

Ophthalmoproblem

Jason Blair, MBA Sanjay Sharma, MD, MSC, MBA, FRCSC

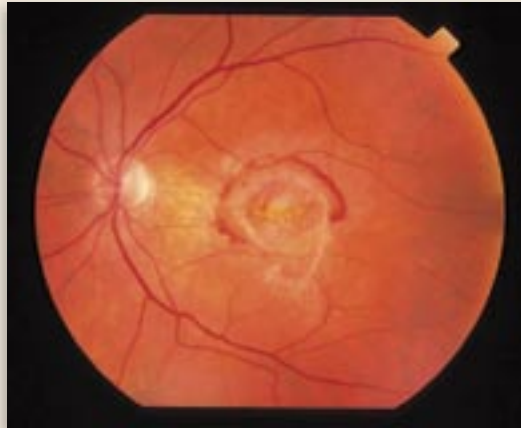


Photo credit: Ophthalmic Photography, Hotel Dieu Hospital, Kingston, Ont.

An alert and coherent 80-year-old woman comes to your office quite upset. She complains of seeing images of trees and water. You know she has bilateral age-related macular degeneration (ARMD) that has progressed rapidly over the last 2 years. Her poor vision recently dictated a move into a nursing home. She says the “scenes” of trees and water started appearing several weeks ago and now occur with increasing frequency. Typically, a “scene” will last from a few seconds to a few minutes. She says she knows the images are not real.

The woman’s medical history shows she has had a transient ischemic attack, but she is otherwise healthy. She takes acetylsalicylic acid daily. She says the nursing home recently started giving her sleeping pills because of her increased agitation.

On examination, her vital signs are within normal limits, and she is afebrile. Results of urinalysis are within normal limits, as is her neurologic examination. Her visual acuity is limited to seeing hand movements with the right eye and light with the left. Anterior segment examination is unremarkable. The fundus of her left eye is shown above; the fundus of her right eye is similar in appearance.

Given these symptoms and signs, the most likely cause of the visual disturbances is:

1. A transient ischemic attack involving the occipital lobes
2. Delirium
3. Vitreous traction related to progression of ARMD
4. Hallucinations secondary to use of sleeping pills
5. Charles Bonnet’s syndrome

Answer on page 1370

Mr Blair is a fourth-year medical student at Queen’s University in Kingston, Ont.

Dr Sharma is an Associate Professor of Ophthalmology and an Assistant Professor of Epidemiology at Queen’s University.

Answer to Ophthalmopuzzle continued from page 1365

5. Charles Bonnet's syndrome

Charles Bonnet's syndrome (CBS) is the most likely cause of well formed visual hallucinations experienced by elderly people with severe visual impairment and no other findings. Patients know the hallucinations are not real. In this case, the fundus photograph serves only to demonstrate the severity of this patient's ARMD. The diagnosis is made clinically.

Charles Bonnet's syndrome is "seen in otherwise intact, elderly individuals with usually binocular visual loss, comprising exclusively visual, well formed hallucinations of people, places, and things, but devoid of emotional or threatening content; these 'scenes' are superimposed on the actual visual environment, usually last only a very few minutes, and are clearly understood by an insightful patient to be hallucinations that do not interfere with normal mental function."¹ The hallucinatory phenomena are thought to be related to sensory deprivation and have been compared to phantom limb pain.

Prevalence of CBS in visually handicapped people is thought to be between 11% and 15%.² It seems, though, that many of these patients (73% in one study³) conceal their extraordinary visual experiences from others, including their physicians.^{3,4} Teunisse et al found that, for most patients, this was because they feared being diagnosed with mental illness.³ Teunisse et al also found that CBS is often misdiagnosed. Of the 16 patients who consulted doctors about their hallucinations, only one patient received the proper diagnosis of CBS.³ The syndrome is more common in elderly people, likely due to the increased incidence of profound visual loss in that age group.²

Hallucinations

A typical CBS hallucination, according to Menon et al,² occurs as a sharply focused, immobile, well formed image that appears when patients are alert and have their eyes open and vanishes

spontaneously after a few seconds.² The most common images seen are disembodied faces, small costumed figures, animals, and branching structures.² In the context of visual impairment, the clarity of the scenes contrasts sharply with the blurred perception of real objects.² Almost all CBS hallucinations are exclusively visual and have no auditory component.²

According to a recent review, most investigators agree that patients with CBS retain enough insight to know that their hallucinations are not real.² This knowledge, however, might not always be immediate. The reviewers also noted that a few studies found that many CBS patients possess only partial or fluctuating insight.²

Diagnosis

Universal diagnostic criteria for CBS have not yet been established. A recent review² highlighted the criteria set forth by German authors Podoll et al.⁵ Translated, the criteria are as follows.

