Migraine Aura Without Headache

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Migraine is described as a familial disorder characterized by recurrent headaches that are variable in intensity, frequency, and duration.[1] Attacks are usually unilateral but can also be bilateral and accompanied by throbbing pain, photophobia, phonophobia, nausea, and vomiting. Some migraines are preceded by, or are associated with, neurological and mood disturbances. All of the above characteristics, however, are not necessarily present in each attack, nor in each patient.[2]

It has been suggested that the prevalence of migraine is probably markedly underestimated. Estimates range from 10% to 34% of the general population, with some authors reporting both age-related and sex-related differences. Prevalence appears to be highest among women and among young adults of both sexes. Patients with acephalgic migraine (migraine aura without headache) may represent as many as 3% of migraineurs.[3]

Case Report

A 40-year-old man presented for evaluation of a 12-year history of periodic visual disturbance. The disturbance, which occurs approximately every 6 months, reportedly begins centrally as a small, bilateral, circular distortion, and then expands, over a 20-minute period, into an enlarging three-quarter circle of brightly colored and flickering lights described as being "similar to multiple small prisms laid side-by-side in semicircular fashion." The disturbance continues to enlarge until it grows out of the patient's field of vision. There is no history of paresthesia, olfactory or auditory disturbance, nausea, vomiting, or headache preceding, during, or subsequent to the disturbance described. The patient also denies antecedent trauma or emotional stress. The episodes are reportedly always similar in nature, with an expanding scintillating scotoma and without subsequent headache. The patient's first episode, his most recent episode, and "a few" of the others have occurred after a 60-minute exercise period, which he performs consistently as a matter of his daily routine. There is a history of myopia, for correction of which soft contact lenses are used, and numerous vitreous floaters have been reported. The patient takes no medication, has no history of illicit drug use, and is otherwise healthy except for a history of mild seasonal allergic rhinitis. There is a family history of migraines.

A physical and neurologic examination of the patient was performed, and the findings were within normal limits. The diagnosis of migraine aura without headache was made. The patient was not placed on any specific therapeutic intervention. Since his episodes of scotoma occur only sporadically (every 4 to 6 months) and are not associated with additional symptomology, initiation of prophylaxis or abortive therapy was deemed inappropriate.

Long-term follow-up of the patient includes annual examination. He has been encouraged to keep a diary of visual phenomena, paying particular attention to activity, diet, and associated symptoms.

Discussion

The terms *acephalgic migraine* or *migraine equivalent* have been replaced within the Classification and Diagnostic Criteria for Headache Disorders, Cranial Neuralgias and Facial Pain by the Headache Classification Committee of the International Headache Society. These previous designations have been replaced by the term *migraine aura without headache*, which describes migrainous events exclusively manifested by one of the neuro-

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logical disturbances that usually precede or accompany the headache of classical migraine.

Approximately 20% of migraineurs may experience acephalgic attacks of migraine at one time or another.[1] Indeed, that various symptoms can occur in the absence of any headache has been noted for some time; in fact, visual phenomena were described as early as the 12th century. It is not, therefore, surprising that vivid accounts of migraine accompaniments exist throughout history, since the scintillating scotoma is the most frequently occurring visual symptom in migraine with or without headache. Terms such as "sparkling, dancing lights," "vibrating light," "wiggly line," "shimmering, like heat off a hot road," "jagged flashes," and "flickering light" are often used to describe this dramatic occurrence. Colors most often reported are red, gold or yellow, green, and blue or purple.[4]

While acephalgic migraine would fall into the category of migraine aura without headache, episodes of migraine aura without headache can occur in individuals with a history of classic migraine. When acephalgic migraine begins after the age of 40, the diagnosis must be suspect, since thrombotic transient ischemia can also be the cause. These patients have a history of classic migraine in younger years, then the headaches cease and the patients begin having what C.M. Fisher has called "late-life migraine accompaniments."[5,6] Since one must be careful to assure no other underlying pathology, extensive studies of the heart and vascular system are frequently needed to confirm this diagnosis.

