Case Report: Visual Snow Syndrome after Repetitive Mild Traumatic Brain Injury

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SIGNIFICANCE: Visual snow syndrome is a recently recognized condition with its own diagnostic criteria, evolving pathophysiologic research, and potential treatment options.

PURPOSE: This report documents a rare but likely underdiagnosed condition called visual snow syndrome. A review of the current literature on pathophysiology and treatments is discussed.

CASE REPORT: A 40-year-old Whiteman started experiencing symptoms of constant pulsating pixels throughout his entire visual field approximately 3 weeks after a series of mild concussions. In addition, he experienced a persistence of images and photosensitivity. The patient had normal eye examination results, visual fields, and retinal imaging result. Brain MRI, magnetic resonance angiography, electroencephalography, and cerebrospinal fluid analysis were unremarkable. A positron emission tomography scan demonstrated hypometabolism in the posterior parietal lobes and left posterior cingulate gyrus. Pharmacological treatment with antiepileptic and migraine medications was unsuccessful. Tinted lenses were essentially ineffective with a 10% reduction in symptoms reported with the use of a custom blue-tinted lens.Vision rehabilitation aids with optical character recognition were used for prolonged reading needs.

CONCLUSIONS: Although rare, visual snow syndrome should be considered in all patients reporting continuous pixelations in their vision for more than 3 months, especially when accompanied by at least two of the following: photosensitivity, palinopsia, enhanced entopic phenomena, or nyctalopia. The pathophysiology is still unclear at this point, with evidence suggesting a link to the secondary visual cortex, specifically the lingual gyrus. More studies are needed to determine the exact cause, especially studies that separate visual snow syndrome patients with and without comorbid migraine. Because the pathophysiology is unclear, the treatment course is also unclear. Anecdotal evidence may suggest that tinted lenses may be of some value.

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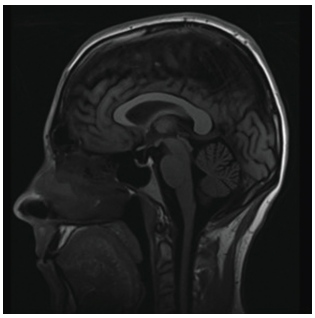

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Visual snow syndrome is a disorder primarily characterized by continuous dots in the entire visual field, often described like TV static. To meet this diagnosis, patients must also report two of the following symptoms: palinopsia, enhanced entopic phenomena, photophobia, or nyctalopia. The pathophysiology of the disorder is not well understood, although there is some preliminary evidence suggesting a link to the secondary visual cortex. The disorder has been reported to arise with and without an inciting incident such as traumatic brain injury as reported in this case. Currently, both pharmacological and optical treatments have limited success in reducing the symptoms of visual snow syndrome.

CASE REPORT

A 40-year-old White man presented to the clinic for a comprehensive eye examination. He was having difficulty reading because of chronic, constant, pulsating pixels, like TV static, throughout his vision. He reported that the pixels would occur with both of his eyes open and closed. His symptoms began approximately 3 weeks after a series of about 50 blows to the head (no loss of consciousness) in 2007. This was reflected in his medical record. He also reported photosensitivity and episodes of palinopsia (an afterimage of an object just sighted or a trailing of that object), which was most prevalent after viewing a backlit screen. His ocular health history was unremarkable. His medical history was positive for anxiety, post-traumatic stress disorder, tinnitus, and a history of testosterone deficiency. He denied any history of migraine or migraine with aura. His medication and social history included herbal supplements, intermittent "detox" and paleo diets, and marijuana with no additional illicit drugs. He had a history of taking Clomid (Patheon Pharmaceuticals Inc., Waltham, MA) intermittently from January 2010 to February 2011 for his testosterone deficiency (Fig. 1).

His best-corrected visual acuity was 20/20 in each eye at distance and near. His binocular evaluation revealed a small exophoria at distance and near with excellent compensating positive fusional vergence ranges. The results of entrance testing including pupils, extraocular motilities, Amsler grid, and monocular color vision were all normal. His ocular health examination was unremarkable, including macula and retinal nerve fiber layer optical coherence tomography, Humphrey visual field 24-2 and 10-2.

