Persistent positive visual phenomena in migraine

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Article abstract—Ten patients with migraine developed persistent positive visual phenomena lasting months to years. The complaints were similar in their simplicity and involvement of the entire visual field and usually consisted of diffuse small particles such as TV static, snow, lines of ants, dots, and rain. Neurologic and ophthalmologic examinations were normal, and EEGs were normal in eight of eight patients tested. MRI was normal in all patients except one who had nonspecific biparietal white matter lesions and another with a small venous angioma. Treatment of this unusual complication of migraine was unsuccessful.

NEUROLOGY 1995;45:664-668

A variety of positive and negative phenomena characterize the visual aura of migraine. These include visual hallucinations such as stars, sparks, flashes, or simple geometric forms[1]; enlarging scintillating scotomas and fortification spectra; and transient amaurosis, hemianopias, or cortical blindness.[1] More unusual and complex visual distortions include metamorphopsia, palinopsia, or alterations in body perception ("Alice in Wonderland syndrome").[2,3]

The International Headache Society defines a typical aura as one that develops over more than 4 minutes, lasts less than 1 hour, and precedes, accompanies, or follows the headache.[4] A "prolonged aura" lasts more than 60 minutes but less than or up to 7 days.[4] When they are of longer duration, or when an infarction is demonstrated radiographically, a diagnosis of "migrainous infarction" is satisfied.[4] Here we report a group of patients with migraine and persistent positive visual phenomena without evidence of infarction.

Case reports. The patients reported herein represent a collection of individuals seen by the authors with migraine, persistent positive visual phenomena, normal ophthalmic and neurologic examinations, and absence of major psychiatric disease. All 10 patients described below have normal visual acuity, color vision, kinetic or threshold perimetry, and direct and indirect ophthalmoscopy. Pertinent clinical, brain MRI, and EEG data are summarized in table 1. The patients are grouped according to the relationship between the persistent positive visual phenomena and their migraine.

Group I. *Patients 1 to 3: Persistent positive visual phenomena definitely secondary to migraine.* These patients had visual aura with headache, followed by persistent visual phenomena.

Patient 1 *(Cleveland Clinic Foundation [CCF]).* A 32-year-old woman had a long personal history of migraine headaches accompanied by typical auras of photopsias. Several family members also had migraine. In 1990, with each migraine some of the photopsias began to persist and accumulate, eventually encompassing her entire visual field all of the time. They have lasted for 4 years, and she describes the photopsias as floating in space, with a flickering quality. Occasionally she experiences purple-and-gold or rust-colored waves with her eyes closed. Headaches without aura responded to verapamil, low-dose aspirin, fluoxetine, baclofen, and buspirone, but these had no effect on the persistent positive visual phenomena.

MRI, EEG, transcranial Doppler, carotid ultrasound, electroretinogram (ERG), and single-photon emission computed tomography (SPECT) were all normal. Estradiol, antinuclear antibody, complement, rheumatoid factor, vitamin B_{12}, folate, anti-cardiolipin antibody, and lupus anticoagulant levels were also all normal.

Patient 2 *(Hospital of the University of Pennsylvania [HUP]).* At age 16, this man had wrestling-related head and neck trauma followed by a "bright flash" with subsequent bilateral leg weakness and transient headache. He also had two seizures treated with phenytoin for 1 year.

At age 26, he saw "a spiral, with spokes" for 20 minutes followed by a headache. The next day he experienced lights throughout his visual field accompanied by

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Received May 27, 1994. Accepted in final form September 14, 1994.

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Table 1. Clinical and laboratory data of patients described in this report with persistent positive visual phenomena (PPVP) and migraine

Pt no.	Age/Sex	Duration of previous headache history before onset of PPVP	Headache with aura	Duration of PPVP	Brain MRI	EEG
		Group I: PPVP definitely related to migraine				
1	32/F	"Years"	+	4 yr	Normal	Normal
2	26/M	1 d	+	>2 mo	Normal	Normal
3	29/F	Same day	+	5 mo	Normal	Normal
		Group II: PPVP probably related to migraine				
4	9/F	4 yr	+	>2 mo	Normal	Normal
5	67/F	4 mo	+	4 mo	Small right parietal white matter lesion, most likely a venous angioma	Normal
		Group III: PPVP possibly a migraine equivalent				
6	37/M	1.5 yr	–	6 mo	Normal	Normal
7	20/F	15 yr	+	2 yr	Normal	Normal
8	30/M	2 yr	+	8 mo	Normal	Not done
9	23/F	1.5 yr	–	3.5 yr	Small biparietal-occipital white matter lesions	Normal
10	36/M	None	–	5 yr	Normal	Not done

Patients are grouped based on the relationship of PPVP to migraine (see text for details).

daily headache. Treatment with amitriptyline resolved the headaches, although the visual phenomena persisted; carbamazepine had no effect. ERG and CSF were normal. SPECT following administration of 4.9 mCi of I-123 spectamine suggested decreased left temporal lobe activity.

