Visual snow case series: review of 248 cases with attention to underlying causes or inciting events

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Background

- Visual snow (VS) is described as similar to seeing black and white or colorful flickering dots like “television static.” Visual snow syndrome (VSS) is diagnosed if symptoms have lasted longer than three months with at least two additional symptoms: photophobia, nyctalopia (impaired night vision), palinopsia (after images or trailing of moving objects), or enhanced entoptic phenomena.
- The mean age of patients that develop VSS is 12.8 +/- 13.2 years.[2]
- The prevalence of VSS is around 2%.[3]
- The most common inciting events reported by patients in the literature have been a change in migraines, head trauma, systemic infection, and drugs (recreational and prescribed).[2]
- We reviewed cases of VS at our institution and gathered details on the most common triggering events, and determined whether prognosis differed in patients with inciting events.

Methods

- This study was a retrospective chart review, and it was approved by our IRB.
- Using a database engine, 449 patients were identified using the term “visual snow.”
- Exclusion criteria: patients who did not consent to research participation (n=59), alternative diagnoses (n=97), and those with previous or outside history of VS, but not corroborating information (n=45).
- Demographic data was gathered as well as details regarding inciting events or secondary causes.

Results

- 248 patients met criteria for VS. 38 of them reported VS as migraine aura and were not included in our analysis of demographics and comorbidities.
- 210 patients were included (89 men and 121 women) with a median age of onset 20 +/- 13.3 years. 22 patients had VS for as long as they could remember. The median length of follow up was 3.6 years after the onset of their symptoms.
- VSS characteristics: persistent photophobia (n=93, 44.3%), nyctalopia (n=58, 27.6%), palinopsia (n=104, 49.5%), floaters/blue field entoptic phenomena (n=128, 60.9%), self-light (n=34, 16.2%), photopsias (n=50, 23.8%), brain fog (n=37, 17.6%), and dissociation/derealization (n=28, 13.3%) Five patients described a family history of VS.
- Of the 210 patients, 181 (86.2%) had brain magnetic resonance imaging (MRI), 165 (78.6%) had ophthalmologic exams, 55 (26.2%) had electroencephalograms (EEG), 51 (24.3%) had ocular coherence tomography (OCT), 20 (9.5%) had positron emission tomography (PET) scans of the brain, 29 (13.8%) had paraneoplastic panels, and 18 (8.6%) visual evoked potentials.
- Comorbidities are in Table 1 and inciting events/secondary causes are in Table 2.

Comorbidities	%/210
Migraine with aura (n=102)	48.6%
Migraine w/o aura (n=56)	26.7%
Anxiety (n=102)	48.6%
Depression (n=58)	27.6%
Tinnitus (n=71)	33.8%
POTS (n=48)	22.9%
Concussion (n=23)	11%
ADD/ADHD (n=30)	14.3%
Fibromyalgia (n=15)	7.1%
PTSD (n=3)	1.4%
Persistent Perceptual Postural Dizziness (n=28)	13.3%

Inciting Events or Secondary Causes	%/210
Post Concussion (n=15)	7.1%
Change in headache/aura (n=14)	6.7%
Post-infectious (n=13)	6.2%
Hallucinogen Persisting Perceptual Disorder (n=10)	4.7%
Medication-related (n=11)*	5.2%
Ocular abnormalities (n=7)	3.3%
Hormonal changes (n=5)	2.4%
Idiopathic intracranial hypertension (n=4)	1.9%
Epilepsy (n=3)	1.4%
Unclassified autoimmune disorder (n=2)	1.0%
Multiple sclerosis (n=2)	1.0%
Other systemic illness (n=2)	1.0%
Neoplastic (n=1)	0.5%
Psychiatric – Onset of schizophrenia (n=1)	0.5%
Degenerative – PCA (n=1)	0.5%

*Amantadine and testosterone also listed in ocular abnormalities and hormonal.

Results (continued)

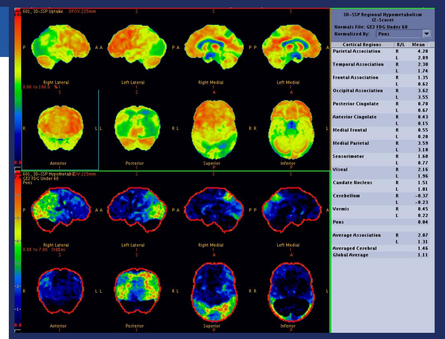
- 23 patients had a partial response to medication with none experiencing complete remission. The most beneficial medications were benzodiazepines (n=6), lamotrigine (n=5), topiramate (n=3), and acetazolamide (n=3).
- Patients with “secondary/abrupt onset” VS were not more likely to respond to medication treatment compared to primary VS. (p=0.1401)

Discussion

- The most common inciting events of VS were head trauma, changes in migraines, post-infectious, recreational drugs, and medication-related which is consistent with a large cohort that was recently described.[2]
- The exact pathophysiology of VS is not clear, but the two prevailing theories describe cortical hyperexcitability with dysfunction of higher order visual processing[4] or thalamocortical dysrhythmia.[4,5]
- One interesting case in our cohort was a patient who developed VS at the onset of posterior cortical atrophy (Figure 1). It has also been reported as a presenting symptom of Creutzfeldt-Jakob Disease.[6] Patients with VS should therefore be evaluated with a full neurologic exam and imaging.
- The study had limitations expected in a retrospective review. Information was limited to what was available in the chart and what was asked by the consulting physician. Due to the nature of our clinic, not all patients pursued follow up with us, and outcome information was sometimes limited.

Figure 1

FDG-PET scan of the brain: Images show hypometabolism in the parietal lobes bilaterally worse on the right, right temporal lobe, and in the occipital lobes bilaterally. Hypometabolism is also seen in the precuneus bilaterally but normal in the posterior and anterior cingulate gyri. This is consistent with posterior cortical atrophy. (PCA)



Conclusions

- VS can present both spontaneously or in association with inciting events or comorbid medical diagnoses. There does not appear to be any significant difference in phenotype or response to medications despite the cause of VS.

References

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Disclosures

Carrie E. Robertson: Advisory Board for Lundbeck, Biohaven, Amgen, and Eli-Lilly. She also receives honoraria as an author for UpToDate.

Ivan Garza: receives honoraria as an author for UpToDate.