipine daily. Amlodipine was increased to 10 mg and her transient visual disturbance has not recurred.

Ischemia as a Cause of Transient Vision Loss

The various causes of transient vision loss are summarized in Table 1. Transient vision loss due to ischemia is also known as amaurosis fugax. Embolic lesions from the heart and carotid arteries, atherosclerotic thrombotic lesions involving ophthalmic, central retinal or short posterior ciliary arteries, vasculitis, hypercoagulable disorders, and vasospasm can all cause monocular or binocular transient vision loss.

Amaurosis fugax, or fleeting blindness, usually lasts for a few minutes, followed by gradual recovery within 24 hours. Vision loss caused by ischemia is characterized by sudden, painless darkening of vision in association with partial or complete visual field defects. Most of the visual disturbances are negative phenomena, such as diffuse loss of vision, a curtain coming down (altitudinal field defect), nasal or temporal vision loss, constriction of the visual field, and dark spots, but the rare patient with transient vision loss from ischemia describes positive phenomena, such as flashing light and scintillating scotomas.[1,2] Patients usually are aware of the time of onset, frequency and duration of the episodes.

In terms of the pathology of ischemia, thromboembolic

causes include cardiac thromboemboli (atrial fibrillation, valvular disease), carotid thromboemboli (atherosclerosis, dissection), aortic arch emboli, and hypercoagulability. Hemodynamic causes include postural hypotension, malignant hypertension, and high blood viscosity. Vascular causes of transient vision loss due to ischemia include vasculitis, vasospasm, arteriovenous fistula and vertebrobasilar insufficiency.

Ophthalmic Causes of Transient Vision Loss Due to Ischemia

Retinal Ischemia

Retinal hypoxia, or ocular ischemia, is signaled by the onset of amaurosis fugax in 15% of patients, and 74% of patients with ocular ischemia have severe carotid artery stenosis.[3] Cardiac and carotid emboli to the retinal vasculature may cause retinal hypoxia and transient vision loss. Sources of cardiac emboli include valvular disease, mural thrombus and atrial myxomas. On funduscopic examination, calcific or cholesterol emboli (Hollenhorst plaque) may be observed. Carotid ultrasound and echocardiogram should be considered.

The treatment of a retinal transient ischemic attack is the same as that for a cerebral transient ischemic attack. The risk of ischemic stroke and death is increased in patients who have had a retinal transient ischemic attack. The short-term risk of ischemic stroke after a transient ischemic attack is 3% to 10% at two days and 9% to 17% at 90 days after the episode.[4] Patients with retinal transient ischemic attack need a work-up for stroke, including brain MRI with diffusion-weighted imaging, electrocardiogram, echocardiogram and vascular imaging by means of computed tomographic angiogram (CTA) and MRA. Brain infarction is demonstrated on MRI in 25% of patients with acute retinal ischemia[5] and in 18% of patients with a retinal transient ischemic attack.[6]

Temporal Arteritis

Temporal arteritis should always be considered in patients over 60 years of age who present with transient vision loss. Therefore, during history taking in this age group of patients, questions should be asked about the presence of constitutional symptoms, such as weight loss, fever, cough, myalgia and fatigue, and other associated symptoms, such as new headache, scalp tenderness and jaw claudication. Episodic vision loss during position changes, such as bending over or getting up from a supine position, may be associated with temporal arteritis. An elevated erythrocyte sedimentation rate, C-reactive protein and platelet count may be helpful in clinical decision-making. These tests should be ordered in all patients with transient monocular vision loss, even in patients who have no other symptoms of temporal arteritis. Up to 20% of patients with temporal arteritis and ocular symptoms have otherwise occult disease.[7] If clinical suspicion of temporal arteritis exists and a temporal artery biopsy for tissue diagnosis is under consideration, systemic corticosteroid therapy should be initiated without delay.

Vasospastic Vision Loss

As shown in the illustrative case, patients with vasospastic vision loss may show a dramatic change in clinical findings in a short period of time. Visual acuity can change from light perception to 20/20 or vice versa during a couple of hours of the clinic visit. Vascular imaging should be performed to exclude stenosis, thrombosis or dissection, but imaging studies are unremarkable in many patients. Diagnostic conventional cerebral angiography could be considered but a calcium channel blocker trial, such as nifedipine or amlodipine, also could be considered for both diagnostic and treatment purposes.