Patient 3 *(HUP).* In mid-1991, this 29-year-old woman twice saw a jagged line with the left eye followed by transient monocular loss of vision and fatigue. In November 1991 she experienced an episode of tingling of her left hand which then involved the tongue and gums but was unaccompanied by headache. Her mother had had migraines related to pregnancy, and the patient was diagnosed with a migraine equivalent.

In early November 1992, she awoke with right arm numbness associated with stars filling the visual field of the right eye. This resolved spontaneously, but on November 10, 1992, she experienced sudden disorientation followed by stars filling the visual field of both eyes, followed by a diffuse, nonpulsating headache. When the headache ceased, she developed persistent constant flashing lights and circles which were worse at night and present with eyelids opened or closed. CSF was normal, but IgM anti-cardiolipin antibodies (15.0 U; normal, 0.0 to 9.0 U) were present. SPECT following administration of 22 mCi Tc-99m–labeled HMPAO showed asymmetric activity in the visual association areas, right greater than left. Phenytoin, carbamazepine, verapamil, and nifedipine had little effect on the visual symptoms, which resolved spontaneously in April 1993. Repeat SPECT demonstrated decreased activity in the left thalamus, left mesial temporal lobe, and left posterior cingulate gyrus.

She was free of persistent positive visual phenomena for 5 months until November 1993, when she experienced a 10-minute episode of "zig-zag" lines for 10 minutes followed by a pulsating headache for 1 day. In February 1994 she had two transient episodes of disorientation and left arm weakness and tingling, lasting 5 minutes. One was followed by a pulsating bitemporal headache lasting 24 hours. Carotid ultrasound, transcranial Doppler, and transesophageal echocardiogram were all unremarkable.

Group II. *Patients 4 and 5: Persistent positive visual phenomena probably secondary to migraine.* These patients had a history of migraine with aura. The temporal relationship between the onset of the persistent positive visual phenomena and migraine events is not as clear as that in group I. However, headaches occurred during the persistent visual phenomena, suggesting some association.

Patient 4 *(Children's Hospital of Philadelphia).* A 9-year-old girl first complained of transient visual loss followed by somnolence at age 2 years. At age 5 she developed a headache preceded by "cloudy vision." She had a third episode at age 7 when she saw colored "blobs" of red and green. The paternal uncle and grandmother had migraine.

At age 9, for 2 months she complained that she saw constant blobs of white and gray, squiggles, clouds, comets, bubbles, "lines of ants," and "a million dots." Headaches occurred every other day, but they had no effect on the visual images, which were persistent and worse at night. Occasionally, she saw blue squares and "people's heads," and once her mother appeared to be far away although she was close by. Phenobarbital and amitriptyline decreased the frequency of headaches and reduced but did not resolve the visual images.

Patient 5 *(Bascom Palmer Eye Institute [BPEI]).* A 67-year-old woman with a 15-year history of migraine with fortification spectra complained of 4 months of constant

"black cracks and lines" in her entire visual field as well as colored dots of light which "bounced like neon lights" with eyelids open or closed. Erythrocyte sedimentation rate, ERG, and visual evoked responses were normal. MRI revealed an abnormal signal in the right posterior parietal white matter consistent with a small venous angioma. SPECT following 9.5 mCi of Tc-99m–labeled HMPAO suggested bilateral decreased parietal activity. Although amitriptyline and ibuprofen relieved her usual headaches, the visual phenomena persisted, and she was reluctant to try other medications.

Group III. *Patients 6 to 10: Persistent positive visual phenomena possibly a migraine equivalent.* These patients had suggestive migraine histories, but the persistent visual phenomena developed without any associated migraine events.

Patient 6 *(BPEI).* This 37-year-old man experienced periodic bilateral retrobulbar headaches for 1½ years, occasionally accompanied by nausea and vomiting. By decreasing his consumption of chocolate, milk, and sweets, he diminished the frequency of the episodes, and aspirin reduced their intensity.

