

Clinical Spectrum of Migraine Aura Without Headache

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Keywords

migraine aura, scintillating scotoma, ocular migraine, late-life migraine accompaniment, acephalgic migraine

Objectives

At the conclusion of this program, participants should be able to:

1. Apply the International Headache Society diagnostic criteria for the appropriate diagnoses of typical aura without headache and retinal migraine.
2. Identify the characteristic features which allow confident diagnosis of typical aura without headache and retinal migraine.
3. List a differential diagnosis for typical aura without headache and retinal migraine and develop guidelines for further investigation to exclude alternative diagnoses.

CME Questions

1. What are some of the characteristic features suggestive of the diagnosis of typical migraine aura without headache?
2. What is the typical attack duration for typical aura without headache?
3. What are the diseases essential to exclude before diagnosing a patient greater than age 50 with ocular migraine?

Introduction

Migraine aura without headache is a clinical condition frequently encountered in daily practice by neuro-ophthalmologists, ophthalmologists, and neurologists. It is generally a benign condition; however, the varied spectrum of visual manifestations overlaps with other conditions such as intracranial ischemia and epilepsy that may not follow a benign course. Distinguishing isolated migraine aura from other pathophysiologic mechanisms of visual phenomena is the foremost task of the clinician in order to prevent serious ophthalmologic and neurologic morbidity.

Aura is defined as transient, fully reversible visual, sensory, or motor symptoms attributable to migraine pathophysiology, which may or may not be associated with headache. Visual auras, the focus of this review, are the most common of the aura types, representing 99% of migraine auras.¹ It has long been recognized that visual auras may occur in the absence of headache and various terms have been used to describe this clinical situation. These include acephalgic migraine, atypical migraine, complicated migraine, migraine equivalent or variant, and late-life migraine accompaniment. These terms generally

apply to binocular visual phenomena localizable to the optic chiasm or, more commonly, to the retrochiasm visual cortex. The 1998 International Headache Society (IHS) diagnostic criteria identify this condition as migraine aura without headache.² The revised 2004 criteria identify it as typical aura without headache.³ The term *typical aura without headache* will be used in this syllabus. In contrast to binocular visual phenomena from intracranial aura, monocular visual phenomena localizable to the anterior visual pathways may also occur. This is typically in the form of monocular visual loss. The terms ocular migraine, ophthalmic migraine, and retinal migraine generally apply to this condition. Both the 1998 and 2004 International Headache Society diagnostic criteria identify this as retinal migraine.^{2,3} The term *ocular migraine* is used in this syllabus, since the pathophysiology does not always involve the retina, but may at times result from a process involving the choroid or optic nerve.

Diagnostic Criteria

The IHS diagnostic criteria for typical aura without headache are as follows: two or more episodes of aura without headache; aura is fully reversible; aura is indicative of focal cortical dysfunction; at least one aura evolves over 5 minutes; two or more symptoms occur in succession; duration of symptoms is less than 60 minutes; and history and examination are negative for other etiologies of the symptoms (Table 1).³ For ocular migraine the diagnostic criteria are as follows: two or more episodes of monocular positive or negative visual phenomena; visual change is fully reversible; headache typical for migraine occurs during visual loss or within 60 minutes; and history and examination are negative for other etiologies of the symptoms (Table 2).³ Despite the clear delineation of diagnostic criteria, the ultimate diagnosis remains a clinical judgment and the extent of diagnostic work-up may vary for individual patients. There is undoubtedly overlap between typical intracranial aura and ocular migraine symptoms, particularly since many patients' perception of a homonymous hemianopic process is that it is monocular, involving only the eye with the temporal visual field abnormality. In fact, in a descriptive analysis of migraine aura, two-thirds of patients reported a monocular aura.¹ Both typical intracranial aura and ocular migraine may involve positive (photopsias, scintillations) and negative (scotomas) phenomena.

