



Charles Bonnet syndrome

Šarl Boneov sindrom

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Abstract

Introduction. Charles Bonnet syndrome (CBS) is a condition that causes visual hallucinations in patients without any mental illnesses. CBS is characterized by the presence of vivid, complex and recurrent visual hallucinations, and do not occur in the setting or as part of delirium or other psychological illnesses. The condition is present in patients who have visual loss due to age-related macular degeneration (AMD), cataracts and/or other ocular diseases that influence vision. **Case report.** A 81-year-old woman reported to ophthalmologist complaining of visual hallucinations that consisted of white pigeons. Hallucinations were present for two years and she was well aware that hallucinations were unreal. Mental illnesses were excluded by the psychiatrist. Complete ophthalmologic examination was performed, and finding revealed visual acuity of 0.3 (right eye) and 0.5 (left eye), in both eyes cataracts and AMD (wet form). Optical coherence tomography confirmed the fundoscopic finding of AMD. The patient rejected treatment of cataracts and AMD due to old age, and hallucinations persisted. **Conclusion.** CBS should be considered in patients with visual hallucinations and ocular diseases that influence vision. It is essential to distinguish CBS from mental illnesses, since patients with CBS are fully aware that hallucinations are not real. Awareness of CBS could help physicians upon referring patients to ophthalmologists instead of psychiatrists, and therefore avoid patients being misdiagnosed.

Key words:

hallucinations; vision, ocular; macular degeneration; diagnostic techniques and procedures; diagnosis, differential.

Apstrakt

Uvod. Šarl Bonetov sindrom (CBS) je stanje koje izaziva vizuelne halucinacije kod bolesnika koji nemaju psihička oboljenja. CBS se ogleda u pojavi živopisnih i kompleksnih vizuelnih halucinacija koje se ponavljaju, ali se ne javljaju tokom delirijuma ili drugih psihičkih oboljenja. Stanje se javlja kod bolesnika koji imaju smanjenu vidnu oštrinu zbog senilne degeneracije makule, katarakte i/ili drugih očnih bolesti koje utiču na vid. **Prikaz bolesnika.** Žena, stara 81 godinu, javila se oftalmologu zbog vizuelnih halucinacija koje su činili beli golubovi. Halucinacije su bile prisutne tokom dve godine i bolesnica je bila svesna da one nisu stvarne. Pregledom psihijatra isključena su psihička oboljenja. Urađen je kompletan oftalmološki pregled i nalaz je pokazao vidnu oštrinu 0,3 (desno oko) i 0,5 (levo oko), obotranu kataraktu i senilnu degeneraciju makule (vlažna forma). Optička koherentna tomografija potvrdila je nalaz senilne degeneracije makule. Bolesnica je odbila lečenje katarakte i senilne degeneracije makule zbog starosti, a halucinacije su ostale prisutne. **Zaključak.** Kod bolesnika sa vizuelnim halucinacijama i očnim bolestima koje utiču na vidnu oštrinu, trebalo bi razmotriti postojanje CBS. Od ključne važnosti je razdvojiti CBS od psihičkih oboljenja, jer su bolesnici sa CBS u potpunosti svesni da halucinacije nisu stvarne. Saznanje o postojanju CBS može pomoći lekarima prilikom upućivanja bolesnika oftalmologu, a ne psihijatru, i tako izbeći postavljanje pogrešne dijagnoze.

Ključne reči:

halucinacije; vid; žuta mrlja, degeneracija; dijagnostičke tehnike i procedure; dijagnoza, diferencijalna.

Introduction

Charles Bonnet syndrome (CBS) is an uncommon condition causing visual nontreatening hallucinations in patients without any mental illnesses. It is characterized by the presence of vivid, complex and recurrent visual hallucinations occurring in psychologically normal patients and not in the

setting of delirium¹. Charles Bonnet, a Swiss philosopher, first described the syndrome in 1760 in a publication describing visual hallucinations experienced by his 90-year-old grandfather who was blind secondary to cataracts².

Although best described in age-related macular degeneration (AMD), CBS phenomenon may occur in any condition causing vision loss³. The symptoms of CBS vary from seeing

geometric figures to experiences of seeing people or animals⁴. Even though the content of the hallucinations in CBS is generally not distressing to the patient, they may cause fear of impending insanity⁵. The prevalence of CBS has been reported to vary from less than 1% to 40% in different populations, but it is likely that this variation is because of differences in inclusion criteria, inconsistent depth of questioning and reluctance of patients to admit to having hallucinations¹. Only about 1/5 of the patients admit to have told others about their symptoms⁶.

