

Visual Snow Syndrome

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INITIAL PRESENTATION

Chief Complaint: Visual snow

History of Present Illness: A 59-year-old pilot with a history of visual aura since his teenage years and anxiety presented to the neuro-ophthalmology clinic with a primary complaint of seeing a "snow storm" for one year. He described constant bilateral "TV static" filling his entire visual field that was worse at night. The symptoms began in association with flickering lights, tinnitus, jumping images, and severe anxiety. The associated symptoms resolved after one month but anxiety exacerbates his visual static.

Past Ocular History: Radial keratotomy, both eyes (OU)

Medical History:

- Visual aura
- Anxiety

Medications:

- Alprazolam 0.5 mg every AM
- Alprazolam 1.0 mg every evening
- Quetiapine 100 mg with dinner
- Sertraline 250 mg qAM

Allergies: None

Family History: Non-contributory

Social History: Former smoker, quit 20 years previously

Review of Systems: Negative beyond HPI

OCULAR EXAMINATION

- Visual Acuity with correction:
 - Right eye (OD): 20/25+2
 - Left eye (OS): 20/20 -2
- Ocular Motility/Alignment:
 - OD: Full, orthotropic
 - OS: Full, orthotropic
- Intraocular Pressure (IOP):
 - OD: 14 mmHg
 - OS: 14 mmHg
- Pupils:
 - OD: 6 mm in dark, 3 mm in light, 3+ light and near reactions, no relative afferent pupillary defect
 - OS: 6 mm in dark, 3 mm in light, 3+ light and near reactions, no relative afferent pupillary defect

- Confrontation visual fields:
 - OD: Full
 - OS: Full
- External:
Normal OU
- Slit lamp examination:
 - Lids/lashes: Mild blepharitis OU
 - Conjunctiva/sclera: Clear and quiet OU
 - Cornea: Well-healed radial keratotomy scars OU
 - Anterior chamber: Deep and quiet OU
 - Iris: Normal architecture OU
 - Lens: Trace nuclear sclerosis and cortical spoking OU
- Dilated fundus examination (DFE):
 - Vitreous: Normal OU
 - Disc: Normal, pink optic nerves OU
 - Cup-to-disc ratio: 0.2 OD, 0.3 OS
 - Macula: Normal OD, few parafoveal drusen OS
 - Vessels: Normal OU
 - Periphery: Normal OU
- Additional Testing:
Goldmann Visual Field demonstrated constriction of the I1e isopter, but was otherwise full.

Optical coherence tomography (OCT) of the optic nerve head and retinal nerve fiber layer (RNFL) performed elsewhere were reviewed. It was normal on the right and showed foveal vitreomacular adhesion and subfoveal drusen on the left.

Magnetic resonance imaging of the brain and orbits with and without contrast was normal.

Differential Diagnosis:

- Visual Snow Syndrome
- Entoptic phenomenon
- Migraine with aura
- Autoimmune retinopathy
- Cancer associated retinopathy
- Posterior vitreous detachment
- [White dot syndrome](#)

CLINICAL COURSE

Given his unremarkable anterior and funduscopic exams, the constancy of the visual phenomena and their diffuse static-like character, his constellation of symptoms were most consistent with visual snow. When shown an online visual snow simulator, he confirmed his symptoms matched the computer simulation.

DIAGNOSIS: Visual Snow Syndrome

DISCUSSION

Introduction

Visual snow is a neurologic condition that manifests with persistent positive visual symptoms consisting of tiny flickering dots covering the entire visual field. The flickering dots resemble 'static' or 'snow' and are interposed between the patient's vision and background [1]. This symptom often persists even with the eyes closed. When accompanied by two of the four

additional visual symptoms: palinopsia (persistence of an image after stimulus), enhanced entoptic phenomena (e.g., floaters), photophobia and nyctalopia (poor night vision), the constellation is known as the visual snow syndrome [2]. Patients can also experience non-visual symptoms, most commonly, bilateral tinnitus [3].

Visual snow (VS) has been given a variety of names, most referring to prolonged or sustained migraine aura dating back to 1982 by Haas [4]. However, Liu, et. al, in 1995 is generally given credit for the initial series of patients with detailed descriptions of the symptoms of visual snow when it was described as persistent migraine aura [5]. Other early studies suggested VS to be a clinical phenomenon secondary to illicit drug use or mental illness, known as hallucinogen persistent perception disorder [3]. Schankin, et al., addressed these common misconceptions in 2014 concluding no associations between illicit drug use and VS. The study also did not find causal relationship between mental illness and VS but did note that anxiety and depression were common long-term comorbidities [3]. In a subsequent study in 2015, Schankin et al., identified visual snow syndrome as a unique clinical entity distinct from migraine aura and proposed diagnostic criteria (see "Diagnosis" section below) [8].

Etiology/Epidemiology

VS tends to occur in younger patients, with an average age of 29 years. There is no sex predilection. Roughly 40% of patients describe having symptoms since childhood, with nearly a quarter reporting sudden onset of symptoms. Comorbid conditions, such as migraine, tinnitus, and migraine aura may also be seen, and are more common among patients with visual snow syndrome compared to visual snow. [9]

Pathophysiology

The neurophysiological mechanism of visual snow is poorly understood and research in this area is in its infancy. However, early neuroimaging studies have helped further our understanding [6]. Two main hypotheses have emerged to explain the pathophysiology of VS based on the clinical description and additional symptoms, although neither has been explicitly cited as the sole explanation.