He developed blurry and "gray" vision in the left lower quadrant of both eyes lasting several weeks. For the next 6 months he had persistent television-set "snow" and grainy vision throughout all fields. Later, he noticed an "energized sparkle to the snow" and the sensation worsened in dim illumination. He perceived objects as if he were closely analyzing a newspaper photograph. Aspirin and amitriptyline were unhelpful.

Patient 7 *(BPEI).* A 20-year-old woman had a 15-year history of headache triggered by orange juice combined with chocolate and preceded by a 20-minute visual prodrome where objects became "fragmented" and "cubed." She also experienced mild tension-type headaches that increased in severity during menses.

She was involved in a motor vehicle accident, during which she struck another car that crossed in front of her. There was no loss of consciousness or head trauma, but afterwards she saw constant white and black dots, "snow," and "TV static" over her entire visual field that worsened when she stared at a white background. Later, persistence of visual images (palinopsia) developed. Reading, watching TV, and driving were possible but could be done only with difficulty. SPECT demonstrated biparietal hypoperfusion. Nortriptyline and carbamazepine resolved only the palinopsia, and the other visual phenomena have persisted over 2 years.

Patient 8 *(BPEI).* A 30-year-old man had a 2-year history of classic migraine characterized by left homonymous streaks of light lasting 30 minutes followed by a pulsating headache associated with nausea and emesis. The episodes resolved with propranolol. His history was significant for marijuana use from ages 18 to 22, and his mother and maternal grandmother had migraine headaches.

He complained of 8 months of "snow" and "flickering" similar to what was "between TV channels." Initially experienced only in dark illumination, eventually the visual phenomena became constant, without headaches. SPECT scan revealed bilateral parieto-occipital hypoperfusion. An EEG was not performed. Nifedipine was unhelpful, but sertraline reduced the visual phenomena by 50%.

Patient 9 *(BPEI and CCF).* Five years prior to evaluation, a 23-year-old woman had a pulsating, throbbing headache associated with nausea, vomiting, and a stiff neck after eating Chinese food. Her medical history was significant for ice cream headaches, mitral valve prolapse, and a "high" antinuclear antibody. The patient's mother and paternal uncle had migraine.

Three-and-a-half years prior to evaluation, the patient developed a constant "rain-like" pattern in front of both eyes, at times appearing like a carpet background, often associated with the illusion of motion. Despite atenolol, the visual patterns intensified and converted to persistent "heat waves" with flickering lights. Although extremely uncomfortable, they did not interfere with visual function. She also reported five episodes of visual "blackout" and bright "daggers and spots" lasting 40 seconds each.

ERG and SPECT were normal, but MRI revealed nonspecific small biparietal-occipital white matter lesions. Verapamil, clonazepam, and aspirin had only modest effect.

Patient 10 *(Scheie Eye Institute).* A 36-year-old man complained of a "bright white snowflake spinning in front of [his] eyes." The image appeared bright and was present with eyes opened or closed and lasted 1 week before disappearing spontaneously. Carotid noninvasive studies and echocardiogram were normal. On three occasions during a 5-year period he had three episodes of "classic" scintillating scotoma and fortification spectra thought to be migrainous although no history of headache or migraine was present.

Subsequently, while viewing brightly lit objects (especially computer or television screens), he noticed constant flickering "like a fluorescent bulb that is about to go out" at the edges of the visual field of both eyes. This persisted for 5 years. Bright objects in the temporal field (left eye > right eye) persisted as bright lines for up to 15 seconds when he looked away, and after looking at a brightly lit wall and moving his head or eyes quickly, the temporal portion of his visual field "glowed with a rim of brightness." If he changed from a light to dark environment, a small ring or "brightness wave" appeared in his temporal field and lasted for several seconds. EEG was not performed.

Discussion. Our 10 patients' persistent positive visual phenomena were extremely similar in their simplicity, quality (table 2), and involvement of the entire visual field. They commonly described diffuse small particles, such as TV static, snow, lines of ants, dots, and rain, that lasted for months to years. Some patients reported greater awareness of the visual phenomena when looking at the sky or at a light-colored wall. Complex phenomena such as palinopsia, micropsia, and formed hallucinations were exceptional. Some characterized these unformed visual hallucinations as bothersome, uncomfortable, or emotionally disabling, but not as interfering with visual function. Other patients were unconcerned by them. For the most part, medications such as tricyclic agents, calcium channel and beta blockers, and analgesics were unhelpful.

