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Expert Opinions

Are Blurred Vision and Short-Duration Visual Phenomena Migraine Aura Symptoms?

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Case 1.—A 29-year-old woman has a 6 year history of headaches 2-3 times a month. She describes a left temporal throbbing with an intensity of 8 out of 10 associated with nausea, light and noise sensitivity but no vomiting. She also reports that her vision is blurred in both eyes during the headache. She takes sumatriptan 100 mg with relief of the headache in 1-2 hours. Stress, lack of sleep, and menses are triggers.

Case 2.—A 38-year-old woman has a 10 year history of headaches described as a generalized pressure and throbbing with an intensity ranging between 5 and 8 out of 10 associated with nausea, vomiting about 10% of the time, light and noise sensitivity occurring about once a week. About 20% of the time, she sees zig-zags in the left or right field of vision, which she has timed and that last 2 to 3 minutes. She takes rizatriptan 10 mg with relief of headaches in about 2-3 hours. Stress is a trigger.

Questions.—Is blurred vision in both eyes alone a migraine visual aura? According to the International Classification of Headache Disorders, 3rd edition criteria, a visual aura should last 5 minutes or more? Is it possible to have a visual aura lasting less than 5 minutes? What visual symptoms other than blurred vision might occur during or before a migraine lasting one minute or less which are not due to migraine aura?

DISCUSSION

(Case 1) According to the International Criteria for Headache Disorders version 3 beta (ICHD-3β), migraine aura is a fully reversible visual, sensory, speech and/or language, motor, brainstem or retinal symptom.[1] At least one symptom should develop gradually over a minimum of 5 minutes and each aura symptom lasts 5-60 minutes. Motor symptoms may last for 72 hours. Although not specifically stated in the diagnostic criteria, the comments in section 1.2 indicate that migraine aura is associated with regional changes in cerebral blood flow and that cortical spreading depression of Leão is the likely

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underlying mechanism. This implies that aura originates in the brain (cortex or brainstem) or in the retina (which is essentially part of the brain).

The visual manifestations of aura are generally divided into (1) positive phenomena, (2) negative phenomena, and (3) perceptual phenomena, with positive phenomena occurring most frequently.[2] Positive phenomena include fortification spectra (teichopsia), with shimmering lights that generally have an angulated geometry ("zig-zags"). The classic scintillating scotoma also includes an area of negative phenomena (scotoma) circumscribed by a shimmering border that gradually enlarges over the visual hemifield and breaks up into the periphery. Other manifestations of positive phenomena include phosphenes (spots of light or a "flash bulb" effect), dark spots, sparkles and dots in the vision. Scotomata, homonymous hemianopia, tunnel vision and complete visual loss are examples of negative phenomena. Perceptual abnormalities include kaleidoscope vision, fragmented vision ("cracked glass"), corona phenomena, achromatopsia, and various forms of distortion (metamorphopsia), such as heat waves, macropsia (things look too large), micropsia (things look too small), telopsia (things look farther away), and pelopsia (things look too close).[3] The experience of metamorphopsia is commonly termed the Alice in Wonderland syndrome, of which migraine is the most common etiology.[4] The visual manifestations of retinal migraine are either positive (spots or flashing lights) or negative (monocular scotoma or total monocular visual loss).[5]

Patients often have difficulty describing their aura symptoms, particularly whether they are binocular or monocular. This is surprisingly true even in individuals who have had aura for years but never analyzed or articulated their symptoms. Positive visual aura phenomena are often still visible with the eyes closed, so asking about monocular viewing may not be informative. A homonymous hemianopia is frequently described as visual loss in one eye, rather than in one hemifield. Asking the patient what they were able to see with both eyes open while looking straight ahead often clarifies the homonymous nature of the visual loss; loss of vision in one eye does not produce hemifield loss with binocular viewing. When suspecting monocular visual loss, asking the patient whether they covered *either* eye – not "one eye" – is helpful; often they have never tried this. Accurate reporting of the aura duration is unreliable unless the patient has actually timed the aura.

Blurred vision is consistently the most frequently reported visual manifestation of migraine, endorsed by 54.1% of 122 patients in our study using a structured questionnaire that included blurred vision as a descriptor.[6] However, patients who had blurred vision as their sole visual aura manifestation were not included in the study. A previous retrospective study of 100 patients identified 27 with "foggy" or blurred vision.[2] A prospective diary study of 216 auras in 72 patients reported "foggy" or blurred vision as an isolated visual aura symptom in 25% of visual auras, and in 11% of all visual auras overall.[7] Many patients with migraine complain of blurred vision during the headache phase of their attacks, often lasting hours, which exceeds the usual accepted duration of migraine aura. Nonetheless, if one accepts the premise that migraine aura arises from the brain or the retina, blurred vision does not qualify as an aura symptom.

