First Reported Case of Alice in Wonderland Syndrome Due to an Arteriovenous Malformation

Veeresh Kumar N. Shivamurthy1*, Prateeka Koul2, Siddharth Gupta4, Jeffrey Kornitzer1,4, David Marks4

1Department of Neurology, Emory University School of Medicine, USA
2Department of Neurology, Northshore-LIJ, USA
3St. Joseph’s Children’s Hospital, Division of Child Neurology, Paterson, NJ, USA
4Department of Neurology, Rutgers New Jersey Medical School, USA

*Corresponding author: Veeresh Kumar N. Shivamurthy, Department of Neurology, Emory University, School of Medicine, 3131 N. Druid Hills Rd, apt 8305, Decatur, GA, 30033, USA; Email: nskveeresh@gmail.com

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Abstract

Alice in Wonderland Syndrome (AIWS) is characterized by unusual perceptual disturbances similar to those experienced by Alice in Lewis Carroll’s novel Alice’s Adventure in Wonderland. These disturbances may be caused by a range of organic, toxic, and psychiatric etiologies. Appropriately treating the underlying condition may reduce and altogether eliminate the sensory disturbances. This is the first reported case of an arteriovenous malformation presenting with symptoms consistent with AIWS.

Keywords: Alice in Wonderland Syndrome; Arteriovenous Malformation; Perceptual distortion; Visual hallucinations

Introduction

Alice in Wonderland Syndrome (AIWS) was first described in 1955, named after Lewis Carroll’s novel “Alice’s Adventure in Wonderland”, which is thought to be inspired by Carroll’s own migraine experiences [1, 2]. This syndrome is characterized by a constellation of visual and perceptual disturbances. Aetiologies of AIWS have been divided into eight main groups: infectious, central nervous system lesions, peripheral nervous system lesions, paroxysmal neurologic disorders, psychiatric disorders, medication, substance-induced, and miscellaneous [3]. In this case report, a patient presents with AIWS as a manifestation of non-convulsive status epilepticus secondary to a right parietal arteriovenous malformation.

Materials and Methods

Case report and Literature review were performed.

Results

A 24-year-old man with known focal-onset epilepsy managed with phenytoin and a known right parietal vascular malformation presented to the Emergency Department (ED) with breakthrough seizures. His history of seizures began in 2011 when neuroimaging revealed a 1.1 x 1.0 cm right parietal vascular malformation. He was prescribed phenytoin 300 mg daily but was then lost to follow-up. On his current encounter, he presented with a breakthrough seizure described by his mother as “repeated leftward head deviation and left shoulder shaking, followed by jerking of his arms and rhythmic kicking of his legs” that lasted one minute. Neurological examination upon initial presentation revealed an intermittent left visual field cut with left-beating nystagmus but an otherwise intact neurological examination. After having another seizure in the ED, he was loaded with levetiracetam 1,500 mg in addition to phenytoin 300 mg. Urine toxicology was negative and basic metabolic panel was unremarkable.

Upon further evaluation, he continued to note the intermittent left visual field cut along with visual hallucinations of a physician coming through a portal to the patient’s left side like the Marvel comic character Doctor Strange. He also reported experiencing visual disturbances for two weeks prior to his breakthrough seizures, such as the same left visual field deficit and bizarre visual and somesthetic disturbances. For instance, he reported seeing cars floating; he walked into his home and found everything new and unfamiliar, the phenomenon otherwise known as jamais vu. During his hospital stay, he endorsed witnessing people morph into other people; he also perceived one physician’s right shoulder and...
right-handed fingers growing bigger and his left hand shrinking in size. His family reported that some of the patient’s hallucinations consisted of him facing an empty part of the room and talking to an imaginary person, his mother floating, moss growing on his relatives, and the nurses walking around at night resembling zombies.

The vascular malformation was re-evaluated with computerized tomography (CT) of the head without contrast, which showed heterogenous hyperattenuation consistent with a right parietal cavernous malformation measuring 2.3 x 1.7 cm. Video EEG recording showed non-convulsive focal status epilepticus with a right temporoparietal seizure focus and a high frequency rhythmic buildup in this region. Concurrently, he showed clinical symptoms of visual hallucinations as described above with ictal left-beating nystagmus and left visual field cut. Magnetic resonance imaging (MRI) with and without contrast also revealed a 2.6 x 1.6 x 1.6 cm vascular malformation. He experienced upward of forty electrographic and clinical seizures while on the dual regimen of levetiracetam and phenytoin. He was also given a one-time injection of lorazepam 4 mg during the EEG recording, but his seizures persisted. Valproate 1000 mg twice a day was added to his antiepileptic drug regimen, but subsequent EEG recordings continued to show frequent seizures.

