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CASE REPORT

CHARLES BONNET SYNDROME IN FUNGAL KERATITIS AND AGE-RELATED MACULAR DEGENERATION: A CASE REPORT

Abstract

The primary symptom of Charles Bonnet Syndrome is the occurrence of chronic visual hallucinations, which are observed in patients belonging to the geriatric population who had recently experienced a significant loss of vision. Charles Bonnet Syndrome is under-recognised owing to its low awareness among clinicians. We report a case of a 78-year-old man with blindness due to keratitis and age-related macular degeneration brought for a psychiatric consultation after the onset of visual hallucinations. Black insects playing on the patient's body along with visuals of the meals others could not see characterised the patient's hallucinations. Physicians are expected to have substantial knowledge of Charles Bonnet Syndrome for its correct diagnosis and management.

Keywords: Charles Bonnet Syndrome; Visual hallucinations; Keratitis; Age-related macular degeneration; Vision disorders.

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OLGU SUNUMU

FUNGAL KERATIT VE YAŞA BAĞLI MAKULA DEJENERASYONUNDA GELİŞEN CHARLES BONNET SENDROMU: BIR OLGU SUNUMU

Öz

Charles Bonnet Sendromu ileri derecede görme kaybı yaşayan özellikle geriatrik hasta popülasyonunda sıklıkla kronik görsel halüsinasyonların ön planda olduğu, çeşitli halüsinasyonlar ile karakterize bir klinik tablodur. Charles Bonnet Sendromu klinisyenler arasında hastalığın farkındalığının ve tanınırlığının az olması nedeniyle az tanı veya yanlış tanı almaktadır. Burada yaşa bağlı makula dejenerasyonu ve fungal keratit tablolarının birlikte olması ile giden ve ciddi görme kaybı yaşaması sonrası yemeklerinin üzerinde gördüğü, diğer insanların görmediği siyah böceklerle karakterize görsel halüsinasyonları nedeniyle psikiyatri bölümünden konsültasyon istenen 78 yaşında herhangi bir nörolojik hastalığı olmayan bir erkek olgu sunulmuştur. Hekimlerin Charles Bonnet Sendromu konusunda bilgi sahibi olmaları ve bu konuda farkındalığı artırmak amaçlanmaktadır.

Anahtar sözcükler: Charles Bonnet Sendromu; Görsel halüsinasyonlar; Keratit; Yaşa bağlı makula dejenerasyonu; Görme bozuklukları.

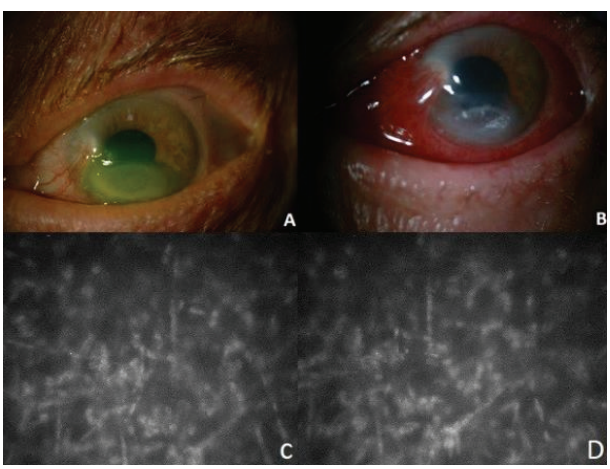
INTRODUCTION

The primary symptom of Charles Bonnet syndrome (CBS) is the occurrence of chronic visual hallucinations, which are observed in patients of the geriatric population who had recently experienced a considerable loss of vision (1). CBS has been primarily observed in patients with an ophthalmic disease; however, stroke patients with hemianopia have also reported its occurrence (2, 3). CBS was first described by Charles Bonnet in 1769, when his grandfather experienced visual hallucinations after the loss of vision due to cataract (3). Bonnet documented the complex visual hallucinations experienced by his grandfather, Charles Lullin and published his findings in a report (3). The condition was later termed as CBS. Ophthalmologists and neurologists use the term CBS to describe visual hallucinations occurring due to an ocular disease or a visual pathway disease.