Pathophysiology

As described above, visual dysfunction is the most frequent manifestation of migraine aura without headache (Table 1). The classic scintillating scotoma, with march of fortification figures and expansion over time, is characteristic of migraine. Typically, some alteration of perception initially occurs either as an indistinct opacity or as a loss of illumination. This sensation is followed, in seconds to minutes, by the appearance of a small central scotoma of only a few degrees that is lined on one side with a luminous, zigzag line, termed the *fortification spectrum*. The spectral lines usually form an acute angle and are nonconfluent. These lines may be colored or gray, and they appear to oscillate in brightness, suggesting a boiling or rolling effect. The fortification spectrum expands in the shape of a horseshoe and is often lined on the inner edge by an absent area of vision termed the *negative scotoma*.[1] The term *buildup* is used to describe the gradual expansion of the fortification spectrum, and occurs in 75% of cases.[2] As the scotoma expands, it typically drifts or "marches" toward the periphery, this process generally lasts from 20 to 30 minutes. There are

Table 1. Neurologic Manifestations Associated with Migraines

Visual disturbances
Scintillating scotoma (fortification spectrum/wavy vision)
Heminopia
Diplopia
Blurred vision
Blindness
Vertigo
Sensory disturbance
Face (when around the mouth and hand: chiro-oral)
Aphasia
Hemiparesis/Hemiplegia
Decreased hearing (very uncommon)
Decreased level of consciousness (rare)
Pupillary abnormalities
Mydriasis

estimates that the cortical disturbance responsible for the scotoma proceeds at a rate of 3 mm per minute through the visual cortex.[7] During the scotoma, mental efficiency may be impaired, and reasoning, reading, writing, or speaking may be affected.[2,8] Men seem to experience more specific, sharp, bright lines in their scotoma, while women more often describe blurring and bright spots.[7]

Theories abound regarding the cause of the migraine phenomenon. The major explanations include vascular reactivity (vasoconstriction-vasodilation),[8] changes in brain chemistry (serotonin), abnormalities of platelet function, and spreading cortical depression. The vasospastic and vasodilatory explanation is perhaps the oldest and most persistent and has enjoyed qualitative support over the years, particularly with regard to the existence of vasoconstriction during the migraine process.[1]

In studying patients with classic migraine, Olsen[9] and Lauritzen[10] found that by intracarotid injection of xenon, regional cerebral blood flow was reduced during the aura, beginning in the occipital area and gradually spreading anteriorly over the entire hemisphere. The reduction of regional cerebral blood flow did not respect vascular territories, but instead seemed to respect architechtonic territories such as the central sulcus and the sylvian fissure.[9,10] The authors were not able to demonstrate regions of increased flow; thus, their findings call into question the role of vasodilation in the migraine process. Furthermore, they could not reproduce similar decreases in flow in patients with common migraine (migraine without aura).

Differential Diagnosis

As Wiley[11] has noted, patients are usually able to adequately describe their symptoms by using their hands to

show the vibrations starting in the center of the visual field and progressing to the periphery over a variable time course of 15 to 30 minutes. Having the patient diagram the aura on paper is also helpful. Clearly, the old adage "the more complete the syndrome, the less difficult the diagnosis" is quite true in the case of acephalgic migraine. In the absence of the classic sick headache, the patient's predominant visual phenomenon must be well described and chronicled in order to avoid diagnostic errors. Thus, when a patient is unable to provide an accurate accounting, the clinician is compelled to search for other causes of photopsia: environmental agents, or specific abnormalities of the eye, including problems with the cornea, lens, vitreous body, and retina, or abnormalities of the brain or vascular system (Table 2).

Workup

The diagnosis of migraine aura without headache should be made only after the possibility of organic disease has been systematically excluded through a detailed patient history. As Hupp[1] notes, even the astute observer may ascribe a monocular origin to the episodes when the disturbance in the contralateral visual field is not appreciated. Indeed, when visual phenomena occur in the peripheral visual field, an occipital source is most likely. It is therefore important to instruct the patient that, during an attack, each eye should be occluded in turn so that the field of the uncovered eye can be carefully observed. Hupp suggests displacing one eyeball using a finger. The visual disturbance will move with the eyeball if it originates in the retina, but will remain fixed if it originates in the occipital cortex.[1]

The diagnosis of migraine aura without headache can be entertained if the patient has the major migraine characteristics, including migration of scintillating scotoma, recurrences of similar episodes of 15 to 30 minutes' duration, a history of similar spells with headache, an eventually benign course, and a normal physical, ophthalmologic, and neurologic examination.

