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Amaurosis Fugax- A Clinical Review

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Abstract
Amaurosis fugax is a transient visual disturbance that is typically caused by a circulatory, ocular, or neurological underlying condition. Patients with amaurosis fugax are at risk for stroke, myocardial infarction, vision loss, and other serious consequences. A thorough case history, careful clinical and ocular examination, and appropriate systemic testing will lead to the best possible outcome for these at-risk patients.

Introduction

Amaurosis fugax is a sudden, temporary, partial or total loss of vision from any cause. Vision loss typically lasts from a few seconds to several minutes before returning to normal. In clinical practice, it is not uncommon for a patient to present with transient visual disturbances described as "waves," "jagged lines," "blurred" areas, "blacked-out" areas, or strange "spots" appearing in their vision. By the time patients seek treatment, the symptoms have usually resolved. This fact complicates the clinical evaluation and management of these patients.

The word "amaurosis" comes from the Greek language and means "to darken or obscure." The ancients used this term to describe all loss of vision that could not be obviously explained. The word "fugax" is also from the Greek and means, "fleeing." It is related to the word- "fugitive." Previously, amaurosis fugax was a "catch-all" diagnosis, but today it must be treated as a serious symptom of an underlying pathology.

The extensive differential diagnosis for amaurosis fugax can be managed efficiently by categorizing the possible etiologies. Wray described four classifications of amaurosis fugax based upon the underlying pathophysiology of the visual disturbances. The four types of amaurosis fugax described were embolic, hypoperfusion, angiospasm, and idiopathic.[1,2] The 1990, The Amaurosis Fugax Study Group defined five causes of transient monocular blindness. The distinct causes that they described were embolic, hemodynamic, ocular, neurologic, and idiopathic.[3] By evaluating these categories, one can see that the common etiologies for amaurosis fugax are circulatory problems (either embolic or hypoperfusion), ocular problems, neurologic problems, or idiopathic causes. Today, with advanced diagnostic evaluation techniques, it is rare that amaurosis fugax is attributed to an idiopathic cause. When amaurosis fugax was described as idiopathic in the past, it was often related to migraine equivalent headaches or psychogenic causes. Therefore, in the simplest terms, three causes of transient visual disturbances exist: circulatory, ocular, or neurologic. Keeping in mind these three diagnostic categories, the clinician will ultimately reach the proper diagnosis and initiate appropriate referral and management of the amaurosis fugax patient. Table 1 presents the differential diagnosis for transient visual disturbances according to the three categories of underlying pathophysiologies.

The clinician's goal must be to determine the etiology of the patient's transient visual disturbances. This is important because the underlying sources of amaurosis fugax could range from life threatening conditions to simply dry eyes. Patients that present with true hemispheric ischemia symptoms such as weakness on one side of the body, confusion, or other neurological symptoms certainly have a higher risk of stroke, but potentially all amaurosis fugax

patients are "at risk" and need to be properly evaluated.[4,5,6] In a previous study, general practitioners responded that they would only refer 72% of patients with transient vision loss for further testing and evaluation by a specialist.[7] This referral rate is too low when as many as 51% of these patients will suffer from a cerebral vascular accident.[8,9] In addition, if amaurosis fugax is found to be associated with carotid artery disease, there is a significant increased risk of death from a myocardial infarction.[10] So, efficient and effective management is required for these patients.

Table 1: Differential Diagnosis of Amaurosis Fugax

Circulatory	Ocular	Neurologic
Embolic	Dry eye syndrome	Optic neuritis
Carotid emboli	Keratitis	Papilledema
Cardiac emboli	Blepharitis	Multiple sclerosis
IV drug use	Iritis	Intracranial tumor
Hypoperfusion	Intermittent angle closure glaucoma	Psychogenic
Coagulation disorders	Optic disc drusen	Migraine[11,12]
Inflammatory arteritis	Vitreous detachment	Lupus (SLE)[17]
Carotid stenosis	Retinal break	
Ophthalmic artery stenosis	Orbital tumor[13,14]	
Cardiac failure or arrhythmia	Intraocular hemorrhage	
Increased blood viscosity	Angiospasm / vasospasm[15,16]	
Migraine[11,12]		

Subjective Symptoms of Amaurosis Fugax

When interviewing a patient with amaurosis fugax, it is important to pay special attention to the medical history. The clinician should attempt to uncover the presence of hypertension, diabetes, previous myocardial infarcts, prior cerebrovascular accidents, hypercholesterolemia, long-standing migraine history, or peripheral vascular disease.

