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CASE REPORT

Complex visual hallucinatory experience in an elderly blind woman with glaucoma: revisiting Charles Bonnet syndrome

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Key Clinical Message

For fear of being ridiculed, individuals with visual hallucinations hide their experiences and thus remain unrecognized and miss treatment. An elderly blind woman secondary to glaucoma experienced visual hallucinations accompanied by gross behavior disturbances. She improved with sodium valproate after haloperidol failed and remained relatively improved upon 3 months follow-up.

Keywords

Behavioral disturbance, Charles Bonnet syndrome, visual hallucinations.

Introduction

Visual hallucination in a person with loss of vision was first described by Charles Bonnet in 1769 when his 89-year-old grandfather, who experienced visual hallucinations after visual loss due to cataract. The condition was later termed Charles Bonnet syndrome after Charles Bonnet himself developed the condition in his old age [1]. Charles Bonnet syndrome (CBS) is usually characterized by vivid and complex visual hallucinations in a patient with visual loss in the absence of any other psychiatric illness and with intact insight into the condition. Gold and Rabins have suggested that the term CBS should be used strictly to describe complex visual experiences with insight [1, 2], although it can be argued that CBS with intact insight and without psychosis is exceedingly rare [3]. Notably, visual hallucinations in CBS have been reported in a variety of psychiatric and neurological disorders: psychosis, schizophrenia, dementia, drug or alcohol abuse (delirium tremens), Alice in Wonderland syndrome (AIWS), epilepsy, Parkinson disease, narcolepsy, brain tumors, migraine, and long-term sleep deprivation [4].

This is a case of an 85-year-old woman with blindness in both eyes who presented with visual hallucinations followed by uncharacteristic behavior and lack of insight. Her visual loss was due to a history of glaucoma; however, her history was also remarkable for a previous stroke. The psychiatric symptoms did not initially respond to antipsychotic medication haloperidol, but the signs and overall insight improved after substitution with sodium valproate.

Case Report

We report a case of an 85-year-old woman with blindness due to glaucoma brought for a psychiatric consultation after the onset of uncharacteristic behavior. The behavioral disturbance was characterized by irritability, aggression, sleep disturbances, irrelevant speech associated with shouting, pacing around aimlessly, accusing others of playing black magic on her and seeing human figures others could not see.

At the time, she was brought for psychiatric consultation, she had had the subjective visual experiences for about 8 months with progressive worsening. It started with isolated incidences of seeing strange figures of extraordinarily tall and miniature humans climbing walls. The symptoms gradually became worse, and she began

complaining that the human figures were following her everywhere, observing her activities, at times fighting each other and even threatening her with snakes.

As symptoms progressed, she became agitated with frequently pacing around the house and disruptive sleep. She became significantly distressed to the point of refusing to undress while taking a shower for fear of being watched by strangers "the human figures," and therefore bathing while fully clothed.

The relatives reported that during the periods of behavioral disturbance, the patient often demanded to be taken to see her mother who passed away many years back, insisting that she is in a dire situation somewhere needing her help. At times, she would also have tantrums about a particular neighbor from her hometown in her younger days planning to harm her and other villagers stealing her belongings.

Furthermore, she often complained that her husband, who was functionally incapacitated from Alzheimer's disease, was conspiring with her sister and playing black magic on her. When asked to clarify, she said, "When those tall people appear, I also see my husband and my sister crafting live babies together and laugh on my face." The experience made her agitated and aggressive toward her husband and sister.

It is reported that the onset of psychotic experience was preceded by an episode of stroke, which left her with paralyzed left arm and mouth deviating to one side for some time, although she had regained her functionality after a while.

Further interviewing revealed a negative history for past psychiatric diagnosis or disturbances in memory and language skills, and it was acknowledged that on the days when she did not have the visual experiences, she was relatively calm and cooperative. She denied episodes of hearing voices others could not hear, a sense of her thoughts being controlled by external influences, or that her thoughts being spoken out loud, actions or feelings were being controlled by external forces.

During the first visit, she was a healthy-appearing senior woman who was fully conscious, alert, and oriented to the voices of her accompanied relatives. She was well-kept, cooperative, and her speech was relevant. Her recent and past memories were intact. She was particularly concerned about the visual experiences and attributed them as demons planning to harm her as they followed her everywhere, even while taking a shower.

Cognitive assessment was performed using Mini-Mental Status Examination (MMSE), on which she scored 18/20, with inability to assess some of the domains due to her blindness, including orientation, naming, reading, writing, and construction. The patient missed one point on recall and another point of attention/concentration.

Laboratory investigations including malaria rapid diagnostic test (MRDT), complete blood count, liver function test, renal function test were all unremarkable. Brain magnetic resonance imaging was advised but was not performed due to financial constraints.

The initial treatment choice of haloperidol 1.5 mg twice daily did not improve the symptoms for the first 2 weeks. After 4 weeks of trial, haloperidol was substituted by 300 mg of sodium valproate given at a dosage of 100 mg in the morning and 200 mg at night. Over the course of 3 months, the patient markedly improved regarding frequency and severity of symptoms.

