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Charles Bonnet Syndrome with Atypical Features: Case Report

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Case Report
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Abstract

Charles Bonnet syndrome (CBS) is a lesser-known condition characterized by recurring or persistent complex visual hallucinations (VHs) that emerge alongside a preceding decline in visual acuity. Individuals with CBS notably maintain their grasp on reality and possess an understanding of the fictitious nature of their VHs. However, due to a general lack of awareness within the medical community; CBS is often erroneously diagnosed as dementia or new-onset psychosis. This syndrome is linked to age-related macular degeneration, glaucoma, and cataracts. Diagnosing CBS hinges primarily on the patient's medical history, clinical presentation, information from collateral sources, and a process of exclusion for other potential explanations. Unfortunately, no definitive cure exists for CBS, with treatment primarily centered on offering reassurance and education; nevertheless, persistent hallucinations containing unsettling imagery might require targeted intervention, employing off-label use of psychotropic medications (such as antipsychotics, antidepressants, and anticonvulsants) to address refractory symptoms. This case report underscores a CBS instance with atypical features, highlighting the diagnostic and therapeutic approach applied to promote accurate identification and treatment.

Keywords: Atypical Features; Charles Bonnet Syndrome

Abbreviations: CBSL: Charles Bonnet syndrome; VHs: Visual Hallucinations; ED: Emergency Department; ADLs: Activities of Daily Living.

Introduction

A 78-year-old African American male presented at the emergency department (ED) accompanied by his family. The primary concern was the sudden exacerbation of his chronic VHs, which now featured more intricate, vivid, and inappropriate content [1]. Alongside this, there was a noticeable progression in troubling behavioral disturbances and episodes of agitation. The patient's family was deeply worried about his deteriorating mental state and behavioral issues, to the extent that they were considering admitting him for inpatient care at a geriatric acute psychiatry hospital [2]. Their concern centered on what they perceived as a progressive psychosis and disruptive behavior. Interestingly, the family mentioned the possibility of these changes being connected to some form of spiritual possession by "evil spirits".

Diagnostic Workup

After conducting a medical assessment, it was discovered that the patient had atrial flutter as indicated by the EKG results. Additionally, there was evidence of acute kidney injury, with a raised creatinine level of 2.4 mg/dL. Once the patient's medical condition was stabilized, the main medical team sought input from the Psychiatry Consult/Liaison

(C/L) service. This consultation was aimed at evaluating the patient's eligibility for inpatient psychiatric admission, considering that his VHs had been responsible for numerous instances of agitation at home in his medical history.

Medical & Psychiatric Assessment

In his psychiatric evaluation, pertinent factors included the absence of any previous history of mental illness, inpatient psychiatric hospitalizations, utilization of psychotropic medications, or substance use disorders/abuse. His medical history encompassed hypertension, chronic kidney disease (stage III), a stroke that had occurred four decades earlier, bilateral cataracts, bilateral glaucoma, coronary artery disease, and arthritis.

Further insights were gleaned from the patient's family members who accompanied him. They indicated that the patient's VHs had likely been present for several years, but there had been a recent increase in both frequency and intensity, with the VHs displaying more graphic and sexually inappropriate content. These VHs manifested sporadically throughout the day, with a heightened prominence during the evenings. Interestingly, engaging in outdoor activities seemed to alleviate and diminish the occurrence of VHs.

The patient conveyed an awareness that these VHs were not shared or experienced by others, which suggested that he wasn't entirely fixed in his belief regarding their reality. Nonetheless, he exhibited significant distress, often to the point of agitation, due to the intrusive and vivid nature of these repetitive images. For instance, he described seeing unfamiliar individuals on the floors of his home while watching his nightly "westerns" TV shows, and these distressing visuals were a cause of significant disturbance.