Neurologic clinical examinations and routine blood testing yielded normal results. Digoxin laboratory results were also normal. His most recent brain MRI from 2018 was unremarkable, as well as his magnetic resonance angiography from 2012. His electroencephalogram from 2018 was unremarkable. His 2018 cerebral spinal fluid analysis of a-beta, p-tau, and total tau was unremarkable, showing no evidence of chronic traumatic encephalopathy. He had a fluorodeoxyglucose–positron emission tomography scan in 2019 that revealed mild

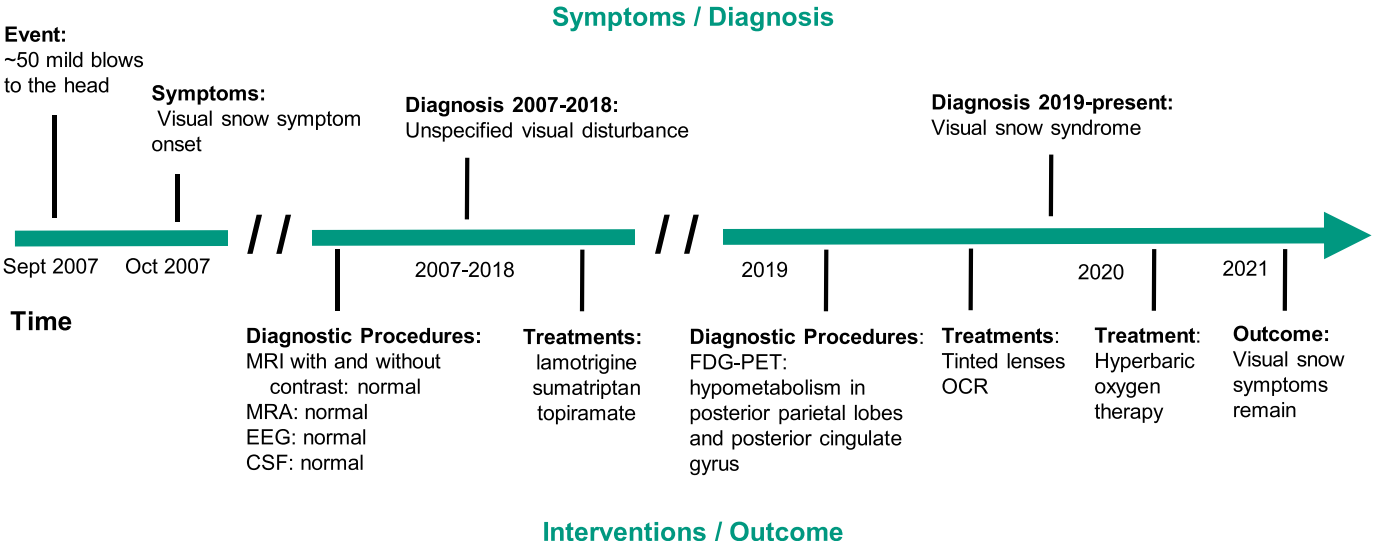

FIGURE 1. Clinical timeline: 40-year-old Whiteman with visual snow syndrome. CSF = cerebral spinal fluid; EEG = electroencephalogram; FDG-PET = fluorodeoxyglucose–positron emission tomography; MRA = magnetic resonance angiography; OCR = optical character recognition.

hypometabolism in the posterior portion of both parietal lobes and in the left posterior cingulate gyrus.

Pharmacologically, he has trialed lamotrigine (generic), sumatriptan (generic), and topiramate (generic) in the past but did not find any remission of symptoms and could not tolerate the adverse effects. He is no longer interested in pharmacological treatment for his visual symptoms. Experimentally, the patient trialed hyperbaric oxygen chamber therapy six times a week for 1 month. He felt his overall well-being was improved after a temporary worsening of his post-traumatic stress disorder symptoms, but the visual snow symptoms were unchanged.