Typical Aura Without Headache

The key to establishing the correct diagnosis is familiarity with the classic features of migraine aura and identifica-

tion of red flags for the diagnosis. Aura accompanies migraine headaches in up to 30% of migraineurs⁴ and, in patients with migraine headache with aura, between 13% and 47% experience episodes of aura without headache.^{5,6} The prevalence of isolated aura without headache is between 1 and 6%^{5,7-9} and onset may be at any age, from childhood into senescence. The majority of information available regarding the characteristic features of typical aura without headache is compiled from case series of patients meeting IHS diagnostic criteria for the disorder. The classic migraine visual aura consists of binocular homonymous hemianopic visual dysfunction in the pattern of a scintillating scotoma, which typically expands slowly and migrates across the visual field. This classic scotoma has been described in detail by many since the 1800s,¹⁰⁻¹² including C. Miller Fisher who described his personal experience with 41 scintillating scotomas between the ages of 59 and 85.¹³ The episode frequently begins with a small line of flickering zigzags, which then expands into an arc and moves toward the periphery before breaking up and disappearing. Often, in the center of the arc is a scotomatous area of blurred or blackened vision.

Although the scintillating scotoma is the most characteristic migraine aura, it may not be the most common. A study examining aura characteristics via questionnaire in 100 migraineurs with aura determined that the most common aura is small bright dots or stars, experienced by 42%.⁶ Eighty-nine percent of patients experienced positive aura phenomena, with only 20% of those experiencing the classic scintillations. Forty-five percent of the cohort experienced negative phenomena, with scotomas being the most common, and 45% experienced disturbed visual perception, with blurred or foggy vision being the most common.⁶ A second study of 156 patients with aura via doctor-directed interview produced alternative results, with over 80% of patients reporting the classic flickering, zigzag lines.¹

Characteristic aura onset is gradual with a build-up and migration of the aura. Duration may vary between 1 and 60 minutes, with the majority of auras lasting less than 30 minutes. A personal or family history of migraine or a personal history of motion sickness in a patient with isolated visual phenomena is strongly supportive of migraine aura as the possible diagnosis. In a young patient (typically accepted as less than 40 years old) with the absence of vascular risk factors, the diagnosis of isolated migraine aura may be made with more confidence and less diagnostic testing than in an older patient. However, episodic visual phenomena compatible with migraine aura have been reported in a number of other conditions, including occipital lobe mass lesions, arteriovenous malformations (AVM), and dural arteriovenous malformations (dAVM) (Table 3).

These conditions are often associated with headache in addition to the visual events. Occipital lobe AVMs may cause aura from vascular steal or from focal seizure, but this is usually followed by a generalized seizure clearly differentiating it from migraine aura.¹⁴ Other causes of occipital seizures cause secondarily generalized seizures in the majority of patients, as well.¹⁵ This makes the diagnosis obvious, but in two-thirds of patients with occipital lobe seizures, severe headache followed the seizure – making differentiation from migraine difficult in patients without seizure generalization.¹⁵ Inter-event homonymous hemianopia suggests a structural lesion, as does a persistent unilateral presentation of symptoms. Visual symptoms from occipital lobe seizures last only seconds, compared to minutes for migraine aura, and they tend to be very elemental, poorly formed phenomena.¹⁵ As noted above, similar elemental phenomena are also common in aura.⁶ None of 26 patients with occipital AVMs experienced a scintillating scotoma characteristic of migraine aura,¹⁴ although 3 of 18 patients with occipital lobe seizures experienced achromatic flickering lights.¹⁵ Sudden onset of stationary visual symptoms is also suggestive of an underlying deficit, although sudden onset visual symptoms occur in 3% of migraine auras.¹ The presence of colored visual symptoms is considered to be more common with occipital lobe seizures, but occurs in up to 33% of migraine auras.^{1,6}

Visual phenomena such as kaleidoscopic images, hemianopic scotomas, and dark spots with or without headache may occur with dAVMs, as well.¹⁶ None of 7 patients with dAVMs had the classic migraine scintillations. Worsening of pre-existing visual auras without headache at the time of diagnosis of acute leukemia was reported in a 41-year-old man, drawing attention to precipitation of aura by systemic abnormalities.¹⁷ The decision of when to pursue neuroimaging or other diagnostic tests in a young person with isolated typical aura without headache is ultimately left to the clinician. No clear recommended guidelines are available, but characteristic features of typical aura without headache are listed in Table 4, and the diagnostic yield of additional testing is likely quite low when several of these features are present. In contrast, diagnostic yield of additional testing such as neuroimaging is greatly increased by the presence of red flags listed in Table 5.