Carotid Artery Dissection

Although a rare cause of transient vision loss, extracranial internal carotid artery dissection may be manifested by transient monocular vision loss. One study showed that 28% of patients with extracranial internal carotid artery dissection had transient monocular visual loss, frequently in association with painful Horner syndrome ipsilaterally.[8]

Postprandial Transient Vision Loss

Postprandial transient vision loss may be associated with severe carotid artery occlusion. Relative hypoperfusion of the retinal and choroidal circulation by increased blood flow to the intestines after eating may be a mechanism for this condition.

Light-Induced Transient Vision Loss

Bright light signal may compromise the regeneration of retinal pigment and induce episodes of transient vision loss in patients with severe carotid artery stenosis.

Transient Cortical Blindness - Ischemic

Patients with cortical transient ischemic attacks may present with episodic transient vision loss. It could be caused by posterior or anterior cerebral circulation insufficiency. This condition needs to be considered in older patients who have cardiovascular risk factors. Vertebrobasilar insufficiency may cause unilateral or bilateral vision loss, and the vision

loss can be isolated or combined with other neurologic symptoms, such as ataxia, dysarthria and/or vertigo. A work-up for ischemic stroke must be done as soon as possible.

Ophthalmic Causes of Transient Vision Loss Not Related to Ischemia

Patients with transient vision loss from nonischemic ophthalmic causes usually have a longstanding history of fluctuating symptoms that have been gradually worsening. They may not know the exact time the symptoms started. Patients may experience fluctuation of symptoms during the day, depending on the time of day and on the direction of gazes, and they may have other symptoms, such as ocular irritation, red eye and headache. Patients with diabetes may have a history of poorly controlled blood glucose levels.

Tear Film Abnormalities

Tear film abnormalities as a result of dry eye syndrome or blepharitis are common causes of transient visual disturbance. In particular, patients using contact lenses and older patients with a history of neurodegenerative disorders such as Parkinson's disease may experience ocular foreign body sensation and conjunctival injection with transient visual disturbance. The duration of episodes may be variable, from several minutes to several hours. Patients may state that blinking or lubricating eye drops help relieve the symptoms. Ophthalmic evaluation with slit lamp may show inflammation along the lid margins, abnormal tear film break up time, superficial punctate keratitis and conjunctival congestion.

Corneal Endothelial Dysfunction

Patients with corneal endothelial dysfunction may develop corneal edema and visual disturbance. The visual disturbance from the edematous cornea is often worse in the morning and improves throughout the day. Patients may experience visual disturbance before the corneal edema becomes noticeable on the slit lamp examination.

Anterior Chamber Abnormalities and Transient Elevation of Intraocular Pressure

Anterior chamber abnormalities that can cause transient vision disturbance include uveitis, hyphema and intermittent angle-closure glaucoma. Patients who have transient visual disturbance and ocular discomfort need prompt ophthalmologic evaluation to rule out intraocular inflammation or elevated intraocular pressure. Patients with uveitis or intraocular inflammation (infectious, noninfectious) may also experience ocular pain and photophobia. Ophthalmologic evaluation with slit lamp may reveal conjunctival or episcleral injection and inflammatory reaction in the anterior chamber and vitreous. Intermittent angle-closure glaucoma may also cause vision loss and ocular pain as well as associated systemic symptoms such as headache, nausea and vomiting. Ophthalmologic evaluation with the slit lamp and gonioscopy may demonstrate a shallow anterior chamber and angle.

Vitreous Abnormalities

Patients who experience transient episodes of vision loss may report that they see floaters, especially on a monotonous background such as a white wall. Vitreous floaters may be a consideration in patients with high-refractive errors or a history of ocular injury or diabetic retinopathy. Floaters may be visualized on slit lamp examination and fundoscopy. Generally, a sudden worsening of floaters and flashing light may portend vitreoretinal traction or retinal detachment. Recently, transient vision loss after intravitreal injection of ocriplasmin was reported.