Underlying organic disease must always be considered, however, if the history does not fit or if the neuologic examination is abnormal. A computed tomography (CT) scan and magnetic resonance imaging (MRI) are necessary to exclude a mass or lesion, and MRI should be performed if a venous thrombosis is suspected (eg, in a postpartum woman with new-onset migraine equivalents). A hematologic evaluation might include a complete blood and differential count and partial thromboplastin time. A vasculitis panel would include erythrocyte sedimentation rate, rheumatoid factor, antinuclear antibody titer, and serum protein electrophoresis. The cardiovascular evaluation includes electrocardiography,

Table 2. Causes of Photopsia

Environmental
Reflections off corrective lenses or windows, lightning flashes and other environmental light sources can, on occasion, be mistaken for a visual phenomenon.
Drugs: digoxin, clomid
Eye
Cornea
Corneal edema can cause halos or rainbows around lights, similar to those caused by acute glaucoma.
Corneal foreign bodies can cause light distortions due to localized edema.
Mucous strands which can accompany a conjunctivitis might result in a disruption of vision.
Lens
Cataract formation—the posterior subcapsular variety often predisposed to light distortions.[11]
Vitreous
Vitreous traction can produce the "lightning flashes of Foster Moore." This phenomenon is generally seen by elderly individuals on moving the eyes rapidly far to the side. They occur only for a second or two. They are benign and eventually disappear, or they are replaced by small black conglomerates in the peripheral fields upon rapidly moving the eyes in extreme ranges.[8]
Phosphene of quick eye motion.
Retina
Retinal microemboli can produce a transient impression of flashes or sparkles of light of short duration.
Retinitis can occasionally cause a distortion described as flashes of light. This is probably due to vitreous traction. Retinal tears can produce transient flashes of light which can be followed by floaters due to intravitreal hemorrhage.
Papillitis
Brain
Concussion may be associated with transient photopsia.
Seizure aura: prior to a generalized seizure can embody a light distortion similar to the scotoma of migraine. Partial seizures have also had primary sensory changes associated with EEG changes.
Masses: tumor, arteriovenous malformations. A rare focal lesion over the visual cortex or involving its connections has been known to simulate exactly the progression of the migrainous hemianopic scotoma.[7]
Psychosis with accompanying visual hallucination may present a confusing picture.
Vascular
Amaurosis fugax (transient monocular blindness) embolic phenomena or ocular hypoperfusion due to atherosclerotic narrowing of nutrient arteries could be confused with a migraine accompaniment.
Involution of the occipital lobe.
Cerebral venous thrombosis. There have been reports of migraine-type visual disturbances associated with venous thrombosis with cerebral infarction.
Vasculitis and lupus anticoagulant have been associated with migraine.[12]
Giant cell arteritis can present with episodic blindness as a prodrome for days to months in almost half the patients prior to persistent visual impairment.[7] These patients often have a variable history of headache.

echocardiography, and possibly cardiac angiography. If extracranial carotid narrowing is suspected, noninvasive ultrasound Doppler can be utilized; however, complete carotid evaluation requires cerebral angiography.[1] An electroencephalogram is needed if a seizure is suspected.

Treatment

There is a paucity of information on the treatment of migraine aura without headache. No therapy is required for rare occurrence; however, with increased frequency of attacks, some form of intervention seems warranted. Prolonged auras or bothersome auras may respond to rapidly acting effervescent aspirin. This is probably the most useful intervention for the case of migraine aura without headache. Other abortive drugs that might be helpful but are currently without Food and Drug Administration approval for this use are the calcium channel blockers, particularly sublingual nifedipine.

Patients using oral contraceptives who develop migraine symptoms have a higher incidence of neurophthalmic sequelae, including cerebral vascular accidents.[10] Any patient with scintillating scotoma who is using an oral contraceptive should be advised to discontinue the medication and to adopt an alternative form of birth control.

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