During the case history, the clinician should consider the patient's age. If the patient is over the age of 45 years, then ischemic attacks are more common causes of transient visual disturbances. Patient age has been shown to be the most important factor in predicting the presence of significant carotid occlusive disease.[19] If the patient is under the age of 45, then benign migrainous transient visual disturbances are the most frequent cause. In fact, 41% of amaurosis fugax patients under the age of 45 will have an accompanying headache to help solidify the diagnosis of migraine.[20] Studies have not shown any patients under the age of 40 suffering a stroke following transient visual disturbances. Additional testing or diagnostic studies are therefore unwarranted in these young patients with transient visual loss, who are otherwise healthy since significant systemic disease is rarely discovered.[19,20]

The clinician should inquire about the frequency of the transient visual disturbances. Repeated events are more likely hypoperfusion secondary to arterial stenosis while isolated events may be due to an embolism. If the patient reports increasing frequency of symptoms, this may be very suggestive of an impending cerebral infarct; therefore the work-up should be undertaken without delay.

The clinician should investigate the onset of the transient vision loss. Hypoperfusion events will have a less rapid onset than the brief, transient visual loss that will be described in attacks of embolic or vasospastic origin. Hypoperfusion problems will develop over a matter of minutes, not seconds.[3]

Next, the clinician should question the patient regarding the duration of their visual disturbances. If the disturbance lasts for 2-30 minutes and resolves, this is suggestive of an ischemic attack.[21] If the disturbance lasts for only seconds, an ocular diagnosis such as vitreous traction or retinal breaks should be initially considered. Orthostatic hypotension typically produces transient visual disturbances lasting up to 45 seconds while a vasospastic or migrainous transient visual disturbance can last from a few minutes to several hours.

The clinician should consider whether the visual symptoms are monocular (one eye) or binocular (two eyes). If the patient reports that the visual disturbance is in one eye, then it is more likely associated with an occlusive retinal or carotid artery condition. If the patient reports that the symptoms occur in both eyes, then one should consider a vertebro-basilar circulatory condition or other posterior circulatory problems. It is often difficult for a patient to state if the symptoms are monocular or binocular, but since transient visual disturbances are usually multiple events, the patient can be educated to cover each eye the next time the symptoms occur to see if it is truly a monocular or binocular event.[22]

Additionally, the clinician should attempt to determine if the patient is describing a negative visual phenomenon or a positive visual phenomenon. A negative visual phenomenon is one where the vision blurs, fogs, dims, or blacks out. These symptoms are closely associated with ischemic events. If, on the other hand, the patient is describing flashing lights, zigzag lines, and colorful patterns in their vision, these positive visual phenomena are more closely related to a migrainous or ocular event.[23]

Pain associated with transient visual disturbances is common in cases of hypoperfusion and vasospasm. Migraine patients may describe severe pressure and pain that lasts for extended periods of time, while patients that describe chronic ocular or retrobulbar aching pains are more likely to suffer from carotid stenosis. [2]

Finally, it is wise to go through a series of direct questions to elicit further clues into the causes of the transient visual disturbances. Examples of these questions include:

- Does blinking or rubbing the eyes modify the symptom? If blepharitis or dry eyes, the answer will be positive.
- Are the symptoms worse with eye movements? If positive, vitreous traction, an orbital tumor, or optic neuritis may be present.
- Does exercise increase the symptoms? If there is underlying demyelinating disease or vasospasm, the answer may be positive.
- Do you have other symptoms like; scalp tenderness, jaw claudication, headaches, fever, or weight loss? All of these associated symptoms would point towards temporal arteritis.
- Do you experience motion sickness or have a history of headaches? If migrianous, the answer may be positive.

Table 2: Key Questions for Patients with Transient Visual Disturbances

1. Age of the patient?
2. How frequently do the symptoms occur?
3. How quickly do the symptoms arise?
4. How long do the visual disturbances last?
5. Are the symptoms in one eye or both eyes?
6. Describe the visual disturbances-
    - Classify as either-positive or negative phenomenon
7. Is there any pain associated with the visual disturbance?
8. Does blinking or rubbing the eyes modify the symptom?
9. Are the symptoms worse with eye movements?
10. Does exercise alter or cause the visual disturbances?
11. Are there any associated symptoms?
    - Such as; scalp tenderness, jaw claudication, malaise, fever, weight loss
12. Is motion sickness experienced?
13. Is there a long history of headaches?

Objective Findings of Amaurosis Fugax

Upon completion of the case history, the clinician can now begin to examine the patient with the three diagnostic categories in mind. The clinician should perform a measurement of blood pressure, heart rate and rhythm, palpation of the temporal arteries, and auscultation of the heart and neck.

