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Case Report

Visual Snow: A Case Series from Israel

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Keywords

Visual snow · Palinopsia · Photophobia · Nyctalopia

Abstract

Our aim was to examine the symptoms and clinical characteristics of visual snow in a group of 6 patients from a Department of Ophthalmology and a Department of Neurology. Visual snow is now recognized as a true physiological disorder. Previously, physicians unaware of this syndrome may have misinterpreted its symptoms as a persistent visual aura. By promoting awareness of this syndrome, greater quantitative and qualitative research may expand our understanding and treatment of this disorder.

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Introduction

During recent years, the condition of visual snow (VS) has accelerated its acceptance as a true syndrome, separate from migraine. These patients experience similar visual symptoms with sensations of seeing “snow,” a persistent positive visual phenomenon comparable to

grainy or pixelated television static [1–5]. This distressing condition impairs daily life, often leading to numerous unnecessary examinations and treatment attempts. Therefore, expanding our knowledge and understanding of this condition is important and beneficial for the patient and caregiver.

Symptoms vary, but can present with persistent, innumerable flickering black and white dots throughout the visual field, though they may occasionally appear as colorful, flashing, or transparent. The intensity may differ in ambient lighting, be more noticeable on plain backgrounds, or more attenuated on textured backgrounds [1–3, 5, 7]. Furthermore, patients may experience additional visual symptoms, such as palinopsia, photophobia, nyctalopia, and enhanced entopic phenomena, which arise from the structure of the visual system itself [1–3].

Case Reports

Case 1

An 18-year-old male presented with a past medical history only notable for asthma, and without a history of headaches or migraine. He complained of a persistent visual disturbance of squiggly worms moving in front of his eyes, without any connection to his head movement. This phenomenon first appeared after drinking alcohol. Prior to his examination, he began to see flashing white small dots and complained of seeing a shadow around moving objects and an orange glowing ball when closing his eyes. He also suffered from tinnitus, more so in the right ear. An electroretinogram showed insignificant changes. He was evaluated by a psychiatrist who diagnosed anxiety related to his symptoms, with no additional findings.

Case 2

A 32-year-old myopic female presented with a past medical history of migraine headaches without aura, familial Mediterranean fever, and polycystic ovary syndrome. Her visual symptoms began during her second trimester of pregnancy. She described the visual disturbance as small circles in front of her eyes lasting seconds, in addition to a constant visual disturbance similar to “snowflakes” in her whole visual field. She also suffered from tinnitus.

Case 3

A 28-year-old male presented with complaints of VS symptoms (VSS) over the past year, since he started using Roaccutane (isotretinoin) for acne treatment. His symptoms consisted of constant flickering white dots encompassing all his visual field, in addition to nyctalopia and a sizeable number of floaters. The patient also suffered from muscle pain, chronic tension-type headaches, concentration difficulties, impotence, and mild depression. Blood tests showed elevated CPK and prolactin, with a low testosterone level. A treatment trial consisting of serotonin-norepinephrine reuptake inhibitors (Duloxetine) and tricyclic antidepressants (Amitriptyline) had failed.

Case 4

A 46-year-old myopic female patient with a past ocular history of keratoconus reported experiencing VSS since childhood, without complaints of headaches or tinnitus. The patient

described the visual disturbances as constant flashing lights, similar to TV static noise, throughout her visual field.

Case 5

A 23-year-old woman, with a past medical history of migraine headaches with aura, experienced since the age of 12, presented with a 4-year history of VS, described as small flashing dots and a “granular” or “hazy” image with a “static”-like background and complaints of missing areas in her visual field. In addition, she complained of photophobia, nyctalopia, and numbness throughout her body. She is currently receiving vitamin B₁₂ injection therapy. Her neurological examination was normal and imaging studies showed only mild sinusitis. The rest of the examination was normal.

Case 6

A 40-year-old male patient presented with a past medical history of depression and chronic treatment with selective serotonin reuptake inhibitors (Vortioxetine). He had no complaints of headaches or tinnitus. Over the past 10 years, the patient suffered from a visual disturbance described as static dots, flickering throughout the visual field, worsening in dim light. Moreover, he described white shadows around moving objects that lasted for a few seconds.

All the patients’ ophthalmic and neuro-ophthalmic examinations were normal. Additional blood tests for all the patients described, including TSH, APLA, and vitamin B₁₂, showed no important findings except for an elevated CPK and prolactin and reduced testosterone in Case 3. All the patients also underwent perimetry, brain MRI, electroretinogram, and visual evoked potentials, which were normal.

Discussion

The first description of VS was termed in 1995 by Liu et al [3]. They described 10 patients suffering from migraines who developed a persistent positive visual phenomenon lasting for an extended period of time. VS was found to be common in 30–60% of patients suffering from migraines. Schankin et al. [1] hypothesized that VS is a clinically distinct entity which should be classified as a separate syndrome in its own right. Patients with “visual snow” suffer from continuous TV static-like tiny flickering dots filling the entire visual field. Most patients described a syndrome with additional visual symptoms which were included in the proposed diagnostic criteria for VSS (Table 1) [1]. It is important to emphasize that VSS is a diagnosis of exclusion. Similar entities that need to be ruled out are migraine, retinal and vitreous diseases, and retinal detachment.

We describe herein 6 cases of patients suffering from VS: 5 without a trigger and 1 where VS was reported after being treated with oral Isoretinoine for acne (Case 3). Isotretinoin (13-*cis*-retinoic acid) has been used for many years in the treatment of severe cystic acne and various other skin disorders. At present, there is no report in the literature connecting this phenomenon to the drug. We assume this to be an incidental finding. CPK, testosterone, and prolactin levels may be affected by the use of this drug [8].

