

Visual Snow Syndrome: A Case Report and Review

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ABSTRACT

We present the case of a 16-year-old male, but with a past medical history of anxiety disorder, allergic rhinitis, and bilateral ear epidermal cysts who also presented for Visual Snow Syndrome (VSS). Patient underwent multidisciplinary evaluation and neuroimaging/studies were all normal. He endorsed photophobia. Although he denied tinnitus, his past medical history included recurrent ear cysts that required drainage. He reported that his anxiety was mainly due to social situations. At the time of exam, his family declined medications for anxiety or VSS. While there is a case report of reduction of VSS symptoms with amitriptyline, we were not able to investigate this further with our case due to the family's wishes. There is evidence that VSS patients may have hypermetabolism in the lingual gyrus and dysfunctional connectivity in the brain. Further studies may be needed to confirm this association and develop appropriate intervention strategies to normalize brain function.

Case

The patient is 16 y/o M with hx of anxiety disorder, allergic rhinitis, bilateral ear epidermal cysts and longstanding visual snow presents for follow up. When looking at objects, patients reports seeing a grainy picture with some white lights. Patient reported that he had seen an ophthalmologist who diagnosed him with Visual Snow Syndrome in the past. MRI brain with and without contrast including evaluation of optic nerves was normal. He wears glasses but denies any other vision changes. He reported no headaches, problems with balance, or difficulties with concentration. He also denied any difficulties with schoolwork. During the physical exam, he preferred the lights off. He stated he feels comfortable in low light environments. Although he denied tinnitus, he had recurrent left and right ear cysts that required drainage. His family history was significant for migraines on his mother's side. He reported no known allergies. Review of systems was largely unremarkable except for anxiety in social situations. His family declined medication treatment for VSS and anxiety at the time of examination.

Physical Exam and Related Studies

Exam within normal limits. Base Eye exam, Slit Lamp, and Fundus exams were normal. MRI with and without contrast indicated low lying cerebellar tonsils consistent with Chiari 1 malformation. The appearance of the orbits was unremarkable.

Discussion

VSS symptoms were initially reported by Liu and his group back in 1995 defined as "persistent positive visual phenomena" [1] and termed as visual snow syndrome by Jager, et al. [2]. Visual snow is an uncommon, chronic condition, sometimes highly disabling, that is in need of collaborative research in order to make progress towards understanding, treatment and cure. The main clinical feature of the syndrome described consistently by patients is an unremitting, positive visual phenomena, present in the entire visual field and characterized by uncountable tiny flickering snow-like dots that resemble the noise of a detuned analogue television interposed between the person's vision and the background. The 'static' is typically black and white but can also be colored, flashing or transparent "snow". Affected

individuals can experience additional visual symptoms such as images that persist or recur after the image has been removed (palinopsia); sensitivity to light (photophobia); visual effects originating from within the eye itself (entoptic phenomena) and impaired night vision (nyctalopia). Entoptic phenomena that are found (either alone or in combination) in visual snow are the blue field entoptic phenomenon, floaters (the perception of which is defined as myodesopsia), self-light of the eye and spontaneous photopsia. In addition, there may be less common symptoms such as migraine-like symptoms, ringing in the ears and fatigue. Tinnitus is extremely frequent in patients with visual snow, with up to three quarters of subjects reporting the symptom [3].

As in these cases, there may be concurrent psychiatric symptoms with VSS [4]. While there are relatively few case reports in the literature of VSS, there are even fewer with reports of psychiatric symptoms. The first-line psychiatric treatment for his psychiatric conditions would have been an SSRI, but there is no evidence of SSRIs being effective in the treatment of VSS. Given the existence of a case report of amitriptyline being successful in the treatment of VSS [5], a case could be made to start treatment with amitriptyline and consider adding an SSRI later. Tricyclic antidepressants such as amitriptyline were previously commonly used to treat depression, OCD and anxiety, so the hope was that there would be reduction in this patient's psychiatric symptoms with amitriptyline monotherapy. However, in accordance with his family's wishes, no medications were given.

