Charles Bonnet Syndrome: Two Case Reports

To the Editor: Charles Bonnet syndrome is a rare disorder with estimated prevalence of 0.8% in low vision group,[1] 0.6% in the elderly,[1] and 0.4% in Asians.[2] True Charles Bonnet syndrome is rare and most cases have Charles Bonnet syndrome plus.[3] These are supposed to be release hallucinations that occur when the brain does not get stimulations from the eyes. Despite many case reports, there is a lot to learn about the syndrome and the exact pathology is still unknown.[4] Here we describe two cases of true Charles Bonnet syndrome that presented with different anatomical lesions.

Case 1

A 60-year-old man presented with a 6-year history of abnormal behavior and staggering gait; illness was progressive and insidious in onset. He was allegedly seeing some insects that others could not and he could clearly demarcate them from the surroundings. The bugs were present everywhere and he became distressed as he tried to remove them from his clothes and food without success. He washed his hands very frequently to get rid of them. His wife complained that in the last few days before the consultation his gait had changed and he had collided with objects on either side of his path. His medical and psychiatric histories were unremarkable. His neurological examination revealed the presence of bitemporal heteronymous hemianopia and a CT scan of the head showed optic chiasmal meningioma. The patient was sent for neurosurgery.

Case 2

A 75-year-old man came to us with the complaint of seeing some faces that other could not see. He did not have sight in his right eye due to an injury during adulthood, but he could see a face with the right eye and was able to describe it in detail. The face was constantly present and was only relieved when he went to sleep. For the previous 5 years he had developed similar visual hallucinations in the other eye, which progressed gradually. In the left eye he began to see a plethora of faces with distorted features that moved toward him and disappeared when they reached him. He could delineate every face from the crowd. He could relieve this vision by widening his eyes, but the problem increased if he blinked or kept his eyes half closed. The images disappeared during sleep. His medical and psychiatric histories were unremarkable and neurological examination revealed the constriction of the visual field in all quadrants. He was sent for ophthalmologic examination and his visual acuity was found to be 20/60 in the left eye. Ophthalmoscopic examination disclosed primary optic atrophy in the left eye. No cause could be identified for this atrophy as the patient declined a neuroradiological examination. The right eye had corneal opacity making fundoscopy impossible.

Discussion

Charles Bonnet syndrome is diagnosed when a person has complex, persistent or repetitive, stereotyped visual hallucinations, and has insight into the disease in the absence of other psychopathology.[5,6] Poor vision and sensory deprivation are common precipitants.[5,6] The syndrome has to be differentiated from the other causes of complex visual hallucinations that occur transiently (e.g., hypnogogic hallucinations, epileptic phenomenon, treated Parkinson's disease, Lewy body dementia, and migraine) or causes of persistent complex visual hallucinations (e.g., peduncular hallucinations that occur due to the brainstem damage, delirium tremens).[7] None of these were present in our patients. Most patients retain full insight into hallucinations and can stop them by closing or opening their eyes,[7] as evidenced by our second case.

Hallucinations in Charles Bonnet syndrome have been reported to occur due to damage to the visual pathway, as in partial blindness, after cataract surgery, and in optic nerve damage caused by sella tursica meningioma.[6,8] These kinds of visual pathologies were present in both of our patients. The hallucination content appears to be stable across cultures; distorted faces with prominent eyes and teeth are commonly reported[9] and hallucinations of insects are also known.[6] Despite an exhaustive search we could not find any literature that could correlate particular types of visual hallucinations with the particular neuropathology. Hence we still need to learn more about Charles Bonnet syndrome and more cases are required to understand the pathology.

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Risperidone Treatment of Complex Hallucinations in a Patient With Posterior Cortical Atrophy

To the Editor: Posterior cortical atrophy is a clinical syndrome in which Alzheimer's disease seems to be the major pathologic cause.[1–4] This progressive dementia initially presents with visual disturbances, including some or all features of Balint's syndrome and Gerstmann's syndrome, with a later onset of cognitive decline associated with typical Alzheimer's disease.[1–4] Postmortem studies show predominant occipitoparietal atrophy generally associated with neuritic plaques and neurofibrillary tangles.[3]

Visual hallucinations occur in up to 25% of patients who are diagnosed with posterior cortical atrophy.[1,4] We report a case of a patient with posterior cortical atrophy who was experiencing intolerable complex hallucinations that were successfully treated with risperidone after an unsuccessful trial of quetiapine.

Case Report

A 66-year-old married, Caucasian woman with a 10-year history of cognitive decline, characterized by early onset of visual impairment not associated with anterior visual pathology, was brought to the psychiatric emergency department by her daughter for severe mental distress caused by visual and auditory hallucinations. The hallucinations consisted of a young, vulgar girl who reportedly stole the patient's clothing, demanded to eat the patient's food, falsely informed the patient that her husband was physically ill, and threatened to kill the family pet. The patient was often awake at night agitated and responding to the hallucinations.

The hallucinations began about 8 weeks earlier. Two weeks prior to the emergency department visit, the patient was prescribed quetiapine, 50 mg b.i.d., by her primary care physician. Her medications also included memantine, 10 mg b.i.d., donepezil, 10 mg daily, and venlafaxine XR, 150 mg daily.

Upon examination, the patient had Balint's syndrome, Gerstmann's syndrome, ideational and ideomotor apraxia, prosopagnosia, fluent aphasia with anomia, and demonstrated paraphasias and neologisms. She had difficulty maintaining and shifting set. A physical exam was unremarkable, but notable for the absence of parkinsonian signs such as cogwheel rigidity, stooped posture, shuffling gait, bradykinesia, and resting tremor. A brain CT showed diffuse cortical atrophy with a pronounced parieto-occipital predomination. The patient was diagnosed with probable Alzheimer's disease with posterior cortical atrophy syndrome.

The patient was admitted to the inpatient medical psychiatry unit and treated with risperidone, 1 mg h.s. Quetiapine was discontinued and her preadmission medications were continued. Initially, the patient was often found verbally responding to the hallucinations, was tearful, and was not able to be redirected. Over the course of 10 hospital days, the hallucinations significantly decreased in frequency and severity. She was transferred to an assisted living facility with no further subjective concerns about the hallucinations.

Discussion

Posterior cortical atrophy is a rare but disabling dementia syndrome. Our patient also had a complex visual and auditory hallucination leading to a delusional system which substantially decreased the quality of her life and was detrimental to her mental health. A literature search of OVID and PsycINFO yielded no reports for treatment of psychotic symptoms in patients with posterior cortical atrophy. Initiation of risperidone, 1 mg h.s., was well tolerated for this patient, significantly reduced her psychotic symptoms, and improved her quality of life.

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