Case report

An 81-year-old woman reported hallucinations during past 2 years. Purely visual hallucinations consisted of white pigeons – walking through the room, sitting on TV set or flying around. At first hallucinations were not upsetting, but later the patient was frightened for her health, since she was aware that the hallucinations were not real. Pigeon hallucinations were mostly appearing in early evening hours and lasted for few minutes. She was not able to specify triggering factors of hallucinations appearance nor their resolution. In the beginning, hallucinations were present approximately few times *per* month, but later they occurred more often, almost on a daily basis. Hallucinations were visual only, she could not feel or hear the manifestations. The patient denied having any other visual problems. The husband of the patient explained that during these hallucinations no behavioral changes were present,

and that she was fully aware of hallucinations and could exactly finger-point where the pigeons were.

The past medical history included arterial hypertension and hip replacement. On physical examination she appeared well, oriented, blood pressure 140/85 mmHg, pulse 84 beats/minute, regular respirations, complete blood count was normal, glucose level of 6.1 mmol/L. The patient was using three antihypertensive drugs and, seldom, painkillers. She was also examined by a psychiatrist, and finding showed no mental pathological changes. Head magnetic resonance imaging (MRI) was done. A finding excluded brain tumor, and only mild cortical atrophy was confirmed.

The ophthalmic medical history showed a previous eyes examination three years before. The findings were: best corrected visual acuity (BCVA) 0.9 in both eyes, cataracts, intraocular pressure (IOP) of 15 mmHg, AMD (dry form), hypertonic fundus gr. II, cup to disc ratio (CDR) of 0.6, but there was no record of optical coherence tomography (OCT) nor Amsler grid testing. Current ophthalmic exam revealed: BCVA of 0.3 (right eye) and 0.5 (left eye), normal pupillary reaction and eye motility, nucleocortical cataracts, IOP of 16 mmHg; fundoscopic findings included: AMD (wet form), hypertonic fundus gr. II, CDR of 0.6; computed visual field analysis showed no glaucomatous nor vascular defects. OCT was performed, and AMD (wet form) was confirmed in both eyes (Figures 1 and 2). Therefore, Amsler grid testing was also done, and showed defects, more apparent in the right eye.

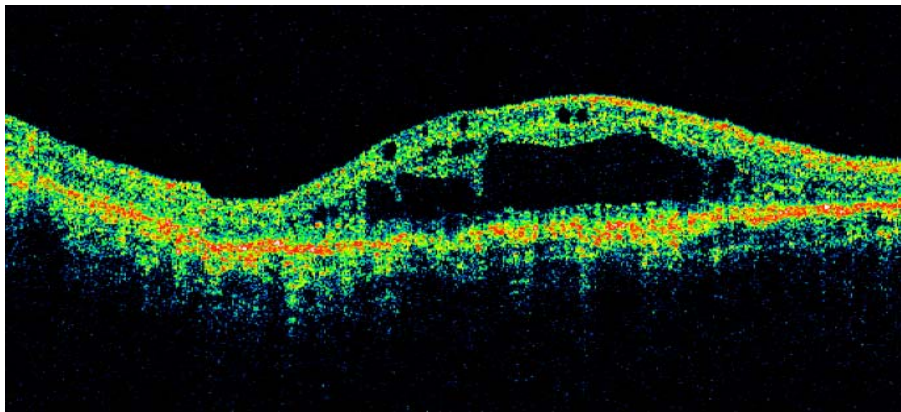


Fig. 1 – Optical coherence tomography finding (right eye): age-related macular degeneration (wet form) – severe pathological changes.

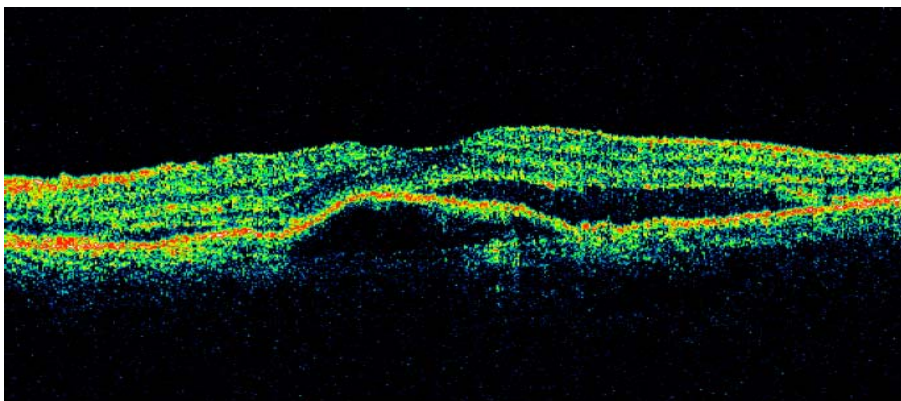


Fig. 2 – Optical coherence tomography finding (left eye): age-related macular degeneration (wet form) pathological changes.

