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CLINICAL COMMUNICATIONS TO THE EDITOR

Visual Hallucinations from Age-Related Macular Degeneration

To the Editor:

Visual hallucinations occur in various settings, including: cerebral ischemia, epilepsy, metabolic and endocrine disorders, psychiatric disease, and other forms of neurologic disease.¹⁻⁴

A considerable proportion of visually impaired individuals often with age-related macular degeneration (ARMD) experience formed hallucinations, which is termed "Charles Bonnet syndrome" (CBS). The term was coined by de Morsier⁵ recognizing the biologist Charles Bonnet, who described the hallucinatory experiences of his grandfather Charles Lullin.

CBS has been described and defined in a multitude of ways, with the most common being the occurrence of complex visual hallucination in individuals with preserved insight and intellectual function without altered consciousness, cognitive or psychiatric disturbances, sleep disorders, or focal neurologic lesions and often associated with ophthalmic pathology. Non-delirious patients who admit to visual hallucinations in the setting of visual impairment from ARMD often undergo potentially unnecessary extensive medical work-up.

CASE ONE

An 83-year-old white female was referred for retinal evaluation and had a known diagnosis of ARMD. Her presenting visual acuity was 20/400 in the right eye and 20/30 in the left eye. Anterior segment examination was remarkable for posterior chamber intraocular lens implants in both eyes. Posterior examination of the right eye on clinical examination and fluorescein angiogram revealed a greater than 50% subfoveal classic choroidal neovascular membrane. Posterior segment examination of the left eye revealed dry ARMD with multiple large drusen. Over a period of 9 months the patient underwent a total of 3 photodynamic therapy treatments for the right eye. She later developed a macular scar in the right eye and remained with dry ARMD in the left eye. Her medical history was significant for non-insulin-dependent diabetes mellitus, hypertension, and emphysema. Her medications included glyburide, Advair (GlaxoSmithKline, Research Triangle Park,

NC) and Combivent (Boehringer Ingelheim Corporation, Ridgefield, CT) inhaler, lisinopril, and valsartan.

During a follow-up visit, the patient noted that for the past year she had episodes in which people suddenly appeared in her house and suddenly disappeared. These episodes lasted approximately 2 minutes and usually occurred weekly. The patient revealed that she had explained this to her internist, who had performed a thorough evaluation including magnetic resonance imaging of the head, magnetic resonance angiography, psychiatric evaluation, and laboratory evaluation including thyroid levels, fluorescent titer antibody, and a drug panel. All testing results were unremarkable, and psychiatric evaluation also was unremarkable. The patient explained that she had been hesitant to reveal these visual hallucinations and was unaware of CBS. After a thorough explanation of CBS, the patient's anxiety over these visual hallucinations was relieved, and her internist and psychiatrist were notified of this syndrome.

CASE TWO

A 79-year-old white female was seen for a second opinion; she had a known diagnosis of ARMD. Presenting visual acuity was 20/100 in the right eye and 20/80 in the left eye. Anterior segment examination was remarkable for posterior chamber intraocular lens implants in both eyes. Posterior segment examination of both eyes revealed large drusen within the fovea, with retinal pigment epithelium stippling.

Her medical history was significant for hypertension and hypercholesterolemia. Her medications included diltiazem and simvastatin. The patient noted that she had intermittent episodes occurring 1 to 2 times per week and lasting 1 to 2 minutes, in which she saw people of Latino descent in her home. She had previously explained this to her general practitioner, who performed a thorough evaluation including magnetic resonance imaging of the head, magnetic resonance angiography, psychiatric evaluation, and laboratory evaluation including electrolytes, blood urea nitrogen, creatinine, thyroid function test, fluorescent titer antibody, vitamin B-12, folate, drug panel, and lumbar puncture. All testing results were unremarkable, as was the psychiatric evaluation.

The patient was given a thorough explanation of CBS, and her anxiety over these visual hallucinations was relieved. Her general practitioner and psychiatrist were notified of this syndrome.

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DISCUSSION

Patients are often reluctant to admit to their visual hallucinations for fear of being diagnosed with a mental illness. Therefore, the prevalence of CBS is likely to be underestimated.⁶⁻⁹ CBS frequently goes unrecognized in clinical practice.¹⁰⁻¹² CBS is often unfamiliar to medical personnel, and there is a tendency to misdiagnose mental illness.^{3,10,11,13} In one series only 16 of 60 patients with CBS had consulted a doctor about their hallucinations, and only 1 case was diagnosed correctly.

The content of these hallucinations has been described in many ways. The most common image is that of a person,⁸ but distorted faces, animals, and figures also have been described.² They are always localized in external space, and the contents are well organized, defined, and clear.^{9,14,15} The typical CBS hallucination has been described variously as a sudden sharply focused, immobile image, most often the face or figure of a person, which occurs when the patient is alert, with eyes open, and vanishes spontaneously after a period of seconds.¹⁶ The hallucinations typically occur independently of any triggering factors or desire to generate the image,^{16,17} although in some individuals they may be triggered by a wide variety of stimuli such as fatigue, stress, or bright light images last for periods ranging from seconds to minutes or hours and subsequently disappear spontaneously or in response to actions such as closing the eyes.^{2,4,12,15,18,19}

Reactions to these visual experiences vary from pleasant indifference to curiosity and even terror.^{14,20} Full insight into the unreality of their hallucinations may not be immediate because patients may have a period of initial deception.^{14,21} The most commonly associated ocular pathology is ARMD,² although CBS has been documented in the context of visual impairment anywhere along the visual pathway including cataract, choroideremia, corneal opacities, retinal detachment, optic neuritis, and so forth. These visual hallucinations do not occur in those born blind but only in the context of acquired visual impairment.²²

Visually impaired individuals are not immune to hallucinations secondary to other neuropsychiatric conditions or emotional disturbances such as Alzheimer disease, delirium, Parkinsonism, and schizophrenia, to name a few.^{2,23} Once hallucinations have been documented, thorough ophthalmic and neurologic examination should be undertaken to determine any potentially treatable causal pathology.²⁴ However, in the nondelirious patient a simple test of cognitive function such as the Mini-Mental State Examination may be adequate, with further psychiatric evaluation perhaps indicated only in the context of cognitive impairment. Because CBS frequently goes unrecognized in clinical practice, it is important for the primary care practitioners to recognize the potential for hallucinations in these patients and that further work-up may not be necessary when these patients have full insight into the unreality of these hallucinations. Recognition of this syndrome could save limited health care dollars, relieve patient anxiety, and save time wasted on unnecessary testing and evaluation.

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