Since a thorough ocular examination will often uncover the diagnosis quickly, the clinician should refer the patient to an optometrist or ophthalmologist if an ocular cause is suspected. The eye care provider should perform biomicroscopy paying special attention to the lid margins, tear film, cornea, and anterior chamber. Conditions such as dry eyes, blepharitis, and iritis can all be "ruled out" from the differential diagnosis if the biomicroscopy is normal. Gonioscopy permits direct observation of the anterior chamber angle of the eye under high magnification. This clinical observation can rule out micro hemorrhage in the anterior chamber or evidence of angle closure glaucoma; therefore it should be performed for patients experiencing transient visual disturbances.

Automated visual field testing should be performed. Ischemic events can produce visual field loss, such as a classic inferior altitudinal defect. An altitudinal defect is a

loss of vision in the superior or inferior aspect of the patient's visual field that respects the horizontal meridian. Careful assessment of pupillary reactions will test the function of a patient's optic pathway. A relative afferent pupillary defect will be present whenever there is a significant unilateral visual deficit. A relative afferent pupillary defect is an indicator of optic nerve disease typically associated with an ischemic etiology.

A dilated retinal examination is mandatory for patients presenting with amaurosis fugax. The retina, retinal vasculature, optic nerve and vitreous will all provide additional information in forming a clinical diagnosis. A vitreous detachment, retinal tear, or retinal detachment should be observable once the patient's eyes are dilated. Unilateral optic disc edema is suggestive of anterior ischemic optic neuropathy or optic neuritis. Bilateral optic disc edema is most always associated with increased intracranial pressure, but is really associated with reduced or disturbed visual acuity. Poor circulation to the optic nerve in any ischemic conditions will present with a pale optic disc. Any finding of a retinal vascular occlusion as seen in Figure 1 is a serious sign of ocular ischemia.

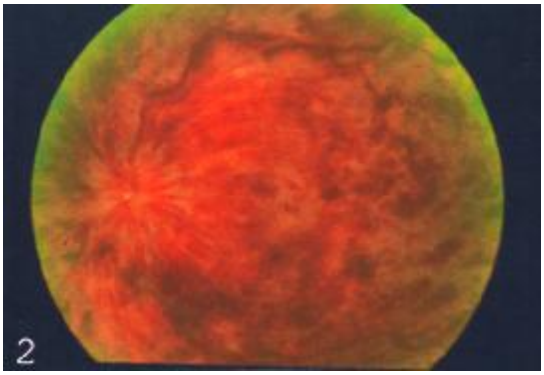

Figure 1: Central retinal vein occlusion

Intravascular retinal emboli may be observed. It is important to know that cholesterol plaques within retinal vessels will have a bright, yellow refractile appearance.

These, so called, Hollenhorst plaques are commonly associated with lesions of the ipsilateral carotid artery and are rarely from the heart. Calcified emboli, which are larger and white in appearance, may originate from the common carotid artery or from cardiac valve disease.[3] In 2002, Mead, et al. postulated a mechanical mechanism that causes emboli from the carotid arteries to enter into the ophthalmic artery causing a transient visual disturbance, while emboli from the heart are more likely to travel past the ophthalmic artery and continue on to cause an ischemic attack in the brain. Their theory is that small emboli break off from the carotid artery walls due to increased speed of blood flow through an area of stenosis. These smaller emboli travel along the wall of the internal carotid artery out of the rapid flow of the central part of the vessel. Because of their small size and positioning along the carotid vessel wall, carotid emboli can easily enter into the first branch that is encountered, the ophthalmic artery. In contrast, cardiac emboli are larger in size and are already moving at a high velocity by the time they enter the common carotid artery. Their fast pace of travel and larger diameters keep them moving in the middle of the vessel, traveling past the narrow branch of the ophthalmic artery.[24]

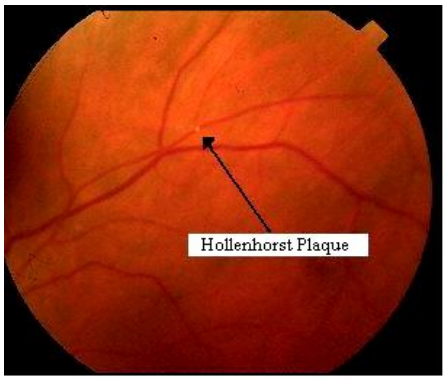

Figure 2: Hollenhorst Plaque within a retinal artery

Studies have shown that orthostatic hypotension in diabetic patients can cause blood flow in the ophthalmic artery to decrease by almost 100% when simply moving from a supine to seated position. This dramatic loss of perfusion to the eye will cause significant transient vision loss.[25] Therefore, any signs of diabetic retinopathy could be linked to amaurosis fugax.