CASE

We report a case of a 78-year-old man with blindness due to fungal keratitis and age-related macular degeneration brought for a psychiatric consultation after the onset of visual hallucinations. The best-corrected visual acuity observed after the ophthalmologic examination was 0.1 on the right eye and 0.05 with Snellen on the left eye. Upon the examination of the biomicroscopic anterior segment, the right cornea was normal but the slit-lamp examination revealed a corneal epithelial defect of 4×4 mm with infiltrate, and the inferior peripheral cornea on the left side was covered due to superficial corneal neovascularisation (Fig. 1A). Corneal scrapings were sent for cultures, such as for bacteria and fungi. The constructed in vivo confocal microscopy (confoscan 3.4, Nidek Co. Ltd., Gamagori, Japan) revealed fungal filaments in the stroma deep (Fig.1C, 1D). The patient was initially administered with topical voriconazole, ciprofloxacin and sefalosporine on an hourly basis. The corneal culture grew nothing prominent. On the follow-up visit, the cornea was found to be cleared (Fig. 1B); hence, there was a slight deterioration in the patient's condition. Since he was diagnosed with zona four months ago, amniotic membrane transplantation was performed to promote re-epithelialisation due to neurotrophic cornea. The right eye had atrophic age-related macular degeneration, which was observed in the fundus examination. Black insects playing on the patient's body along with visuals of the meals others could not see characterised the patient's hallucinations. During the time when the patient was brought for psychiatric consultation, he was having subjective visual experiences for about one week with the conditions worsening progressively. It started two weeks after the keratitis treatment. The symptoms gradually became worse. As the symptoms progressed, he became agitated. During the first visit, he was fully conscious, alert and oriented to the voices of his accompanied relatives. He was cooperative, and his speech was appropriate. His recent and memories were intact. Cognitive assessment was performed by using Mini-Mental Status Examination, on which he scored 18/20, with an inability to assess some of the domains, such as orientation, naming, reading,

Figure 1. Fungal keratitis in the left eye A. Before the antimicrobial therapy, B. On the follow-up visit, C, D. Fungal filaments in the corneal stroma with IVCM.



writing and construction, due to his blindness. The patient missed one point on recall and another point of attention/concentration. His neurological examination showed no acute intracranial findings and was negative for any acute changes. CBS was confirmed, and differential diagnosis was performed. Supportive care was provided, and the patient was discharged for home.

CONCLUSION

The purpose of this case report is to examine the characteristics of CBS with ophthalmic diseases. The prevalence of CBS has different variabilities and ranges from 0.4% to 30% (4, 5). Khan et al described the highest prevalence of CBS in their study (4). Ophthalmologists and neurologists use the term CBS to describe the visual hallucinations occurring due to an ocular disease or a visual pathway disease. In our case report, the patient had visual hallucinations; however, hearing or smelling hallucinations with CBS have also been reported. Vale CT et al presented eight patients with CBS and visual hallucinations. Aetiologies of these patents were severe glaucoma, optic neuropathy and age-related macular degenerations (3). Leandro JE et al calculated and found an increased prevalence of CBS in patients with the age-related macular degenerations (5). However, in our study, our patient was also diagnosed with fungal keratitis, and this diagnosis is uncommon in the literature. Diagnostic criteria for CBS is still not clear (6). The currently accepted theory propounds that vision loss leads to visual sensory deafferentation of the visual association cortex, further raising disinhibition and later spontaneous alerting of the visual cortical areas (3, 4). The brain activity in the absence of visual input has been compared with what occurs in phantom pain syndromes (7). Another theory is the release phenomenon, where the lost input to the primary visual areas suggests a disinhibition of visual association regions, further contributing to a release of visual hallucinations (6). Among the pathological causes, hepatic disorders, toxic-metabolic reasons, uraemia, encephalopathy associated with cardiac insufficiency, endocrine disorders, vitamin deficiency, inflammatory and infectious diseases must be considered and differentiated accordingly. All patients should be subjected to neurologic examination and brain MRI for organic reasons. The visual hallucinations of patients were revealed as either simple geometric patterns or complex recognisable shapes such as pictures or faces. Anticonvulsants, antidepressants, neuroleptic and cholinesterase inhibitors have been tried with positive and convenient results. CBS diagnostic criteria include full or partial retention of sight into the unreal nature of the hallucinations, the presence of formed, complex, persistent or repetitive stereotyped visual hallucinations, the absence of hallucinations in other sensory modalities, and the absence of primary or secondary delusions (6). CBS is under-recognised owing to the low awareness among clinicians. The symptoms can persist for years. We must rule out any other pathological causes and referral to psychiatrist for the evaluation of the cognitive function. Therefore, the substantial knowledge of CBS allows for its correct diagnosis and management.

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