CASE REPORTS
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Complex visual hallucinations with retention of insight: four cases of Charles Bonnet syndrome

Složene optičke halucinacije kod bolesnika sa očuvanim uvidom: četiri slučaja Šarl Boneovog sindroma

Vesna Tepšić Ostojić*†, Zagorka Gojković*, Bratislav Živić*†

***Military Medical Academy, Psychiatric Clinic, Belgrade, Serbia; †University of Defence, Faculty of Medicine of the Military Medical Academy, Belgrade, Serbia**

Abstract

Introduction. Complex visual hallucinations with retention of insight due to visual impairment are key symptoms of Charles Bonnet syndrome. The syndrome is a standalone diagnosis in the International Classification of Diseases, 11th Revision. Nevertheless, in clinical praxis, it is often misdiagnosed as psychosis or early stages of dementia, and it goes underreported by patients because of the fear of being diagnosed with a mental illness. **Case report.** We presented four elderly patients, who were referred for psychiatric consultation due to visual hallucinations, with preserved insight, but with impaired vision. All four patients had complex, vivid, and colorful hallucinations consisting of realistic objects, people, animals, or scenery that tend to recur. Their emotional response and impact on quality of life differed, and psychopharmacotherapy was determined according to their psychological symptoms. Empathic explanation of the symptoms' origin and reassurance of the absence of mental illness for patients and caregivers were of vital importance in all cases. **Conclusion.** With the aging of the population, the number of patients with impaired vision also increases, and the importance of a multidisciplinary approach in the diagnostic procedures and treatment of Charles Bonnet syndrome is emphasized. Increased awareness of clinical characteristics and therapeutic approaches is required among all physicians who are in contact with elderly and/or impaired vision patients.

Key words:
charles bonnet syndrome; diagnosis, differential; hallucinations; vision disorders.

Apstrakt

Uvod. Složene optičke halucinacije sa očuvanim uvidom kod bolesnika sa oštećenjem vida predstavljaju ključni simptom Šarl Boneovog sidroma. Ovaj sindrom je uvršten u jedanaestu reviziju Međunarodne klasifikacije bolesti kao samostalna dijagnoza. Ipak, često se u kliničkoj praksi bolesnicima sa tim sindromom pogrešno dijagnostikuje psihoza ili početna faza demencije, a sindrom retko prijavljuju i bolesnici, zbog straha od postavljanja dijagnoze mentalnog oboljenja. **Prikaz bolesnika.** Prikazali smo četiri bolesnika starijeg životnog doba, koji su bili upućeni na psihijatrijsku konsultaciju zbog optičkih halucinacija, uz očuvani uvid, a sa oštećenjem vida. Sva četiri bolesnika imala su složene, žive halucinacije u boji, koje su predstavljale realne predmete, ljude, životinje i pejzaže i koje su se ponavljale. Emocionalni odgovor i uticaj na kvalitet života tih bolesnika bio je različit, a psihofarmakoterapija je bila određena u skladu sa kliničkom slikom. Pokazivanje empatije u objašnjavanju porekla simptoma i razuveravanje da se ne radi o psihijatrijskom poremećaju, bili su od velikog značaja kod svih bolesnika, kao i kod njima bliskih osoba. **Zaključak.** Starenjem populacije povećava se i broj bolesnika koji imaju oštećenje vida i naglašava se značaj multidsciplinarnog pristupa u dijagnostici i terapiji Šarl Boneovog sidroma. Potrebno je povećati svest o kliničkim karakteristikama i terapijskom pristupu kod svih lekara koji su u kontaktu sa starijim bolesnicima i/ili bolesnicima sa oštećenjem vida.

Ključne reči:
šarl boneov sindrom; dijagnoza, diferencijalna; halucinacije; vid, poremećaji.

Introduction

Charles Bonnet syndrome (CBS) is a medical condition named after a Swiss philosopher and naturalist, Charles Bonnet (1720–1792), who first documented the phenomenon of complex visual hallucinations in visually impaired patients with no mental illness in the 1760s [1]. Patognomonic CBS hallucinations are complex, vivid, and mostly colorful

Correspondence to: Vesna Tepšić Ostojić, Military Medical Academy, Psychiatric Clinic, Crnotravska 17, 11 000 Belgrade, Serbia.
E-mail: vesna.j.tepsic@gmail.com

and are comprised of realistic objects, people, animals, or scenery that tend to recur [2]. They occur involuntarily and retain insight that what is observed is not real. Hallucinations are rarely simple photopsias consisting of lights and flashes [3]. Hallucinatory content is often congruent with the emotional response and is usually not disturbing, but, in some cases, it can be anxiety-provoking and can affect the quality of life [4, 5]. They can manifest regardless of the level of the lesion of the visual system (eyes, optic nerve, or brain), but typically, there is a loss of central visual acuity [2]. Older age, social isolation, and sensory deprivation are the triads that are usually present in the CBS etiology [4, 6].