Prevalence

The prevalence of visual snow in the general population is currently unknown. Nearly 200 documented cases of visual snow worldwide have been reported [6]. The available data tells us that there is possibly a higher prevalence of the disease in the male population and that the average age of affected subjects is relatively young [1]. This early onset, static disease may be underestimated due to lack of knowledge about VSS.

Pathophysiology

Pathophysiology is also unclear, but it is likely to involve how the brain processes vision [7]. This is mostly due to the characteristic of the chief symptom of the syndrome, i.e., the visual static, which is a whole-field visual disturbance. This makes a localization of the problem in the visual pathway or primary visual cortex extremely unlikely. Furthermore, additional symptoms such as palinopsia, which can be considered an inability to suppress the just seen, and the enhanced entoptic phenomena in the context of normal ophthalmological tests, also point in the same direction towards a central neurological disorder of the visual pathway, due to causes yet to be determined. Given a combination of tinnitus and visual symptoms, this has led some researchers to hypothesize a possible relationship between the two conditions, which both possibly represent a similar dysfunction in sensory processing, respectively of the visual and auditory system in the brain. Research has been limited because of issues of case identifi-

cation and diagnosis, the latter now largely addressed, and the limited size of any studied cohort. Initial functional brain imaging research suggests visual snow is a brain disorder.

A neuroimaging study using [18F]-FDG PET seems to have confirmed these hypotheses. The study demonstrated, in patients affected by visual snow, a hypermetabolism of the lingual gyrus [8]; this is an area of the visual cortex involved in several other conditions such as photophobia. The lingual gyrus is also a key element of complex physiological functions such as visual memory, perception of color and identification of facial expressions. Visual cortical hyperexcitability and thalamo-cortical dysrhythmia have also been hypothesized as possible causes for the pathophysiology underlying visual snow [9-11] Further studies on larger numbers of patients are needed to confirm these initial hypotheses. The pathophysiology of psychiatric symptoms is also unclear. While it may be that the distress of persistent VSS and/or tinnitus could lead to low mood and anxiety, it is not clear that psychiatric symptoms are exclusively a consequence of this distress. In one study, patients with lifelong VSS reported lower levels of distress from visual symptoms but were equally likely to have psychiatric symptoms [12].

Diagnosis

Visual Snow is a clinical diagnosis that comes from the fulfillment of a set of criteria and the exclusion of secondary causes of similar visual disturbances, such as underlying ophthalmological and neurological diseases. The differential diagnosis of visual snow includes prolonged migraine aura. The relationship between migraine and visual snow is a complex one; it has been in fact proven that migraine can aggravate the clinical presentation of the visual snow syndrome and that it has a higher prevalence in this disorder than in the general population [13]. Visual snow should also be distinguished from hallucinogen persisting perception disorder [14,15].

Treatment

Treatment is focused on managing the symptoms [16-18]. The lack of knowledge on the basic biology of visual snow syndrome has caused a general deficiency of effective treatment strategies for most patients. No clinical and systematic trials have been performed to date, and all available data on treatment comes from single patients or case reports. The current evidence seems to show that commonly used medications such as migraine preventives, antidepressants or pain medication do not consistently improve or worsen visual snow. There have been single positive experiences with drugs such as lamotrigine [19] which need to be contemplated within their low level of evidence.

Conclusion

Currently VSS seems to have overlapped with migraine, and with no clear identifiable pathophysiology, making the diagnosis is purely clinical. VSS has most often been refractory to variety of antidepressant/antiseizure medicines. In the meantime, we are left with Class

IV evidence that a minority of patients may profit from a trial of oral lamotrigine and with the confidence that most patients cope very well with reassurance [20]. While there is a case report of reduction of VSS symptoms with amitriptyline, we were not able to investigate this further with our case due to the family's wishes. As there is hypermetabolism in the lingual gyrus and growing evidence of dysfunctional connectivity in the brain, single region real-time fMRI neurofeedback may be a promising intervention strategy to normalize brain function in the future [20].

Disclosure

We have no conflicts of interests to disclose, and all authors have approved the final document.

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