Optic Nerve Abnormalities

Transient visual obscurations are commonly associated with papilledema from elevated intracranial pressure. Patients with increased intracranial pressure may experience transient vision loss with positional changes and Valsalva maneuver, and may also experience headache in the morning and positional headache. Transient visual obscuration from papilledema can be monocular or binocular, typically lasting for seconds only, and can be described as a gray, black or white out of vision.

Transient visual obscurations are also associated with optic disc anomalies, such as optic disc drusen, Fuchs coloboma and optic nerve sheath meningioma.[9] These conditions may be associated with transient elevation of intraocular pressure, which needs to be considered and monitored.

Orbital Mass

Patients with an orbital mass may experience vision loss in a particular direction of gaze, called gaze-evoked amaurosis. The vision loss only lasts for seconds and baseline vision returns on straight-ahead gaze. Orbital meningiomas, optic nerve gliomas, cavernous hemangiomas, orbital osteoma and orbital fractures have been reported to cause gaze-evoked amaurosis.[10] Interference of axoplasmic flow by direct compression of the optic nerve and transient ischemia by compression of the vascular supply to optic nerve may be underlying mechanisms for gaze-evoked amaurosis caused by an orbital mass. In addition to orbital computed tomography and MRI imaging, real-time color Doppler studies of

the optic nerve vasculature may be useful in revealing a reduction of blood flow on eccentric gaze when the patient is symptomatic.[11]

Nonophthalmic Causes of Transient Vision Loss Not Related to Ischemia

Fluctuating Blood Glucose

Patients with diabetes mellitus and fluctuating blood glucose levels may experience transient visual disturbance from osmotic change in the crystalline lens and from myopic shift by increased thickness of the lens. The visual disturbance may last from several minutes to several hours. On ophthalmologic examination, visual acuity may improve on pinhole testing.

Uhthoff Phenomenon in Demyelinating Conditions

Patients with demyelinating conditions may experience Uhthoff phenomenon, a transient vision loss associated with elevation of the body temperature after exercise or a hot shower. Typically the visual disturbance lasts until the body temperature normalizes.

Epilepsy and Transient Cortical Blindness - Nonischemic

Transient vision loss has been reported in patients with occipital lobe epilepsy. The vision loss occurs during ictal and postictal periods and improves with antiepileptic medications.[12]

Migraine

Migraine-associated visual aura may be characterized by positive visual symptoms, such as photophobia, scintillating scotomas, zigzag lines and flickering colorful lights, and by negative symptoms, such as dimming, focal scotoma and vision loss. Most transient forms of hemianopic vision loss in younger patients with no cardiovascular risk factors are likely the result of migraine. In migraine-associated vision disturbance, positive visual phenomena usually precede negative visual phenomena. Migraine-related transient vision loss develops gradually over several minutes and usually lasts from 30 minutes to several hours. The vision loss in migraine can be followed by headache but also may occur without headache. The patient may have a history of migraine or motion sickness or a family history of migraine. Avoidance of migrainous triggers and a migraine prophylaxis regimen may help reduce and treat the transient vision loss, but it's important to exclude other causes of transient vision loss before attributing the vision disturbance to migraine.

Conclusion

No standard classification system and no guidelines exist for the diagnosis and management of transient vision loss, but a systematic process of differential diagnoses and management can be facilitated by determining initially whether the vision loss stems from an ischemic or a nonischemic condition. The patient often does not know whether the episode of vision loss was monocular or binocular, therefore, the initial work-up should basically proceed in the same manner for monocular and binocular transient vision loss. If ischemic transient vision loss is a concern, the patient should undergo a work-up for ischemic stroke, including brain MRI, carotid imaging with Doppler, CTA and MRA of the head and neck, echocardiogram and a lipid panel. Ischemic transient vision loss can precede stroke in about 18% to 25% of patients.[5,6] If carotid artery stenosis is discovered during the work-up, early intervention with carotid endarterectomy may be considered for asymptomatic or symptomatic severe stenosis (over 70%) and symptomatic moderate stenosis (50% to 69%).[13] If vasospastic transient vision loss is a concern, a calcium channel blocker can be tried for diagnosis and management, as in the illustrative case in the article.

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Disclosure

None reported. **MM**