REVIEW

CME CREDIT

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Migraine aura without headache: Benign, but a diagnosis of exclusion

ABSTRACT

Migraine aura without headache should be considered as a diagnosis in anyone who has recurrent episodes of transient symptoms, especially those that are visual or neurological or involve vertigo. Visual and neurological symptoms due to migraine are not unusual and most commonly occur in older persons with a history of migraine headaches. Migraine aura without headache should be diagnosed only when transient ischemic attack and seizure disorders have been excluded.

KEY POINTS

Migraine auras are reversible and recurrent episodes of neurological symptoms that resolve within 1 hour. They are associated with migraine but may not precede a headache.

Auras almost always involve visual symptoms. Images are usually bright, shimmering, and dynamic, and may form geometric shapes. Ischemic symptoms, on the other hand, tend to be dark and not moving.

Auras usually need no treatment. If desired, a short-acting agent, such as a beta-agonist inhalant, sublingual nitroglycerin, meclofenamate, or naproxen sodium, can be tried. Verapamil or antiepileptic drugs may be used as prophylaxis.

Triptans should not be used to treat an aura. Oral triptans do not act fast enough to affect an aura, and the rapid-acting injectable sumatriptan, if given during the aura, may not abort the subsequent headache.

This paper discusses therapies that are experimental or that are not approved by the US Food and Drug Administration for the use under discussion.

MIGRAINE AURAS can occur alone, without being followed by a headache. This usually benign condition occurs more often in older people with a history of migraine, but it may also occur in others.

This article covers both common and unusual symptoms attributed to migraine aura without headache and how to distinguish them from other conditions. The pathophysiology, epidemiology, and treatment of migraine without headache are also discussed.

DEFINITION

The classification of headache disorders, published by the International Headache Society in 1988 and updated in 2004, defines aura as "a recurrent disorder manifesting in attacks of reversible focal neurological symptoms that usually develop gradually over 5 to 20 minutes and last for less than 60 minutes."[1]

"Migraine aura without headache" is now the accepted term for "migraine equivalents" or "acephalgic migraine": episodic symptoms believed to be migrainous auras but not followed by a headache.

VISUAL SYMPTOMS NOT UNCOMMON

Migraine aura without headache can occur at any age and in people who never had a migraine headache.[2–5] However, it is most common in older people, especially in those who had auras accompanied by migraine headaches when younger.[6] Patients may continue to have migraine headaches in addition to auras without a subsequent headache.

Transient visual disturbances are not uncommon in the older population. On the

TABLE 1

Transient episodic symptoms of migraine aura without headache

Visual
- Photopsia
- Scotoma
- Hemianopsia
- Diplopia
- Blindness
- Metamorphopsia

Neurological
- Paresthesias
- Vertigo
- Amnesia
- Confusion
- Alteration of mood
- Hemiparesis
- Hearing loss

Other
- Cyclic vomiting
- Recurrent abdominal pain
- Coronary artery spasm
- Raynaud disease

Visual aura symptoms are common in older people; nonvisual aura symptoms are less so

other hand, nonvisual symptoms that may be migrainous are so uncommon that very few studies exist on their prevalence and epidemiology.

The Framingham study reported that migrainous-type visual symptoms occurred in 1% to 2% of the elderly participants.[4] Of those reporting such symptoms, 77% said they occurred for the first time after age 50 years, 42% had no history of migraine, and 58% said episodes were never associated with a headache.

Mattsson and Lundberg[5] compared 100 women with migraines in a headache clinic in Sweden with 245 women in the general population and found that the lifetime prevalence of visual disturbances without a headache was 37% in those with migraines and 13% in the general population. Undoubtedly, some of those in the general population had migraines as well.

Ziegler and Hassanein[6] found that 44% of patients diagnosed as having migraine with aura reported having had an aura occur without a headache at some time.

Fisher,[2] in a series of 120 patients over age 40 years with transient episodes resembling migraine auras, found that most had visual symptoms and 20% had nonvisual neurological symptoms.

PATHOPHYSIOLOGY OF MIGRAINE AURAS

Migraine is a very complex inherited disorder, and its pathophysiology is not well understood.

A migraine attack apparently starts in the brainstem and involves activation of the trigeminal nerve and areas of the cortex. Branches of the trigeminal nerve innervate the anterior cerebral vessels as well as the structures of the anterior head and face. Various peptides and other vasoactive substances are released at the neurovascular junction, causing mild vascular constriction followed by dilation and perivascular inflammation.[7] The decreased blood flow does not seem to cause cerebral ischemia.

In addition, a wave of depolarization may spread across the cortex, especially in the occipital lobe. This so-called "spreading depression" accounts for the slowly evolving nature of the visual symptoms typical of a migraine aura. Nonvisual and non-neurological symptoms that are believed to be migrainous (eg, variant angina, vertigo, and abdominal migraine) are likely due to dysfunction of the autonomic nervous system.

The vascular system in general is much more reactive in patients with migraines. They have vasomotor instability and are more prone to conditions such as Raynaud disease, livedo reticularis, vasomotor rhinitis, cardiac arrhythmias, syncope, urticaria, and flushing.

