



A 69-Year-Old Man with Complex Nocturnal Visual Hallucinations

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A 69-year-old man with diabetic retinopathy presented with a 6-month history of nightly sleep-related hallucinations. He awakened in the middle of the night to recurrent vivid images that he realized were not real: a beautiful blonde woman standing across the room, large colorful spiders moving across the wall, or a Christmas tree rustling in his closet. Images were never frightening, persisted for seconds to minutes, and vanished after putting on his glasses or turning on the lights. Hallucinations had no auditory or tactile component, and there were no symptoms or signs of a psychiatric disorder. He had no seizure history or epilepsy risk factors such as febrile seizures, head injury, meningoencephalitis, or family history of seizure disorders. He denied tremor, cognitive concerns, or symptoms suggesting dysautonomia. He

denied excessive daytime sleepiness, and there were no ancillary symptoms suggesting narcolepsy. There was no history suggesting dream enactment behavior or other parasomnia behaviors.

Neurologic examination demonstrated poor visual acuity and mild length-dependent sensory loss in the extremities consistent with peripheral neuropathy. Short Test of Mental Status¹ score was normal although he missed 3 of 4 short-term recall items. No parkinsonian features were present on examination. MRI of the brain and screening overnight oximetry were normal. Recent blood work demonstrated hemoglobin A1C 10.3%.

QUESTION: What is your diagnosis?

ANSWER: Charles Bonnet Syndrome: complex visual hallucinations in the visually impaired.

Charles Bonnet Syndrome (CBS) is characterized by complex, recurrent visual hallucinations in patients with poor vision. Patients maintain insight into the false nature of the hallucinations.

CBS can occur with pathology at any level of the visual system. Common causes include: macular degeneration, cataracts, retinopathy, and glaucoma. Risk increases with severity of visual loss and increased age. Reported prevalence ranges from 11% to 27% in elderly patients with visual loss. The pathophysiology of CBS is thought to stem from external sensory deprivation (from vision loss). This results in a cortical release phenomenon, allowing endogenous activation of the visual cortex, leading to hallucinations.

CBS may initially present to the sleep physician. Reductions in visual input are exaggerated in the dark, causing events to occur primarily at night. Reassurance and optimization of vision are the cornerstones of management. Pharmacologic intervention may be used in cases where hallucinations are associated with fear or have a negative impact on quality of life.² Given a link between CBS and incipient dementia, long-term neurologic follow-up is important.³ Lapid et al. followed 77 patients with CBS; 26% eventually developed dementia, most commonly dementia with Lewy bodies.⁴ The cardinal features of dementia with Lewy bodies are: dementia with fluctuating cognition, formed visual hallucinations, and parkinsonian features.⁵

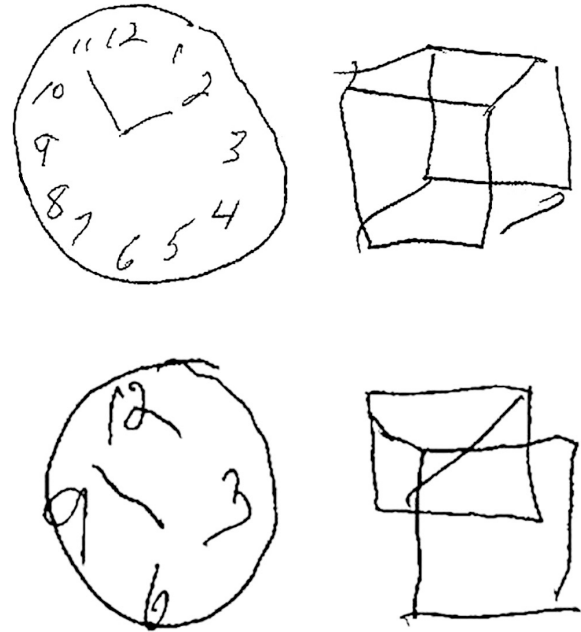
FOLLOW-UP

Our patient was advised on measures to optimize vision through use of corrective lenses and adherence to glycemic control. Use of a nightlight led to cessation of events. CNS Vital Signs and Useful Field of View computerized neurocognitive testing demonstrated performance in the 19th percentile, with reduced domain scores in verbal memory, psychomotor processing speed, attention, and reaction time.⁶ Serial evaluations revealed further reductions in cognitive abilities. At 3 years following symptom onset, the patient had developed emergence of daytime hallucinations, and neuropsychologic testing demonstrated visuospatial impairments suggestive of evolving dementia with Lewy bodies (**Figure 1**).

SLEEP MEDICINE PEARLS

1. CBS is relatively common in older individuals with visual loss. Hallucinations typically occur at night; patients may present initially to the sleep medicine physician.
2. Reassurance and optimization of vision are cornerstones of management.
3. Neurologic follow-up over time is crucial given association between CBS and dementia, particularly dementia with Lewy bodies.

Figure 1—Progressive visuospatial impairments.



The patient was asked to draw a clock (placing hands at “10 after 11”) and to copy a cube. (Top) Baseline evaluation is normal. (Bottom) Evaluation 18 months later demonstrates deterioration in visuospatial and construction abilities. Combined with evolving daytime hallucinations and cognitive decline, this suggests a diagnosis of dementia with Lewy bodies.

CITATION

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DISCLOSURE STATEMENT

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