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CASE REPORT

Seeing the unseen: Charles Bonnet syndrome revisited

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Abstract

Charles Bonnet syndrome (CBS) is a rare condition that encompasses three clinical features: complex visual hallucinations, ocular pathology causing visual deterioration, and preserved cognitive status. Common associated ocular pathologies include age-related macular degeneration, glaucoma, and cataracts. Several theories have been proposed to try to explain the visual hallucinations. However, the pathophysiology remains poorly understood, and treatment is largely based on anecdotal data. The lack of awareness of CBS among medical professionals often leads to inappropriate diagnosis and medication. In a country like India, where awareness of mental health is not widespread, cultural myths and stigma prevent patients from seeking professional help. Here we describe two cases of CBS and revisit different ocular morbidities that have been reported to occur in conjunction with CBS. Psychiatrists and ophthalmologists alike must be sensitive to this clinical condition to ensure prompt diagnosis and treatment.

INTRODUCTION

Charles Bonnet syndrome (CBS) is a rare clinical condition that encompasses three clinical features: complex visual hallucinations, ocular pathology causing visual deterioration, and preserved cognitive status. Common associated ocular pathologies include age-related macular degeneration, glaucoma, and cataracts. While symptoms of CBS could herald the onset of dementia in the elderly, patients are often neurologically normal.[1] Furthermore, an intraocular cause alone may not necessarily lead to CBS; a lesion anywhere in the visual pathway can be associated with the syndrome.[2]

CASE PRESENTATION

Case I

Mrs J, a 75-year-old widow, was referred for a psychiatric consult by her physician for history of abnormal behaviour for the past 7 months. She had been evaluated by her ophthalmologist and diagnosed as having open-angle glaucoma in both eyes; 7 years earlier, she had been diagnosed with age-related macular degeneration. Best-corrected visual acuity in her right eye was counting fingers at 1 metre, and her left eye was 20/400 (1.3 logMAR). Anterior segment evaluation showed nuclear sclerosis grade II in both eyes. On gonioscopy, the angles in both eyes were found to be open. At presentation, the patient had been using timolol maleate/brimonidine eye drops twice daily and dorzolamide eye drops three times daily in both eyes for the past 4 years. Intraocular pressure in both eyes while the patient was on antiglaucoma medication was 12 mmHg. Posterior segment evaluation showed bipolar notches in both eyes and geographic atrophy at the macula in both eyes.

The patient's family members had noticed strange, abnormal behaviour over the past 7 months. The patient complained of seeing patterns on walls, faces of unknown people, and non-existent objects. Despite the visual impairment, the patient started seeing geometrical patterns on the wall, which would turn into idols of deities. Caretakers found the patient trying to touch the wall and pray. She soon started enquiring to her relatives about the new men in the house, and she described seeing naked men of Indian and foreign

origin roaming around the house and walking through walls. They were accompanied by women at times and often indulged in sexual acts. The patient said that these people stood around her; sometimes, they just smiled, and sometimes, they provoked her by making faces at her. The patient realized that the visuals were unreal and felt distressed by them. Furthermore, she said that she had never heard these people and they would never reply despite the patient hitting them, shooing them, or requesting that they go away. Soon, the patient became extremely distressed; she started complaining of reduced sleep and developed crying spells. The hallucinations would start in the afternoon and worsen in the evening. She was prescribed lorazepam tablets 2 mg at night by her family physician, but there was no improvement.

A thorough medical evaluation was performed to exclude any organic causes. On neurological examination, there were no signs of cogwheeling, bradykinesia, or resting tremor. On mental status examination, the patient was well oriented, conscious, cooperative, and communicative. All routine haematological investigations, electrolytes, blood and urine cultures, and toxicology screen were within normal limits. Brain magnetic resonance imaging showed signs of age-related change. She had no family history of dementia and her memory was intact during mental status examination.

She was prescribed 2.5-mg olanzapine to take for 1 week at night. At 6-month follow-up, she felt there had been a significant improvement in her sleep and moderate reduction in her hallucinations, with only one episode of hallucinations in the interim period.

CASE II

Mr R, a 71-year-old single man, was referred by his family physician for disturbed sleep and visual hallucinations. On ophthalmological examination, best corrected visual acuity in his right eye was 20/400 (1.3 logMAR) and hand movements close to the face in the left eye. The patient had been diagnosed with bilateral chronic angle closure glaucoma, and he underwent combined phacoemulsification with an intraocular lens implantation and trabeculectomy with mitomycin-C in both eyes 3 years earlier. Postoperatively, he had an uneventful recovery. The intraocular pressure in both eyes was 9 mmHg, with well-functioning filtering blebs. The right eye had a 0.9:1.0 cup-to-disc ratio, and the left eye showed total glaucomatous cupping.

