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Case Report

Boundaries of visual phenomena in migraine: the Charles Bonnet's Syndrome - report of two cases

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Introduction
Two patients were referred to a tertiary headache center due to complex visual phenomena that might be related to migraine aura. Eventually, it turned out that they were Charles Bonnet syndrome (CBS) patients. The syndrome is characterized by vivid visual hallucinations in the absence of other psychiatric symptoms, often in patients with optical morphophysiological disorders. The pathophysiology of CBS remains uncertain, and the syndrome lacks robust diagnostic criteria. So far, reports on CBS are scarce in Brazilian neurological literature, leading to challenges in identification and diagnosis. The condition is primarily recognized through anamnesis and clinical examination.

Objective
To report two cases of CBS to draw the attention of headache specialists to this syndrome.

Case reports
The first case involves a 93-year-old male patient, retired, who reported experiencing vivid hallucinations, sometimes of an old white car and sometimes of a group of children. Despite the clarity of these visions, the patient remained aware that they were inaccurate. In the second case, a 70-year-old male farmer began experiencing hallucinations involving scratches, scribbles, letters, stones, and women in his visions.

Conclusion
Here, two cases of CBS were reported concerning a not-so-rare condition, but for sure very uncommon to headache specialists, whose visual phenomenology does not seem to be related to migraine phenomena.

Keywords:
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Introduction

Headache Medicine is far from a naîf field in Medicine. In addition to the existence of more than 300 medical conditions associated with headache, migraine being a protagonist (ICHD-3), there is an intriguing set of visual manifestations that may or may not be related to migraine, namely snow vision and persistent aura without infarction (ICHD-3). Two elderly patients with complex visual symptoms were referred to a tertiary headache center, and they fit the diagnosis of Charles Bonnet syndrome (CBS).

CBS consists of sudden and vivid visual hallucinations, in which the patient is aware that their perceptual experiences are unreal, with no psychological or intellectual changes, in patients with profound visual disabilities (1). In general, it is a benign disorder. The visual phenomena consist of objects, people, animals, and even scenes with vivid images during the episodes (2).

CBS was first reported in the 18th century by the Swiss naturalist and philosopher Charlie Bonnet when observing his grandfather, who had visual hallucinations. There is a predominance of involvement in elderly individuals (3). There is an association with a set of diseases characteristic of senility of vision, such as cataracts (4) and degenerative diseases of the retina (5).

Visual hallucinations can appear in simple forms, such as photopsies and myodesopsis, or complex ones, such as cars and people, including color figures and movements. Usually, episodes occur daily, lasting 3 to 5 minutes, and evolve over 6 to 12 months. These hallucinations may cause significant disturbances exacerbated by visual deterioration due to ophthalmological diseases.

The exact neurophysiological basis of CBS remains uncertain (6). However, the prevailing hypothesis suggests that the phenomenon is related to visual deafferentation. This refers to decreased visual input caused by changes in the visual system, such as cataracts, glaucoma, and macular degeneration. These changes in visual input are thought to disrupt neuronal communication in the visual cortex, potentially awakening or hypersensitizing dormant areas, which then give rise to visual hallucinations (7). This hypothesis can be seen as expanding Cannon's Law of Supersensitivity of Denervated Structures. The concept of denervation supersensitivity suggests that disruption of the regular nerve supply to the organ results in homeostatic compensation, which may have both pre-and post-synaptic components, to restore the normal functioning of that organ (8).

Another topic that deserves to be mentioned is the lack of clear and widely accepted diagnostic criteria for CBS. Currently, there is the French-Canadian questionnaire for the multidimensional analysis of visual hallucinations (9), and the one that I use the most, the Teunisse (10) CBS diagnostic criteria, is depicted in Table 1.

Table 1 - Teunisse's Diagnostic criteria for CBS

Diagnostic Criteria	Description
1.Detailed and complex visual hallucinations	The patient should report detailed visual hallucinations, which may include vehicles, people, walls, and other scenes.
2. No cognitive and psychiatric deficits	The patient cannot have degenerative or psychiatric diseases that alter cognitive function.
3. Awareness of the fantasy nature of visual hallucinations	The patient must be aware that the hallucinations are fictitious, at least for some period.
4. Do not present any other type of hallucination	The patient must present only visual hallucinations, not reporting other sensory types.
5. Presence of visual déficits	The patient must have significant vision loss due to a disorder or disease.

Source: Teunisse, 1996

The prevalence of Charles Bonnet Syndrome (CBS) is up 11 to 15% of the population with retinal degenerative processes or disorders leading to vision impairment (11).

Nevertheless, reports on CBS in Headache Medicine are still scarce, prompting the motivation to write this case report.

Case Reports

The following cases were sent to a tertiary headache service and examined by one of the authors (CAB) to assess the visual phenomena and their relationship to migraine.