After ophthalmological examination the patient was advised to treat cataracts and AMD, but she rejected to do so due to old age. Follow-up was done in three months. At the follow-up exam ophthalmological status was unchanged, and hallucinations persisted.

Discussion

Increasing life expectancy in population, many of whom show significant visual loss secondary to AMD or glaucoma, is causing an increase in the incidence of CBS⁷. Visual hallucinations in CBS patients are simple or complex in nature, and may be composed of photopsias, simple shapes, grid-like and branching patterns or may be presented by complicated images of people, faces, animals, flowers⁸. The majority of visual hallucinations are strange or bizarre to patients, but are seldom distressing, and do not involve hallucinations in other sensory modalities^{2,9}.

The true mechanism in the formation of visual hallucinations in CBS is not exactly known^{2,10}. There are a few theories explaining CBS.

The theory of deafferentation is a hypothesis, accepted most commonly to explain CBS. Deafferentation means the loss of visual input into the brain, which leads to change in the excitability of the visual association cortex¹¹. When the sensory visual input into the cortex is removed (with ocular pathology or visual pathway damage) spontaneous neuronal discharge in the visual association cortex occurs, increasing the excitability within the visual association cortex, resulting in visual hallucinations^{2,9}.

CBS has been associated with social isolation, cognitive defects, sensory deprivation, as well as low-quality social interaction⁵. Santos-Bueso et al.⁷ have concluded that visual loss and advanced age are the two main triggering factors in CBS. Visual deficits are generally the result of macular degeneration, cataracts, glaucoma or diabetic retinopathy. Although macular degeneration is most commonly associated with visual impairment, any condition that diminishes vision can cause CBS, but little or no correlation has been confirmed between the severity of visual impairment and prevalence of CBS⁵. The majority of patients with CBS are elderly (70–85 years), but cases have been reported in all age groups¹². In our patient, the possible trigger for visual hallucinations, besides age, could be the moment of transition of AMD from dry to wet form, along with progressing cataracts, and together causing vision reduction. Unfortunately, we have no previous OCT recordings to confirm our hypothesis.

It's been well documented that many drugs can be associated with visual hallucinations, such as antibiotics, anticonvulsants, antidepressants, antiparkinsonian drugs, hallucinogenes, stimulants, hormonal and cardiovascular drugs¹³. Of cardiovascular drugs, adverse effects of commonly used angiotensin-converting enzyme (ACE) inhibitors are numerous.

Doane and Stults¹⁴ have reported that in older patients visual hallucinations are very common side effect of ACE inhibitors. The presented patient was also treated for hypertension, but ACE inhibitors were not included in the therapy.

Diagnostic work-up with a patient suspected to have CBS should include a calm, nonjudgmental approach, since clinicians who demonstrate lack of awareness can cause further distress in patient. A full general physical examination should be conducted, with the focus on excluding acute illness as a cause of possible delirium. It is very important to perform ophthalmologic examination¹⁵. If a patient has an ocular disease that the diminishes vision, and reports visual hallucinations, but is aware that these hallucinations are not real, it is most likely that diagnosis is CBS. In unclear cases it is highly recommended to consult a psychiatrist.

The differential diagnosis that should be considered in patients with visual hallucinations are, among others, Lhermitte's hallucinosis (LH), Parkinson's disease and Lewy body dementia (LBD). In LH visual hallucinations are also vivid, well formed, and recognized as unreal, but in contrast to CBS, LH is associated with dementia, lesions and midbrain infarcts, while in Parkinson's disease and LBD, deficits in higher visual cortex may be present^{12,13}.

Currently, there is no universally accepted treatment for CBS. Visual hallucinations often resolve once the underlying cause of vision loss is treated, therefore in CBS confirmed patients with diminished vision, the easiest therapeutic approach is to improve patients' vision. Such measures might include improving lighting at home, to wear glasses or contact lenses, remove cataracts and treat any other eye condition that could improve vision¹³. The hallucinations associated with CBS may diminish and/or resolve as vision worsens or completely fades⁵. Roever et al.¹⁵ recommend increased social contact with patients having visual hallucinations. Pharmacological treatments are largely reserved for patients truly distressed by the hallucinations. Medications are only modestly effective, and they only attenuate the patient's emotional reaction and fail to eliminate the hallucinations^{5,10}. Antipsychotics have shown benefit in individual patients, but there is little evidence to support this therapeutic approach in CBS, since there is a high risk of side effects¹⁶.

Conclusion

Charles Bonnet syndrome should be considered in patients with visual hallucinations and ocular diseases that diminish vision. It is very important to distinguish this syndrome from mental illnesses and delirium, since CBS patients are fully aware that visual hallucinations are not real. Increased awareness of CBS can help physicians in referring the patient to ophthalmologists instead of psychiatrists, and doing so, avoid patients being misdiagnosed.

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