Given the progressive growth of his cavernous malformation and its focal electrographic correlate, the patient underwent a gross total resection of the malformation during his fourth day of hospitalization. A right parieto-occipital craniotomy with microdissection was performed to circumferentially separate the vascular malformation and surrounding hematoma and hemosiderin ring from the normal brain parenchyma. The gross specimen was a soft red and brown tissue; histologically, the specimen showed malformed vessels such as capillaries, arteries, and venules with abundant parenchyma between the vessels, consistent with an arteriovenous malformation (AVM).

Post-operatively, the patient had a non-focal neurological examination and no longer experienced the aforementioned perceptual disturbances. A subsequent EEG did not reveal any epileptiform activity. He was tapered off the phenytoin and discharged home on valproate 1000 mg twice a day and levetiracetam 1000 mg twice daily. At a follow-up visit two weeks later, he reported having no subsequent hallucinations or seizures. After several months, he was also tapered off the valproate and has been symptom and seizure-free while maintained solely on levetiracetam.

Discussion

Our patient presented with symptoms consistent with AIWSS secondary to seizures given their periodic nature caused by a progressively enlarging arteriovenous malformation. He had numerous visual and sensory distortions, such as dysmetropsia, macropsia, and kinetopsia, consistent with a clinical diagnosis of AIWS. Lanska and Mastria et al propose three categories of symptoms that further characterize AIWS [2, 4]. Category A consists of perceived changes in the patient’s own body (aschematia, macroemiatosognosia), while Category B consists of such changes external to the patient (macropsia, micropsia), and Category C is a combination of the two. Our patient falls into Category B based off his symptoms such as macropsia and micropsia [4]. From a systematic review of 169 published case reports of AIWS, Blom has found neurological problems to underlie the clinical manifestations amongst adults, especially migraines, in 27.1% of the total cases [3]. In the same systematic review, only five cases of 169 total cases manifested with epilepsy [3].

Interestingly, our institution reported a right temporoparietal cavernoma that caused seizures and concurrent visual distortions consistent with Category B Alice in Wonderland Syndrome [4, 5]. AVMs and Cerebral Cavernous Malformations (CCMs) are known to manifest as seizures [6, 7]. There are reported cases in the literature of vascular malformations in the parietal and temporal cortices as established foci for such seizures [7, 8]. However, there are no reported cases of epileptogenic arteriovenous malformations to-date that also demonstrate perceptual disturbances as our patient experienced [5, 9]. Lesions in the parietal lobe correspond to changes in visual and spatial perception, depending upon the specific region affected; for instance, a lesion in Brodmann Area 5 may lead to astereognosia, while a lesion in Brodmann area 7 affects the perception of movement [10]. There is evidence in the literature of some patients with parietal lobe epilepsy having aurae of distortions of body images and visual illusions [11].

Our patient’s most likely seizure focus is contained in an area known as the Temporal-Parietal-Occipital Carrefour (TPO-C). These cortical areas are involved in integrating visual, somatosensory, and vestibular inputs. In the TPO-C, the parietal and temporal streams of visual system are integrated with somatosensory and vestibular inputs. Hence, the TPO-C is where visual and somatosensory information are integrated to generate the inner and external representation of self. These inputs may be further broken down into different streams that aid a person to perceive an object’s movement and position (parietal inputs), as well as its size, form, and color (temporal stream). As our patient had an arteriovenous malformation in the parieto-temporal region, one would expect the patient to develop perceptual abnormalities involving the distortion of an object’s relative size and position, which he did as previously described [8]. In addition, given that the patient’s visual symptoms were episodic, it is more likely that they represented an ictal phenomenon rather than a direct manifestation of the arteriovenous malformation itself.

The current case report and our previously published report...
present vascular malformations as underlying aetiologies for the clinical manifestations of Alice in Wonderland Syndrome [5]. Both may present with signs of headaches, intracranial bleeds, and seizures, but their underlying anatomical features differentiate the severity and management of the presenting symptoms [12]. Discernible lesions in the central nervous system make up a relatively small fraction of total AIWS cases, but this case report adds arteriovenous malformation to the list of potential underlying aetiologies [3]. Although there are a limited number of cases in the literature of Alice in Wonderland Syndrome secondary to vascular malformations, proper management can be curative and relieve the patient of disabling and troubling symptoms [3]. As such, neuroimaging and electroencephalogram should be considered in a patient presenting with AIWS.

Figure 1: A) T2 flair without contrast axial: within the right parietal lobe, lobulated T2 hyperintense mass with peripheral T2 hypointense rim. B) T1 pre-contrast sagittal. C) T1 post-contrast sagittal: right parietal lobe with hyperintense mass with contrast enhancement.

Figure 2: EEG on admission demonstrated - Sustained rhythmic discharges from right centro-temporal and right centro-parietal regions.
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References