Ophthalmologically, an extensive medical filter evaluation of varying compositions was trialed to help alleviate his visual symptoms of visual snow and photosensitivity. He preferred a 10% transmission gray tint in a wraparound frame for photosensitivity in bright light. He preferred a 20% transmission blue tint for daily distance glasses and a 60% transmission blue tint for his reading glasses. The medical filters were able to alleviate his photosensitivity. Regarding the visual snow, he reported a reduction of visual snow symptoms by only 10% with the blue medical filters. Of the >100 filters trialed, he notably had a strong negative response to all tints in the yellow spectrum. In addition, he trialed binasal occlusion without any relief of symptoms. In addition to the tints, the patient was also managed with an optical character recognition device as a reading aid and a smartpen, which can record notes, as a writing/reading aid.

The possible etiologies for his visual disturbance included Clomid use, a neurologic structural lesion, epilepsy, persistent aura without infarction, and visual snow syndrome. Clomid can cause visual disturbances including palinopsia, auras, shimmering of vision, and photophobia. It is unlikely that Clomid is the culprit because he was started on these medications in 2010, but his records showed he began complaining of symptoms in 2007. He did not have a neurologic structural lesion or epilepsy because his MRI, magnetic resonance angiography, and encephalography were unremarkable. Persistent aura without infarction is a type of migraine. This occurs in patients who have previously had a migraine with aura and does not classically have any additional symptoms such as palinopsia, continuous light sensitivity, or nyctalopia. The symptoms the patient was experiencing support the diagnosis of visual snow syndrome.

DISCUSSION

The first description of visual snow syndrome was reported by Liu et al.[1] in 1995. The diagnostic criteria for visual snow syndrome were outlined by Schankin et al.[2] in 2014 as "dynamic, continuous, tiny dots in the entire visual field lasting longer than 3 months" along with at least two of the following: palinopsia, enhanced entopic phenomena, photophobia, or nyctalopia. The visual snow phenomenon has many descriptions including "TV static," "bubbles," "rain-like patterns," "clouds," "squiggles," and "carpet background."[3] The dots are most commonly black and white or transparent; they have also been described as colored.[4] The dots are more noticeable on a blank background or while reading.[3] Enhanced entopic phenomena can include excessive floaters, excessive blue field entopic phenomenon, self-light of the eye, or spontaneous photopsia. Nyctalopia is impaired night vision.

The diagnosis of visual snow syndrome requires that the symptoms not be consistent with migraine aura or better explained by any other disorder.[2] It is important to obtain MRI to rule out compressive lesions and encephalography to rule out epilepsy. Visual snow can be a symptom of neurodegenerative disease, such as Creutzfeldt-Jakob disease[5] or glycine receptor antibody syndrome.[6] A thorough eye examination must be performed to rule out retinal pathology that could attribute some of the symptoms described previously. It is possible to experience visual snow phenomenon without having visual snow syndrome.[7]

The prevalence of visual snow syndrome is rare. An online self-report study in the United Kingdom found "visual snow" (visual snow without two additional symptoms) to have a prevalence of 3.7% and visual snow syndrome to have a prevalence of 2.2%.[7] Visual snow has been reported in both adults and children.Studies that reviewed age at symptom onset show 17.2% to 45% as having visual snow symptoms for as long as they could remember.[8,9] For

participants who developed visual snow onset later in life, the average age at onset is 18.4 to 24.5 years.[8–10] This disorder affects males and females equally, although females are 30% more likely to have an increased number of symptoms.[4] Visual snow syndrome is often seen with other comorbidities such as migraine (51 to 80%)[8,9,11] and tinnitus (50 to 59.1%)[7–9]; the presence of migraine aura can make this diagnosis even more difficult. These patients are 2 to 2.5 times more likely to have an increased number of symptoms, and those with comorbid migraine are more likely to experience photopsias.[4] Patients with visual snow syndrome also commonly complain of concentration problems, irritability, and lethargy, which may suggest involvement of the limbic system.[9]

One study evaluated photosensitivity of patients with visual snow versus controls and found increased photosensitivity in patients with visual snow syndrome independent of comorbid migraine on the Leiden Visual Sensitivity Scale. Interestingly, the visual snow syndrome scores were similar to values found in patients with chronic migraine with aura during an attack.[12]

