Psychiatria Danubina, 2024; Vol. 36, No. 3-4, pp 394-396 https://doi.org/10.24869/psyd.2024.394 *Case Report*

FROM DIALYSIS TO HALLUCINATIONS: CHARLES BONNET SYNDROME AS A SEQUELA OF BILATERAL NON-ARTERITIC ANTERIOR ISCHEMIC OPTIC NEUROPATHY

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received: 9. 9. 2024; revised: 1. 10. 2024; accepted: 2. 10. 2024

Keywords: *charles bonnet syndrome, non-arteritic anterior ischemic optic, hemodialysis, chronic kidney disease, hypotension*

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INTRODUCTION

Despite being described over 250 years ago the Charles Bonnet Syndrome (CBS) remains poorly understood (Carpenter et al. 2019, Pang 2016). CBS typically involves complex visual hallucinations, although simple ones may also occur, in cognitively intact individuals (Arda 2021). It commonly affects elderly patients with significant visual impairment (Pang 2016, Schadlu et al. 2009). The pathophysiological mechanism is unknown, but sensory deprivation in the visual cortex may lead to the projection of hallucinations through compensatory mechanisms of brain excitability (Sawant & Bokdawala 2013).

Anterior ischemic optic neuropathy (AION) is the second most common cause of optic neuropathy in older and middle-aged adults (Sawant & Bokdawala 2013). AION results from either temporary or persistent hypoperfusion of the anterior segment of the optic nerve, leading to acute loss of visual acuity (Sawant & Bokdawala 2013). AION can be subdivided into two types: arteritic AION (A-AION), where ischemia is typically secondary to giant cell arteritis (GCA), and non-arteritic AION (NA-AION), where ischemia is due to non-inflammatory small vessel disease. NA-AION constitutes approximately 85% of AION cases, with bilateral involvement occurring in 15% of patients over a 5-year period (Banik 2013).

CASE

We report a 56-year-old male with chronic kidney disease (CKD) and multiple cardiovascular risk factors. He has been on maintenance hemodialysis (HD) for approximately 30 years following two failed renal transplants.

Hours after a HD session, he experienced a sudden, painless decrease in visual acuity in the right eye, without headache or ocular pain. During the session, his blood pressure was consistently low, with a recorded minimum of 79/36 mmHg. He was referred to the ophthalmologic emergency department (ED) and presented with a corrected visual acuity (VA) of 7/10 in the right eye (RE) and a VA of 6/10 in the left eye (LE). Within 24 hours, VA rapidly declined in the RE, with significant sequential involvement of the LE.

Acute-phase laboratory evaluation revealed an erythrocyte count of 2.61 x 10^{12}/L, hemoglobin of 8.2 g/dL, hematocrit of 25.3%, platelet count of 251 x 10^{9}/L, fibrinogen of 853 mg/dL, D-dimers of 0.34 µg/mL, creatinine of 6.4 mg/dL, and urea of 58 mg/d.

A Cranial Magnetic Resonance Imaging (MRI) with contrast of the brain and orbits, along with cerebral MR Venography, excluded the diagnosis of papilledema secondary to intracranial hypertension

Within 48 hours, the patient's vision deteriorated to no light perception (LP) in the RE and only the perception of large shapes in the LE. Intravenous methylprednisolone was initiated at 1 mg/kg/day for 3 days. Despite treatment, vision loss progressed, resulting in no LP in either eye by the 6th day, accompanied by a bilateral absolute afferent pupillary defect (Figure 1).

A diagnosis of unilateral NA-AION was considered, likely due to cardiovascular risk factors and severe hypotension during the HD session (Huang et al. 2013). A superior temporal artery doppler ultrasound and biopsy ruled out A-AION (Banik 2013).

Genetic testing for Leber's optic neuropathy and additional tests, including a thrombophilia panel (anti-cardiolipin and anti-beta-2 glycoprotein antibodies), and serologic tests for toxoplasmosis and syphilis (Dimitriadis

