CLINICAL VIGNETTE

Down the Rabbit Hole: Visual Distortions after Ischemic Stroke

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Introduction

Alice in Wonderland Syndrome (AIWS) is a rare neurological condition that includes distorted personal perception of somatosensory and visual input, as well as perception of time.[1] It was originally described by Caro Lippman in 1952 and the current moniker was coined by the psychiatrist John Todd in 1955, who was inspired by the experiences of the titular character in Lewis Carroll's story, Alice's Adventures in Wonderland.[2] In the story, Alice undergoes extreme changes to her body size, from shrinking down to 10 inches tall and enlarging to over 9 feet tall, after ingesting various Wonderland snacks. Classically, patients with AIWS experience two classes of symptoms: metamorphopsias, or visual distortions, and aschematias, or altered perceptions of self and time.[3,4] Due to the rarity of the syndrome, reaching a correct diagnosis requires clinicians to be vigilant of the different etiologies that may cause AIWS.

Case Presentation

A 52-year-old man initially presented to his local emergency room with right sided weakness and confusion. His past medical history includes type 2 diabetes mellitus, end stage renal disease on peritoneal dialysis, coronary artery disease, heart failure with preserved ejection fraction and obstructive sleep apnea. Magnetic resonance head imaging (MRI) revealed an acute infarct involving the left occipital, posterior parietal, and posterior temporal lobes (see Figure 1). His hospital course was complicated by sepsis due to Clostridium difficile and a witnessed seizure for which he was started on levetiracetam. Electroencephalogram (EEG) captured an electrographic seizure originating from the left frontal pole. As his encephalopathy resolved, the patient reported various visual disturbances that were distressful and frustrating to him. Examples included: seeing text and body parts enlarge and shrink, seeing objects suddenly appear closer and further to him while ambulating, seeing persistent trails after movement, and illusory movements in his peripheral vision. He also reported bizarre visual hallucinations such as grass growing out of his sink and a landscape in the corner of his hospital room. He retained insight into these hallucinations. The distortions were paroxysmal, typically lasting about 30-60 seconds, with no identifiable triggers. He denied any changes in his visual acuity, or diplopia. He had no headaches, or changes in perception of time or sensations.

His general physical examination was unremarkable. Neurological examination was notable for rare word finding difficulties but otherwise fluent speech output. He had full visual fields, intact pupillary reflexes, but impaired color vision perception in his right eye on Ishihara color plates. There were no signs of visual neglect, and extraocular muscle movements were normal. Strength testing revealed mild weakness in his right leg, 5- out of 5, as measured on the Medical Research Council Scale. The remainder of his neurological examination was normal.

The patient was discharged to acute rehabilitation. Over his two weeks in acute rehab, he noted decreasing frequency of his visual symptoms. Formal vision testing at rehab discharge was unchanged. He had no further seizures after starting levetiracetam.

Discussion

AIWS now includes 42 visual symptoms and 16 somesthetic and non-visual symptoms. These are all considered distortions of sensory perception rather than hallucinations or illusions.[2] This patient's presentation includes the most common feature of AIWS; metamorphopsias. Metamorphopsias has on many forms but the most common include: micropsia and macropsia (seeing things smaller and larger), pelopsia and teleopsia (seeing things closer and farther). The second most frequently reported features are aschematias, (altered perceptions of self), which was not present in this patient. The most reported aschematias are derealization, depersonalization, micro- and macrosomatognosia, feeling a part of or the entire body is smaller or larger than actual), and time distortion, with sensation of time acceleration or deceleration.

AIWS has been described as an epiphenomenon in migraines, epilepsy, central nervous system infections, particularly Epstein-Barr Virus (EBV), and cerebrovascular disease in both ischemic and hemorrhagic lesions. In the pediatric population, EBV is most frequently associated with AIWS, whereas in adults, migraines are the most common. It is important to distinguish the symptoms of AIWS from primary psychiatric disorders that may also feature visual hallucinations and symptoms of depersonalization and derealization. These include schizophrenia spectrum disorders and other hallucinatory syndromes.[2] AIWS has also been reported with illicit drug use, including lysergic acid diethylamide (LSD), and prescribed medications including dextromethorphan and topiramate.[2,3] Interestingly, this patient had experienced a seizure, but EEG

suggested a frontal lobe seizure focus, not in the area affected by his ischemic stroke.

The incidence of AIWS varies, as there are no universally accepted diagnostic criteria.[2] It is generally assumed to be rare, but some studies report a high prevalence, of greater than 15%. Individual symptoms may be common in the general population.

Nuclear medicine studies during episodes of AIWS have demonstrated decreased perfusion in all lobes of the brain either individually or in combination.[4] Pathophysiology from structural and functional imaging have implicated areas downstream in the visual pathway, visual cortical areas V4-5. Lesions of the temporo- parieto – occipital junction seems to have a dominant role in AIWS.[1] Theoretically, this would make sense as somatotopy is primarily controlled by parietal cortex, whereas higher order vision processing occurs in the anterior portions of the occipital. Lesions of the left and right cortices have been reported. It is postulated that decreased perfusion to the visual pathway and center may be responsible for this syndrome. This patient suffered an ischemic stroke in the left temporal, parietal, and occipital cortices, consistent with the most frequently noted locations.

Treatment of AIWS is primarily supportive and to treat any underlying conditions. Due to the paroxysmal nature of symptoms, most patients report an inconvenience rather than disability from the symptoms. Patients should be cautioned against driving or performing duties that require fine motor control as the visual distortions may impede their ability to perform tasks. Most cases of AIWS, have self-limited symptoms which improve over time. Although some cases, have persisted for years.[3]

Conclusion

AIWS manifests primarily with bizarre visual distortions and altered sensation of self and time. In adults, it is most often seen in migraineurs although other underlying conditions such as epilepsy and CNS infections may lead to AIWS.[5] The temporo-parieto – occipital junction is typically involved. The prognosis is generally good, with most patients experiencing full resolution of their symptoms. It is important to distinguish AIWS from other organic brain and psychiatric disorders to guide treatment. Further research is needed for better understanding treatment.

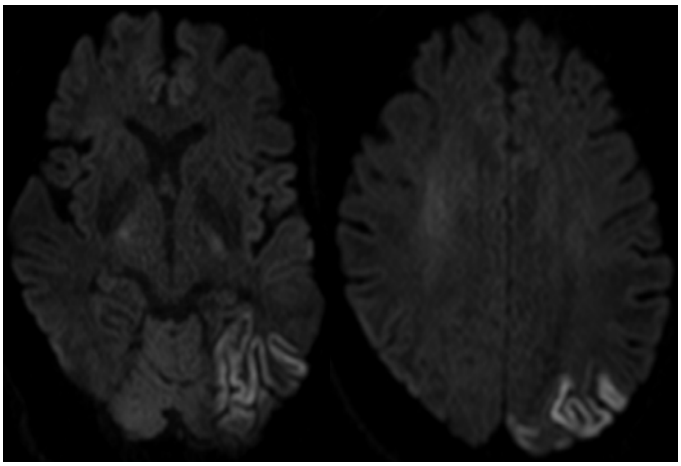

Figure 1. Representative axial slices from MRI brain: diffusion weighted imaging demonstrates hyperintensities involving the left posterior temporal, parietal, and occipital lobes with corresponding hypointensities on apparent diffusion coefficient sequences (not shown).

REFERENCES

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