CHARLES BONNET SYNDROME

Suzanne Collier, MD
ETSU Psychiatry PGY-IV
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CASE PRESENTATION

• 87 year old Caucasian female

• Patient seen as consult within hours of admission

• Admitted by her PCP, whom she’d seen as outpatient earlier that day

• Reason for admission and consult: “psychosis, altered mental status”
HISTORY OF PRESENT ILLNESS

• Patient accompanied by her daughter, who assists her mother in providing HPI and also provides collateral information.

• Visual hallucinations for 3 years.

• Initially VH occurred only at nighttime.

• VH were of animals that appeared smaller than they actually are, such as small cows, cats, horses, bears.

• Lilliputian hallucinations.

• Not frightening to patient.
HPI, CONTINUED

• In the last 2 to 3 months prior to admission, VH have been occurring in the daytime as well as nighttime

• In the last 3 to 4 weeks, she has begun seeing VH of men in her apartment (day and night)
  • Had good insight that they were not real, but felt frightened nonetheless

• More recently, she’s begun having VH of more and more people
ADDITIONAL HISTORY

• Patient’s son died 3 years ago around the time that VH began
• Patient diagnosed with macular degeneration 3 to 4 years ago
• Managed by PCP for depressive disorder and anxiety
  • Prescribed Xanax 0.5mg PO QHS and Zoloft 50mg PO Qday
  • Does not take either of these medications regularly
• No other past psychiatric history
DIFFERENTIAL DIAGNOSIS FOR COMPLEX VISUAL HALLUCINATIONS

• Hypnagogic hallucinations
• Peduncular hallucinosis
• Delirium tremens
• Parkinson’s disease and Lewy body dementia
• Migraine coma
• Visual field disturbance and Charles Bonnet syndrome
• Focal epilepsy
# Complex Visual Hallucinations

<table>
<thead>
<tr>
<th>Disease</th>
<th>Special features of hallucinations</th>
<th>Duration</th>
<th>Consciousness</th>
<th>Insight</th>
<th>Sleep</th>
<th>Lesion</th>
</tr>
</thead>
<tbody>
<tr>
<td>Hypnagogic hallucinations</td>
<td>On falling asleep</td>
<td>Seconds to minutes</td>
<td>Drowsy</td>
<td>Usually preserved</td>
<td>Associated with narcolepsy</td>
<td>Brainstem in secondary cases</td>
</tr>
<tr>
<td>Peduncular hallucinosis</td>
<td>More often in evening. Any part of visual field. Rarely polymodal.</td>
<td>Often prolonged</td>
<td>Normal</td>
<td>Usually preserved</td>
<td>Disturbed</td>
<td>Brainstem or thalamus</td>
</tr>
<tr>
<td>Delirium tremens</td>
<td>Very variable hallucinations may be polymodal. Autonomic instability.</td>
<td>Often prolonged</td>
<td>Agitated and confused in later stages</td>
<td>Often reduced in later stages</td>
<td>REM overflow, with little sleep</td>
<td>None</td>
</tr>
<tr>
<td>Parkinson's disease and Lewy body dementia</td>
<td>Often in the evening. Any part of visual field. Rarely polymodal.</td>
<td>Minutes</td>
<td>Not unconscious, normal, or drowsy/inaccessible</td>
<td>Usually preserved</td>
<td>Reduced REM sleep</td>
<td>Widespread, cortex and brainstem</td>
</tr>
<tr>
<td>Migraine coma</td>
<td>Noted during recovery from coma</td>
<td>Up to 2 days</td>
<td>Usually normal at time of hallucination but may be depressed</td>
<td>Preserved</td>
<td>Unknown</td>
<td>Ataxia in some cases</td>
</tr>
<tr>
<td>Visual field disturbance and Charles Bonnet syndrome</td>
<td>Localized to disturbed visual field and often in early morning or evening</td>
<td>Prolonged</td>
<td>Normal</td>
<td>Preserved</td>
<td>Normal</td>
<td>Visual pathway from retina to striate cortex</td>
</tr>
<tr>
<td>Focal epilepsy</td>
<td>Brief, stereotyped. May be localized to part of visual field. May have other epileptic features. Normal between episodes</td>
<td>Usually seconds</td>
<td>Often impaired</td>
<td>Usually preserved</td>
<td>Normal</td>
<td>Posterior temporoparietal</td>
</tr>
</tbody>
</table>
IMAGING AND OTHER STUDIES

• **MRI brain without contrast:** No evidence of acute intracranial process. White matter foci that are nonspecific and most commonly associated with chronic small vessel ischemic change.