Importantly, the patient faced notable challenges in administering his glaucoma eye drop medications due to compromised dexterity, likely due to his history of arthritis, which also affected his visual acuity. Approximately four weeks prior to arriving at the ED, he reported breaking his eyeglasses. This timeframe correlated with both the family's account of the escalating severity of his visual hallucinations (VHs) and the recent surge in behavioral disturbances. Nonetheless, he was able to manage his Activities of Daily Living (ADLs) at home, where he lived independently. According to his family, there were no glaring signs of memory impairment or significant functional decline compared to his usual baseline.

During the physical examination, bilateral cataracts were noticeable, and he exhibited reduced visual acuity, particularly at night. In the mental status examination, he demonstrated alertness and orientation to person, place, and time. He described his mood as "fair," maintaining a mostly euthymic emotional state. His affect remained stable and congruent, and his thought process was logical and linear. He denied experiencing VHs during his hospital stay, as well as any history of auditory hallucinations or delusional thoughts. While his memory assessment indicated intact registration and immediate recall, delayed recall was impaired, with him recalling only one out of the three items tested. His digit span and concentration/focus were intact, and there were no signs of paranoia or self-harm tendencies. Additionally, his behaviors during the hospital stay were considered appropriate and markedly different from his behaviors at home. This divergence was noted in his interactions with both family members and medical staff.

After receiving IV fluids for Acute Kidney Injury, resulting in a decrease in creatinine levels, and with the cardiology team introducing Warfarin anticoagulation for his atrial flutter, the patient's condition was sufficiently stable for discharge. The decision was made for him to return home with a prescription for home healthcare assistance, particularly for managing his glaucoma medication and other medical needs. His follow-up plan included a referral to ophthalmology for an assessment regarding potential bilateral cataract surgery, as well as for an eye examination and obtaining a new pair of eyeglasses [3]. Furthermore, he was recommended to undergo an outpatient neurocognitive evaluation to explore the possibility of a multifactorial origin for his emerging neurocognitive disorder, especially given his historical stroke and impaired memory recall. The C/L team initiated the patient on a trial of Escitalopram at a daily dose of 5 mg, along with Risperidone at 0.25 mg twice daily. These medications aimed to address his anxiety, agitation, and behavioral disturbances stemming from his VHs. Additionally, he was prescribed off-label vitamin E at an oral daily dose of 200 mg as a memory enhancement measure.

The patient's family received education regarding the underlying pathophysiology of the illness and was encouraged to reevaluate their initial belief of the VHs being connected to "evil spirits." They expressed agreement in coordinating referrals and adhering to follow-up care. They also pledged to enhance the lighting in the patient's home environment and to make regular check-ins and visits to minimize his time spent alone at home; with these plans in place, the patient was discharged to continue his care and recovery in a supportive setting.

Discussion

The diagnosis and management of CBS remain intricate, hindered by its limited recognition within the medical field despite the increasing prevalence of ocular issues among the elderly. Research has yielded varying prevalence estimates

for CBS among those with visual impairment, spanning from 0.4% to 12% [4,5]. Remarkably, epidemiological investigations indicate that as many as 63% of older adults with substantial visual deficits might be susceptible to CBS [6]. The underlying pathophysiology of CBS originates from visual impairments affecting any portion of the visual pathway, extending from ocular structures to the cerebral visual cortex. These impairments result in sensory deafferentation, prompting uncontrolled firing of visual cortical regions through a disinhibitory mechanism and this process gives rise to an array of hallucinations and visual distortions. In the elderly demographic, a predisposition towards cognitive impairment can exacerbate CBS, coinciding with the high prevalence of concurrent visual deficits [7].

Unlike hallucinations seen in psychotic conditions, where individuals firmly believe in the authenticity of their perceptual experiences, CBS patients are aware of the illusory nature of their hallucinations and acknowledge that others do not share their perceptions. In contrast to individuals with delirium or dementia, those with CBS maintain their cognitive abilities and functional capacities. However, despite their awareness, these patients can experience emotional distress due to the recurring and vivid nature of their hallucinations, occasionally leading to behavioral disturbances.