- The predominant symptom is the occurrence of visual hallucinations in elderly people in normal mental health.
- There is no evidence of delirium, dementia, negative effect on intellectual capacity, deterioration as in affective syndromes, paranoid developments, psychosis, intoxication, or neurologic disease.
- Loss of vision as a consequence of ocular disease is found in most cases as a specifying factor, but is not obligatory for diagnosis.

The article² also pointed to diagnostic criteria proposed by Gold and Rabins.⁶

- Visual hallucinations are formed, complex, persistent or repetitive, or stereotyped.
- Insight is fully or partially retained.
- Primary and secondary delusions are absent.
- Hallucinations in other modalities are absent.

In diagnosing CBS, it is important to exclude other possible causes of formed visual hallucinations. Some of these are delirium, Alzheimer disease, parkinsonism and levodopa-induced hallucinations, dementia with Lewy bodies, schizophrenia,

cerebral peduncle disease, epilepsy, medication effects, alcohol withdrawal, and hallucinations during sleep-wake transitions.^{2,4}

While CBS patients have insight into the unreality of the hallucinations, the emotional effect of the images is a separate matter. Several studies reported that most patients had a negative emotional response to the hallucinations that was compounded by being unaware of CBS.^{3,4} Most patients were relieved when reassured that the condition is not a psychiatric disorder, but a recognized phenomenon.³ Indeed, education and reassurance are the cornerstones of treatment.

Management

Patients diagnosed with CBS need to be informed that hallucinations are a common and benign phenomenon in visually impaired people. They should be reassured that they are not mentally ill. Better lighting, increased social activity, and pursuing interests might reduce hallucinatory activity.^{2,3} Eventually, in most people, the hallucinations cease spontaneously, usually as vision deteriorates further.² For frequent, nonresolving, or distressing hallucinations, pharmacotherapy has been used, but with mixed results. Carbamazepine, clonazepam, valproic acid, and gabapentin are among the agents that have been tried.²

Recommendation

Patients with deteriorating vision should be asked whether they are having hallucinations. Those not having symptoms should be warned about the possibility of hallucinations. Those having formed hallucinations should be screened for causes other than CBS; follow up is important to exclude causes such as Lewy body dementia. Those with unformed visual hallucinations, such as light flashes, vertical streaks, and new floaters, should be immediately referred to an ophthalmologist to rule out treatable retinal disease. Once CBS has been diagnosed, the condition should be explained clearly to patients and their families.



References

1. Tasman W, Jaeger EA, editors. *Duane's clinical ophthalmology*. Rev ed. Philadelphia, Pa: Lippincott-Raven Publishers; 1996.
2. Menon GJ, Rahman I, Menon SJ, Dutton GN. Complex visual hallucinations in the visually impaired: the Charles Bonnet Syndrome. *Surv Ophthalmol* 2003;48:58-72.
3. Teunisse RJ, Cruysberg JR, Hoefnagels WH, Verbeek AL, Zitman FG. Visual hallucinations in psychologically normal people: Charles Bonnet's syndrome. *Lancet* 1996;347:794-7.
4. Brown GC, Murphy RP. Visual symptoms associated with choroidal neovascularization: photopsias and the Charles Bonnet syndrome. *Arch Ophthalmol* 1992;110:1251-6.
5. Podoll K, Osterheider M, Noth J. [The Charles Bonnet syndrome.] *Fortschr Neurol Psychiatr* 1989;57:43-60.
6. Gold K, Rabins PV. Isolated visual hallucinations and the Charles Bonnet syndrome: a review of the literature and presentation of six cases. *Compr Psychiatry* 1989;30:90-8.

—*—*—