Comparable to these patients, studies have shown a relatively high prevalence of migraines in patients with VSS. One prospective study of 120 patients found a history of

migraines with or without aura in 58% of VS patients and migraines with typical aura in 31% of the patients [1]. As in previous large case series published, our patients' visual evoked potentials were normal [1, 4]. Eren et al. [6] found that VSS patients showed a unique pattern of increased N145 latency and reduced N75-P100 amplitudes. Tinnitus appears to be common in this population as well, occurring in up to 63% of patients, though the frequency and persistence of tinnitus has, as yet, not been described [1, 4].

Half of the patients suffered from VSS. The distribution of visual complaints is presented in Table 2. Three of the patients suffered from migraines (2 without aura), and 2 patients suffered from tinnitus. VSS prevalence is unknown; it is more frequent in males with a ratio of 1:2.2 [7]. Many believe that VS begins in young adulthood; however, some cases of childhood onset have been described [8, 9].

Several reports have described different treatment possibilities, yet treatment effectiveness varies, and thus far, no randomized controlled trials have been performed. One such example is the case report of Unal-Cevik et al. [10] of a 25-year-old female successfully treated with lamotrigine [see also 11]. The best evidence we have of investigating a greater cohort is the retrospective analysis of van Dongen et al. [12] where Lamotrigine was prescribed to more than half of the patients, with ~20% experiencing a partial remission of symptoms. Other treatments described (i.e., valproate, topiramate, acetazolamide, and flunarizine) have not led to any improvement except for topiramate in one patient, who discontinued the medication due to adverse side effects [12, 13].

Although the pathophysiology of VSS is unknown, the description of the TV static pattern in the visual field points to a higher level of processing beyond the lateral geniculate body. A number of theories have been suggested such as a hyperactive visual cortex leading to a detection of visual stimuli that healthy individuals do not detect, or alternatively, an impaired processing of simultaneous afferent information projecting to the cortex, leading to the perception of visual stimuli in the absence of such, possibly due to decreased neuronal inhibition. Brain imaging studies have shown no specific structural abnormalities [1, 4, 5] and no regional functional changes in cerebral water diffusion and perfusion [14]. A recent functional brain imaging study performed on VSS patients reported that ¹⁸F-FDG PET lingual hyper metabolism, compared with healthy controls after adjusting for the presence of a typical migraine aura [15], strengthens the theory of visual cortex involvement. Recently, Lauschke et al. [4] formed a new hypothesis regarding the pathophysiological process of VS. The authors presumed that VSS is part of a thalamocortical dysrhythmia, with hyperexcitability playing a role, and is also associated with other disorders of sensory processing such as tinnitus, tremor, and migraine, possibly also involving the magnocellular pathway [1–4, 15]. An improvement in symptoms was also observed when using yellow-blue colored filters. McKendrick et al. [16] compared 16 patients with VSS with healthy controls and demonstrated suprathreshold processing of contrast and luminance, pointing to an imbalance between visual cortical inhibition and excitation, consistent with elevated spontaneous, cortical excitability in the primary visual cortex.

Conclusion

By promoting awareness of this syndrome, the fears of those who suffer from VSS will be alleviated and the initial labeling of these patients as mentally ill will be eliminated. Since the patients had constant complaints, without any visible cause, they had undergone a multitude of tests and examinations. We believe that if the complaints and history are typical according to the VSS syndrome criteria [17], further investigation is redundant, saving unnecessary and costly examinations. On the other hand, only after excluding other diagnoses can the doctor's and patient's fears be alleviated. In addition, greater quantitative and qualitative research may expand our understanding and treatment of this disorder, which appears to be connected to sensory processing, with no structural pathology found thus far. Patient education regarding this syndrome may possibly lead to faster improvement and recovery, better coping, and the minimization of the effect of this condition.

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Statement of Ethics

The study was approved by the Institution's (Hillel Yaffe Medical Center) Helsinki Committee. Informed consent was not needed. All subjects' identifications have been omitted/avoided.

Disclosure Statement

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Author Contributions

Eran Berkowitz: design, literature search, and manuscript preparation. Yaron River: data acquisition, manuscript editing, and manuscript review. Kathleen Digre: literature search, manuscript preparation, manuscript editing, and manuscript review. Beatrice Tiosano: design, manuscript editing, and manuscript review. Anat Kesler: data acquisition, literature search, manuscript preparation, manuscript editing, and manuscript review.

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Table 1. Criteria for the definition of the visual snow syndrome

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- A.** Visual snow: dynamic, continuous, tiny dots appearing in the entire visual field, lasting for more than 3 months; the dots are usually black/gray on a white background or gray/white on a black background; they can also be transparent, white flashing or colored; visual field and electroretinogram are usually normal; not caused by a previous intake of psychotropic drugs
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- B.** Presence of at least two additional visual symptoms of the following four categories:
- (i) Palinopsia
 - (ii) Enhanced entoptic phenomena; at least one of the following: excessive floaters in both eyes, an excessive blue field entoptic phenomenon, self-light of the eye, or spontaneous photopsia.
 - (iii) Photophobia
 - (iv) Nyctalopia
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- C.** Symptoms are inconsistent with the typical migraine visual aura as defined by the International Headache Society (Headache Classification Committee of The International Headache Society, 2004).
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- D.** Diagnosis of exclusion: normal ophthalmology tests (best corrected visual acuity, dilated fundus examination, visual field, and electroretinogram), no intake of psychotropic drugs.
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Table 2. Additional visual complaints of patients suffering from visual snow

Visual complaint	n (%)
Palinopsia	2 (33)
Entopic phenomena	3 (50)
Nyctalopia	2 (33)
Photophobia	1 (16)