It's snowing! Seven cases of persistent migrainous visual aura

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INTRODUCTION:

☐The visual disturbance symptom described as "visual snow" has been present in the literature since 1982 (10). In the past 25 years, other reports have been published using different names, such as prolonged migraine aura status, sustained visual aura, persistent positive visual phenomena, persistent migrainous visual phenomena, and persistent aura without infarction (1-10) to describe the same entity. The second classification of the International Headache Society named it persistent migrainous visual aura (PMVA) and describes it as persistent visual phenomena lasting more than seven days without evidence of infarction. The time frame differentiates persistent migrainous visual aura from prolonged auras, the latter characterized by visual symptoms not exceeding seven days. Complications such as infarcts are usually seen in migraines, but not in PMVA. In general, patients suffering from this problem complain of positive visual symptoms varying from simple, unformed disturbances to complex ones. The classical complaint is seeing "snow flakes" or "TV static", but other manifestations were described. The pathophysiology is unknown, although it is thought to involve neurovascular changes.

CASE REPORTS:

Patient #1:
24 yo woman
1 ½ yrs "TV static", sudden onset & constant.
"Heat waves" & tiny flashes of light.
Most visible against plain background (wall or sky).
Associated palinopsia, blurry vision OU, & photophobia.
Chloroquine recently started for malaria prevention.
New onset migraine headaches one month prior to exam.
PMH negative.
Neuro-ophthalmologic examination completely normal.
HVF, MRI/MRA of brain, EEG, VEP, & ERG were normal.
ANA positive, no other abnormal rheumatologic investigation.
Sx persisted after d/c of chloroquine and BCP.
Sumatriptan alleviated the h/a, but no change in the visual complaints.

Patient #2:
62 yo woman
7-8 yrs visual complaints.
Blue and red "Chinese characters" involving entire VF & positive white scotoma.
Sxs resolved for a month, then one recurrent similar episode.
Sx-free many yrs until 6 months prior to her visit.
Sx infrequent initially but became daily.
Intermittent "TV static" in a very specific horizontal pattern.
FH positive for migraine h/a in mother, brother & one daughter.
Neuro-ophthalmologic examination normal.
MRI of brain normal; carotid doppler showed 60-79% stenosis bilaterally.
Little improvement on trial of Vytorin.

Patient #3:
21 yo woman
Abrupt onset of "moving, flickering or jumping" of images in VF OU.
"TV snow" in front of all images with eyes open or closed.
Problems started 2 weeks after initiating Prozac.
No benefit with d/c of all OTC medicines, caffeine & Prozac.
Reported occasional head discomfort.
H/o panic attacks and anxiety.
Neuro-ophthalmologic examination normal.
Single WM periventricular hyperintensity over R parietal lobe, unenhancing, on MRI of brain. MRI of orbits, TSH, B12, ANA, ESR were normal.

Patient #4:
47 yo man
2 yrs continuous "static" or "snow", entire VF OU.
Pinpoints of light flickering on and off, progressively worse.
Lightening bolts in periphery of VF OU, come and go.
H/o migraine headaches w/o visual aura, last h/a 20 yrs previously.
12 yr h/o photo- & phonophobia w/balance problems & vertigo.
Neuro-ophthalmologic examination normal.
MRI/MRA of brain, MRA of neck, VEP, EEG, TEE, HVF, B12, CRP, RPR, TSH ANA, RF all normal or negative.
No relief with trial of Medrol, topiramate, & verapamil, but photopsias stabilized on latter.

Patient #5:
26 yo man
Abrupt onset "snowy vision" @ age 19.
Constant "static-like" "snow," very fine in size, variable in intensity.
Tinnitus, centrally inside head rather than in ears.
H/o head injury age 13 yrs w/ one isolated seizure.
H/o anxiety and depression; no migraines.
Neuro-ophthalmologic examination normal.
MRI of brain normal.
Clonazepam & Lexapro for anxiety and depression; no change in his visual sx.

Patient #6:
24 yo man
2 yr h/o progressive, bilateral & unremitting "visual snow".
20-30 min of migrating, scintillating scotoma starting in the R lower periphery expanding to involve his central vision.
Alcohol exacerbates symptoms.
Neuro-ophthalmologic examination normal.
MRI of brain, EEG, carotid Doppler, VEP, ERG, lyme titers were normal.
Trial of gabapentin, acetazolamide, topiramate, valproic acid and clonazepam. Clonazepam reduced intensity of sx, but he d/c'd it after experiencing drowsiness.