The patients' ages varied from 9 to 67 years; six were women, four were men, and five had a family history of migraine. Seven of our patients had typi-

Table 2. Persistent positive visual phenomena described by our patients

Simple, unformed
A million dots
Black cracks and lines
Blobs of white and gray
Blue squares
Bubbles
Carpet background
Circles
Clouds
Comets
Dots
Black and white
Colored
Grainy vision
Heat waves
Lights
Flashing
Flickering
Lines of ants
Photopsias
Rain-like pattern
Snow
Squiggles
TV static
Complex
Micropsia
Palinopsia
People's heads

cal migraine with aura (range: 0 days to 15 years) before the constant phenomena developed. Patient 1's visual aura (photopsias) persisted, but patients 2 to 5, 7, and 8 had persistent visual complaints that were different from their prior auras. Patients 6, 9, and 10 did not have previous migraine with aura. Visual symptoms and headaches did not crescendo prior to the onset of the persistent phenomena, and no unifying prodromal event was apparent.

We are unaware of previous reports of full-field persistent positive visual phenomena, although three patients with hemifield symptoms have been described. Haas[5] reported two individuals with "prolonged migraine aura status"; the first was a 70-year-old man with a long-standing history of migraine headaches who developed a constant "pinwheel of bright whirling color, mainly yellow and red" in the left homonymous hemifield accompanied by left hand paresthesias and clumsiness. The episode lasted 5 weeks and resolved with aspirin and cyproheptadine. The second patient was an 18-year-old man with a history of migraine with aura who experienced 7 months of "concentric gray circles like ripples in a pond" and "clustered sets of concentric circles in the right visual field," unaccompanied by headache. Both patients had normal neurologic and ophthalmologic examinations, but only the first had CT of the brain and EEG, both of which were normal. Luda et al[6] reported a 65-year-old woman with "sustained visual aura" who had a 50-year history of migraine with aura and who then developed 12 months of "scintillating geometric figures (in the shape of either rings or chains)" in the right visual hemifield. Carbamazepine, diazepam, flunarizine, nimodipine, and citicoline were unhelpful. Examination, EEG, and brain MRI were normal, but SPECT demonstrated relative hypoperfusion of the left parieto-occipital and frontal areas.

Lessell[7] defines visual hallucinations as a "symptom in which the patient claims to see something or behaves as if he sees something that the observer cannot see." The usual causes of visual hallucinations were absent in our patients. There was no evidence of digoxin use[8] or alcohol intoxication or withdrawal, and none reported abuse of drugs such as cocaine,[9] LSD, marijuana (except a distant history in patient 8), or mescaline,[7] but we did not perform toxicology screens. None of our patients had an established psychotic psychiatric disorder, and usually in this setting visual hallucinations are more complex with occasional auditory components.[7] Positive visual phenomena may occur in patients with visual loss due to disruption at any level in the afferent visual pathway,[10,11] but our patients had normal ophthalmic examinations including visual fields.[12] Neuroimaging disclosed no occipital[13] or midbrain lesions.[14] The normal EEGs failed to confirm epileptogenic foci, and several patients received anticonvulsant therapy without response. Examinations adequately excluded ocular or retinal causes, the normal ERGs (in patients 1, 2, 5, and 9) made photoreceptor dysfunction unlikely, and collective awareness of their own retinal circulation (entoptic phenomena) seems improbable.[7] The qualitative similarity of the patients' descriptions made functional disorders unlikely.

West[15] theorized that in the normal waking state, the constant bombardment of external and internal stimuli inhibits the emergence of "previous perceptions." However, "impairment of information (sensory) input then permits the emergence or release of previously recorded percepts which can be woven into hallucinations." Cogan[16] similarly distinguished continuous "release" visual hallucinations from irritative (convulsive) ones that were episodic. He presented patients with various ocular, optic nerve, chiasmal, optic tract, and occipital lesions who developed formed and complex visual sensations, and Cogan, like West, concluded "a major factor in releasing the hallucinations is loss of the normally inhibiting visual control through blindness, hemianopia, or loss of cognitive functions."