The term late-life migraine accompaniments was coined by C. Miller Fisher and lends validity to the concept that aura without headache may certainly occur in the elderly, perhaps even with a higher frequency than in the young.^{13,18,19} Compared to the 1-6% prevalence of aura without headache in the general population, the prevalence over age 55 is 16%.¹⁸ Two large case series, the first with 120 patients and the second with 85, characterize late-life auras in the absence of headaches in

patients thoroughly evaluated for systemic thromboembolic disease. These studies are strongly supportive of the presence of migraine aura without headache with onset over age 40 and they even document the occurrence of episodes of total blindness and sudden onset homonymous hemianopia in patients with no evidence of thromboembolic disease; however C. Miller Fisher himself underscores the importance of the fact that this diagnosis is one of *exclusion* in an older patient. Patients over 40 with isolated classic scintillating scotomas were excluded from these studies, with the comment by C. Miller Fisher that this entity is clearly accepted as migrainous. Based on these two studies, a new set of criteria for establishment of the diagnosis of migraine accompaniments was outlined as follows:

1. Scintillations or other visual display followed by other neurologic symptoms
2. Build-up of scintillations
3. "March" of paresthesias
4. Two or more similar spells
5. Headache (present in 50%)
6. 15-25 minute spells (95% of TIAs last less than 15 minutes)
7. "Flurry" of spells around age 50
8. Benign course
9. Normal angiography
10. Exclusion of cerebral thrombosis, embolism, dissection, epilepsy, thrombocytopenia, polycythemia, and thrombotic thrombocytopenia

Criteria 9 and 10 underscore the impression that the diagnosis of any aura other than the simple classic scintillating scotoma is one of exclusion. Other features emphasized in these studies are that late-life migraine auras often occur without a past history of migraine and that they may occur in a patient with a migraine history after many years of being headache-free. These features are supported by a review of the Framingham study cohort, in which the prevalence of visual migrainous phenomena was identified as 1.33% in women and 1.08% in men, and episodes of typical visual aura without headache occurred for the first time after age 50, in the absence of headache or a history of recurrent headache.⁷ The benign prognosis of the diagnosis of migraine aura without headache in patients meeting many of Fisher's diagnostic criteria has been supported by other studies.^{20,21}

Ocular Migraine

Ocular migraine is defined as a transient or permanent monocular visual disturbance during migraine or in a patient with a strong migraine history.²² Clinically, it generally presents as transient painless monocular visual loss lasting less than 30 minutes in patients less than age

40 and accompanied or followed by headache. The pathophysiologic mechanism of the visual loss is considered to be vasospasm resulting in transient ischemia of the choroid or retina. The patient may describe central complete or incomplete visual loss, visual blurring or dimming, or scotomatous visual deficits. Retinal vasospasm and ocular hypoperfusion have been directly observed on funduscopic examination and documented by fluorescein angiography during attacks.²²⁻²⁷ Permanent sequelae such as central or branch retinal artery or vein occlusion, retinal infarction, anterior and posterior ischemic optic neuropathy, and vitreous or retinal hemorrhage are infrequently documented.^{23,28-31}

Headache occurs at the time of visual loss or shortly after in 41% of patients and at other times in 25%.²² **The IHS criteria underscore the difficulty of determining a migrainous nature in patients in the absence of headache and caution the clinician to ensure that the diagnosis is one of exclusion in this setting.**

