

Case report

Seeing the unseen in the sane; Charles Bonnet Syndrome, an overview with case presentations

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Introduction

Charles Bonnet Syndrome (CBS) is a condition characterized by the occurrence of complex visual hallucinations in individuals with significant vision loss but without cognitive impairment. First described by Charles Bonnet¹ in 1760, visual hallucinations seen primarily in elderly patients with visual impairment, often occur in patients with age-related eye diseases like macular degeneration, glaucoma, or diabetic retinopathy. Although the hallucinations experienced by patients are vivid and can be distressing, CBS² is not linked to psychiatric disorders and generally does not involve other senses or cognitive functions³. Diagnosis of the disease can be challenging as the patients may not be willing to report symptoms due to various reasons. Awareness of this rare but important condition is pivotal in managing patients with severe visual impairment.

We report two patients from an eye clinic in Sri Lanka for the first time with features of Charles Bonnet Syndrome highlighting the importance of comprehensive management of patients with severe visual impairment.

Case report 1: Mrs. A, Age 82

Background: Mrs. A is a 82-year-old woman who presented with poor vision of gradual onset in both eyes for more than 2 years duration. On examination visual acuity was 6/60 in the right eye and 1/60 in the left with cataract in both eyes. Dilated funduscopy showed age related macular degeneration (ARMD) in both eyes. She underwent cataract extraction with lens implantation and was managing well with remaining peripheral vision.

About three months after Mrs. A began to see visual hallucinations. She described them as small children flocked around her bed with faces clear and recognizable. Sometimes they were tiny objects of

similar patterns. They were seen for few minutes and then disappeared instantly after several minutes. They occurred several times a day, especially in low-light environments.

Insight and impact: Mrs. A was fully aware that the images were not real, but the frequency and vividness of the hallucinations were distressing to her. She initially hesitated to report these symptoms, fearing they might be mistaken for a psychiatric illness.

Management and outcome: After a thorough examination and reassurance regarding the nature of CBS, Mrs. A was taught techniques to interrupt the hallucinations, including looking away or turning on brighter lights. With improved understanding, her anxiety about the hallucinations decreased. They continued intermittently for about 8 months. No pharmacological intervention was deemed necessary, as Mrs. A was managing well with behavioral adjustments. After about one year when she was seen at follow up visits she was completely free of symptoms.

Case presentation 2: Mr. B, Age 75

Background: Mr. B, was a 75-year-old unmarried man living in a retirement home, with advanced glaucoma, and diabetes with nonproliferative diabetic retinopathy. He presented with a progressive loss of peripheral vision. His visual acuity was 6/60 in the right eye and 6/36 in the left.

Presenting symptoms: Mr. B reported seeing geometric shapes, particularly in his peripheral vision, which sometimes appeared to be moving. On a few occasions, he saw animals, including cats, dogs and birds, which were not actually present. These hallucinations generally occurred during quiet times when he was resting or alone. Particularly when he got up in the night to go to the washroom with the dim night light in the room.

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Insight and impact: Mr. B recognized that these images were not real, but he became increasingly anxious, worrying that this might be an early sign of dementia or another psychiatric illness. He was hesitant to speak about these experiences until he was specifically asked about his visual symptoms in a follow-up consultation.

Management and outcome: Upon diagnosing CBS, Mr. B was reassured of the benign nature of his condition. Adjustments to his living space, such as increasing lighting and engaging in more activities, helped reduce the occurrence of the hallucinations. His anxiety was alleviated through education and counseling, and he no longer feared that the hallucinations were signs of cognitive decline.

Discussion

Both cases highlight common features of CBS: significant visual impairment, awareness of the unreality of hallucinations, and associated anxiety or distress. They also underscore the importance of physician awareness of CBS, as patients may not voluntarily disclose their symptoms due to stigma. Early diagnosis and reassurance can significantly improve patients' quality of life, reducing both distress and anxiety.

Charles Bonnet Syndrome (CBS) is a condition characterized by the occurrence of complex visual hallucinations in individuals with significant vision loss but without cognitive impairment. This phenomenon was first noted by Charles Bonnet in 1760. CBS first described by George de Morsier in 1967, is primarily seen in elderly patients with various degrees of visual impairment⁴, often due to age-related eye diseases like macular degeneration, glaucoma, or diabetic retinopathy. Although the hallucinations⁵ experienced by patients are vivid and can be distressing, CBS is not linked to psychiatric disorders^{6,7} and generally does not involve other senses or cognitive functions.

Pathophysiology

The exact cause of CBS remains unclear. However, a widely accepted theory suggests that when the brain is deprived of normal visual input, as seen in patients with significant vision loss, it compensates by generating visual images⁸. This phenomenon of deafferentation leads to the spontaneous activation of visual centers in the brain, resulting in hallucinations. Patients are often aware that the images are not real, distinguishing CBS from psychosis.

Diagnosis

Diagnosing CBS can be challenging, as patients may be reluctant to discuss their symptoms for fear of being labeled as mentally ill. A thorough history and clinical examination, with a focus on visual acuity and cognitive assessment, are essential. The absence of auditory or other sensory hallucinations and the patient's insight into the unreality of the images are key indicators. Imaging studies or neuropsychological evaluations are generally unnecessary unless there is a need to exclude other potential causes of hallucinations. Visual hallucinations due to Charles Bonnet Syndrome are generally formed, complex, persistent or repetitive and stereotyped. Insight is fully or partially retained and primary or secondary delusions are absent. No hallucinations in other modalities are seen.

Treatment

There is currently no definitive treatment for CBS. Management primarily involves educating patients and providing reassurance about the benign nature of the condition. In some cases, improving the underlying visual impairment, if possible, may reduce hallucinations. Medications like anticonvulsants or anti-psychotics have been used in severe cases, though with limited success. Cognitive-behavioral therapy (CBT) and techniques to distract or "interrupt" the hallucinations, such as blinking rapidly or moving the eyes, and changing the light intensity can be helpful in some cases.

Conclusion

Charles Bonnet Syndrome is a relatively under-recognized yet common condition in patients with significant visual impairment. Understanding and addressing CBS are crucial in providing holistic care to visually impaired patients, emphasizing the importance of patient education and supportive management. Although no definitive treatment exists, educating patients on the nature of CBS and offering practical techniques can alleviate much of the associated distress⁹. Further research into therapeutic interventions could offer more options for managing CBS in the future.

References

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