Making the Diagnosis

In many patients presenting with amaurosis fugax, the diagnosis will be established after the clinical or ophthalmic examinations. The examinations may have uncovered dry eye syndrome, a posterior vitreous detachment, buried disc drusen or some other ocular condition that has caused the transient visual disturbances. The eye care provider can manage these conditions. But, if the diagnosis has still not been established, further evaluation is warranted.

Laboratory testing must be part of the initial diagnostic work-up in amaurosis patients.[26] Clinicians should order a complete blood count for patients over the age of 45 with transient visual disturbances looking specifically for anemia or other hematological disorders. A complete chemistry panel will provide information regarding diabetes, electrolytes, and liver enzymes. Additionally, a thyroid screening and a coagulation profile should be ordered. An immediate sedimentation rate (ESR) and C-reactive protein test should be ordered if any suspicion of temporal arteritis exists. For patients whose symptoms of amaurosis fugax are monocular, a non-invasive evaluation of carotid circulation such as a carotid duplex study is needed in association with radiology.[27] Internal carotid artery stenosis and carotid artery plaques are the most frequent findings

causing this circulatory type of amaurosis fugax.[28,29] Carotid Doppler, carotid duplex, computerized tomographic angiography, or magnetic resonance angiography may all be successfully utilized to evaluate the carotid circulation.[30] The importance of carotid imaging can not be overstated. In fact, 53% and 55% of patients referred for carotid duplex studies from eye care providers tested positive for significant internal carotid artery stenosis.[31,32]

If the amaurosis fugax patient's symptoms are binocular, then a CT scan or MRI should be ordered. This study should be done with specific interest in the areas of the occipital lobe and along the optic pathways. Imaging should be done immediately if the history and examination point toward a posterior circulatory condition.

An echocardiogram could be ordered if the history and clinical evaluation suggests that cardiogenic emboli may be the source of the amaurosis fugax, but studies have shown that less than 4% of these patients will have cardiac involvement. So, the echocardiogram should be considered as a secondary test.[33]

Invasive investigations such as temporal artery biopsy, carotid arteriogram, and timed fluoroscein angiography may be necessary as the evaluation progresses, but referral to a medical specialty will have already occurred at that point.

Management of Amaurosis Fugax

Management of the amaurosis fugax patient varies as widely as does the pathophysiology of their underlying conditions. Patients with amaurosis fugax due to a circulatory etiology should be managed by the appropriate medical or surgical specialist, such as a vascular surgeon. Commonly, carotid artery stenosis is uncovered and the treatment varies according to the amount of stenosis. In 2003, Rothwell, et al. pooled the data from 3 major clinical trials regarding management of carotid stenosis. Their results have led to the current standard-of-care where a patient with 30-49% carotid occlusion will likely be treated with blood thinners. A patient with 50%-69% carotid occlusion may be treated with carotid endarterectomy or blood thinners to restore the carotid circulation. Patients with 70%-99% occlusion will most likely be treated with surgery and those patients with complete obstructions will be treated with medication only.[34] Cardiogenic emboli will need evaluation and treatment from a cardiologist or vascular surgeon. These patients may require surgery to replace a malfunctioning heart valve or medication to control atrial fibrillation.

The patient's primary health care provider may treat migraine equivalent headaches that are found to be causing transient visual disturbances. Treatments of migraines vary from avoidance of "trigger" mechanisms, to abortive medications, to systemic medications, to biofeedback. Common systemic medications for migraine prevention include beta-adrenergic blockers, antidepressants, neurostabilizers, calcium channel blockers, nonsteriodal agents, and serotonin receptor antagonists. Various health care specialties may be able to provide symptomatic and preventative relief for migraine patients.

Management of ocular conditions such as dry eye syndrome, iritis, vitreous detachment, or retinal disease can be managed by optometrists and ophthalmologists. Most of these conditions can be medically managed with good visual outcomes. Neurologists, neurosurgeons, or psychiatrists should handle the management of any neurological conditions that cause amaurosis fugax. Only through coordination of care and careful follow-up, will the amaurosis fugax patient have the best possible medical outcome.

Conclusion

The diagnosis and management of a patient with amaurosis fugax or transient visual disturbance presents as a significant clinical challenge. A systematic approach to the patient's work-up will narrow the almost overwhelming volume of differential diagnoses each step of the way. Being mindful of the possible circulatory, ocular, or neurologic underlying causes will direct the evaluation. A careful case history, problem-focused examination that includes a dilated retinal evaluation, appropriate ordering of additional studies and laboratory tests will lead to appropriate care and management of patients with amaurosis fugax.

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