Ocular migraine may have onset at any age. In young patients, transient monocular blindness in the absence of headache not attributable to embolic, rheumatologic, or hypercoagulable disease is often called ocular migraine by exclusion. Diagnostic testing for carotid and cardiac embolic disease in such patients was found to be of very low yield in a study of 100 young patients with amaurosis fugax, but care must be taken with the diagnosis in the absence of headache.³² Many experts, however, prefer to label this as *idiopathic vasospastic amaurosis fugax*³³ since the migrainous nature cannot be determined. The course is typically benign; however, vasospastic amaurosis fugax may be an indication of local or systemic inflammation.²⁴ Appropriate bloodwork and evaluation for collagen vascular disease, vasculitis, and hypercoagulable states is essential if history is not absolutely characteristic of recurrent attacks in the setting of classic migraine headache.²³ In the older patient, carotid and cardiac emboli and temporal arteritis must be definitively excluded (Table 3). There are no visual loss characteristics definitive enough to accurately differentiate *embolic amaurosis fugax* from ocular migraine.³⁴ Attacks of amaurosis fugax from carotid atherosclerotic stenosis are frequently painless, shorter than ocular migraine (less than 5 minutes versus 15-20 minutes), and were considered in the earlier literature to be unassociated with positive visual phenomena; however, a review of 25 patients with amaurosis fugax from carotid stenosis revealed that nearly 50% experienced positive visual phenomena including scintillations and colored images similar to migraine aura and nearly 20% experienced amaurosis lasting between 16 minutes and 12 hours.³⁴ It is known that in 20-30% of older patients with amaurosis fugax, an embolic source is not identified. Ocular migraine is certainly in the differential diagnosis.

Treatment

The decision to treat typical aura without headache and ocular migraine should be based on a discussion with the patient regarding the effect of the visual symptoms on patient function. Do the symptoms interfere with the patient's ability to function normally? Often, once a patient's anxiety over what the symptoms may represent is alleviated, the patient determines that treatment is unnecessary.

If treatment is required, medications useful for migraine prevention are often the mainstay of therapy. Few clinical trials have specifically addressed the efficacy of migraine medications for aura prevention in the absence of headache. However, a few studies have assessed medication effect on the duration of migraine aura. Acutely, magnesium administration may decrease aura duration.³⁵ Sumatriptan, nimodipine, and nifedipine have no effect on aura duration.³⁶⁻³⁸ With regard to the prophylactic medications, metoprolol has no effect on the frequency of migraine auras and even increases auras in some patients.³⁹ An open-label trial of topiramate for migraine with aura resulted in no effect on duration or frequency of auras.⁴⁰ Four studies have assessed the efficacy of lamictal, with mixed results. Three studies demonstrated a decrease in aura duration and frequency,⁴¹⁻⁴³ but one double-blind, placebo-controlled study showed no effect.⁴⁴ For ocular migraine, calcium channel blockers,^{45,46} amyl nitrate, nitroglycerin, aspirin, naproxen, and pentoxifylline may be beneficial.⁴⁷

Conclusions

With regard to the diagnosis of typical aura without migraine and ocular migraine, there is a clinical spectrum. At one end of the spectrum is the young patient with a long history of classic migraines and recurrent, stereotyped episodes of the classic scintillating scotoma, sometimes occurring with the typical headache and sometimes without. There is little question in this patient that migraine is the mechanism behind the visual events and few clinicians would proceed with diagnostic investigations. At the other end of the spectrum is a patient over the age of 50 with vascular risk factors, no history of headaches, and episodes of transient monocular blindness or homonymous hemianopia. It is possible that migraine may be responsible for the symptoms, but it would be dangerous to come to this conclusion without extensive investigations to exclude carotid disease, cardiac disease, and temporal arteritis. With regard to patients who fall between the two ends of the spectrum, few diagnostic guidelines exist regarding the necessity or yield of diagnostic evaluation. One should keep in mind that the IHS criteria for both typical aura without headache and retinal migraine render these conditions as diagnoses of exclusion. The conclu-

sion is that "...the clinician, with his history, remains the only judge of whether a person has migraine."⁴⁷

CME Answers

1. Positive visual phenomena, build-up and migration of the visual abnormality, recurrent stereotyped events, personal or family history of migraine.
2. Fifteen to thirty minutes.
3. Temporal arteritis, carotid atherosclerotic stenosis or occlusion, cardiac emboli.