One hypothesis suggests that VS is caused by dysfunction in the secondary or supplementary visual cortex [1] surrounding the primary cortex. [6] The bilaterality, diffuse involvement of the entire visual fields, and the constancy of symptoms support the concept that the pathology is cortical. The lack of monocular or homonymous visual field defect makes dysfunction in the anterior visual pathway, optic radiations or primary visual cortex unlikely [1]. Instead, the associated symptoms of palinopsia and enhanced entoptic phenomena suggest a problem with visual processing in the supplementary visual cortex. In one study, 17 patients with VS and 17 age- and gender-matched healthy controls underwent a [18F]-2-fluoro-2-deoxy-D-glucose ([18F]-FDG) positron emission tomography (PET). Patients with visual snow had significant hypermetabolism in the area of the right lingual gyrus and the left anterior lobe of the cerebellum [7]. Hyperactivity in the lingual gyrus, the area responsible for modulating visual processing, may explain the positive visual symptoms observed in VS [8]. Similar imaging studies performed for patients with photophobia, common to both visual snow syndrome and migraine, also suggest that the lingual gyrus is involved in the perception of photophobia during migraine [8]. This shared pathophysiology might thus in part explain the comorbidity of migraine and VS [2].

Another proposed hypothesis for the etiology of VS is cortical hyperexcitability, or reduced habituation in the visual cortex, which allows patients to visualize normally subthreshold stimuli [6]. This is supported by enhanced perception of entoptic phenomena in VS, thought to be caused by a disorder of habituation and sensory processing [9]. Studies using electrophysiologic testing have demonstrated cortical hyperexcitability or hyperresponsivity in VS patients compared to control healthy patients. In these studies, when control subjects were presented repetitive stimulation using visual evoked potentials, physiological habituations occurred while visual snow patients had potentiation of the stimuli [10]. Hyperexcitability affecting widespread neuronal networks has been shown to drive thalamocortical dysrhythmia, which has been linked to migraine auras and tinnitus and, thus, may also explain the co-incidence of VS and tinnitus [2].

Symptoms and Signs

Patients with visual snow experience unremitting visual symptoms characterized by numerous flickering dots resembling 'static' or 'snow' covering their entire visual field [3]. Furthermore, patients often have other characteristic symptoms such as after-images or palinopsia, the perseveration of a visual stimulus manifest as either trailing of images or a stationary image superimposed on a new visual scene [3]. Patients may also experience enhanced entoptic phenomena, or see their own normal structures within the eye, These structures include floaters (microscopic collagen fibers in the vitreous) and blue field entoptic phenomena (white blood cells in the retinal blood vessels) [8]. Although these structures may be visualized by healthy patients, patients with visual snow have an accentuated awareness of these symptoms and are often disturbed by their presence [8]. Other common symptoms in VS include photophobia, photopsias, and nyctalopia.

Below is a simulator for this condition which may replicate patients' visual symptoms.

- <http://visionsimulations.com/visual-snow.htm>

Diagnosis

Visual snow syndrome is diagnosed based on clinical signs and symptoms. Diagnostic criteria was proposed by Schankin, et al., in 2015 to help identify patients with this syndrome. The criteria for diagnosis of the syndrome states that patients must have symptoms of visual snow lasting longer than 3 months. They must also have two of the four additional symptoms: palinopsia, enhanced entoptic phenomena, photophobia and nyctalopia. The symptoms must be different from a visual

migraine aura. Lastly, symptoms should not better be explained better by another disorder. Ophthalmologic and neurologic exams performed on VS patients are normal and there are currently no recommendations for pursuing diagnostic imaging for such patients. [8]

Treatment/Management/Guidelines

There is currently no standardized treatment proposed for visual snow syndrome. Migraine treatments are often unhelpful and may worsen symptoms [6]. There have been several treatment trials that had varying successes in individual patients [3]. Several medications, including lamotrigine, sertraline, nortriptyline and carbamazepine, have shown some improvement of visual snow symptoms in individual cases but have not been consistently effective [6]. Further studies are needed to further understand this syndrome and identify treatment options.

Patient Resources

- https://www.youtube.com/watch?time_continue=1&v=O9l0_oJ110M
- <https://www.visualsnowinitiative.org>
- visualsnowsyndrome.com/resources

<p>EPIDEMIOLOGY/PATHOPHYSIOLOGY</p> <ul style="list-style-type: none"> ◦ No gender predilection ◦ Onset typically in childhood ◦ Pathophysiology – hypermetabolism in the supplemental visual cortex proposed (possible, potential) 	<p>SIGNS</p> <ul style="list-style-type: none"> ◦ Normal neurologic and ophthalmologic exams
<p>SYMPTOMS</p> <ul style="list-style-type: none"> ◦ "Static" or "snow" in both visual fields ◦ Palinopsia ◦ Enhanced entoptic phenomena ◦ Nyctalopia ◦ Photophobia ◦ Photopsia ◦ Migraine and tinnitus are common comorbidities 	<p>TREATMENT</p> <ul style="list-style-type: none"> ◦ Reassurance

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