This particular case of CBS stands out due to its atypical characteristics, notably the presence of reduced cognitive faculties. The patient's memory assessment showed intact registration and immediate recall, but delayed recall was impaired, with the ability to recollect only one out of three tested items. Intriguingly, his digit span and concentration remained unaffected, and no signs of paranoia or self-harm tendencies were evident. This distinctive cognitive profile introduces intriguing possibilities in the context of CBS. Although CBS is primarily linked with primary visual deficiency, the concurrent presence of neurocognitive impairment adds a unique dimension to this case. This coexistence could potentially differentiate this case from others, highlighting the interplay between sensory deficits and cognitive functioning in CBS.

In the pursuit of accurate diagnosis and heightened healthcare provider awareness, adopting established diagnostic criteria, comprehensive patient interviews, collateral insights, and thorough analysis of contributing factors is crucial. A defining characteristic of CBS, a disorder marked by hallucinations, is its occurrence in elderly patients grappling with age-related visual changes such as macular degeneration, glaucoma, and cataracts.

While a definitive remedy for CBS remains elusive, various interventions can alleviate symptoms, including pharmaceutical agents, behavioral strategies, patient education, ophthalmic interventions, and reassurance. The chosen approach hinges on symptom severity. Alongside assuring patients about the benign nature of their hallucinations, both non-pharmacological and pharmacological interventions have proven effective in managing CBS. In cases of distressing and unrelenting hallucinations, especially those that resist treatment, considering the off-label use of psychotropic medications may be prudent. However, such a decision requires meticulous evaluation of potential risks, benefits, and alternatives, particularly in the elderly population with comorbidities. Notably, atypical CBS presentations may occasionally align with underlying neurocognitive disorders, a common occurrence in the elderly. Moreover, atypical CBS tends to affect elderly patients who are experiencing age-related cognitive decline, which can erroneously lead to diagnoses such as Lewy Body dementia or a primary psychotic disorder. It is important to be aware of the presentation of hallucinations in CBS, which occurs solely without delusions or paranoia, versus a psychosis presentation in early dementia, which may present with command auditory hallucinations, delusions, or paranoia [8].

Conclusion

This case serves as a valuable illustration of the diagnostic approach and management strategies applied to CBS, a condition often linked with age-related macular degeneration, glaucoma, and cataracts. The patient's clinical presentation and the timeline of symptom development align consistently with CBS, underscoring the complexity involved in diagnosing this syndrome. Indeed, arriving at a CBS diagnosis can be intricate due to the overlap of symptoms with several potential differential diagnoses. In this specific case, the list of differentials encompassed psychotic disorders, dementia, and hallucinations induced by substances. Nonetheless, through a meticulous assessment, distinctive features emerged that pointed toward CBS as the most plausible explanation for the patient's visual hallucinations. Notably, his reality testing remained intact, he exhibited an awareness of the hallucinatory nature of his experiences, and his visual impairment was apparent. These critical insights guided the diagnosis. The patient's presentation was aligned with the hallmark symptomatology of CBS, characterized by a history of visual impairment and reported visual hallucinations that abated in response to increased illumination or a change in surroundings.

While the initial presentation exhibited atypical behavioral disturbance and agitation at home, the absence of psychotic features and the normalization of his behavior during his stay on the inpatient medical floor suggested that these issues were precipitated by the VHs experienced in his home environment. A distinct facet of this case was the

potential presence of an underlying neurocognitive disorder, as indicated by the patient's impaired recall during the initial neurological assessment. It is plausible that this unique CBS presentation was influenced by sensory deprivation, stemming from his history of cataracts and compromised visual acuity when not using glasses, particularly in dimly lit settings. His challenges in appropriately administering his glaucoma eye drops further emphasized the impact of his visual impairment. This case not only showcases the nuances of diagnosing CBS but also highlights the importance of considering both classic symptomatology and atypical features, such as concurrent neurocognitive disorders, when evaluating patients with visual hallucinations associated with visual impairment.

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