DOI:10.4274/tnd.25986
Turk J Neurol 2016;22:184-187

Charles Bonnet Syndrome: Three Cases

Charles Bonnet Sendromu: Üç Olgu

Aygün Akbay Özşahin, Zeynep Çiğdem Diler, Sevinç Çelik, Gülay Kenangil, Füsun Domaç

Erenköy Mental Health and Neurological Diseases Training and Research Hospital, Clinic of Neurology, İstanbul, Turkey

Summary

Charles Bonnet syndrome (CBS) is characterized by the presence of visual hallucinations due to the decrease in visual acuity or loss of vision without any pschiatric problem. We report three patients of aged 73, 76, and 75 years with a severe loss of vision due to ocular diseases and visual hallucinations. Case 1 was a man aged 73 years who presented with visual hallucinations that completely recovered after cataract surgery. Case 2 was woman aged 75 years with severe visual impairment from end-stage age-related macular degeneration, thus visual hallucinations continued to persist. Case 3 was legally blind with CBS-type visual hallucinations and amnestic dementia syndrome. CBS may terminate spontaneously or continue for months or years. Ideally, the patient should be considered for ophthalmologic intervention. Although CBS is benign, the visual hallucinations can cause great anxiety for patients. Patients should be warned about the adverse effects of pharmacologic therapy. Cognitive decline may occur at early stages or later.

Keywords: Charles Bonnet syndrome, visual hallucination, dementia

Öz

Charles Bonnet sendromu (CBS) görme kaybı veya görme keskinliğinin azalmasıyla ortaya çıkan ve herhangi bir psikopatolojinin eşlik etmediği görsel halüsinasyonlarla giden sendromdur. CBS tanısı ile izlenen 73 ve 76 yaşında erkek ile 75 yaşında kadın üç ayrı olgunun klinik özellikleri incelenmiştir. Tipik CBS'ye ait görsel halüsinasyonları olan birinci olgu katarakt operasyonundan sonra tamamen düzelmiş, maküler dejenerasyonu olan ikinci olgunun şikayetleri devam etmiş ve tedaviye bağlı ekstrapiramidal yan etkiler gelişmiştir. Üçüncü olguda katarakt operasyonuna bağlı ileri derecede görme bozukluğu ve görsel halüsinasyonlarla birlikte amnestik demans tablosu da mevcuttur. CBS kendiliğinden iyileşebilir veya aylarca, yıllarca sürebilir. Görme bozukluğunun düzeltilebilmesi önemli bir tedavi aşamasıdır. İyi prognozlu kabul edilse de bazı hastalarda ciddi anksiyete yaratabilir. Hastalar tedavi arayışlarında yan etkiler konusunda da uyarılmalıdır. Bilişsel fonksiyonlar hastalığın başlangıcında veya ileri döneminde kısmen etkilenebilmektedir.

Anahtar Kelimeler: Charles Bonnet sendromu, görsel halüsinasyonlar, demans

Introduction

Charles Bonnet syndrome (CBS) is characterized by visual hallucinations without accompanying psychopathologies, in patients with visual loss or visual acuity loss due to any lesion of the eye, optic nerve, chiasm, optic tractus, optic radiation or visual cortex. The syndrome was first described by the Swiss philosopher and naturalist Charles Bonnet (1720-1793) in 1769 upon expression of live colored hallucinations by his grandfather Charles Lullin, who had ocular problems with intact cognitive function following a cataract operation. Charles Bonnet himself was also reported to experience similar visual hallucinations due to ocular problems at advanced age. Lullin's notebook was found in 1902 and published by Flournoy. In 1936, George de Morsier

Address for Correspondence/Yazışma Adresi: Aygün Akbay Özşahin MD, Erenköy Mental Health and Neurological Diseases Training and Research Hospital, Clinic of Neurology, İstanbul, Turkey
Phone: +90 533 224 10 99 E-mail: ayguun@hotmail.com
Received/Geliş Tarihi: 07.09.2015 **Accepted/Kabul Tarihi:** 26.11.2015

