Retinal Migraine

Brian M. Grosberg, MD, Seymour Solomon, MD, and Richard B. Lipton, MD*

Address
*Department of Neurology, Albert Einstein College of Medicine and The Montefiore Headache Unit, Montefiore Medical Center, 111 East 210th Street, Bronx, NY 10467, USA.
E-mail: bgrosber@montefiore.org

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Retinal migraine is a primary headache disorder, clinically manifested by attacks of transient monocular visual loss associated with migraine headache. Although isolated reports suggest that retinal migraine is rare, it likely is under-recognized. Retinal migraine usually is reported in women of childbearing age who have a history of migraine with aura. It typically is characterized by negative monocular visual phenomena lasting less than 1 hour. More than half of reported cases with recurrent transient monocular visual loss subsequently experienced permanent monocular visual loss. Although the International Headache Society diagnostic criteria for retinal migraine require reversible visual loss, our findings suggest that irreversible visual loss is part of the retinal migraine spectrum, likely representing an ocular form of migrainous infarction.

Introduction

In 1882, Galezowski [1] introduced the term "ophthalmic megrim" to describe four patients with migraine and temporally related, irreversible monocular visual loss. In two of these patients, prior attacks of migraine headache were associated with brief bouts of ipsilateral visual obscuration. Subsequent publications have emphasized similar cases of monocular visual loss attributable to migraine, a phenomenon known in current parlance as retinal migraine [2–5,6•,7–45]. The term "retinal migraine," coined by Carroll [46] in 1970, paradoxically described patients with episodes of transient and permanent monocular visual impairment, specifically in the absence of migraine headache. Most subsequent observers have used the term "retinal migraine" for those cases of monocular visual impairment temporally associated with attacks of migraine. Some have noted that unilateral visual loss was not restricted exclusively to the retina and advocated the term "anterior visual pathway migraine" or "ocular migraine" [6•,47]. The authors think that the latter term is most appropriate. The term "ocular migraine" allows for the distinction between the loss of vision in the homonymous fields of both eyes and that of one eye and includes sites other than the retina, such as the choroid or the optic nerve, that may be affected. The mechanism of these monocular visual defects is uncertain. Some think that the visual defects associated with migraine are the result of vasospasm within the retinal or ciliary circulation [48]. Others, including the authors, think that the defects are due to spreading depression of retinal neurons comparable with spreading depression in the cerebral cortex associated with the typical auras of migraine.

Diagnostic Criteria

The first edition of the International Classification of Headache Disorders [49] outlined diagnostic criteria for retinal migraine. The criteria required at least two attacks of fully reversible monocular visual defects lasting less than 60 minutes and occurring before, during, or after the headache (type unspecified). Other causes of this phenomenon were to be ruled out. In the revision of these criteria (2nd edition, 2004) [50], the reversible monocular defects could be positive or negative visual phenomena, but the associated headache now was to be migraine without aura beginning during the visual symptoms or following them within 60 minutes (Table 1). Based on studies, the authors think that the associated migraine should be with or without aura, that the visual defects occur before, during, and after the headache, and that permanent monocular visual loss (PMVL) be part of the syndrome comparable with the syndrome of migrainous infarction. Our suggestions for revision of the criteria of retinal/ocular migraine are noted in Table 2.

In a review of the English medical literature, 73 cases were found and four new cases were added from the Montefiore Headache Unit that fulfilled the criteria of transient monocular visual loss (TMVL) or PMVL [51,52] (Table 2). Because most cases were described before the publication of International Headache Society (IHS) criteria, some of the details of the headache and visual loss were incomplete. We used our best judgment in classifying these cases.

Clinical Features

In most cases of TMVL and PMVL, the migraine headache was ipsilateral to the visual loss. In contrast to the current IHS criteria for retinal migraine, almost all of the patients had a history of migraine with aura prior to their attacks of retinal migraine or during attacks of monocular visual loss. In almost all of the cases, the cerebral auras were visual. There was great variability in the temporal relationship of the

Table 1. 2004 International Headache Society criteria for the diagnosis of retinal migraine

A. At least two attacks fulfilling criteria B and C
B. Fully reversible monocular positive and/or negative visual phenomena confirmed by examination during an attack or by the patient's drawing of a monocular field defect during an attack
C. Headache fulfilling criteria B through D for 1.1 Migraine Without Aura begins during the visual symptoms or follows them within 60 minutes
D. Normal ophthalmologic examinations between attacks
E. Not attributed to another disorder

Data adapted from Headache Classification Subcommittee of the International Headache Society [50].