Visual snow syndrome can be classified as primary or secondary. Primary visual snow syndrome is when the symptoms arise without an inciting incident. Secondary visual snow syndrome is when the symptoms arise secondary to an event such as traumatic brain injury, hallucinogenic drugs, or prescription drugs.[3] Although reports of visual snow onset after minor head injury are not common, it has been reported. Liu et al.,[1] Metzler and Robertson,[3] and Schankin et al.[2] all discuss cases of visual snow onset immediately after a minor head injury. Bessero and Plant[13] reported a couple of patients with a delayed onset of visual snow occurring one and many weeks after mild head injury. The prevalence or relationship of visual snow syndrome after traumatic brain injury has not been established.

The pathophysiology for visual snow syndrome is unclear at this point, and there are different theories debating where the issue originates. The ocular examination of visual snow syndrome patientsshould be unremarkable for the cause of visual snow and its associated symptoms.Some authors argue for overactive neuronal structures within the primary visual cortex. They evidenced this with an initial case report that demonstrated lack of habituation and even potentiation on visual evoked potentials in a patient with visual snow symptoms and migraine.[14] Concerns were raised as to whether this was from the visual snow syndrome itself or migraine, as lack of habituation and hyperexcitability on electrophysiological testing has already been demonstrated in migraine patients and patients with persistent visual aura.[14] A subsequent study found statistically significant loss of habituation in visual snow syndrome subjects with and without migraine in the right eye, but only the visual snow syndrome with migraine group for the left eye.[10] The occipital lobe was also looked at with phosphene detection using transcranial magnetic stimulation as a measure of occipital lobe excitability.[10] Phosphene thresholds only reached significantly lower thresholds in patients with visual snow syndrome and migraine when compared with healthy controls.[10] It is difficult to determine if these findings are truly from visual snow syndrome or from comorbid migraine. Overall, throughout the currentliterature, there is a paucity of support for the primary visual cortex as the origin of visual snow syndrome.Metzler and Robertson[3] also state that, because there is an even distribution of pixels throughout the visual field, it is unlikely that the issue stems from the primary visual cortex; if this were true, the pixels would likely be denser centrally and spread out peripherally.

Enhanced brain imaging suggests a hyperexcitation of the secondary visual cortices, especially at the lingual gyrus. Fluorodeoxyglucose–positron emission tomography scans have demonstrated hypermetabolism of the lingual gyrus, especially on the right, in patients with visual snow syndrome.[9,11] A visual evoked potential study found increased N145 latency and reduced N75-P100 amplitude in visual snow syndrome patients. The authors link their findings back to hypermetabolism of the lingual gyrus, as the late P100 phase may localize to this area of the brain.[15] Voxel-based morphometry studies demonstrate a cortical gray matter volume increase at the junction of the right lingual and fusiform gyrus, as well as increased gray matter volume in of the right lingual gyrus.[9,16] A study by Puledda et al.[17] looked at the blood oxygenation level–dependent response of visual snow syndrome patients against healthy controls and found an increased response in the area of the right lingual gyrus. Furthermore, magnetic resonance spectroscopy was performed at the level of the lingual gyrus, and findings revealed an increase in metabolic lactate levels ($P < .001$) and a trend for increased excitatory glutamate ($P = .06$) in visual snow syndrome patients.[17] The lingual gyrus has been noted to be involved with visual processing and photosensitivity in migraine sufferers. It seems that the lingual gyrus may be a promising component to visual snow syndrome, although further research, including larger sample sizes with and without migraine subgroups, is needed to confirm this observation.

Other cortical areas/characteristics addressed in research studies include the insular cortices, parietal or parieto-occipital junction, and occipital bending.

The blood oxygenation level–dependent functional MRI study by Puledda et al.[17] also noted reduced activity in the anterior insular cortices in visual snow syndrome patients. This area helps the brain determine which visual information to pay attention to. It can either determine the visual information to be important and disseminate signals along to the other areas of the limbic system or determine that visual information is background noise and should be ignored; these signals are not propagated along.