named this syndrome as CBS (1). Hallucinations might be simple or complex, or start as simple and become complex. They might either be periodic or continuous, as well as being well organized and clearly distinguishable. Hallucinations might occur in the form of photopsia, the perception of seeing light, geometric figures, motifs, well-formed or half-formed faces, people, animals, landscape, vehicles, buildings, or Lilliputian (tiny people) images. The hallucinations occur while the patient is conscious and they are out of the patient's control. Hallucinations occur as blurred images and can be differentiated from normal vision. The clinical picture often develops in acquired visual impairments. The most commonly reported ocular pathology is age-related macular degeneration. Light flashes might be colored, particularly in senile macular degeneration, or black and white in glaucoma or diabetic retinopathies. Hallucinations might be colored, constant, mobile, or mobile within the visual field. Sometimes the same hallucination might repeat in a single patient. Hallucinations occur more commonly on a white background, wall, or a piece of paper. The duration to develop hallucinations might range from a few hours or days in acute events including optic neuritis, stroke, or surgery, to a year in chronic ocular diseases. Although bilateral loss of visual acuity might be a trigger, the phenomenology of visual hallucinations often does not correlate with the underlying ocular disease. Also, the pathology has rarely been reported in patients with unilateral loss of vision. Patients might not necessarily have a wide deficit in visual field; bilaterally impaired vision, reduced visual acuity, cerebral injury, cognitive deficits, social isolation and lack of stimulation constitute the strongest risk factors. There is no consensus on the threshold value for visual acuity. In 1995, Teunisse et al. (2,3,4) reported that visual acuity of the better eye less than 0.3 constitutes a risk factor for the disorder. Trigger factors might include fatigue, lighting on low surfaces, bright lights, and stress. The prevalence rate has been reported as 10-60%; and no significant relationship has been established with the presence of CBS and age, sex, or underlying ocular pathologies. Nevertheless, the majority of patients' ages range between 70-85 years. There are contradictory reports of male or female predominance of the disorder (5). There are no defined diagnostic criteria; however, the recommended diagnostic criteria include acquired visual impairment, at least one complex visual hallucination in the last four weeks, a minimum of a 4-week interval between the first and last hallucinations, complete or partial insight with regard to the unreality of hallucinations, absence of hallucinations in other sensory modalities, and absence of delusions (5,6). Mild cognitive impairment is thought to be a predisposing factor. Overall, hallucinations tend to occur on the side of visual field loss. Additionally, elementary hallucinations might be isolated or accompanied by complex hallucinations, despite not being listed among the recommended diagnostic criteria (7).

In this manuscript, our objective was to present three cases demonstrative of different aspects of CBS.

Case Reports

Case 1

A man aged 73 years presented to our outpatients clinic claiming to have seen strangers inside his house, children, and guards in uniforms. He had no cognitive impairments other than mild memory problems that did not affect daily life. Cranial magnetic resonance imaging (CMRI) and electroencephalography (EEG) were insignificant. Ocular examination revealed that visual acuity was 0.4 on the right and 0.5 on the left, and there was bilateral corticonuclear cataract and age-related fundus degeneration. Hallucinations disappeared completely following cataract operation of both eyes.

Case 2

A right-handed woman aged 75 years presented to our outpatients clinic and reported seeing strangers inside her house, her late husband, faces on the curtains or walls, and ox head blasting out of television. The patient was aware of the fact that these images were unreal, and afraid of becoming mentally ill. She also described praying against these potential demons. She lived with her daughter, who was a healthcare worker, and no pathologies were found in her CMRI, EEG, and sleep EEG. Her daughter reported that she too felt nervous watching her mother constantly staring at walls or curtains; she knew that her mother had preexisting visual impairment due to macular degeneration. The patient had quite an apprehensive premorbid personality and had long since feared being close to cemeteries in fear of demons or spirits. Her neurologic examination was normal and the patient was able to count fingers at four-meter distance. Both the patient and her daughter were informed of CBS and treatment with quetiapine 25 mg/day was recommended. However, the patient felt utterly stricken by the hallucinations and presented to other centers in search of a solution and was prescribed risperidone. The latter treatment provided a benefit for a while, but six months later the patient was referred back to our center with the onset of Parkinsonism. Risperidone therapy was ceased and extended-release quetiapine 50 mg/day was recommended. Also gabapentin, and various selective serotonin reuptake inhibitor (SSRI) therapies were tried as recommended in literature; however, the patient benefited from none and continues to suffer from the symptoms.

Case 3

A right-handed man aged 76 years presented to our outpatients clinic with symptoms of amnesia, nervousness, hallucinations, and a behavior disorder. He was reported to have significantly diminished vision following a cataract operation. He had been suffering from hallucinations, harming both himself and others because he believed the dreams to be real, nervousness, and behavior impairment for one and a half years. He reported seeing doors on walls, cars and trees inside the house and falling off the chairs or tables while trying to reach them, hitting tables or chairs in an attempt to fix them because they appeared bent, or breaking the wall to get through door images on the wall. His neurologic examination revealed an intact person and place orientation, and partially preserved time orientation. Also, no motor deficits were determined or extrapyramidal signs were observed. He scored 19 out of 28 in the mini mental test due to severely impaired vision. Other tests could not be performed because he was illiterate. Computed brain tomography showed cortical atrophy, increased third and lateral ventricle sizes, and hypodense ischemic gliotic foci in periventricular white matter. EEG proved normal. The patient lived in next door to his daughter and was able to pursue his daily living partially on his own and do basic shopping in the

neighborhood. He was able to perform most daily living activities of unaided. He accepted that these hallucinations were partially imaginary, but also thought that some were real.