Patient #7:
22 yo man
6 yr h/o "sparkles" in VFs, present 24/7.
Sx accompanied by severe h/a, nausea, vomiting, neck pain, photophobia, phonophobia.
Neuro-ophthalmologic examination normal.
MRI of brain normal.
Pamelor, valproic acid, lorazepam, baclofen & steroids provided partial relief.
Propranolol & sumatriptan did not help.

SUPPORTED IN PART BY RESEARCH TO PREVENT BLINDNESS

DISCUSSION:

Persistent migrainous visual aura (PMVA) is characterized by the presence of positive visual phenomena described as lines, dots, flashes of light, TV static and snowflakes. Occasionally, complex patterns, such as micropsia or palinopsia, are seen. Symptoms last longer than seven days. Our patients have a timeframe variation from 11 months to 8 years. Jager et al. (8) reported a case with more than 28 years of symptomatology. The pattern of visual field involvement is also heterogeneous. Initial descriptions localized visual symptoms to hemifields, but subsequent reports involve the entire field. Our patients all fit this pattern, although some of them developed both isolated and diffuse symptoms. Most of the patients have a history of migraine headaches, although it is not always present. Amongst our patients, only two have a consistent history of headaches. Interestingly, patients with migraine who developed PMVA do not necessarily have a history of aura despite longstanding migraine headaches. Other patients developed the visual symptoms with new onset headaches. Family history of headaches is also reported. Hann et al. (5) described two first-degree related patients with PMVA.

There are no abnormalities in the neuro-ophthalmological exam in the patients with PMVA. Investigations including MRI/MRA of head and neck, MRI of orbits, EEG, ERG and VEP are usually normal or unremarkable. Two cases described by Haan et al. (5) demonstrated slow wave abnormalities on EEG in the occipital area of unclear significance. Slow waves are commonly seen in patients with migraine headaches. Two studies in 2004 report functional imaging in PMVA. Relja et al. (9) described the first case studied with both brain single photon emission computed tomography (SPECT) and perfusion MRI. In this case the studies performed one month following onset of symptoms reflected decreased blood perfusion over the left fronto-parietal-occipital and right occipital areas on SPECT while perfusion MRI showed decreased perfusion over the left hemisphere compared to the right. Seven months later with improved symptoms, repeat perfusion MRI showed normalization of the area of hypoperfusion. Jager et al. (8) reported four cases of PMVA studied with diffusion and perfusion-weighted MRI and failed to show significant difference in perfusion or diffusion properties within the cortex of the patients studied.

The pathophysiology of the phenomenon is unknown. It is thought that it involves a mechanism similar to the cortical spreading depression of Leao and changes in cerebral blood flow without evidence of ischemia on diffusion images. Spreading depression is a neuronal depolarization wave with subsequent suppression of electrical activity across the cortex after a mechanical or chemical perturbation. No triggering event for the development of PMVA was identified. Some papers suggested a relationship between lysergic acid diethylamide (LSD) use and the persistence of visual abnormalities for several years (4). This abnormality was called hallucinogen persisting perception disorder (HPPD). Two of our patients developed symptoms after initiation of new medications. However, there was no improvement after their discontinuation. Therefore, a clear relationship cannot be established.

The treatment of PMVA is not well established. It is known that the symptoms are refractory to several medications used to treat or prevent migraine. Anticonvulsants, antidepressants, calcium channel blockers, diuretics, benzodiazepines have been tried. The patients responded poorly or not at all to the trials. There are some reports of patients successfully treated with furosemide, acetazolamide, valproic acid and lamotrigine (3,5,6,7). However, the general experience, as in case of our patients, has been disappointing.

CONCLUSIONS:

1-Persistent migrainous visual aura (PMVA) is characterized by a constellation of positive visual symptoms.
2-Migrainous changes in neuronal and vascular function are proposed as a mechanism of the disease.
3-Tests of brain imaging and retinal function are consistently normal.
4-The treatment with several anti-convulsants, diuretics, calcium channel blockers and steroids has been disappointing.
5-The patients needed several visits to neurologists and ophthalmologists without a clear diagnosis established.
6-The fact that seven patients were seen in a short time raises the possibility that it can be more prevalent than it is thought.

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