We present four cases of CBS that were referred for psychiatric consultation due to the presence of hallucinations. Permission was given from the patients, and the personal information was de-identified to protect their anonymity.

Case report

Case 1

A 64-year-old male patient diagnosed with diabetes mellitus at the age of 27 with multiple complications listed in his medical record, including diabetic neuropathy and retinopathy, was referred to psychiatric consultation after reporting visual hallucinations to his endocrinologist. Hallucinations started after a few days in the hospital and were recurring. He saw his patron saint, St. John, sitting on his bed, walking through the room from patient to patient, sometimes holding a candle or talking to the favorite female saint of the patient's wife, St. Petka. They were both smiling, with warmth in their eyes and grace in their movements. He described them as very pleasant and comforting. The patient had no previous history of psychiatric symptoms or treatment and had mild cognitive impairment. He had full insight into the quality of his visions. Even though he knew they were not real, he enjoyed the comfort they provided him in his sickness and loneliness. No psychopharmacotherapy was prescribed as he had full insight and was not afraid of them, and no follow-up was suggested.

Case 2

A 78-year-old female patient, hospitalized for hip surgery, presented with severe anxiety and sleep disturbance for the last several days and was alone in the room because of the COVID-19 preventive measures. Psychiatric evaluation showed no history of mental illness and no cognitive decline. Medical history showed she was diagnosed with hypertension and glaucoma, but no ophthalmological report was available. The patient reported disturbing images she had been seeing for the last few days. She described an ugly female person sitting on the window sill that scared her. That woman's head was shrinking and growing, sometimes looking familiar, but the patient was unable to recognize her. Sometimes the woman she saw had the head of an unpleasant insect, a wild beast, or even of a nurse that the patient did not like because she found her cruel and lazy. Occasionally, the same head would be there for several minutes, and at other times, different heads would change rapidly. The patient was terrified and angry and had a level of anxiety that interfered with all aspects of her functioning, including sleep. Since the detailed explanation of her visual experiences and reassurance showed no benefit, she was prescribed olanzapine 2.5 mg in a single evening dose during her stay in the hospital. Even though her hallucinations were still present, they were less frequent, and she did not report fear, anxiety, or sleep disturbance. She was advised to continue outpatient psychiatric follow-up.

Case 3

A 77-year-old female patient was referred by her general practitioner for outpatient psychiatric consultation accompanied by her daughter. She was a widow and had been living alone for two years; her two daughters and grandchildren visited often. The patient had severe rheumatoid arthritis with restricted mobility and hypertension. She had left eye cataract surgery six months ago and was listed for the right eye surgery as well, as explained by her daughter. She described her daily "afternoon experiences" that started a few weeks after the cataract surgery, including seeing very colorful bucolic scenery in her living room, plants with big leaves, flowers, butterflies, and small animals like rabbits, squirrels, bees, etc. It was like a magical garden with mixed colors – leaves were rarely green and instead were blue, white, etc. The flowers were of all colors, sometimes floating in the air so that animals could ride on them. Flowers and animals were often smiling or playing to entertain her. The patient understood her visual experience was not real and that there were hallucinations that did not frighten her, but she was afraid of going mad. Detailed explanations and reassurance were offered to her and her daughter, and the patient decided to calmly wait for the other eye surgery. Therefore, no psychopharmacotherapy was prescribed.

Case 4

An 84-year-old female patient came with two grandchildren, with whom she lived, to the Emergency Room after reporting seeing a little girl calmly sitting on the kitchen floor and playing with her doll. The little girl neither talked to her nor wanted anything from her. The patient had no emotional response to her vision whatsoever, but her grandchildren were afraid that this was a sign of dementia. Her grandchildren knew her eyesight was declining, and after explaining the probable nature of hallucinations, they were advised to make an appointment with an eye specialist, neurologist, and psychiatrist if necessary.