Case 1

Man, 93 years old, mathematics teacher and poet. No history of migraine. He is lucid, alert, and active. He has had retinitis pigmentosa for 20 years, his left eye has subtotal amaurosis, and his right eye has 10% visual acuity. Two years ago, he began to experience visions of a white car in the distance, small and parked, completely dissociated from the environment where he found himself, for example, in his living room. The vision appeared in episodes lasting 15 minutes to 30 minutes, three to four times a day, at night. The image was clearer compared to the previous vision loss. The patient had the perception that the image was not real. The visual hallucinations disappeared after 15 days.

A day ago, he reported that he started seeing children playing without recognizing them. The vision presented two episodes, one during the day and one at night, with the nighttime period having clearer images. Once again, the patient was aware that the images were fantasy.

Despite his advanced age and his amaurosis, he manages his own business without cognitive deficits. Uses Apixaban and Nebivolol for atrial fibrillation. Computed tomography

showed no evidence of infarcts or significant brain injuries.

Case 2

A male, 70 years old, farmer has had macular degeneration for ten years. He has less than 10% vision in both eyes. He is on treatment for arterial hypertension, with losartan 50 mg twice daily, with satisfactory response.

About a year ago, he started seeing letters, scratches, sometimes dots, and flashing. He used to draw (Figure 1).

On occasion, the images were complex, with a wall built with stones, sometimes animals like a frog, and sometimes an unknown woman. Images could last for hours, sometimes recurring every day. They might be present for up to six days. Notably, the image of the woman caused him embarrassment by accompanying him during the bath.

They were clearer at night and perceived as clearer than natural vision. They appear regardless of whether they are alone or in their emotional state. The CT scan showed no significant data.

Figure 1. Drawings that the patient represented his hallucinations
Source: prepared by the author (2020)

Discussion

The reported cases meet the diagnostic criteria for CBS as outlined by Teunisse. The visual hallucinations in patients with profound visual impairment may be consistent with the deafferentation theory, as previously reported (12).

Considering that none of the patients had a history of migraines and given that the visual symptoms are pretty dissimilar to visual auras, combined with the fact that migraines very rarely start in later phases of life, it turns out quite unlikely that these are migraine phenomena.

An important aspect to highlight is the differences in diagnostic criteria that can lead to doubts about the character of the aura in cephaliatric medicine. The description of Migraine with aura, according to ICHD-3, can be identified by the following signs: "Recurrent attacks, lasting minutes, of unilateral fully reversible visual, sensory or other central nervous system symptoms that usually develop gradually and are usually followed by headache and associated migraine symptoms" having the following diagnostic criteria (13):

Table 2 - diagnostic criteria for migraine with aura by ICHD-3

Diagnostic criteria:
A. At least two attacks fulfilling criteria B and C
B. One or more of the following fully reversible aura symptoms: 1. visual 2. sensory 3. speech and/or language 4. motor 5. brainstem 6. retinal
C. At least three of the following six characteristics: 1. at least one aura symptom spreads gradually over 5 minutes 2. two or more aura symptoms occurin succession 3. each individual aura symptom lasts 5–60 minutes 4. at least one aura symptom is unilateral 5. at least one aura symptom is positive 6. the aura is accompanied, or followed within 60 minutes, by headache
D. Not better accounted for by another ICHD-3 diagnosis.

Source: The International Classification of Headache Disorders, 3rd edition

Migraine with aura is often accompanied by intense migraine attacks, occurring unilaterally and throbbing, lasting from 5 to 60 minutes, associated with symptoms such as nausea, vomiting, photophobia, and phonophobia, which occur with variable frequency, often during migraine episodes, but do not occur during the visual hallucinations present in Charles Bonnet Syndrome (CBS), which can last from a few minutes to several hours, with fluctuating frequency.

It is worth noting that the nature of the visual phenomena witnessed in Charles Bonnet Syndrome, due to reduced visual capacity, is complex and detailed, contrasting with the auras of migraines that manifest themselves as variable scotomas and zigzag patterns. In this context, it is essential to emphasize that, as mentioned in Teunisse's diagnostic criteria, the visions experienced in SCB are restricted to visual phenomena, not extending to other sensory modalities. This contrasts with the symptoms that may be present in migraine attacks with an aura, such as paresthesia and sensory, auditory, and motor changes (13).

With the advent of this classification, the significant difference between the two pathological conditions becomes evident when the diagnostic criteria presented in Table 3 are compared. The migraine crisis criterion establishes the need for the presence of migraine—before, during, or after the crisis—as reported in criterion C of Table 2.

It is essential to mention the factor of the visual perception of the images. During CBS, patients perceive that the images are not authentic, even though they are vivid. However, in migraine attacks with aura, patients tend to report altered visual perception, which does not involve detailed images or true visual hallucinations.

In this way, it is possible to understand the distinction between the two neurological phenomena, allowing the mentioned cases to serve as examples for other headache doctors and clarifying any uncertainties that may arise concerning possible diagnostic ambiguities.

