

Alice in Wonderland syndrome: “Who in the world am I?”

Síndrome de Alice no País das Maravilhas: “Quem sou eu no mundo?”

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ABSTRACT

Alice in Wonderland syndrome (AIWS) is a paroxysmal, perceptual, visual and somesthetic disorder that can be found in patients with migraine, epilepsy, cerebrovascular disease or infections. The condition is relatively rare and unique in its hallucinatory characteristics. **Objective:** To discuss the potential pathways involved in AIWS. Interest in this subject arose from a patient seen at our service, in which dysmetropsia of body image was reported by the patient, when she saw it in her son. **Methods:** We reviewed and discussed the medical literature on reported patients with AIWS, possible anatomical pathways involved and functional imaging studies. **Results:** A complex neural network including the right temporoparietal junction, secondary somatosensory cortex, premotor cortex, right posterior insula, and primary and extrastriate visual cortical regions seem to be involved in AIWS to varying degrees. **Conclusions:** AIWS is a very complex condition that typically has been described as isolated cases or series of cases.

Keywords: Migraine disorders; epilepsy; body image; stroke.

RESUMO

Síndrome de Alice no País das Maravilhas (SAPM) é uma condição paroxística visual perceptiva e somestésica que pode ser encontrada em pacientes com enxaqueca, epilepsia, doença cerebrovascular ou infecções. A condição é relativamente rara e tem características alucinatórias peculiares. **Objetivo:** Discutir as potenciais vias envolvidas na SAPM. O interesse pelo assunto surgiu com um caso de nosso serviço, onde a distropsia da imagem corporal foi relatada pela paciente, que via isto em seu filho. **Métodos:** Os autores revisaram e discutiram a literatura médica de casos relatados de SAPM, possíveis vias anatômicas envolvidas e estudos de imagem funcional. **Resultados:** Uma complexa rede neural incluindo junção temporoparietal direita, córtex somatossensitivo secundário, córtex pré-motor, região posterior da ínsula direita, e regiões do córtex visual primário e extra-estriatal têm diferentes graus de envolvimento na SAPM. **Conclusão:** SAPM é uma condição complexa que tipicamente foi descrita apenas com casos isolados ou séries de casos.

Palavras-chave: Transtornos da enxaqueca; epilepsia; imagem corporal; acidente vascular cerebral.

In 1952, Lippman provided the first description of patients experiencing sensations of becoming remarkably tall or short before or during migraine attacks¹. In 1955, Todd named the condition “Alice in Wonderland” while describing six patients with macrosomatognosia or microsomatognosia, among whom four were migraineurs². Alice in Wonderland syndrome (AIWS) is not specific to migraine or epilepsy and is a perceptual disorder, principally involving visual and somesthetic integration systems³.

Alice in Wonderland syndrome was named after the strange experiences described by Charles Lutwidge Dodgson

(best known as Lewis Carroll) in the Alice in Wonderland books¹. Like Alice, individuals affected with AIWS can experience paroxysmal alterations in their perception of the size of their own body parts, characterized by aschematia and dysmetropsia⁴. Some typical illustrations from Lewis Carroll's book are shown in Figure 1.

Over time, the expression “Alice in Wonderland” has become (mis)used to describe cases of derealization, depersonalization, somatopsychic duality, altered judgment of time, akinetopsia, auditory hallucinations, verbal illusions, dyschromatopsia, zoopsia and complex visual hallucinations^{1,5}.

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Figure 1. Original figures from Lewis Carroll's book, *Alice in Wonderland*, showing her size in relation to animals and her own appearance with a long neck, a large head or thin and small arms.

Although the concept of AIWS seems to have evolved far beyond the original description, abnormal somesthetic symptoms leading to self-body image distortions are the core of the syndrome. In summary, AIWS is a “self-experienced paroxysmal body image illusion” and the definition should remain restricted to these descriptions⁶. From the original Lewis Carroll's book⁷, Figure 1 shows Alice in her adventure through Wonderland.

Nonetheless, it seems to be valid to speculate what are the neurological pathways involved in self- and not-self body dysmorphia in Alice's eyes.

A case of an otherwise healthy 80-year-old woman with a history of sudden visual distortion of her son's right upper

limb caught our attention. In this case, there was a progressive increase in the size of the shoulder and arm and a decrease in the size of the hand that lasted for 15 minutes, and this was followed by visual haze and pulsatile headache. This patient had been a migraineur all her life, although the frequency and severity of headache attacks had diminished over the last few decades. She occasionally had fortification spectra aura preceding her headache, but had never previously experienced this visual distortion.

Her neurological examination showed homonymous hemianopsia after this aura and headache, and she reported having a moderate degree of holocranial pain and sensitivity to light. Figure 2 shows an axial cranial FLAIR brain magnetic resonance image with hemorrhage in the right occipital lobe. Her electroencephalogram did not show any abnormalities, but it should be noted that this examination was performed some hours after the visual episode. Further investigation confirmed the diagnoses of cerebral amyloid angiopathy in this patient and no aneurysm was identified. The patient was treated clinically and progressed well with complete remission of the hemorrhagic stroke. She did not have any further visual symptoms or headache attacks.