• **CT head without contrast:** No acute intracranial abnormalities appreciated.

• **Chest x-ray:** No acute change.

• **EEG:** 20 minute EEG, no evidence of epileptiform or seizure activity.
WHO WAS CHARLES BONNET?
HISTORY OF CHARLES BONNET SYNDROME

• 1769 - Charles Bonnet documented VH experienced by his 89 year old grandfather, Charles Lullin, who had cataracts
  • Bonnet was a Swiss naturalist, philosopher, and biologist.

• 1936 - Georges de Morsier, a neurologist, coined the eponym Charles Bonnet syndrome in recognition of Bonnet.
  • De Morsier defined CBS as visual hallucinations that occur in older people with otherwise intact mental functioning.
  • Vague diagnostic criteria
 CBS DIAGNOSTIC CRITERIA

- Diagnosis of CBS involves visual hallucinations that occur in older individuals who
  - (1) have intact mental function;
  - (2) do not have dementia, delirium, psychosis, or neurological diseases; and
  - (3) may or may not have ocular diseases

- The majority of criteria require that the person have insight.

- Other proposed/debated criteria: absence of other hallucinations (auditory, olfactory); absence of control over hallucinations; disappearance of hallucinations upon closing the eyes; with no additional delusions
PATHOPHYSIOLOGY

• VH can occur in patients with visual acuity loss or visual field loss from any cause, affecting any part of the visual pathway from the eye to the visual cortex

• Common underlying conditions include age-related MD, glaucoma, diabetic retinopathy, and cerebral infarction

• CBS does not occur with congenital blindness
• Hallucinations occur when visual sensory deafferentation leads to disinhibition of visual cortical regions, which then fire spontaneously

• Similar hallucinations have been reported by individuals subjected to visual deprivation experiments

• fMRI studies offer some support for this theory:
  
  • A 1998 fMRI study revealed that active hallucinations were associated with spontaneous activity in the ventral occipital lobe
  
  • The content of the hallucinations was associated with specific regional activation that correlated with the known specialized function of that area of the visual cortex
  
  • The neurobiology underlying VH in CBS has not been elucidated - possible denervation hypersensitivity?
PREVALENCE OF CBS

- Menon's 2003 study found that the prevalence of complex visual hallucinations in people with visual impairments is between 11% and 15%.

- In 2008 Kahn, using the criteria established by Teunisse et al. (1995), reported that the prevalence of CBS hallucinations in persons with end-stage AMD was as high as 27%.

- Scott, Schein, Feuer, and Folstein's (2001) study on visual hallucinations in persons with retinal disease found that hallucinations were common among those with retinal disease; were underdiagnosed; and were not related to abnormal personality traits, cognitive deficits, or histories of personal or familial psychiatric morbidity.
A PERSONAL ACCOUNT
A PERSONAL ACCOUNT
CLINICAL FEATURES

• Diagnosis of ocular disease is generally established for at least one year before hallucinations emerge

• The likelihood of release hallucinations increases with lower visual acuity
  • Prevalence of hallucinations appears to increase with acuity worse than 20/60
  • More likely to occur with binocular versus monocular disease
CLINICAL FEATURES

- Release hallucinations
  - Can be simple, nonformed images such as lines, light flashes, or geometric shapes
  - Can be complex, formed images of people, animals, or scenes
- Content can be variable, even for individual patients
DESCRIPTION OF VH

• Images are usually colored and may be static, animated, or move en bloc across the visual field

• Usually do not have emotional impact or personal meaning

• No associated auditory or other sensory modality hallucinations

• Occur more often with the eyes open than closed

• Most complex VH occur in the setting of sensory deprivation
DIAGNOSIS

• Complete neurologic evaluation

• In absence of known eye disease, complete ophthalmologic evaluation with visual field testing

• Brain MRI if there is visual field deficit or other focal neurologic deficit

• If atypical features for CBS, further evaluation is needed
TREATMENT

• Reassurance

• Teach patients how to suppress hallucinations
  • close eyes
  • look away from the hallucination
  • increase visual stimuli (increased illumination)
  • increase social interaction
TREATMENT, CONTINUED

• Patients with continuous hallucinations or disturbing imagery may need specific treatment

  • **SSRIs**
  
  • antipsychotic medications (low dose olanzapine or quetiapine)

  • cholinesterase inhibitors

  • antiepileptic medications
A SUMMARY VIDEO
A SUMMARY VIDEO
REFERENCES


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