Table 3- Differentiating the clinical presentation of migraine with aura from Charles Bonnet Syndrome

Clinical Aspect	Migraine with Aura	Charles Bonnet Syndrome (CBS)
Type of pain	Intense, throbbing, unilateral headache	Absence of headache
Associated symptoms	Nausea, vomiting, photophobia, phonophobia	Generally no associated symptoms
Duration of manifestations	Minutes to an hour	Persists from minutes to several hours, with fluctuating frequency
Visual aspects	Visual disturbances, such as bright lines, blind spots, flashing lights	Vivid, detailed, and colorful visual hallucinations (objects, people, scenes)
Patient's perception	Altered visual perception, without detailed hallucinations	Recognition of the unreality of hallucinations
Origin of manifestations	Neurological changes associated with cerebral circulation	Lack of visual stimuli in patients with vision loss
Recurrence of episodes	Associated with migraine episodes	Occurs in individuals with severe visual impairment
Periodicity between crises	Variable frequency, typically during migraine episodes	Recurrent episodes, with no defined periodicity

Source: prepared by the author

Conclusion

Two cases of Charles Bonnet Syndrome were reported, and the visual phenomena present in patients with CBS manifest differently from migraines with auras. Thus, this case report aims to raise awareness among healthcare professionals about CBS, highlighting the importance of recognizing complex visual hallucinations in patients with visual deficits without any impairment of consciousness. Although rare, the diagnosis of CBS can relieve patients by clarifying their condition and guiding appropriate treatment, improving their quality of life. It is essential to consider CBS as a differential diagnosis, investigating the possibility after ruling out other conditions that cause hallucinations, such as delirium, schizophrenia, and dementia, through complementary examinations.

References

1. Menon GJ, Rahman I, Menon SJ, Dutton GN. Complex Visual Hallucinations in the Visually Impaired. Surv Ophthalmol. 2003 Jan;48(1):58–72. Doi: 10.1016/S0039-6257(02)00414-9.
2. Hamedani AG, Pelak VS. The Charles Bonnet Syndrome: a Systematic Review of Diagnostic Criteria. Curr Treat Options Neurol. 2019 Sep 25;21(9):41. Doi: 10.1007/s11940-019-0582-1.
3. Gonçalves CM, Figueirôa FL da S. Síndrome de Charles Bonnet: Uma Revisão Integrativa. Revista Brasileira de Neurologia e Psiquiatria. 2018 Dec 22;302–21.
4. Rojas LC, Guarnani B. Charles Bonnet Syndrome. In: StatPearls [Internet]Treasure Island (FL): StatPearls. StatPearls Publishing; 2024.
5. Holroyd S, Rabins P V., Finkelstein D, Nicholson MC, Chase GA, Wisniewski SC. Visual hallucinations in patients with macular degeneration. American Journal of Psychiatry. 1992 Dec 1;149(12):1701–6.Doi: 10.1176/ajp.149.12.1701.
6. Carpenter K, Jolly JK, Bridge H. The elephant in the room: understanding the pathogenesis of Charles Bonnet syndrome. Ophthalmic and Physiological Optics. 2019 Nov 8;39(6):414–21. Doi: 10.1111/opo.12645.
7. Ffytche DH. Visual hallucinations in eye disease. Curr Opin Neurol. 2009 Feb;22(1):28–35. Doi: 10.1097/WCO.0b013e32831f1b3f.
8. Yarbrough GG, Phillis JW. Recent Progress: Supersensitivity of Central Neurons — A Brief Review of an Emerging Concept. Canadian Journal of Neurological Sciences / Journal Canadien des Sciences Neurologiques. 1975 Aug 18;2(3):147–52. Doi: 10.1017/S0317167100020187.
9. Cantin S, Duquette J, Dutrisac F, Ponton L, Courchesne M, de Abreu Cybis W, et al. Charles Bonnet syndrome: development and validation of a screening and multidimensional descriptive questionnaire. Canadian Journal of Ophthalmology. 2019 Jun;54(3):323–7. Doi: 10.1016/j.jcjo.2018.05.008.
10. Teunisse RJ, Zitman FG, Cruysberg JRM, Hoefnagels WHL, Verbeek ALM. Visual hallucinations in psychologically normal people: Charles Bonnet's syndrome. The Lancet. 1996 Mar;347(9004):794–7. Doi: 10.1016/S0140-6736(96)90869-7.
11. Boller F, Birnbaum DS, Caputi N. Charles Bonnet Syndrome and Other Hallucinatory Phenomena. In 2018. p. 117–24. Doi: 10.1159/000475702.
12. NORTON-WILLSON L, MUNIR M. Visual Perceptual Disorders Resembling the Charles Bonnet Syndrome. A Study of 434 Consecutive Patients Referred to a Psychogeriatric Unit. Fam Pract. 1987;4(1):27–31. Doi: 10.1093/fampra/4.1.27.
13. Headache Classification Committee of the International Headache Society (IHS) The International Classification of Headache Disorders, 3rd edition. Cephalalgia. 2018 Jan 25;38(1):1–211. Doi: 10.1177/0333102417738202.

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