Blurred vision, defined as seeing images out of focus, arises from an ocular cause. It may be due to the size of the globe (myopia, hyperopia), or an abnormality in the cornea, lens, the aqueous humor or the vitreous. Common corneal conditions causing blurred vision include dry eyes, astigmatism (irregular corneal contour) and corneal edema. Cataracts and accommodation changes (eg, presbyopia) arise from the lens. Inflammation within the aqueous or vitreous may produce blurred vision, usually with extreme photophobia. However, conditions that affect the retina, the optic nerve, and the brain do not produce purely blurred vision as a symptom. Acute retinal problems such as retinal detachment, retinal traction, or macular edema cause flashes, floaters, scotomas, and visual distortion. Migrainous visual symptoms arising from the brain include homonymous hemianopia, metamorphopsia, achromatopsia, tunnel vision, complete visual loss, heat waves, fractured and kaleidoscope vision, as well as formed hallucinations.

With the very rare exception of a unilateral lesion affecting the unpaired nasal fibers

representing the extreme temporal visual field of the contralateral eye in the anterior occipital lobe ("temporal crescent" or "half-moon syndrome"), cortical abnormalities always produce binocular symptoms.[8] The fact that patients with cluster headache and other trigeminal autonomic cephalgias (TACs) with blurred vision or photophobia often report that it is monocular and ipsilateral to their headache argues against a central etiology for this symptom. A prospective study of patients with migraine, various TACs and other primary headaches found unilateral photophobia in 2 of 54 patients with episodic migraine, 6 of 48 patient with chronic migraine and 3 of 22 patients with new daily persistent headache.[9] Unilateral photophobia was more common in patients with TACs, experienced by 4 of 5 with cluster headache, 5 of 9 with SUNCT, 5 of 6 with probable TACs, 5 of 11 with hemicrania continua, and 3 of 6 with chronic paroxysmal hemicrania.[9] The photophobia was ipsilateral to the headache in more than 50% of patients with TACs.

Functional imaging sheds little light on the topic. There is one case report of a 21-year-old woman with a history of migraine without aura who experienced a spontaneous migraine headache during participation in a research study of cerebral blood flow using positron emission tomography.[10] During the course of the scan, which required fixating on line drawings on a computer screen, she developed her usual migraine and bilateral changes in blood flow in the occipital lobe were present after the onset of pain. The blood flow changes spread across the cortical surface at a relatively constant rate, spanning four different cerebral artery territories, consistent with cortical spreading depression. The participant indicated inability to focus clearly on the line drawings during one 15-minute segment of the scan. The authors commented on her "hazy vision" as "less typical" aura symptom, noting that these symptoms had not been previously associated with blood flow abnormalities. In a different circumstance, a report of a 29-year-old man with psychogenic visual loss (measured visual acuities of 20/1000 in the right eye and 20/2000 in the left eye and "spiral" visual fields) found marked reduction of cerebral blood flow in visual association areas bilaterally, sparing primary visual cortex.[11] The interpretation was that suppression of the visual association area is associated with the development of a psychogenic visual disturbance. As there is no indication that that normal acuity could be demonstrated by various maneuvers on the examination, it is uncertain whether or not this patient truly had blurred vision.

I propose that blurred vision is a trigeminal autonomic symptom, likely referable to the parasympathetic nervous system. Within the pterygopalatine fossa, the parasympathetic secretomotor fibers travel with the zygomatic nerve and then join the lacrimal branch of the ophthalmic division of the trigeminal nerve. They supply sensory innervation to the lacrimal gland as well as the eyelid and conjunctivae. The parasympathetic component, derived from the facial nerve, is mediated by the greater petrosal nerve. Activation of parasympathetic system produces lacrimation and rhinorrhea.[12] The greater petrosal nerve, ophthalmic (V1) and maxillary (V2) nerves are in close proximity to each other, which explains why lesions of the trigeminal nerve may cause impaired lacrimation. There is also sympathetic innervation to the lacrimal gland, arising from the superior cervical ganglion. The sympathetics do not synapse in the pterygopalatine fossa but travel with the parasympathetic fibers innervating the lacrimal gland.

Trigeminal autonomic symptoms arise from an imbalance between sympathetic and parasympathetic inputs. For example, parasympathetic overactivity produces nasal congestion via the muscarinic receptors in the muscosal epithelium, whereas the interaction between parasympathetic reflexes and sensory axon responses are the likely mechanism of rhinorrhea.[13] Similarly, decreased parasympathetic input or sympathetic overactivity during migraine produces decreased lacrimation which, in turn, causes corneal dryness. Blurred vision is one of most common manifestations of dry eye, along with ocular discomfort and photosensitivity. Although corneal edema produces blurred or hazy vision, there are no examples of idiopathic episodic or paroxysmal corneal edema in the literature, which makes this an unlikely mechanism. Ipsilateral involvement of the sympathetic and parasympathetic fibers in the pterygopalatine fossa, as occurs in cluster headache and other trigeminal autonomic cephalgias, would account for ipsilateral visual symptoms in these disorders.