There are fewer than 200 cases of AIWS in the medical literature, and these have all been reported because of the curious features of each case. For example, AIWS has now been described in association with infections due to the Zika virus⁸, varicella⁹, or H1N1 influenza¹⁰, among others. Few functional imaging studies have reported on the image representation of body parts or have discussed the potential pathways involved in AIWS. A complex neural

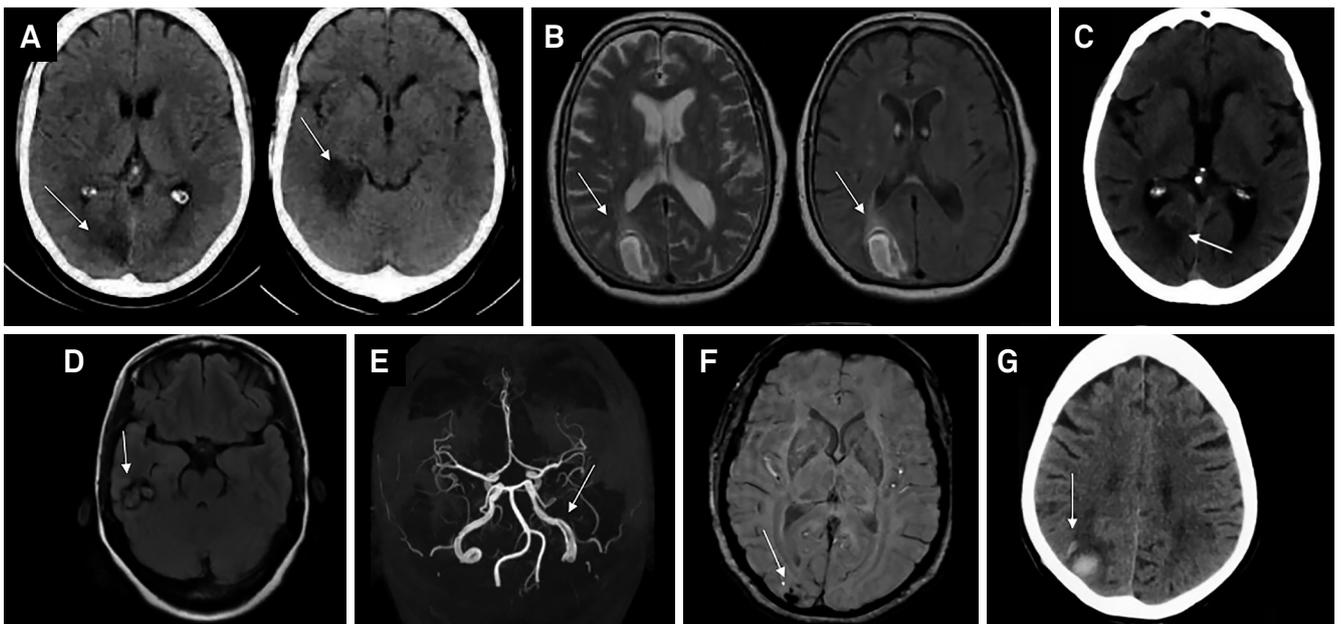


Figure 2. Brain images of patients described with Alice in Wonderland syndrome (AIWS). Patients A and B were described by Camacho Velasquez et al.¹⁴, patient C was published by García-Cabo et al.¹⁶, patient D by Philip et al.¹⁷, patient E shows the arterial dissection associated with AIWS symptoms reported by Mullagari et al.¹³, patient F was described by Morland et al.¹⁵ and patient G is described in the present paper.

network comprising the right temporoparietal junction, secondary somatosensory cortex, posterior parietal cortex, ventral premotor cortex and right posterior insula is involved in the subjective experience of body image and organ ownership^{2,11,12}. Only a few patients with cerebrovascular disease and AIWS have been presented in the medical literature^{13,14,15,16}. Some images from these patients are shown in Figure 2. Interestingly, these vascular lesions were predominantly in the right posterior occipital lobe areas, as seen in our patient. On the other hand, a temporoparietal cavernoma has been described as the origin of AIWS in one patient¹⁷. Aberrant activity in the primary and extrastriate visual cortical regions and in the parietal cortices have been described in AIWS episodes during functional imaging studies^{18,19}. Even occurrences of frontal lobe epilepsy have

been correlated with AIWS²⁰. While the majority of structural lesions leading to AIWS symptoms were on the right side of the brain, this was not exclusive (Figure 2).

Throughout the Wonderland adventures, Alice seems to be a spectator rather than a perpetrator⁷. There is a clear sense of “not belonging”, perhaps best expressed by the Mad Hatter, who declares that Alice can sit at the table, despite not being invited. Moreover, in addition to her metamorphosis, Alice certainly has an eating disorder, in that she binges on whatever food and drink comes her way.

Whatever the complex brain pathways involved in Alice-like behavior and hallucinations are, these rare cases among patients with migraine, epilepsy, cerebrovascular and various infectious diseases are fascinating. “Curiouser and curiouser!” cried Alice in Wonderland.

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