Discussion

Visual hallucinations might be caused by diverse etiologic causes. Other than CBS, potential causes include dementia syndromes and various degenerative diseases, hemispheric or brain stem lesions, metabolic disorders, drug use or substance abuse. Patients' medical history, phenomenology of the hallucinations and their relationship with sleep, as well as concurrent findings are of utmost significance. An ophthalmologic examination, laboratory tests for a metabolic analysis, and cranial imaging should be performed. In our series, the first two cases conformed to the CBS criteria with the presence of complex visual hallucinations, impaired vision, and preservation of insight. In case 1, a complete correction of vision with a cataract operation ended the hallucinations, in line with typical CBS. However, case 2 continues to see images both due to the ocular disorder that cannot be completely cured and because of her pre-existing apprehensive personality. Hallucinations of CBS have been reported to amuse patients; however, it is common for these hallucinations to cause restlessness and anxiety. They might affect the quality of life negatively, although insight has been preserved (6). Cox and Ffytche (2) examined patients with macular disorders and reported that CBS hallucinations did not affect the daily living in 60% of patients, 7% of the patients found the hallucinations amusing, and 32% reported being negatively affected. Studies have demonstrated that the negative effect was associated with the fear factor, frequency and prolongation of hallucinations, not with the sex or ophthalmologic disorder of the patients. It has also been shown that hallucinations might last for five years or more in 75% of patients. Patient's education level, cultural and religious beliefs, and the presence of mild cognitive impairment also affect how hallucinations are interpreted. In case 2, we could not create adequate relaxation with persuasion of the benign nature of the symptoms, and recommended supportive methods regarding lighting and stimulation.

Both the content of hallucinations and the accompanying impairment of vision were in line with CBS hallucinations in case 3, despite the amnestic dementia picture. Visual hallucinations of Alzheimer dementia are often seen in moderate and severe disease stages, but rarely in early stages. Patients do not describe very definite images. However, hallucinations with a live character might be seen in the early stages of Lewy body dementia (LBD). Our case conformed to Alzheimer's dementia rather than LBD due to the presence of Parkinsonism and absence of significant impairment in visual-spatial abilities. Terao ve Collinson (8) reported that patients followed up with CBS might later be diagnosed as having LBD, or the vice versa might be valid-patients with LBD and complex visual hallucinations might later be diagnosed as having CBS because of the absence of accompanying delusion or other hallucinations. Authors have suggested that both the presence and absence of insight into the visual hallucinations should be interpreted within the same clinical spectrum, and elderly patients who are diagnosed as having CBS should also be checked for potential LBD in the follow-up. Pliskin et al. (9) proposed the potential indicator role of CBS for dementia in the early stages. Also, Menon et al. (5) reported that insight might not be complete in all the cases and that some patients have partial or fluctuating insight. Our patient with partial insight was interpreted as having CBS and early Alzheimer's dementia owing to the presence of visual impairment and content of hallucinations; however, the potential to develop LBD was also kept in mind in the follow-up.

Two different mechanisms have been recommended to explain visual hallucinations in CBS. The first one is the release phenomenon, which suggests that diminished visual perception due to problems of vision results in reduced suppression by higher cortical centers and emergence of endogenous activity of visual cortex. Hallucinations might even be triggered by eye closure for a few days. Diminished sensory stimuli and/or vision-stimulated intracerebral perceptions similar to phantom pain syndrome result in "phantom visions". The second hypothesis is the irritative foci theory. According to this, "deafferentation-situation" theory, visual sensory stimuli diminish due to visual impairment, which leads to increased spontaneous discharges of neurons at visual association cortex, and thus visual hallucinations (3,5). Most patients with CBS are in social isolation, which further supports the visual sensory loss phenomenon. Colored hallucinations seen in patients with senile macular degeneration also supports the deafferentation theory. Color vision is impaired in the early period in senile macular degeneration due to the macular localization and early impairment of coni cells responsible of colored vision. Therefore, colored visual hallucinations often seen in senile macular degeneration have been associated with localized hyperexcitability due to selective deafferentation in the colored visual field. Ffytche et al. (10) asked four patients with CBS to describe their visual hallucination experiences and performed functional MRI studies during hallucinations, which revealed increased cerebral activity at ventral extrastriate cortex. Tonic increase in brain activity was also observed in specialized visual cortex at non-hallucinated times in patients with CBS compared with control patients of matching ages with similar visual problems. Together, these results demonstrated that each specialized visual cortical area led to visual hallucinations associated with its own localization, and the pathophysiology of hallucinations might be related with localized increase in cerebral activity. Ganglion cells are significantly reduced in patients with severe glaucoma, which results in physiologic deafferentation without altering visual acuity. However, in age-related macular degeneration, the central retinal ganglion cell loss and diminished visual acuity leads to CBS. Consequently, Madill and Ffytche (7) suggested that CBS should also be considered in ocular pathologies with deafferentation in the absence of reduction in visual acuity.