Table 2. Proposed modification of criteria for retinal (or preferably monocular) migraine

Monocular migraine with transient monocular visual loss or defect
- A. At least two attacks fulfilling criteria B and C
- B. Fully reversible monocular positive and/or negative visual phenomena confirmed by examination during the attack or by the patient's drawing of a monocular field defect during an attack
- C. Headache fulfilling criteria B through D for 1.1 Migraine Without Aura or 1.2 Migraine with Aura begins during the visual symptoms or precedes or follows them within 60 minutes
- D. Normal ophthalmologic examination between attacks
- E. Not attributed to another disorder

Monocular 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 that attack
- C. Headache fulfilling criteria B through D for 1.1 Migraine Without Aura or 1.2 Migraine with Aura begins during the visual symptoms or precedes or follows them within 60 minutes
- D. Abnormal ophthalmologic examination
- E. Not attributed to another disorder

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

visual loss and headache. In most patients, the onset of visual loss preceded or accompanied the headache; in a small number of cases, the visual loss followed the attack of migraine. Attacks of monocular visual disturbances recurred on the same side in most of the patients; only a small number experienced alternation of ocular symptoms during different attacks and these usually were patients who had TMVL rather than PMVL. The duration of the transient visual symptoms was as short as a few seconds (approximately 13% of patients), but usually lasted for many minutes to 1 hour. Approximately one third of patients experienced repeated attacks of TMVL lasting less than 30 seconds and more than 1 hour. A few cases had prolonged but fully reversible monocular visual loss, sometimes lasting hours, days, or even weeks [12,14,25,27,44]. As one may expect, ophthalmologic examination during an attack was infrequent. When such examinations were performed, they usually were normal. Severe narrowing or occlusion of retinal arteries was observed rarely [1–5,8,12,17,18,22,27,31,38,42]. The diagnoses of anterior or posterior ischemic optic neuropathy were reported in a dozen or so cases [6•,7,10,21,24,28,29,34,36,37,45]. Other findings included retinal pigmentary change [4], central retinal venous occlusion [5,13,19], central serous retinopathy [48], optic nerve atrophy [13], optic disc edema [21], and hemorrhages of the optic nerve, retina, or vitreous [1,9,15].

Teichopsias limited to one eye were reported as flashing rays of light, zigzag lightning-like patterns, bright-colored streaks, or halos. Retinal migraine infarction rarely evolved from a progressively enlarging spot that engulfed the entire field of vision [34]. The negative visual losses included blurring, "grey-outs," and "black-outs," causing partial or complete blindness. Visual field defects may be altitudinal, quadrantic, central, or arcuate. Complex patterns of monocular visual impairment, such as the appearance of "black paint dripping down from the upper corner of my left eye," were noted rarely. Most patients who developed PMVL experienced only negative rather than positive features alone or in combination with negative ones. Only approximately 50% of the cases met the current IHS criteria for TMVL and, of those, approximately 50% subsequently developed PMVL. The authors think that irreversible visual loss is part of the spectrum of retinal migraine representing an ocular form of migrainous infarction.

Retinal migraine is primarily a disorder of young women. The gender and age ranges are noted in Table 3. The gender difference was striking in patients with PMVL. That condition occurred 16 times more commonly in women than in men. Those with PMVL also tended to be approximately 10 years older than those with TMVL (37 vs 26 years, respectively). A family history of migraine was noted in approximately 25% of the patients with TMVL and in almost one third of patients with PMVL. Because many cases did not include information on family history, these numbers may be underestimated. Only two patients had familial retinal migraine [35].

Epidemiology and Prognosis

Retinal migraine is thought to be a rare disorder, but its true prevalence is unknown. Recent studies with automated perimetry have demonstrated subclinical visual field defects in patients with migraine [53•]. There was a direct correlation between these findings and duration of disease and advancing age. Although retinal migraine usually has been viewed as a benign condition, it appears that subclinical precortical visual dysfunction and permanent attacks of partial or complete monocular visual loss occur in patients with migraine more often than commonly appreciated [54].