Although the patient did not return to the clinic after the third follow-up for further assessment, her prognosis is likely unfavorable due to several factors. In the majority of CBS cases, the symptoms usually resolve after the correction of vision, which is not possible for this patient due to her irreversible retinal damage secondary to glaucoma. Furthermore, her history of stroke with subsequent psychotic symptoms suggests an organic brain pathology, which has been argued as a predictor of dementia process to follow [5].

Discussions

The diagnosis of CBS is based on several sources of cross-sectional observational data, majority of which agree on the CBS diagnostic criteria to include (1) The presence of formed, complex, persistent or repetitive, stereotyped visual hallucinations, (2) Full or partial retention of insight into the unreal nature of the hallucinations, (3) Absence of hallucinations in other sensory modalities, and (4) Absence of primary or secondary delusions [2]. Others in the field, however, have not included lack of insight as central for CBS diagnosis and reports have shown that the presence of psychiatric symptoms is not rare in CBS [5, 6].

The clinical presentation of our patient may suggest an atypical case of CBS with lack of insight to the unreality of the visual experiences accompanied by behavior change and distressing persecutory ideas during the visual experiences.

The presence of psychotic symptoms and lack of insight in this patient pose a diagnostic dilemma as to whether she exhibits CBS or a primary psychotic disorder. However, we noted that the age of onset at which the first psychotic episode occurred, the nature of visual hallucinatory experience without auditory hallucination, and the overall progression of the symptoms does not favor the diagnosis of a primary psychotic condition such as schizophrenia or schizoaffective disorders. It is argued that a typical case of CBS with visual hallucinations and intact insight without accompanied neuropsychiatric symptoms is exceedingly rare [3]. One of the argument is

that neuropsychiatric manifestations in CBS are missed because many of the reported cases are primarily selected from ophthalmologic as opposed to psychiatric services; therefore, closer scrutiny of cases would likely reveal a higher prevalence of mental manifestation and lack of insight within CBS clinical presentation [3].

This case provokes the continued, unresolved debate about the etiology of Charles Bonnet syndrome. Ophthalmologic, psychiatric, and cortical disturbances have all been implicated in the etiological theories of CBS. The clinical presentation of this case supports the de-afferentation theory, whereby vision loss leads to visual sensory de-afferentation, which causes disinhibition and later spontaneous firing of the visual cortical regions, which leads to visual hallucinations [7]. Furthermore, this hypothesis is supported by functional MRI studies of visual deprivation experiments that have demonstrated an association between visual hallucinations and spontaneous activity of ventral occipital lobe in patients with CBS [8].

Psychoanalytic mechanisms are also thought to account for the onset of Charles Bonnet syndrome, as loss of vision triggers the ego to create a substitutive world full of entertaining scenes to compensate for the visual loss. However, the distressing nature of the index case may not favor this theory. Another psychoanalytic approach argues that failing eyesight reduces the perception of reality, which is thought to be the initial stages of visual hallucinations [9, 10].

From a neurophysiologic perspective, the brain is considered to have censorship mechanisms, which remove all irrelevant impulses from a conscious perception that depends on sensory inputs to reach a certain threshold. It appears that in CBS, at sub-threshold of sensory inputs, the brain allows the subconscious perception to surface as visual hallucinations [11].

The onset of visual hallucination after an episode of stroke highlights the role of a vascular event on the etiology of CBS. Ischemia is a known underlying etiology of cortical lesions, in which the majority of visual hallucinations after stroke due to occipital infarcts rather than middle cerebral infarcts [12]. One hypothesis is that stroke activates visual association cortex indirectly through the loss of direct cortico-cortical inputs and loss of cortex control of thalamic inputs to visual association cortex [13] as opposed to epilepsy which stimulates the visual association area directly [12].

Pharmacological treatment of CBS is usually initiated when reassurance fails and/or the patients are in severe distress [14, 15]. Although there is no specific recommended pharmacological treatment guideline, several agents including antipsychotics, anticonvulsants, anti-anxiety, and selective serotonin inhibitors are widely used for the treatment of CBS with variable results [14–16]. The evidence is limited to support the effectiveness of valproate monotherapy in the treatment of psychosis, and it is typically used as an adjuvant particularly for the symptoms of aggression, excitement, and tardive dyskinesia [17, 18]. Nonetheless, valproate was chosen for this patient due to a more favorable side effect profile compared to carbamazepine, which would also require benzodiazepine administration to maximize its therapeutic effect and thus pose many potential risks due to her geriatric status [14, 16]. The efficacy of valproate in the management of CBS is thought to be through its inhibitory action on the hyperactive extrastriate neuronal pathway, which is considered to be responsible for visual hallucinations in CBS [14, 19].

Conclusion

This case evokes the debate regarding the nature and diagnostic criteria of Charles Bonnet Syndrome and highlights the relationship of CBS visual hallucinations with the development of psychosis symptoms. The case also demonstrates that an atypical form of CBS could easily be confused with a functional psychiatric disorder. A high index of suspicion is vital for providers to identify CBS and initiate appropriate and effective management for patients with similar presentation.

Consent

Written informed consent was obtained from the patient for publication of this case report. A copy of the written consent is available for review by the Editor-in-Chief of this journal.

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Authorship

AAN and IRM: managed the patient from the first contact. AN: did the literature search and wrote the manuscript in correspondence with IM. All authors have read and approved the final document.

Conflict of Interest

The authors have no conflict of interest to declare.

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