There are no widely accepted treatment agents for CBS. Isolated case reports have described benefit with various anti-psychotics including atypical antipsychotics and anti-convulsives including carbamazepine, valproate, clonazepam, gabapentin, pregabalin, cicapride, and SSRIs (11). Anti-convulsives are thought to be effective by diminishing the abnormal neuronal excitations produced by the release mechanism. Gabapentin at a daily dosage of 300 mg might improve phantom vision in the same way it improves phantom pain (12).

CBS might resolve spontaneously or last for years. Generally, visual hallucinations disappear in patients with complete improvement of visual loss. However, symptoms often persist in patients without complete improvement including those with

macular degeneration. Supportive devices aimed at patients with diminished visual capacity, improved social environment, increased lighting and music are recommended in adjunctive therapy. Patients should be warned about the adverse effects they might encounter in their search for 'better' treatment. CBS support groups have been founded in other countries. Patients might be also be recommended to try eye closure and opening, rapid up-down movements, forward-backward movements, gaze fixation, walking towards or away from the image, touching, hitting, calling out the image, or turning the lights on, and focusing on something else (13). It is also important to obtain a detailed anamnesis in an emphatic manner and inform the patients of the benign nature of the condition.

Ethics

Informed Consent: Consent form was filled out by all participants.

Peer-review: Externally and internally peer-reviewed.

Authorship Contributions

Concept: Aygün Akbay Özşahin, Design: Aygün Akbay Özşahin, Zeynep Çiğdem Diler, Data Collection or Processing: Aygün Akbay Özşahin, Sevinç Çelik, Gülay Kenangil, Füsun Domaç, Analysis or Interpretation: Aygün Akbay Özşahin, Sevinç Çelik, Literature Search: Aygün Akbay Özşahin, Writing: Aygün Akbay Özşahin, Zeynep Çiğdem Diler.

Conflict of Interest: No conflict of interest was declared by the authors.

Financial Disclosure: The authors declared that this study received no financial support.

References

1. Draaisma D. Aklın Çıkmazları. Yapı Kredi Yayınları: 2012;17-44.
2. Cox TM, Ffytche DH. Negative outcome Charles Bonnet syndrome. Br J Ophthalmol 2014;98:1236-1239.
3. Güngör Ö, Kalaycı D, Şahin M, Orhan N, Hasıripi H. Bilateral anterior iskemik optik nöroopati ile ilişkili Charles Bonnet sendromu. Ret-Vit 2005;13:157-159.
4. Teunisse RJ, Cruysberg JR, Verbeek A, Zitman FG. The Charles Bonnet syndrome: a large prospective study in The Netherlands. A study of the prevalence of the Charles Bonnet syndrome and associated factors in 500 patients attending the University Department of Ophthalmology at Nijmegen. Br J Psychiatry 1995;166:254-257.
5. Menon GJ, Rahman I, Menon SJ, Dutton GN. Complex visual hallucinations in the visually impaired: the Charles Bonnet Syndrome. Surv Ophthalmol 2003;48:58-72.
6. Jan T, Del Castillo J. Visual hallucinations: charles bonnet syndrome. West J Emerg Med 2012;13:544-547.
7. Madill SA, Ffytche DH. Charles Bonnet syndrome in patients with glaucoma and good acuity. Br J Ophthalmol 2005;89:785-786.
8. Terao T, Collinson S. Charles Bonnet syndrome and dementia. Lancet 2000;355:2168.
9. Pliskin NH, Kiolbasa TA, Towle VL, Pankow L, Ernest JT, Noronha A, Luchins DJ. Charles Bonnet syndrome: an early marker for dementia? J Am Geriatr Soc 1996;44:1055-1061.
10. Ffytche DH, Howard RJ, Brammer MJ, David A, Woodruff P, Williams S. The anatomy of conscious vision: an fMRI study of visual hallucinations. Nat Neurosci 1998;1:738-742.
11. Jackson ML, Ferencz J. Cases: Charles Bonnet syndrome: visual loss and hallucinations. CMAJ 2009;181:175-176.
12. Paulig M, Mentrup H. Charles Bonnet's syndrome: complete remission of complex visual hallucinations treated by gabapentin. J Neurol Neurosurg Psychiatry 2001;70:813-814.
13. Pang L. Visual Hallucinations: Identifying Charles Bonnet syndrome. Int J Ophthalmol Eye Res 2015;4:14-22.