Charles Bonnet Syndrome: Visual Perception as Deception

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Case

A 100-year-old white female presented to eye clinic with complaints of recent onset visual hallucinations. She claimed that she was seeing three young children, all of whom were well-dressed and looking "cute". There was a smaller boy who is persistently hopping next to two girls. She related to occasionally seeing herself (as a young girl) walking with these children, but she knew that they were not real. She is unafraid of these images and believes that the children were there to "protect" her and watch over her.

The patient's previous ocular history included end-stage glaucoma OU. She was using timolol 0.5 % BID and latanoprost 0.005% QHS OU. She had SLT OU in the past. One year ago, her VA was 20/80 OD and 20/400 OS. Current VA, HM OD and CF OS. There is no reported history of psychiatric illness or of hearing voices. The patient was oriented X 3.

IOP 15 mmHg OD and 16 mmHg OS. The patient's slit lamp exam was unremarkable. She was pseudophakic OU. Both optic nerves showed cup-to-disc ratios of 0.9. The fundus OS was unremarkable except for macular mottling and few small scattered drusen. Fundus OD showed the typical "blood and thunder" as seen in a central retinal vein occlusion (CRVO) (Figure 1).

What is the cause of this patient's visual hallucinations?
New onset CRVO causing Charles Bonnet Syndrome.

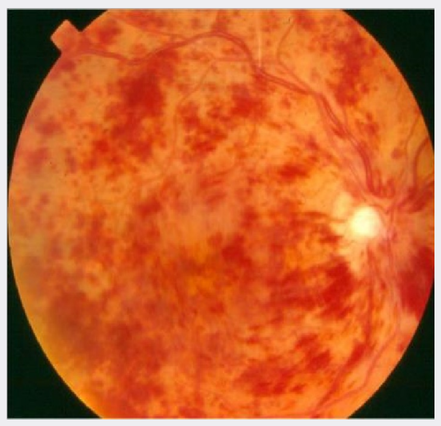

Figure 1: Fundus Exam of Right Eye

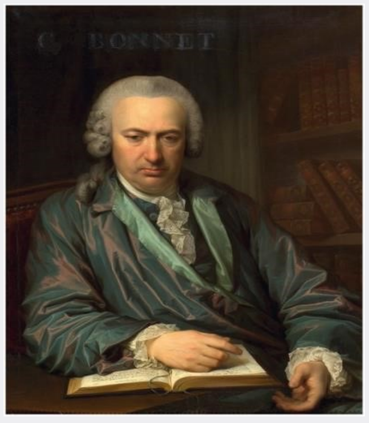

Figure 2: Charles Bonnet[9]

Overview and History

Charles Bonnet Syndrome (CBS): a phenomenon in which an individual with poor vision begins to see images that do not exist – visual hallucinations.

- Often confused for a mental illness or psychosis in a patient. They may visualize various patterns, nature, objects, and even people.[1]
- Patients tend to only complain of visual disturbances. They are unable to smell, hear, or touch these images. Images may be recurrent and can be sometimes complex to describe.
- Eponym was derived from a Swiss philosopher and writer- Charles Bonnet (Figure 2).
- In 1760, Bonnet's grandfather had vision loss after bilateral cataract surgery.
- Shortly afterwards, his grandfather reported seeing random "visions" - described as seeing patterns, buildings, and people. He was cognitively sound and intact and realized that these hallucinations were not real.
- Charles Bonnet deemed his grandfather's visual hallucinations as mere "fictions" of the brain[1] and he also experienced similar visual hallucinations later in his own life as his own vision deteriorated.
- In 1967, another Swiss scientist, George De Morsier, then eventually named these complex visual hallucinations in the setting of severe visual impairment as Charles Bonnet syndrome.[2]

Proposed Etiologies of CBS

Phantom-Vision Theory (also called deafferentation theory): lack of sensory input due to severe vision loss causes spontaneous visual cortex activity that results in visual hallucinations.[3,4]

Release Theory: sensory deprivation (in this case vision loss), may lead to a release of subconscious perceptions.[3,4] These perceptions are known as engrams, which are physical and/or chemical changes that occur to help with new memory associations.[5] This theory states that simply correcting a patient's vision may stop the visual hallucinations.

Irritative Theory: there are spontaneous electrical discharges from the brain's visual cortex which cause hallucinations; however, there has not been much research or results on neuroimaging which can validate this theory.[3,4]

Neuromatrix Theory: CBS may be due to a network of neurons imparting a pattern when visual information is received, like a "neuro signature".[3,4] This theory also states that any change in a human's sensory input will modulate that person's sensory output, potentially leading to hallucinations.[3,4]

Presentation/Symptoms

- Images seen may last seconds-hours
- Episodes can range from days-years with variable frequencies[3,4]
- Most common presentation: patient complains of seeing people
- Images seen: animals, places, patterns, colors
- Patients may also see images of themselves – autoscopia[6]

Prevalence and Associations

- Average Age of Incidence: 75-80 years but can occur at any age
- Prevalence: 0.4% - 30% (reluctance to disclose; misdiagnosis)[7]
- Triggers: fatigue, stress, low levels of illumination, general sensory reduction, social isolation

Management

Dealing with a CBS episode may be frightening and difficult at first, but there are ways to help minimize the hallucinatory effect.

- Certain actions, or "inhibitors" for CBS do exist such as: opening or closing the eyes, blinking multiple times, turning on a light, looking for a distraction, hitting the hallucination, and shouting at the hallucination may allow patients to alleviate the increasing anxiety surrounding their episode.

There are several ways to manage hallucinations when they present in different contexts.[1]

- Hallucinations of Space: visions that can change the shape of rooms and streets, making ambulation difficult for a person. Reaching out and checking the area around is helpful, as well as using a cane or walking stick.
- Hallucinations of Animals: decreased by touching the images of animals seen.
- Hallucinations of People: controlled by understanding that if a person seen in a hallucination is wearing a costume or something different than "normal" clothes, then it is most likely not real. Also, knowing when people are supposed to be visiting a patient's home will help make it easier for a patient to recognize what is real and what is not. Most CBS patients recognize that the images they are seeing are not real.

Treatment

While there is no definitive treatment for CBS, treatment strategies include: correcting the underlying ocular pathology such as cataract extraction to improve vision and overall brightness perception, optical correction, pharmacological agents such as anti-convulsants and neuroleptics, and social interaction for patients.[3,6]

The diagnosis code for Charles Bonnet Syndrome would fall under "psychophysical visual disturbances". The ICD-10 code to support this diagnosis is H53.16.8

References

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