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Table 1. 2004 IHS Diagnostic Criteria – Typical Aura Without Headache (1.2.3)

Description: Typical aura consisting of visual and/or sensory symptoms with or without speech symptoms. Gradual development, duration no longer than one hour, a mix of positive and negative features, and complete reversibility characterize the aura which is not associated with headache.

Diagnostic criteria.

- A. At least 2 attacks fulfilling criteria B-D.
- B. Aura consisting of at least one of the following, with or without speech disturbances but no motor weakness.
 - 1. Fully reversible visual symptoms including positive features (eg. flickering lights, spots, or lines) and/or negative features (ie. loss of vision).
 - 2. Fully reversible sensory symptoms including positive features (ie. pins and needles) and/or negative features (ie. numbness).
- C. At least two of the following:
 - 1. Homonymous visual symptoms and/or unilateral sensory symptoms.
 - 2. At least one aura develops gradually over > 5 minutes and/or different aura symptoms occur in succession over > 5 minutes.
 - 3. Each symptom lasts > 5 minutes or < 60 minutes.
- D. Headache does not occur during aura nor following aura within 60 minutes.
- E. Not attributed to another disorder.

Notes:

- 1. Additional loss or blurring of central vision may occur.
- 2. History and examination do not suggest other disorder or history and examination do suggest other disorder but appropriate tests exclude the disorder.

Comment:

Only a few patients have isolated 1.2.3. More commonly patients with migraine with aura change to a diagnosis of 1.2.3, especially as they get older. In the absence of headache, precise diagnosis of aura and its distinction from mimics that may signal serious disease (eg. TIA) become much more important. This distinction may require investigations. Especially when aura begins after age 40, when negative features (eg. hemianopia) are predominant, or when aura is very long or very short other causes should be ruled out.

Table 2. 2004 IHS Diagnostic Criteria – Retinal Migraine (1.4)

Description: Repeated attacks of monocular visual disturbance, including scintillations, scotomata, or blindness, associated with migraine headache.

Diagnostic criteria.

- A. At least 2 attacks fulfilling criteria B and C.
- B. Fully reversible monocular positive +/- negative visual phenomena (eg. scintillations, scotomata, or blindness) confirmed by exam during attack or (after proper instruction) by the patient's drawing of a monocular visual field defect during an attack.
- C. Headache fulfilling criteria for migraine without aura begins during visual symptoms or follows them within 60 minutes.
- D. Normal ophthalmological examination between attacks.
- E. Not attributed to another cause.

Note:

- 1. Appropriate investigations exclude other causes of transient monocular blindness.

Comment:

Some who complain of monocular visual loss actually have binocular visual loss. Cases without headache occur but the migrainous nature cannot be ascertained. Diagnosis of exclusion.

Table 3. Differential Diagnosis of Migraine Aura

Binocular aura

Focal occipital epilepsy
Occipital lobe mass lesion
Arteriovenous malformation
Dural arteriovenous malformation
Transient ischemic attack

Monocular aura

Vasospastic amaurosis fugax from an underlying systemic condition
 Temporal arteritis
Carotid embolic amaurosis fugax
Cardiac embolic amaurosis fugax

Table 4. Characteristics Suggestive of Typical Aura Without Headache

Positive visual phenomena – especially classic scintillations
Build-up and migration of the visual abnormality
Aura duration 15-30 minutes
Event associated with nausea, photophobia, phonophobia
Recurrent stereotyped events, especially if alternating sides
Normal inter-event examination
Absence of vascular risk factors
Personal or family history of migraine
Personal history of motion sickness

Table 5. Red Flags for Diagnosis of Typical Aura Without Headache

Onset over age 40
Predominance of negative features (i.e., scotomas)
Brief duration (eg. seconds)
Sudden onset without build-up
Static location without migration
Persistent unilateral location
Inter-event homonymous hemianopia