

Expert Opinion

CME

Retinal Migraine: Migraine Associated With Monocular Visual Symptoms

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In 1970, Carroll introduced the term “retinal migraine” in describing 15 patients with transient and persistent monocular visual loss but no associated headaches.¹ He stated, “The term retinal migraine, although a controversial one, could possibly be applied to uncommon cases of this disorder. . . .”

CASE

This 25-year-old man reports a 12-year history of similar headaches occurring about one or 2 times monthly. He develops a left or right temple throbbing which is mild at first and later becomes a 10/10 associated with nausea, vomiting, light and noise sensitivity. About 30 minutes after the onset of all of the headaches, he develops sudden total darkness where he cannot see in the eye contralateral to the headache lasting about 4 hours. The headache is severe for about 5 hours and then mild for 24 to 36 hours.

Aspirin or acetaminophen is of mild help. He tries to go to bed. He had never seen a physician for the headaches before.

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Past medical history was negative. There was no family history of migraine. Neurological examination was normal.

Questions—What is the diagnosis? Is any diagnostic testing indicated? What is the prognosis? What treatment would you recommend?

EXPERT COMMENTARY

This patient reports recurrent, stereotyped episodes of visual loss in one eye in association with migraine headaches. The possibility of alternating hemifield loss should be carefully considered but for this discussion we assume the history is accurate. The diagnostic work-up of this patient should include careful medical, neurologic, and ophthalmologic examinations. When evaluating a headache patient with the complaint of visual impairment, it is important to establish whether one or both eyes are affected. Retrochiasmatic lesions usually cause homonymous hemianopias with varying degrees of congruence, whereas prechiasmatic lesions (ie, optic nerve, retina, or media) produce monocular visual loss. Patients often have difficulty distinguishing between the loss of vision in one hemifield and the loss of vision in one eye. To accurately make this distinction, the patient must alternately cover each eye and compare their views. Once unilateral visual loss is confirmed in this manner, the clinician needs to identify or exclude secondary causes of transient

monocular blindness, because retinal migraine is a diagnosis of exclusion.

The most likely cause of recurrent stereotypical episodes of transient monocular visual loss in association with headaches is retinal migraine. Secondary causes of transient monocular visual loss are less likely to be found in cases that have been recurring for a long period of time. However, if atypical features are present in a patient's history or general physical, ophthalmologic, or neurologic examinations, imaging studies or other diagnostic testing are warranted. Features that should prompt concern for an underlying secondary cause of headache with transient monocular blindness include absence of history typical for migraine, onset after age 50 years, incomplete resolution of monocular visual loss, concomitant medical problems that can precipitate attacks of transient monocular blindness, and the presence of atypical neurologic signs or symptoms.² All cases with persistent monocular visual loss should be fully investigated. To exclude the possibility of a cardioembolic source, investigations such as electrocardiography, echocardiography, and holter monitoring need to be performed. Diagnostic testing in patients with suspected ischemic disease of the eye or brain should include duplex scanning, computed tomography or magnetic resonance imaging and angiography, fluorescein angiography, and in uncertain cases, conventional angiography. Neuroimaging can exclude an orbital or intracranial mass. Other diagnostic possibilities such as vasculitis, hypercoagulable states, illicit drug use, and rheumatologic disorders require a complete laboratory evaluation.

The features in this case are most consistent with a diagnosis of retinal migraine (Table 1),³ once monocular visual is confirmed and other causes of visual loss have been excluded. Other terms that have been used to describe this condition include "ophthalmic migraine," "ocular migraine," and "anterior visual pathway migraine." My colleagues and I have suggested the term "migraine associated with monocular visual symptoms," because it distinguishes between the loss of vision in one hemifield and that of one eye and includes sites other than the retina, such as the choroid or the optic nerve. We have also proposed changes to the current criteria in the sec-

Table 1.—2004 IHS Criteria for the Diagnosis of Retinal Migraine

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| 1.4 | Migraine with transient monocular visual symptoms |
| A. | At least 2 attacks fulfilling criteria B and C |
| B. | Fully reversible monocular positive and/or negative visual phenomena (eg, scintillations, scotomata, or blindness) confirmed by examination during an attack or (after proper instruction) by the patient's drawing of a monocular field defects during an attack |
| C. | Headache fulfilling criteria B-D for 1.1 <i>Migraine without aura</i> begins during the visual symptoms or follows them within 60 minutes |
| D. | Normal ophthalmologic examinations between attacks |
| E. | Not attributed to another disorder |
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IHS = International Headache Society.

ond edition of the International Classification of Headache Disorders (Table 2).⁴