The subject had a history of visual hallucinations for 18 months prior to seeking help. He would awaken multiple times in the middle of the night, and on looking out of the window, he would see people crowded outside a large hall opposite his house. Initially he had shrugged it off as his mind playing tricks on him. However, as his sleep deteriorated over the course of a few months, the subject recounted how he had seen people climbing through the windows into his house and sitting around his bed. By morning, they would pack their belongings and leave. During the day, he would complain of seeing patterns on the walls, which would transform into a man's bearded face. This face resembled a well-known religious leader. At night, he often saw naked women coming into his room. The patient described the people in his house as friendly, as they would always smile at him. The subject never heard them speak to him or talk among themselves.

The hallucinations were not very intense in the morning and gradually worsened as the day progressed. However, the subject realized what he was seeing was unreal. The patient went on pilgrimage every 2 years, but as the date of his next departure approached, the patient realized that travelling with his condition would be extremely difficult and that he needed medical treatment. Despite having these hallucinations, he continued his daily routine without any trouble. On evaluation, his Mini-Mental State Examination score was 28/30. Magnetic resonance imaging showed chronic ischemic changes in bilateral frontoparietal white matter. Routine haematological investigations, electrolytes, and toxicology screen were normal, but Vitamin D3 and B12 levels were low. On neurological examination, there were no signs of cogwheeling, bradykinesia, or resting tremor. On mental status examination, the patient was conscious, cooperative, communicative, and well oriented. He was prescribed gabapentin 100 mg at night and also treated for his vitamin deficiencies.

At his first follow-up, he perceived a 25% reduction in his hallucinations and his sleep had significantly improved. The dose of gabapentin was increased to 200 mg following which he perceived a resolution of his symptoms. At the 3-month follow-up, the patient reported no similar episodes and his sleep remained largely undisturbed.

Summary

In both cases I and II, the patients exhibited the classical triad of CBS: complex visual hallucinations, ocular pathology causing visual deterioration, and preserved cognitive status.

DISCUSSION

CBS is an uncommon cause for visual hallucinations in the presence of visual impairment. The name 'Charles Bonnet syndrome' was introduced by de Morsier in recognition of Charles Bonnet, who had written in detail about the visual hallucinations of his grandfather Charles Lullin.[3] It has been reported that the specific risk factors for CBS include age (>64 years), social isolation, low cognitive function, a past history of stroke, and poor bilateral visual acuity[2,4,5]

The well-described criteria for CBS include the following:[6,7]

- the presence of formed and complex hallucinations
- persistent or repetitive visual hallucinations
- full or partial retention of insight
- the absence of delusions
- the absence of hallucinations in other sensory modalities.

In CBS, patients are aware that their symptoms are unreal, but the hallucinations often seem to fit in logically with the surrounding scenario.[8]

Our patients had been experiencing visual hallucinations for a significant period (case 1 for 7 months and case 2 for 18 months) prior to seeking treatment. Although the visual hallucinations persisted, there were no reported fluctuations in levels of attention and alertness during this time, and on neurological examination, neither patient showed any parkinsonian features, which are the other core features of dementia with Lewy bodies. Supporting features of dementia, such as repeated falls, syncope, systematized delusions, and hallucinations in other modalities (e.g. auditory, tactile), were also absent.

It is important to mention the content of the hallucinations; both patients complained of seeing faces, religious idols, and naked human bodies. In an Indian context, religion plays an important role in day-to-day life, so experiencing hallucinations with religious imagery may be considered as divine intervention. Therefore, caregivers and relatives may also not view such hallucinations as a symptom. Any religious behaviour in response to the hallucinations would also usually be tolerated, unless it was so severe that it interfered in a patient's functioning or affected a caregiver's routines.

Additionally, sexual content is usually met with a lot of guilt, as matters of sexual content are seen as taboo in orthodox Indian society. This is especially the case among religious elderly people, who may not openly discuss such hallucinations for fear of being ostracized, labelled, or ridiculed. Thus, in both cases presented here, societal attitudes may have led to a delay in seeking medical treatment.