Regarding the duration of aura symptoms (Case 2), there is no explanation given in the ICHD-2 or 3β rationalizing the cut-off points of 5-60 minutes for typical aura. Most of the recent literature regarding aura focuses on long-duration aura and I was not able to find any studies specifically investigating aura lasting less than 5 minutes. A systematic review of aura symptoms published in 2013 found one abstract reporting visual aura duration less than 10 minutes in two of 20 patients.[14,15] A prospective diary study of 54 patients recording three consecutive auras found that 6% experienced visual aura lasting less than 10 minutes.[16] A questionnaire-based study of migraine visual aura reported symptom duration from 1 to over 60 minutes but did not further characterize patients with symptoms lasting less than 5 minutes.[2] Our study of migraine visual aura used a lower limit of 5 minutes when querying about duration.[6] A prospective diary study found a duration of visual aura ranging from 5-860 minutes but only included patients meeting ICHD-2 criteria with aura duration of at least 5 minutes.[16] The 5 minute lower limit cut-off in ICHD-3β is presumably to distinguish migraine visual aura from occipital seizures or transient ischemic attacks (TIAs). Occipital seizures are rare, generally described as colored circular patterns with or without additional shapes that move, spin or rotate.[17] The duration is usually less than one minute and they tend to be frequent, often occurring daily. There is an EEG correlate with epileptiform occipital discharges. Brief episodes of blindness or hemianopia have been reported.[17] Treatment, consisting of either carbamazepine or valproate, eliminates the clinical seizure activity.

TIAs consist of transient episodes of neurologic dysfunction cause by focal brain, spinal cord or retinal ischemic without acute infarction. They are no longer defined by duration, as duration is not a reliable indication of infarction. Infarction is tissue-based on neuropathologic, neuroimaging, or clinical evidence of permanent injury.[18] A study of patients coming to the emergency department with episodes of transient neurological dysfunction undergoing extensive neurovascular evaluation compared 32 patients with migraine with aura with 32 patients with isolated migraine aura (without headache) and 32 patients diagnosed with TIA using ICHD-3β criteria. Fisher criteria were employed if the event was the first episode or there were motor symptoms.[19,20] Patients with known epilepsy were excluded. Patients with TIA were older than those with migraine (mean age 65 years, vs 50 for migraine with aura and 40 for isolated migraine aura) and were more often male (82% vs 6% migraine with aura and 19% isolated aura). Patients with TIA were 10 times more likely to have a history of stroke, hypertension, and dyslipidemia (all $P < 0.05$ compared to migraine groups). Visual symptoms were present in 63% of those with migraine aura and headache, 41% of patients with isolated migraine aura, and 10% of patients with TIA. Patients with migraine often had at least two types of symptoms (eg, somatosensory, motor, speech, vestibular) evolving over a longer duration than with TIA, whose symptom onset duration was less than one minute. Transient neurologic deficits tended to be briefer (<1 hour) in TIA patients and lasted longer in patients with migraine (1-24 hours) but no specific clinical or laboratory feature effectively discriminated between TIA and migraine.

A narrative review of late-life migraine accompaniments endorsed Fisher's initial descriptions, indicating that migraine aura usually starts gradually over minutes, in contrast to vascular events or seizures that start suddenly.[21] The march of symptoms with sequential evolution of deficits arising from multiple vascular territories is a hallmark of neurological migrainous accompaniments. A previous history of migraine with aura is often present. In contrast, the visual symptoms of TIA are often negative (ie, hemianopic), begin suddenly and last 3-10 minutes. However, the homonymous hemianopia of migraine most often starts abruptly as well.

Brief, positive visual phenomena lasting less than a minute may arise from the retina. Vitreous traction on the retinal photoreceptors may produce a flash of light resembling lightning.[22] Momentary phosphenes with rapid eye movements may occur in normal individuals. Single discrete or flickering phophenes can be generated by the retina, choroid or optic nerve.

Patient 2 presented by Dr. Evans saw zig-zags in one hemifield lasting 2-3 minutes associated with a typical migraine headache responding to rizatriptan.

The pattern of her visual disturbance is characteristic of migraine and she has a history of previous, stereotypical events. Despite their short duration, her visual aura symptoms are most certainly migrainous. The ICHD definition of aura duration was developed primarily for research purposes, to ensure relative homogeneity among patients participating in studies. In clinical practice, an unusually short or long duration of aura prompts concern for a secondary cause, which is a practical consideration.[23] There are no data in medical literature to "disqualify" patients with aura symptoms lasting less than 5 minutes or longer than one hour from being diagnosed with migraine in the appropriate clinical circumstance. Therefore, I am comfortable diagnosing her visual symptoms as aura.

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