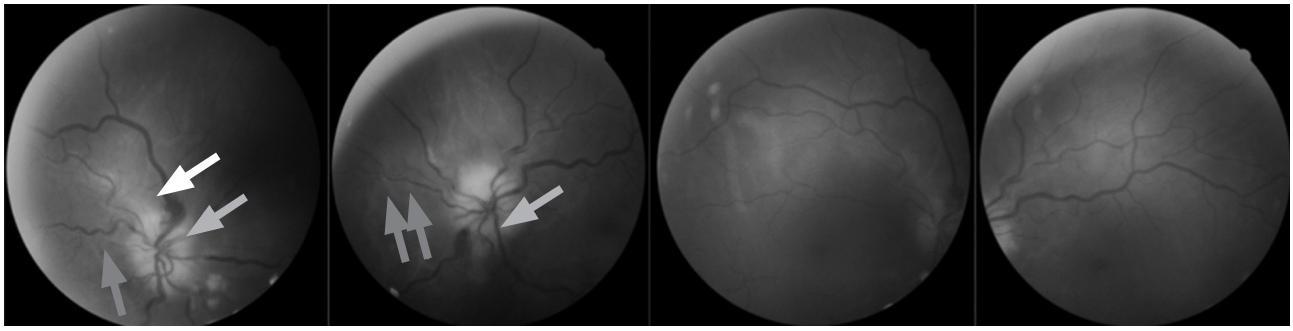

Figure 1 – Retinography of the LE 48 hours after the onset of the NA-AION episode identified peripapillary wrinkles (dark gray arrow), optic disc edema (bright gray arrow) and optic nerve swelling (white arrow)

et al. 2014, Vale et al. 2014), were all negative. After discharge, a follow-up on the 12th day revealed no LP in either eye. Given the unlikely recovery and severity of bilateral amaurosis, he was referred to physical medicine and rehabilitation for gait training.

Seven months after the episode, the patient reported experiencing visual phenomena, including white light and complex shapes, which are indicative of CBS (Carpenter et al. 2019, Pang 2016, Cox & Ffytche 2014). He expressed contentment stating, "At least I could see something, whether real or not, such as horses, dogs, children, circles, and squares". Therefore, treatment was not initiated, and the patient was referred for a psychiatry consultation, which confirmed the diagnosis of CBS. Similarly, treatment was not started because the patient expressed satisfaction with the figures and shapes, which allowed him to "see again".

DISCUSSION

As mentioned in the introduction, medical literature suggests a strong association between CBS and visual impairment (Carpenter et al. 2019, Pang 2016). In cases where the patient is not distressed by the hallucinations or they occur infrequently, treatment may not be necessary, as highlighted in our case (Cox & Ffytche 2014). However, about one-third of patients find the visual hallucinations disturbing (Chen 2014). The first step should be addressing the underlying cause of vision loss (Carpenter et al. 2019, Pang 2016).

Secondly, maximizing residual vision through optometric interventions (such as glasses, contact lenses, optical aids, and/or visual rehabilitation) or surgery (Darija et al. 2018) is usually considered the ideal medical approach. If these measures are ineffective, pharmacological treatment might be initiated. Despite inconsistent results, antipsychotics, anticonvulsants, anxiolytics, and selective serotonin reuptake inhibitors (SSRIs) have been shown to reduce or eliminate visual hallucinations. Some authors also suggest that social isolation may contribute to CBS, hypothesizing that increased social interaction, such as through local support groups, might reduce hallucinations (Carpenter et al. 2019, Pang 2016, Chen 2014). Therefore, it is essential to integrate education and counseling on visual hallucinations into elderly care, while also addressing social isolation and minimizing vision loss when possible (Carpenter et al. 2019, Pang 2016).

As illustrated by our case, a thorough investigation of etiology was crucial for diagnosing both bilateral NA-AION and CBS. Although it was not possible to effectively prevent involvement of the contralateral eye in our case, prompt diagnosis and management of blood pressure and anemia would likely be the only way to mitigate the risk of affecting the other eye. Despite its recognition in psychiatry, CBS is still significantly underrepresented in medical literature. Other clinicians should be adequately informed and sensitized to reassure and educate patients who develop CBS following vision loss about the benign nature of visual hallucinations.

CONCLUSIONS

Although CBS is well-recognized in psychiatry, there is a notable lack of awareness in other medical specialties. Raising awareness of CBS among clinicians, at-risk groups, and the public will likely improve self-reporting of symptoms, expanding the CBS cohort and facilitating large-scale studies to better understand the syndrome.

Our case underscores the importance of a multidisciplinary approach. Greater awareness of CBS among clinicians and at-risk groups will most likely improve the quality of care.

Further studies aimed at understanding the functional changes underlying CBS may pave the way for the development of effective pharmacological treatments.

ACKNOWLEDGMENTS

The author sincerely thanks Dra. Rita Couceiro for her pivotal role and guidance throughout my master's thesis, which served as the inspiration for this article.

Ethical Considerations: Does this study include human subjects? YES

Authors confirmed the compliance with all relevant ethical regulations.

Conflict of interest: 'No conflict of interest'

Funding sources: 'The authors received no funding from an external source.'

Authors cotributions: Daniela Jeremias: Study design, data collection, first draft, approval of the final version. Leonor Brito-Santana: Study design, first draft, approval of the final version. Bruno Vidal: First draft, approval of the final version. Bárbara Mesquita: First draft, approval of the final version. Mário Cunha: First draft, approval of the final version. All authors have read and approved the manuscript.

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