Eye-related visual hallucinations: Consider ‘Charles Bonnet syndrome’

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The Charles Bonnet Syndrome (CBS) is typically characterized by visual hallucinations in elderly people without cognitive defects. This article presents the case of an 80-year-old male patient with a one-year history of visual hallucinations, secondary to glaucoma, in both eyes. Neither a dopamine agonist nor cholinesterase inhibitor therapy improved his symptoms. In this case, the hallucinations were gradually improved after administration of a GABAergic drug, pregabalin, for diabetic polyneuropathy. Placebo-controlled clinical trials would be needed to support this effect of pregabalin, as suggested by this association.

Key words: Charles Bonnet syndrome, dementia, hallucinations, pregabalin

Charles Bonnet Syndrome (CBS) is defined by a triad of complex visual hallucinations, oculopathy causing visual deterioration, and normal cognitive status. The strongest risk factors for CBS are bilateral visual system impairment, reduced visual acuity, social isolation, and sensory deprivation. We report a case of CBS who was admitted to our clinic with severe bilateral visual loss because of bilateral glaucoma and complex and repetitive visual hallucinations, which occurred acutely and localized in the external space. CBS is not uncommon and clinicians should consider this syndrome in patients with hallucinations. Once CBS has been diagnosed, the condition should be explained clearly to the patients and their families.

Case Report

An 80-year-old, right-handed man, without personal or familial neuropsychiatric history was brought by his family to our clinic for his hallucinations. The patient had a one-year history of visual hallucinations, including different types of children such as colorfully dressed boys who caress his arms, small girls with red colored nails near him, or riding on a bicycle around him.

He always gave detailed descriptions of his hallucinatory episodes and explained all the scenes as if they were real. He retained a full insight into the unreality of his hallucinations and could stop them by closing or opening his eyes. He said that the ‘scenes’ of children started appearing two years ago and now occur with increasing frequency. Typically, a ‘scene’ would last from a few seconds to a few minutes. He said that although he knew the images were not real, he was not able to control them and this caused distress. He reported that sometimes he felt anxious when he saw these objects. His medical history was significant for diabetes, hypertension, benign prostatic hyperplasia, and hypertension. He was alert, oriented, and cooperative. His pupils were equally round and reactive to light bilaterally. Extraocular movements and cranial nerves were intact and no other neurological deficit could be detected. General physical findings were unremarkable, except his visual deficit. Five years ago he had been diagnosed with glaucoma in both eyes. His vision had gradually deteriorated for one year, necessitating full assistance from his wife and children in his daily activities. He obtained 26 points on the Mini-Mental State Examination (MMSE), excluding items requiring intact vision. This score was indicative of intact cognition. His laboratory test results such as complete blood count and chemistry panel were within normal limits. Brain magnetic resonance imaging (MRI) suggested mild cortical atrophy and electroencephalography (EEG) was found to be within normal limits. He had been diagnosed with primary dementia before he was admitted to our clinic. Although donepezil had been started for dementia (at the dose of 5 mg/day) and quetiapine had been added for hallucinations (at the dose of 50 mg/day), he showed no change in his symptoms. We started pregabalin at a daily dose of 75 mg for diabetic neuropathy. After 15 days of pregabalin therapy, his hallucinations gradually disappeared. Donepezil and quetiapine treatment was discontinued and during the six months of follow-up, the hallucinations did not recur.

Discussion

The Charles Bonnet Syndrome is characterized by complex visual hallucinations in otherwise psychologically normal people. Estimates of the prevalence of CBS in different samples vary from a small percentage (around 0.4%), to a relatively large percentage (about 27%). Although Charles Bonnet first described the hallucinatory experiences of his grandfather Charles Lullin in the eighteenth century; the term Charles Bonnet Syndrome was first used by De Morsier in 1967. De Morsier defined the condition as the occurrence of visual hallucinations in elderly people with intact cerebral function.

Many risk factors have been associated with hallucinations including social isolation, decreased quality of life, stress and fatigue. These risk factors seem to be the precipitating factors of CBS, in those who have visual disturbance. In an article by Jackson et al. it is reported that hallucinations can diminish with improved acuity, increased social contact or individual medications. However, in their study, Khan et al. could not demonstrate a clear association with living alone. Our patient was not alone, he was living with his wife and children.
The Charles Bonnet Syndrome is more common in the elderly, although it can occur in patients of any age. If physicians cannot recognize it in clinical practice, this syndrome can be interpreted as mental illness. Our case was an 80-year-old man, with normal neurological examination, Electroencephalography (EEG), magnetic resonance imaging (MRI), and mini-mental state examination (MMSE), who had been started on antidiemntal and antipsychotic drugs, without change in the report of hallucinations. The mechanism by which visual hallucinations are produced is unclear. It is suggested that reduced or absent stimulation of the visual system leads to decreased responsiveness in the occipital cortex. Studies of cerebral perfusion during actual hallucinatory experiences suggest that the temporal cortex, corpus striatum, and thalamus are the regions most likely to be responsible for the genesis of visual hallucinations.

The patients with CBS retain full insight into the unreality of their hallucinations. Our patient also experienced complex hallucinations and he described his hallucinations as pleasant; similarly de Morsier has stated that most of the patients found the experience to be pleasant.

Patients with CBS have often ophthalmic pathology, such as, glaucoma, central retinal artery occlusion, and optic neuritis. Our patient had glaucoma in both eyes for five years. When his hallucinations started to occur last year, he had poor vision. Toxic and metabolic disorders are also causes of visual hallucinations. Intracranial tumors, epilepsy, psychiatric disorders, delirium, and neurodegenerative diseases, such as, Parkinson’s disease, Lewy-body dementia, Alzheimer’s, and frontotemporal dementia can be neurologic causes for hallucinations in geriatric age groups. Our case was first diagnosed as possible primary dementia and the patient was given antidemential and antipsychotic drugs. However, he did not have other clinical signs of dementia. Furthermore, the antipsychotic treatment did not produce a significant change in his hallucinations. On account of the pattern, the etiology of hallucinations was attributed to CBS.

There is no effective pharmacotherapy for CBS. First, non-pharmacological interventions like improving the home lighting in the evening and reducing social isolation should be useful. After the non-pharmacological options are exhausted, pharmacological agents such as anticonvulsants (carbamazepine, valproic acid or gabapentin), antipsychotics, antidepressants or cholinergic agents are reported to be effective in some case reports. Pregabalin is the first drug to receive approved labeling from the Food and Drug Administration (FDA) for the treatment of painful diabetic neuropathy and postherpetic neuralgia and is the first antiepileptic agent to receive FDA-approved labeling since 1999. Pregabalin is a more specific α2-δ ligand with a six-fold higher binding affinity than gabapentin. In our patient pregabalin was started at a daily dose of 75 mg for diabetic neuropathy. His symptoms of hallucinations gradually disappeared after that treatment. To support this effect of pregabalin, placebo-controlled clinical trials are needed.

In conclusion, after the other serious life-threatening causes of complex visual hallucinations are excluded, CBS should be kept in mind. We can prevent the patient from unnecessary laboratory testing or hospitalizations if an accurate clinical diagnosis can be made by the physician.

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