SYMPTOMS INVOLVED IN AURAS

According to the International Headache Society, migraine auras gradually develop over a few minutes and last less than 60 minutes. A number of symptoms have been reported as being migraine aura without headache (TABLE 1).

Visual symptoms—bright and dynamic

Any symptom may be involved in an aura, but visual symptoms occur in 99% of migraine auras[8] and tend to accompany any other neu-

rological symptoms that may also occur.

The most common visual symptom in an aura with or without a headache is *photopsia* (unformed flashes of light). *Teichopsia*, also known as fortification spectrum, is believed to be the most diagnostic of migraine. It involves a bright, zigzag border that looks like an aerial view of an old fortress.

Other common symptoms include scotoma (partial loss of sight or a "blind spot" that is often crescent-shaped), visual distortion, "heat waves," blurring, and hemianopsia.

Much less common are diplopia (double vision) and metamorphopsia (altered or distorted objects).

A typical visual aura is not static but grows and moves across the visual field. This dynamic quality may help differentiate migraine symptoms from symptoms of a transient ischemic attack (TIA).

Migraine visual defects are often bright and shimmering—even a blind spot usually has a bright, shimmering border. Defects may also be multicolored or form a geometric pattern. Dark defects or dimming of the vision suggests ischemic disease.

Visual migrainous auras usually last 15 to 30 minutes. In contrast, TIAs with visual symptoms are usually shorter (3–10 minutes). Partial seizures are also usually short, lasting less than 5 minutes. An arteriovenous malformation involving the occipital lobe may cause transient visual symptoms, which are also short in duration.[9] If visual symptoms are recurrent and last longer than 60 minutes, an underlying disorder such as retinal disease, recurrent emboli, a coagulopathy, or vasculitis should be considered.

The visual symptoms of migraine are homonymous—occurring in the corresponding vertical halves of the visual fields of both eyes—due to occipital lobe cortical dysfunction. However, it is often difficult for patients to determine this because they tend to notice a problem on only one side of their vision. A strictly monocular defect suggests retinal disease or ischemia due to carotid artery disease. Ocular migraine, also known as retinal migraine, is very rare and causes strictly unilateral visual symptoms. It usually occurs in young people with a history of migraine.[9] Some people use the term "ocular migraine" generically for any visual events of a migrainous nature occurring without a headache.

Neurological symptoms

Neurological symptoms are the second most common aura symptoms that occur without a headache and, as with other migraine auras, tend to last 10 to 30 minutes.

Neurosensory symptoms, ie, numbness, tingling, or paresthesias, are most common. The paresthesias typically slowly spread up or down the limbs: most commonly, the sensation starts in the fingers, then spreads to the hand and slowly up the arm. This "march" or spreading sensation may go on for several minutes and may progress to the face and mouth area, cross over, and descend the other arm. Episodic numbness of the tongue without other symptoms is thought to most likely be due to migraine.[2]

Mild weakness of the limbs, with the hands and arms most often involved, may also occur. In migraine, the area where the symptoms first appear (usually the hand) clears first, in contrast to ischemic symptoms, in which the area initially involved usually clears last.

A typical visual aura grows and moves across the visual field

Total global amnesia is most often seen in patients with migraine, usually without a headache. Typically, it occurs only once or twice in a lifetime, but some people have repeated spells. The amnesia usually lasts 1 to 2 hours but may be briefer. People appear to function normally during the period of amnesia and are able to carry on normal conversations and activities, such as driving or shopping, but cannot afterwards recall anything that happened. The preamnesia memory is intact when the episode is over.

Speech disturbances, ie, expressive aphasia or dysarthria (disturbed articulation), sometimes occur during migraine attacks, occasionally without an accompanying headache.

Vertigo not uncommonly accompanies migraine, sometimes without a headache.[10] Vestibular studies are normal, and the attacks are much shorter than usually seen in Meniere disease. Migraine aura without headache should be considered in anyone with recurrent attacks of vertigo with normal auditory and vestibular testing.

TABLE 2

Visual symptoms of migraine aura vs transient ischemic attack (TIA)

FEATURE	MIGRAINE AURA	TIA
Duration	15–30 minutes	3–10 minutes
Quality	Dynamic, bright, multicolored Forms geometric patterns	Static, dark Dimming of vision

In children, benign paroxysmal vertigo is believed to be migrainous; most children diagnosed with it eventually develop migraines.[11] The attacks involve sudden vertigo that lasts for a few minutes to a few hours: the child may suddenly stop playing and fall to the ground. Recovery is complete, with no residual auditory or vestibular abnormalities evident.

Confusion can accompany migraine, especially in children, and may occur without headache. Patients with migraines are more likely than the general population to develop confusion after a mild head trauma, such as a sports injury. Adolescents with spells of confusion independent of a migraine headache may be suspected of drug abuse. A history of migraine headaches may be helpful in this situation.

Seizures come on much more rapidly than migraine auras

OTHER POSSIBLY MIGRAINOUS CONDITIONS

A few recurring conditions that are very rare are also thought to be migrainous.