Retinal migraine is thought to be a rare entity, but its true occurrence is unknown. Retinal migraine is most common in women in the second to third decade of life who have a history of migraine with aura.⁵ It is characterized by episodes of fully reversible positive and/or negative visual disturbances within one eye associated with migraine headache. Typical descriptions of positive visual phenomena include flashing rays of light, zigzag lightning patterns or perceptions of bright colored streaks, halos or diagonal lines. The negative visual losses include blurring, blank areas, black dots or spots in the field of vision, causing partial or complete blindness. Complex patterns of visual impairment, such as the coming together of spots and "tunnel vision" (not being able to see items in the periphery of one's visual field), are less common. Unlike this particular case, the visual disturbance often occurs on the same side of the migraine headache and may precede, accompany, or rarely, follow it. The duration of the visual symptoms may be as short a few seconds but usually lasts many minutes to 1 hour. Prolonged but fully reversible visual loss of one eye may rarely occur, sometimes lasting hours, days, or even, weeks.⁶ Nearly half of reported cases (43%) in one series of patients with retinal migraine and recurrent transient monocular visual loss subsequently experienced permanent monocular visual loss.⁴ No specific factor was identified to account for this occurrence. The authors recommended caution in

Table 2.—Proposed Criteria for Migraine with Monocular Visual Symptoms

1.4.1	Migraine with transient monocular visual symptoms
A.	At least 2 attacks fulfilling criteria B and C
B.	Fully reversible monocular positive and/or negative visual phenomena confirmed by at least one of the following:
a.	Examination during the attack showing signs of ipsilateral retinal or optic nerve abnormality
b.	The patient's drawing of a monocular field defect experienced during the attack
c.	A description of positive or negative visual symptoms confirmed by the patient by occluding each eye individually during the attack
C.	Headache fulfilling criteria B-D for 1.1 <i>Migraine without aura</i> or 1.2 <i>Migraine with aura</i> begins during the visual symptoms or precedes or follows them within 60 minutes
D.	Normal ophthalmological examination between attacks
E.	Headache and monocular visual symptoms not attributed to another disorder
1.4.2	Migraine with permanent monocular visual loss or defect†
A.	At least one attack fulfilling criteria B and C
B.	Irreversible monocular positive and/or negative phenomena (eg, scintillations, scotoma, or blindness) confirmed by examination during or following the attack
C.	Headache fulfilling criteria B-D for 1.1 <i>Migraine without aura</i> or 1.2 <i>Migraine with aura</i> begins during the visual symptoms or precedes or follows them within 60 minutes
D.	Abnormal ophthalmological examination confirming retinal vascular abnormality or optic neuropathy
E.	Headache and monocular visual loss not attributed to another disorder

†Most patients experienced only one attack of permanent monocular visual loss.

generalizing this finding, as it may simply be selection bias rather than an accurate representation of this entity.

The underlying pathophysiology of retinal migraine remains largely unknown. In some cases, vasospasm of the retinal or ciliary circulation may have caused retinal or optic nerve ischemia;⁷ this may explain the amaurosis and rare fundoscopic findings during acute attacks of retinal migraine. An alternative theory is spreading depression of retinal neurons, a phenomenon that has been demonstrated in the chick retina.⁸ Similarly, it is possible that those rare cases with prolonged monocular defects associated with migraine headache could have a mechanism similar to

that seen in the cerebral cortex of migraineurs who have persistent aura without infarction.

Prolonged and permanent monocular visual loss appears to occur more commonly in patients with retinal migraine than in cases of prolonged typical aura or migrainous infarction in those with conventional migraine.⁴ The high number of patients with transient monocular visual loss, who eventually develop permanent monocular visual loss, makes retinal migraine a less benign condition than migraine with conventional visual aura. Therefore, although there are no data to determine the efficacy of preventative treatment for this entity, prophylactic drug therapy seems prudent, even if attacks are infrequent. There is currently insufficient clinical information to support specific recommendations for acute and preventive medical therapy in the treatment of retinal migraine. Therapy of the acute attack of retinal migraine should probably not include triptans or ergots because of their vasoconstrictive properties. Similarly, use of oral contraceptive pills should be avoided, if possible. Prophylactic medications that have been tried with anecdotal benefit include calcium-channel blockers, tricyclic antidepressants, beta-blockers, and neuromodulators. Aspirin is a logical agent because of its antiplatelet activity. Personally, I favor antiepileptic drugs (ie, topiramate or divalproex sodium) and tricyclic antidepressants (ie, amitriptyline or nortriptyline). Although some patients respond to beta-blockers, I do not usually recommend them as first-line agents because of their theoretical potential for arteriolar constriction. Although episodes of vasospastic amaurosis fugax appear to have been successfully treated with calcium-channel blockers,⁹ they were not usually effective in the few patients treated at the Montefiore Headache Center.

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