Discussion

Even though CBS was described 250 years ago, a search of the PubMed electronic database shows an upward CBS publication activity, with nearly 60% of publications

within the last ten years [4]. Unsurprisingly, the increase in the number of published papers coincides with the introduction of CBS as a standalone diagnosis by the World Health Organization in the International Classification of Diseases, 11th Revision (ICD-11). CBS is classified as a disease of the visual system as 9D56 Visual release hallucinations [7]. The CBS incidence is not easy to determine, but one can hypothesize that it is far more common than it is diagnosed. The case reports available from the literature propose that CBS is rarely recognized by clinicians and, therefore, misdiagnosed as psychosis or early stages of dementia [6, 8]. Furthermore, visual hallucinations are mostly not reported by the patients themselves because of the fear of "madness" or being diagnosed with a psychiatric disease [9]. Interestingly, the results of the Age-Related Eye Disease Study 2 Research Group [10] showed that when visual hallucinations are solicited in elderly patients with impaired sight, 11% to 15% of them reported experiencing visual hallucinations. The results of this study and by other authors also pointed out that the majority of patients did not report their symptoms to their doctor or family members [5, 10, 11]. We presented four cases that were referred to a liaison psychiatrist as a visit consultation. Two out of the four of our patients reported their hallucinations, and two were recognized by the hospital staff. CBS can be found at any age, but the the age of the vast majority of the diagnosed cases was between 70 and 85 years. Three out of the four presented patients are in this age group. CBS tends to be more frequent in the elderly, and important factors are the different conditions of visual impairment [3, 12].

Common etiologies, except for congenital blindness, include age-related macular degeneration, glaucoma, diabetic retinopathy, cataract, optic neuritis, visual cortex cerebral infarction, retinal vein, or arterial occlusion [3, 12, 13]. The etiology of presented cases is among the most common listed above. As liaison psychiatrists, we had no detailed medical data regarding the exact visual acuity and duration of eyesight impairment. A patient with no previous knowledge of eye disorder was referred to an eye specialist. The most accepted explanation for typical CBS hallucinations is that visual sensory deafferentation leads to disinhibitions of visual cortical regions [4, 14]. Hallucinations are, therefore, spontaneous activation of the disinhibited visual cortex. Neuroimaging supports that theory, showing that the visual cortex activates spontaneously during hallucinations [15]. Hallucinations can be defined as perceptions of stimuli in the absence of an external stimulus and, therefore, can be found in all sensory modalities. In this phenomenon, the hallucinations have been described as formed and vivid, of realistic objects or people, with a recurring tendency. Hallucinations described by our patients are typical for CBS. Given that our patients were from the liaison service, we had no information regarding the course of the CBS. In CBS, the patient retention of insight is present, and in most cases, they do not find these images disturbing. Over time, a decrease from 38% to 8% in emotional input described as disturbing, frightening, or horrifying was reported [14]. CBS hallucinations can affect the quality of life – 60% of patients did not experience any change, 33% had a negative impact, and 7% reported the hallucinations had a good effect [12]. Interestingly, the patient described in the first case finds his hallucinations comforting. Any patient with *de novo* hallucinations should undergo a detailed evaluation to rule out the causes. The patient should go through a detailed assessment by an ophthalmologist, neurologist, and psychiatrist. A differential diagnosis involving visual hallucinations is complex and can include neurodegenerative conditions such as Alzheimer's or Parkinson's disease, migraine aura, epileptic seizures, sensory and/or sleep deprivation, narcolepsy, hypnagogic hallucinations, metabolic encephalopathy, drugs and alcohol withdrawal, delirium, and psychosis [16]. Different common non-psychotropic medicines and over-the-counter medications (including ephedrine and synthetic cannabinoids) [16, 17], psychotropic medications and drugs of abuse must also be considered as the possible cause of hallucinations [16].

Treatment of CBS is challenging and should be planned for each individual separately. Optimizing visual functions at the maximum level possible and explaining that symptoms are not due to mental illness is crucial. The severity of symptoms is the part where therapy comes in. In most cases, clarification and reassurance with empathy are all that is needed, as was the case in three out of the four of our presented patients; only one patient required medication for intensive anxiety and sleep disturbance. In more severe cases, some kind of psychological support or treatment should be made necessary, along with medications. There is no causal medication for CBS. Atypical antipsychotics [16, 18] at low doses like olanzapine or quetiapine are the first choices because of their side effect profile, as well as cholinesterase inhibitors such as donepezil [16]. Other medications with positive effects in a modest series of patients include venlafaxine and escitalopram, as well as valproate and carbamazepine [18].

The prognosis of CBS is directly connected to the cause of vision impairment [15, 16]. The prognosis is far more promising in conditions where visual impairment can be corrected, like cataracts [15, 19], but also less favorable in those with continuous stable or progressing visual impairment [15, 16, 19].

Conclusion

We presented four cases of visual hallucinations with retained insight in elderly patients with impaired vision referred for psychiatric consultation. The aging of the population consequently increases the number of patients with impaired vision, which emphasizes the importance of the multidisciplinary approach to diagnostic procedures and treatment. CBS is a standalone diagnosis in ICD-11, but in clinical praxis, it is often misdiagnosed as psychosis or early stages of dementia and underreported by patients because of the fear of being diagnosed with a mental illness. Empathic reassurance and explanation for patients and caregivers are of vital importance.

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