Pathophysiology

The exact nature of the pathophysiology of CBS is unknown, and many theories have tried to explain the phenomenon of visual hallucinations. It has been postulated that some lesions of the visual pathway lead to the transmission of abnormal signals to the visual cortex, and when added to the existing normal visual cortex activity, these signals are thought to cause the varied, complex visual hallucinations that CBS patients experience.[8,9]

Another theory suggests that severe visual impairment leads to the production of de novo images from the visual cortex, thus causing visual hallucinations.[8] The sensory deprivation/phantom vision theory suggests that this phenomenon is akin to phantom limb syndrome.[10,11]

The neuromatrix theory suggests the existence of a network of neurons – called the neuromatrix – that extends throughout the brain, which is inherently able to generate sensory phantoms. The neuromatrix then gives off a pattern, or the neurosignature, on all afferent inputs, so sensory experiences may have a quality of self and possess affective tone and cognitive meaning. Furthermore, this neuromatrix subserves bodily sensation and has a genetically determined substrate that is modified by sensory experience.[10,12]

Other factors that may cause visual hallucinations in CBS include impaired cerebral perfusion, social isolation, and other psychological factors.[13,14]

Ocular conditions associated with CBS

Age-related macular degeneration and glaucoma are the most common conditions affected by CBS. However, other ocular morbidities have also been reported to be associated with CBS.[10] Table 1 provides a list of some ocular pathologies known to

Table 1 Ocular morbidities associated with Charles Bonnet syndrome that have been reported in literature

1. Cataract[7]
2. Age-related macular degeneration[7]
3. Glaucoma[7]
4. Diabetic retinopathy[7]
5. Corneal opacity[15]
6. Retinal detachment[15]
7. Choroideremia[16]
8. Post-enucleation[17]
9. Multiple sclerosis with optic neuritis[18]
10. Herpes simplex encephalitis[19]
11. Neurosarcoidosis[9]
12. Occipital infarction with homonymous hemianopia[20]
13. Retinitis pigmentosa[21]
14. Bilateral loss of vision[22]
15. Vertebrobasilar insufficiency[23]
16. AIDS with cytomegalovirus retinitis[24]
17. Post macular translocation surgery[25]
18. Suprasellar meningioma with visual loss (surgery resulted in cure)[26]
19. Cranial arteritis[27]
20. Pituitary tumours[28]

This list is not exhaustive.

cause profound visual impairment that have been reported in the literature in relation to CBS.

Treatment

While no clear guidelines exist regarding the management of CBS, it is essential that the underlying ophthalmic pathology be addressed. In cases of cataract or corneal scarring, part or full treatment may require surgical management of the condition. Conditions such as diabetic retinopathy and subretinal haematoma may also need vitreoretinal surgery to improve vision.[8] However, for conditions such as geographic atrophy or end-stage glaucoma, surgery may not offer any visual rehabilitation. In such cases, low vision aids and sight enhancement devices may help.

The cause of visual impairment may be more psychologically afflicting than the hallucinations themselves, and therefore, counselling, reassurance, and supportive care go a long way in improving the general well-being of patients with CBS. It has also been noted that the progression of visual impairment towards total blindness often reduces the frequency of visual hallucinations.[8]

On the issue of pharmacotherapy, the extrapolation of the use of antipsychotics to control visual hallucinations in schizophrenia has led to their use in CBS. Anecdotal evidence indicates the resolution of symptoms in CBS patients when treated with donezepil, olanzapine, venlafaxine, and gabapentin.[29–32] We used olanzapine because lorazepam, which was used initially in one of the cases, did not improve the patient's sleep. Low-dose olanzapine provides sedation and a reduction in anxiety without any extrapyramidal side-effects. Gabapentin's broad spectrum of indications coupled with its safety profile and fewer interactions with other medications makes it an acceptable choice for treatment of CBS. Also, as compared to other anti-epileptics, gabapentin has fewer side-effects such as marked sedation or cognitive impairments.[32]

Conclusion

CBS is probably under-diagnosed given the demographics it tends to affect. Often times, patients do not report or seek help for visual hallucinations. The preserved insight often prevents them from admitting the hallucinations for the fear of being diagnosed with a psychotic disorder. In a country like India, awareness of mental health is not widespread, so myths and misconceptions about mental illness contribute to the stigma, which further leads many people to be ashamed and to avoid seeking professional help.[33] Furthermore, the low level of awareness of CBS among medical professionals means that there is a risk of an incorrect diagnosis, such as delirium, dementia, or psychosis, and inappropriate therapy. The important differentials that need to be ruled out include Alzheimer's disease, delirium, parkinsonism, and schizophrenia.[10]

Despite the presence of visual impairment, CBS presents with symptoms that make it more likely for a patient to seek the help of the psychiatrist than an ophthalmologist. Therefore, it is important that treating psychiatrists and ophthalmologists are sensitive to this clinical entity so that a prompt diagnosis and subsequent treatment can be instituted. This helps avoid the risk of diagnosing and treating non-existent psychiatric illness.

DISCLOSURE

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