There are two case reports demonstrating bilateral parietal hypoperfusion on single-photon emission computed tomography imaging, one with bilateral parietal hypoperfusion and one with bilateral parieto-occipital hypoperfusion.[1]

Lastly, left occipital bending has been reported in a case report[14] and a case series in 4 of 14 patients[10] with a history of visual snow. Occipital bending is the protrusion and bending of one of the occipital lobes toward the other across the anteroposterior axis.[14] The significance of the occipital bending in visual snow syndrome is unclear, but it is thought less likely to be a significant factor because occipital bending is present in a large portion of the general population without visual snow symptoms.

There is no proven treatment for visual snow syndrome. Pharmacological interventions include antiepileptic and migraine medications, with the most reported success in lamotrigine. A study by van Dongen et al.[8] looked at the treatment effect of lamotrigine, valproate, topiramate, acetazolamide, and flunarizine on visual snow syndrome. They found that no pharmacological intervention provided a complete remission of symptoms. The most successful intervention was lamotrigine, which showed a partial remission in 19% of patients. A case report by Unal-Cevik and Yildiz[14] showed a potentiation on reversal visual evoked potential on a patient with visual snow syndrome before treatment with lamotrigine and then an improvement on reversal visual evoked potential after treatment. It is unclear if the potentiation was truly from visual snow syndrome or from comorbid migraine. The patient reported palinopsia reduction by 80%; floaters, frequency of dots, and photopsias reduced by 50%; blue field entopic phenomenon and nyctalopia reduced by 70%; and migraines reduced from three to three times per week to two times per month.[14] Fluoxetine

was reported to have no effect, and sertraline was reported to worsen the symptoms in a 2014 study of 120 patients.[11] There have been no randomized clinical trials to prove efficacy for any pharmacological intervention of visual snow syndrome. It is important to keep in mind the side effect profile of antiepileptic and migraine medications such as mental slowing, dizziness, or excessive daytime sleepiness.[8]

Previous studies have correlated a thalamocortical dysrhythmia with tinnitus.[18] The question was raised whether visual snow syndrome could also be caused by a thalamocortical dysrhythmia, but this has not been directly measured to date.[18] The magno/parvocellular pathways connect the thalamus/lateral geniculate nucleus to the primary visual cortex, whereas the koniocellular pathway connects the thalamus/lateral geniculate nucleus to the primary and secondary visual cortices.[18] Blue/yellow filters have been proposed as a potential treatment option because they may alter the balance between the koniocellular and magno/parvocellular pathways. Although the hypothesis is unproven, activating the koniocellular pathway may reduce or eliminate low-frequency brain rhythms that are speculated in this premise to be causing the visual snow symptoms.[18]

Colored lenses have received anecdotal support in a few case reports and a small case series. A case report by Liu et al.[19] reported a subjective improvement of visual snow syndrome symptoms with dark green–colored medical filters. The patient reported an improvement in 7 of her 11 visual snow syndrome–related symptoms. A case series by Ciuffreda et al.[20] reported subjective improvement of visual snow with a pinkish-purple filter and a 70% reduction of palinopsia while reading with a turquoise-green filter; a second patient found reduced visual snow and photosensitivity with a 40% BPI-IR (Brain Power Inc., Miami, FL) blue tint. A study by Lauschke et al.[18] reported a subjective improvement of visual snow syndrome symptoms with colored medical filters. Ninety-two percent (11 of 12) of patients who underwent colorimetry testing reported subjective improvement of symptoms with a colored filter; 82% of those patients chose colors on the yellow-blue spectrum. A large barrier to these studies is the subjective nature used to determine treatment response.

CONCLUSIONS

Visual snow syndrome is a clinical entity with newly defined diagnostic criteria. The criteria are based on subjective complaints from the patient. The pathophysiology is still unclear at this point, with evidence suggesting a link to the secondary visual cortex, specifically the lingual gyrus. More studies are needed to determine the exact cause, especially studies that separate visual snow syndrome patients with and without comorbid migraine. Because the pathophysiology is unclear, the treatment course is also unclear. If we can understand more about visual snow syndrome, we may be able to come up with a treatment plan that provides full remission of symptoms. Randomized controlled clinical trials are needed.

ARTICLE INFORMATION

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