Cardiac migraine, described in 1974, is believed to be due to coronary artery spasm in patients with migraines, at least in some cases.[12] The prevalence of migraine in a series of patients with variant-type angina (coronary artery spasm) was found to be 26%, vs 6% in patients with typical coronary artery disease and 10% in noncoronary controls.[13]

Two rare conditions, cyclic vomiting and abdominal migraine, arise mainly in children, especially those with a family history of migraine. Most children diagnosed with either of these conditions develop migraine later in life. Before diagnosing these conditions, any possible underlying disease must be excluded by examination and testing.

Cyclic vomiting involves severe, self-limited vomiting spells that may last for several hours. The child is typically irritable and may be photophobic. Episodes end after the child sleeps.

Abdominal migraine involves episodic midline abdominal pain lasting 1 to 8 hours. It is associated with nausea, vomiting, often pallor,[14] and sometimes a mild nonmigrainous headache.

Abdominal migraine can occur in adults but is extremely rare. One woman patient of mine had a history of migraine and recurrent epigastric pains. Several workups revealed no abnormality. The attacks ceased when she was treated with a beta-blocker, a drug commonly used to prevent recurrent migraines.

DIFFERENTIAL DIAGNOSIS

If other causes are eliminated, any recurrent, transient, and episodic symptoms that are fully reversible and last less than 1 hour should be considered migrainous. The diagnosis of migraine aura without headache should be made only when other possible causes have been excluded.

TIAs must be ruled out, since migraine aura without headache commonly affects older persons, in whom vascular disease is more prevalent. Visual symptoms of TIAs can often be distinguished from those of migraine auras (TABLE 2). Ischemic eye symptoms are usually shorter, do not move and spread across the visual field, and generally result in dimming of vision.

Retinal disease can be manifested as flashes of light but tends to linger for long periods of time. Ischemia and retinal diseases cause symptoms that are strictly monocular, not homonymous as in migraine.

Partial seizures may cause repetitive stereotypic symptoms similar to auras but do not occur initially in an older person unless a brain lesion exists (eg, from trauma, vascular disease, neoplasm). A history of months or years of repetitive transient symptoms without evidence of permanent deficits would suggest that such episodes are migrainous.

Seizures hit very quickly, and if there is numbness or tingling, it very rapidly spreads over a limb in a few seconds. Migrainous

paresthesias generally take a few minutes to spread up a limb. The neurological symptoms in a seizure disorder are much shorter than those of migraine.

Recurrent emboli to the brain can cause transient visual or neurological symptoms, but it would be unusual for the same pattern of symptoms to recur many times. Clotting disorders, polycythemia, thrombocytosis, and vasculitis may cause transient visual or neurological disturbances and need to be excluded by the appropriate blood tests.

EVALUATION

If a patient has had recurring symptoms for some time that are typical of migraine aura but has no deficits found on the physical or neurological examination, a complete workup with laboratory and imaging tests is probably not necessary.

However, a complete evaluation should be done if a patient is seen after having only one or two attacks.

Magnetic resonance imaging should be done to rule out a cerebral infarction or a mass lesion.

Magnetic resonance angiography and **vascular ultrasonography** should also be done to evaluate the intracranial and extracranial vessels for stenotic lesions, due either to arteriosclerosis or a vasculitis.

Electroencephalography is needed if the attacks are not typical of migraine aura and might be due to epilepsy.

Laboratory testing for clotting disorders and inflammatory vascular disease may also be necessary.

TREATMENT

Generally, auras without headache do not occur frequently and require no treatment. Once the diagnosis is made, patients can be reassured that the condition is benign.

For patients who want treatment, a few medications can be tried. However, it is difficult to shorten attacks because they are so brief and require very fast-acting agents.

Isoproterenol, an inhaled beta agonist, may shorten the aura.[15]

Vasodilators. With the recent evidence that aura is caused by spreading neuronal depression rather than significant cerebral ischemia, it is possible that vasodilating drugs have other effects in addition to their effects on vessels. Although sometimes useful for treating migraine headache, vasodilators often exacerbate the pain of migraine if taken at times other than very early in the aura.[16]

Sublingual nitroglycerine has been useful in a few of my patients who travel a lot and feared an attack of visual loss while driving.

Sublingual nifedipine has also been helpful but is no longer used because of the risk of profound hypotension.

Rapid-acting nonsteroidal anti-inflammatory drugs such as meclofenamate or naproxen are occasionally effective in shortening the duration of symptoms.

Triptans should not be used to treat an aura. Oral triptans do not act fast enough to affect an aura, and if the rapid-acting injectable sumatriptan is given during the aura, it may not abort a subsequent headache.[17] Because they may cause vascular constriction, triptans need to be used with great caution in older patients who may have vascular disease, hypertension, or other cardiovascular risk factors.

Generally, auras without headache require no treatment

Preventive therapy

Preventive therapy should be offered if attacks are frequent enough or severe enough to cause disability.

Calcium channel blockers, particularly verapamil, are often very effective.

Antiepileptic drugs such as valproic acid, gabapentin, and topiramate are also used and are effective in reducing the frequency and severity of auras without headache.[18]

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