On MRI, patient 5 had a small venous angioma, and patient 9 had small posterior white matter lesions. However, in the setting of normal visual examinations, in both instances it would be difficult to relate these abnormalities to the persistent visual complaints. Nonspecific white matter signal abnormalities have been noted in patients with migraine with aura, but their significance is uncertain.[17]

Traditional theories proposed vasoconstriction-induced cortical ischemia caused migraine aura, and recent experiments measuring regional cerebral blood flow have confirmed decreased regional cerebral blood flow during the aura phase.[18] More recent theories, however, attribute the hypoperfusion to the decreased metabolic demand of cortical neurons resulting from uncharacterized neuronal and biochemical triggers.[19] Four of our patients (numbers 3, 5, 7, and 8) and the patient of Luda et al[6] demonstrated parietal-occipital hypoperfusion on SPECT, and one other had a temporal lobe defect (number 2). However, the inconsistent localization of the SPECT abnormalities and the subjectivity of SPECT interpretations without normative data did not allow us to draw any definite conclusions from the studies in our patients.

Although speculative, our patients' positive visual phenomena may have resulted from spontaneous cortical discharges. Experimental isolation of cortex may result in spontaneous neuronal activity.[7] Thus, selective dysfunction of inhibitory and modulating neurons in extrastriate areas may have allowed spontaneous discharges from the lateral geniculate or visual cortex, resulting in the release visual hallucinations as suggested by West[15] and Cogan.[16]

Acknowledgments

The authors would like to thank Drs. Joan Mollman and Lynn Konchanin for their referrals and Dr. Robert B. Daroff for bringing patient 1 to our attention.

References

1. Sacks O. Migraine aura and classical migraine. In: Migraine: understanding a common disorder. 2nd ed. Berkeley: University of California Press, 1985:55-110.
2. Hachinski VC, Porchawka J, Steele JC. Visual symptoms in the migraine syndrome. Neurology 1973;23:570-579.
3. Rolak LA. Literary neurologic syndromes: Alice in Wonderland. Arch Neurol 1991;48:649-651.
4. Headache Classification Committee of the International Headache Society. Classification and diagnostic criteria for headache disorders, cranial neuralgias and facial pain. Cephalalgia 1988;8(suppl 7):1-96.
5. Haas DC. Prolonged migraine aura status. Ann Neurol 1982;11:197-199.
6. Luda E, Bo E, Sicuro L, et al. Sustained visual aura: a totally new variation of migraine. Headache 1991;31:582-583.
7. Lessell S. Higher disorders of visual function: positive phenomena. In: Glaser JS, Smith JL, eds. Neuro-ophthalmology, vol. 8. St. Louis: C.V. Mosby, 1975:27-44.
8. Piltz JR, Wertenbaker C, Lance SE, et al. Digoxin toxicity. J Clin Neuro-ophthalmol 1993;13:275-280.
9. Siegel RK. Cocaine hallucinations. Am J Psychiatry 1978;135:309-314.
10. Lepore FE. Spontaneous visual phenomena with visual loss: 104 patients with lesions of retinal and neural afferent pathways. Neurology 1990;40:444-447.
11. Siatkowski RM, Zimmer B, Rosenberg PR. The Charles Bonnet syndrome: visual perceptive dysfunction in sensory deprivation. J Clin Neuro-ophthalmol 1990;10:215-218.
12. Lewis RA, Vijayan N, Watson C, et al. Visual field loss in migraine. Ophthalmology 1989;96:321-326.
13. Kölmel HW. Complex visual hallucinations in the hemianopic field. J Neurol Neurosurg Psychiatry 1985;48:29-38.
14. McKee AC, Levine DN, Kowall NW, Richardson EP. Peduncular hallucinosis associated with isolated infarction of the substantia nigra pars reticulata. Ann Neurol 1990;27:500-504.
15. West LJ. A general theory of hallucinations and dreams. In: West LJ, ed. Hallucinations. New York: Grune & Stratton, 1962:275-291.
16. Cogan DG. Visual hallucinations as release phenomena. Albrecht von Graefes Arch Klin Ophthalmol 1973;188:139-150.
17. Fazekas F, Koch M, Schmidt R, et al. The prevalence of cerebral damage varies with migraine type: a MRI study. Headache 1992;32:287-291.
18. Olesen J, Friberg L, Olsen TS, et al. Timing and topography of cerebral blood flow, aura, and headache during migraine attacks. Ann Neurol 1990;28:791-798.
19. Kaufman DM, Solomon S. Migraine visual auras: a medical update for the psychiatrist. Gen Hosp Psychiatry 1992;14:162-170.