

Alice in Wonderland syndrome as the initial and sole manifestation of ischaemic stroke[☆]



Síndrome de Alicia en el País de las Maravillas como primera y única manifestación de un ictus isquémico

Dear Editor:

Ischaemic stroke is the leading cause of death in women and the third in men in our setting.^{1,2} On occasion, diagnosis is hindered by the presence of atypical signs and symptoms, which may confuse the neurologist or clinician attending the patient for the first time.

We present the case of a patient with ischaemic stroke with atypical hallucinations as the sole manifestation.

Our patient is a 95-year-old woman who lives alone and is totally independent in the activities of daily living and does not present cognitive impairment; her medical history only reports hypercholesterolaemia, controlled with low doses of statins.

She consulted due to a one-month history of sudden-onset visual hallucinations limited to the left hemifield. She saw ‘‘people with tiny hands,’’ distorted images, ‘‘[her] relatives’ heads became huge when they approached [her] from the left’’; she also saw animals approaching her from the left. The patient was aware at all times that these were hallucinations but found them highly disturbing. The examination also revealed left homonymous hemianopsia, with no further neurological deficits.

A brain CT scan showed a hypodensity in the right posterior cerebral artery (PCA) territory, affecting the calcarine sulcus (Fig. 1).

Aetiological studies included a Doppler ultrasound of the supra-aortic trunks, which obtained normal results; a transcranial duplex ultrasound, which showed a normal circle of Willis; Holter monitoring, which ruled out emboligenic arrhythmia; and a blood test, with normal results. An electroencephalography revealed slow activity in the right posterior area (coinciding with the area of the ischaemic lesion).

The patient was diagnosed with cryptogenic ischaemic stroke in the right PCA area, manifesting as Alice in Wonderland syndrome (AWS); treatment was started with low doses of levetiracetam in addition to antiplatelets for secondary prevention of vascular events; this achieved good outcomes.

AWS was first described in 1955 by the British psychiatrist John Todd³ and included a group of symptoms ‘‘intimately associated with migraine and epilepsy, although not confined

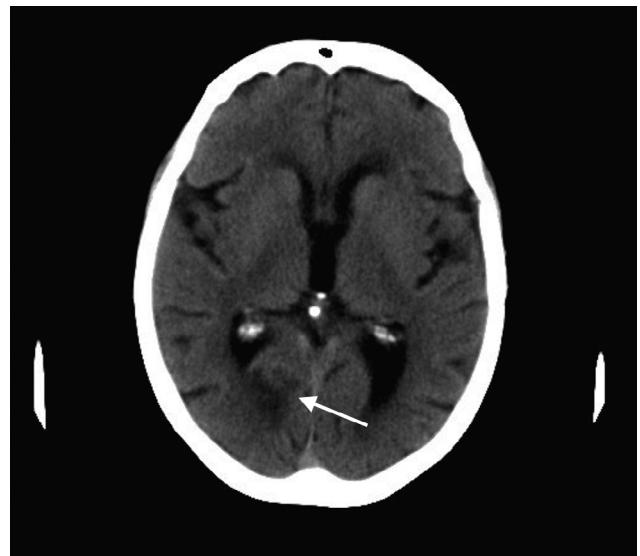


Figure 1 Simple CT scan; the arrow indicates an occipital hypodense lesion, compatible with ischaemic stroke in the sub-acute/chronic stage.

to these disorders’’. It owes its name to the similarity of symptoms with Lewis Carroll’s story, in which Alice experiences changes in body size. As described in the book *Alice’s Adventures in Wonderland*,⁴ the group of symptoms includes visual illusions which make the patient perceive alterations in the dimensions of objects (micropsia and macropsia), repeated images of stimuli after their disappearance (palinopsia), altered shapes (metamorphopsia), or animals (zoopsia).

This syndrome is occasionally accompanied by depersonalisation and altered perception of time. The most remarkable feature of this syndrome is that patients are aware that they are experiencing hallucinations.⁵

Symptoms of AWS are attributed to functional and structural abnormalities in the visual perception system in the occipital lobe⁶; the most frequent causes include space-occupying lesions, migraine episodes, seizures, or hallucinogenic substances.

In some series, cerebrovascular aetiology accounts for 3% of AWS cases, predominantly in patients older than 18.⁷

Most cases are benign, but if there is an underlying cause (as in our patient), symptomatic treatment may be necessary and useful, with antiepileptics being the most frequently used drugs.⁸

Our patient’s progression with low doses of antiepileptics was optimal.

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Charles Bonnet syndrome in a young patient with a history of epilepsy[☆]



Síndrome de Charles Bonnet en paciente joven con antecedentes de epilepsia

Dear Editor:

Charles Bonnet syndrome is a rare condition which usually affects elderly patients, especially women, due to ophthalmological involvement, whether due to retinal or anterior segment disease.^{1,2} We report a case of Charles Bonnet syndrome in a young patient with bilateral atrophy of the optic nerve secondary to intracranial hypertension due to cerebral venous thrombosis.

Our patient was a 44-year-old man who started to present multiple cerebral venous sinus thrombosis of unknown origin at the age of 30. Smoking was the only risk factor he presented at the time of thrombosis. Sequelae of thrombosis were partial seizures with secondary generalisation and sensory symptoms and intracranial hypertension with bilateral oedema of the optical nerve. Treatment was started with acenocoumarol, carbamazepine, a lumboperitoneal shunt, and fenestration of both optical nerves, which did not resolve atrophy. At the age of 40, as partial seizures decreased and 3 electroencephalography studies revealed normal results, we decided to suspend antiepileptic treatment.

At the age of 42, he developed visual hallucinations of serpentine elements, lasting more than 10 minutes; we

requested a magnetic resonance imaging (MRI) study to rule out occipital alterations, due to suspicion of sinus rethrombosis. No changes were detected with regards to the previous MRI studies. We requested an additional electroencephalography; results were considered normal for the patient's age. Since then, the patient has reported seeing a yellow car recurrently appearing at different points of the visual field, in both monocular and binocular vision. Episodes lasted from seconds to hours, with no associated symptoms, disorientation, or headaches. The patient did not associate the visions with fatigue, any particular activity, or with changes to light conditions or position. He was fully aware of the phenomenon and reported that hallucinations caused him no discomfort; therefore, no new treatment was started. Visual acuity was 0.4 in the right eye and 0.25 in the left. He presented severe atrophy of both optic nerves (Fig. 1) and severely restricted visual field (Fig. 2).

Charles Bonnet syndrome is described as visual hallucinations in patients with vision loss. The syndrome has classically been described in elderly patients, especially women.³ It consists of a type of hallucinosis, since the patient is fully or partially aware of the phenomenon, which usually involves a high level of anxiety; hallucinations manifest suddenly, with no voluntary control. Duration ranges from seconds to days.

Most case series report more severe visual impairment than in our case, with the syndrome manifesting in patients with macular degeneration, cataracts, or glaucoma.⁴ It has also been associated with such eye treatments as photodynamic therapy, laser retinal photocoagulation, and injection of anti-angiogenic agents. Before establishing a diagnosis, and given the patient's history, it is important to rule out such other causes of visual hallucinations as inflammatory, toxic-metabolic, infectious, and especially neurological diseases, such as epilepsy, migraine, or Lewy body dementia.⁵ In this case, there was high suspicion of epilepsy due to occipital lobe involvement, given the appearance of simple hallucinosis; with more complex hallucinations, it becomes more improbable.

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