Table 3. Demographic and clinical features of the patients with retinal migraine analyzed in our review (*n* = 77)

Characteristics	TMVL (*n* = 21)	Transformed PMVL (*n* = 20)*	PMVL (*n* = 36)†
Men	11	5	1
Women	10	15	33
Age at onset, *y*	27	25	37.1
Range of ages, *y*	12–53	8–54	15–64
Subtypes of headache			
Migraine with aura	8 women, 8 men	9 women, 4 men	21 women, 1 man
Migraine without aura	2 men	2 women	9 women
Probable migraine	1 woman	4 women, 1 man	1 woman

*Transformed PMVL refers to patients who experienced TMVL and subsequently developed PMVL.
†In PMVL, the gender and subtype of migraine were not reported in two and four, respectively.
PMVL—permanent monocular visual loss; TMVL—transient monocular visual loss.

Minor risk factors for vascular disease were noted in only a few patients with TMVL and PMVL. They included hypertension, hyperthyroidism, pregnancy, diabetes, oral contraceptive use, smoking, and increased levels of Factor VIII. These conditions were not thought to be the main cause of the visual loss.

Differential Diagnosis

The first and primary step in evaluating a patient with a complaint of visual impairment associated with migraine is to determine whether one or both eyes are affected. Retrochiasmatic lesions usually cause homonymous defects with varying degrees of congruence. Prechiasmatic lesions affecting the optic nerve, retina, and associated structures cause monocular visual loss. Many, if not most patients have difficulty distinguishing the loss of vision in one hemi-field versus the loss of vision in one eye. Because a physician usually is not present when the attack occurs, it is essential that the patient be instructed to alternately cover one eye and then the other and compare the two views.

If unilateral visual loss is confirmed, one must attempt to exclude all causes of transient or permanent monocular visual loss; retinal/ocular migraine is a diagnosis of exclusion. One author claimed that "there are no distinguishing symptoms to separate retinal migraine from amaurosis fugax due to retinal embolization" [55••]. However, visual loss associated with retinal migraine usually is of longer duration and the evolution is slower in onset than with microembolization. Moreover, the typical shade dropping over one visual field described by many patients who have microembolization has not been described in patients with retinal migraine.

Diagnostic studies are almost always warranted. Cardioembolic investigations may include electrocardiography, echocardiography, and holter monitoring. Ischemic disease of the eye and brain may be studied by duplex scanning, computed tomography, magnetic resonance imaging and angiography, fluorescein angiography, and conventional angiography in search of an orbital or intracranial lesion. Vascular diseases including vasculitis, hypercoagulable states, and rheumatologic disorders may be evaluated by a complete blood count, prothrombin time, partial thromboplastin time, lupus anticoagulant, anticardiolipin antibody level, erythrocyte sedimentation rate, rheumatoid factor, antinuclear antibody titer, antiphospholipid antibodies, protein C and S and antithrombin III levels, and serum protein electrophoresis. A toxic drug screen should rule out illicit drug use.

Management

There are no clear guidelines for the management of patients with retinal/ocular migraine. Nonpharmacologic measures include avoiding potential migraine triggers. Patients should stop smoking and discontinuation of oral contraceptives may be advisable. Maintenance of healthy habits and eliminating stress are obvious recommendations, but are not followed easily. Because TMVL may lead to PMVL, prophylactic drug therapy is warranted, even though the frequency of attacks may be low. However, there are no data to determine the efficacy of any preventative agents for retinal/ocular migraine. Daily aspirin is recommended. Calcium channel blockers are warranted and all of the other migraine prophylactic agents may be tried. Given the potential risk for monocular complications, those medications with vasoconstrictive properties, such as the ergots and triptans, should be avoided.

Conclusions

The spectrum of monocular visual impairment in migraine is broad. Although the IHS criteria require reversible visual loss, only 53% of candidate patients met these criteria and of those, nearly 50% went on to develop ocular infarction. Most of the remaining patients did not meet IHS criteria because visual loss was permanent. Based on these observations, we suggest amending the IHS criteria for retinal migraine to include two subtypes: retinal migraine with TMVL and retinal migraine with PMVL.

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