

Vivid Visual Hallucinations in Visually Impaired: Charles Bonnet Syndrome – An Analog to “Phantom-Limb Phenomenon”

Sir,

The appearance of visual hallucinations in visually impaired elderly people opens a “pandora’s box” of possibilities. Where the patient and family get unnecessarily concerned about the possibility of insanity, the physicians who are unaware of this condition may also prescribe elaborate investigations. The phenomenon of simple or complex visual hallucinations occurring in patients with vision loss was first described by Charles Bonnet, a Swiss philosopher in the eighteenth century.^[1] He saw his grandfather who suffered from complex visual hallucinations after sustaining vision loss due to cataract. The condition is not uncommon but it is greatly underdiagnosed possibly due to underreporting by Indian patients to avoid a label of insanity. Herein, we describe an elderly male who described a recent onset of vivid visual hallucinations despite having profound vision loss due to long-standing glaucoma.

CASE REPORT

A-79-year-old male complained of visual hallucination for last 1 year. The visual hallucinations included sentences written on the wall, bright coloured flowers in a garden, unfamiliar people moving around him, and the dead people waving at him. For the last few months, he was specifically seeing a Japanese lady waving at him. He described that the image changes with an eye blink. The visual hallucinations included static and moving objects. The images disappeared on closing eyes. Till 2 months back, visual hallucinations were infrequent and lasted for 1–2 min. However, for the last 3 months, the visual hallucinations occurred many times each day and lasted for 15–30 min. He was fully aware that these images were unreal. However, due to sleep deprivation and no respite despite resorting to sleeping pills and with the feeling that he was taken to a surreal world through his blindsight, he requested for medical help. There was no history of auditory or tactile hallucination, delusion, forgetfulness, and change in personality, behavioural problems or any involuntary movements. He had open-angle glaucoma for 25 years (operated 22 years back) that led to progressive bilateral vision loss for the last 5 years. He had hypertension for 2 years that was well controlled with 5 mg of amlodipine.

On examination, his visual acuity was the perception of hand movements at 1 m with either eye. He had intraocular pressure of 14 mmHg in the right eye and 16 mmHg in the left eye. His fundus examination revealed secondary optic atrophy. His higher mental functions were normal. He was aware of the fact that his visual hallucination was unreal. He did not have a mood disorder or any other psychiatric disease.

His blood counts, hepatic, renal and thyroid function tests, electroencephalography, and cranial MRI was normal. In view of complex visual hallucination with preceding visual impairment and normal neurological/psychiatric evaluation, he was diagnosed with Charles Bonnet syndrome (CBS). He was prescribed oral pregabalin 75 mg twice daily and clonazepam 0.5 mg at bedtime. His formed visual hallucination improved over the next 4 weeks. He was last seen 1 month back. He still sees coloured shapes. However, he can sleep normally.

DISCUSSION

Charles Bonnet syndrome occurs in patients suffering from complete or near-complete vision loss. Most patients are elderly people suffering from age-related macular degeneration. However, CBS can affect patients with vision loss secondary to cataract, corneal opacities, retinal abnormalities such as retinitis pigmentosa, diabetic retinopathy, disorders of the optic nerve, optic tract, or visual cortex.^[2] CBS has also been reported among patients with congenital blindness.^[3] The prevalence of CBS among patients with vision loss is higher in the western population ranging from 11–63% as compared to 0.4–1.4% in East Asian countries.^[4,5] A recent study from India showed a prevalence of CBS to vary from 6.7–8.1%.^[6] The exact reason for the higher prevalence of CBS in the western population is not known. Age-related macular degeneration is an important cause of vision loss in the Caucasian population as compared to the Asian population where the cataract is the commonest cause.^[6] The CBS associated with cataract tends to improve after regaining vision following the cataract surgery. Asian patients with visual hallucination do not seek medical attention. One possible reason described is that they fear of getting a label of being insane. Poor awareness of the condition could also be another possible cause of underdiagnosis of CBS in the Asian population.

The exact pathogenesis of CBS is not clear. The most widely accepted theory for CBS is visual deafferentation theory.^[7] Sensory deprivation due to a visual impairment produces spontaneous activation of visual cortices. It is akin to the phantom limb syndrome in which amputees continues to experience pain, discomfort, or other sensory symptoms in the amputated limb due to spontaneous firing of signals in the sensory cortex responsible for sensing the amputated limb. Similarly, in CBS, the primary and secondary visual cortices substitute images when it lacks input from the macula. Functional neuroimaging studies also offer some support to this theory. It has shown that active hallucinations were associated with spontaneous activity in the ventral occipital lobe. Another proposed explanation for CBS is perceptual release theory. When the afferent visual input is decreased below a threshold

level, the brain may allow previously registered subconscious perceptions to surface into consciousness.

There is no definite treatment for CBS. Visual hallucinations of CBS may gradually disappear once the visual deficit is corrected as with cataract surgery.^[8] However, persistent visual hallucinations can be quite disturbing and may affect the quality of life. Most patients do get fearful of having a psychiatric illness. Therefore, clinicians should reassure the patients that the hallucinations are not a sign of mental illness. Non-pharmacological measures such as frequent eye blinking, rapid eye movement from one target to another, or simply looking away may help avoid prolonged visual hallucination. Reassuring patients also helps them to avoid social isolation which is very helpful in most of the cases. In more severe cases pharmacological treatment is necessary. Atypical antipsychotics, antidepressants, anxiolytics, and anticonvulsants have been tried with varied results.^[9] We used pregabalin because, in our experience, it is a potent drug to improve visual hallucinations of CBS. Gabapentin is known to be effective in CBS possibly due to its GABA-ergic action.^[10]

Our patient clearly knew that his visual hallucinations were unreal. He sought medical attention only when the visual hallucination was imposing enough to disturb his sleep. The diagnosis of CBS was made due to the absence of any neurological and psychiatric abnormality.

CONCLUSION

Charles Bonnet syndrome is a relatively benign disorder that usually affects elderly patients with preceding vision loss and presents with simple or complex visual hallucinations. CBS is a diagnosis of exclusion and one should exclude neurodegenerative disorders and psychiatric illnesses before considering CBS. Reassurance is usually enough for mild cases. Atypical antipsychotics, antidepressants, or antiepileptic medications may take care of frequent and prolonged visual hallucinations.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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