CHARLES BONNET SYNDROME: AN ENIGMATIC CONDITION

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ABSTRACT

Charles Bonnet Syndrome (CBS) is an intriguing condition which is often misdiagnosed as psychosis, delirium and early dementia. CBS is essentially described as visual hallucinations in the visually impaired. Various prevalences have been reported for CBS; from less than 1% to 40% which is accounted for by differences in history taking, varying definitions used and the unwillingness which patients show to report their hallucinations. There are various explanations for this mysterious disease namely the ego defense mechanism, the differentiation theory, sensory deprivation, defect in brain censors and a fall of sensory input below threshold level. It has been proposed that isolated visual hallucinations in the older adults may be indicative of the early stages of dementia since the associated neuropsychological changes are similar to the early stages of dementia. There is no specific drug treatment for CBS. Reassurance about the nature of the illness, programmed and intermittent eye closure and opening, with effective treatment of the eye disorder, may all that may be needed. Since it is a very distressing condition, it is important for doctors to seek for hallucinatory experiences by asking the visually impaired patients about it.

Keywords: Intriguing, Visual Hallucinations, Visually impaired, Various explanations, Reassurance, Distressing.

Introduction:

He had started seeing the visions 6 months earlier. Most evenings, the 69 years old saw faces, odd shapes, animals – including a spider which he tried to kill, friends and acquaintances sitting in his armchair.[1]He realized these were hallucinations; he suffered from Charles Bonnet Syndrome (CBS), one of the most enigmatic, fascinating and ambiguous neurological diseases.

As Douwe Draaisma remarks, the enormous variation in the type of images seen by CBS patients is striking.[2]

A 73 years old woman saw a girl who appeared from nowhere and passed through closed doors.[3]

Once regarded as extremely rare, it is now known that up to 40% of people with impaired vision develop CBS[4], although up to 60% may keep the experiences to themselves.[5]

Misdiagnosis, often as psychosis, delirium and early dementia is common[5] and some people with CBS have been almost confined to mental institutions. So, clinicians need to remain vigilant for this intriguing condition.

History:

Charles Lullin was a distinguished and well loved magistrate in Geneva for many years. He was the grandfather of Charles Bonnet, who documented the experiences of the old man when latter was 89 years of age: The experiences were visual hallucinations. The elderly man was seeing things that were not there. He developed a variety of hallucinations, ranging from simple to complex forms. The simplest hallucinations he described were of an infinity of whirling atomic particles swirling around his field of view. He also had hallucinations of simple lines and dots, coloured blobs and geometrical patterns. He saw figures in his room- typically females in elaborate flowing silk robes, but bizarrely, wearing inverted tables or caskets on their heads. In his room, the walls would become adorned with paintings in beautiful golden frames, landscapes, portraits- things that he had never seen before.

Charles Lullin was normal sighted until about 80 years of age, after which his vision began to deteriorate. At first this was partly attributed to a cataract in one eye.

In the late 80s', Lullin's vision deteriorated and a second cataract did not help. By the age of 89, he was unable to read because he had lost his central vision, but with a preserved peripheral field. In that state, he suddenly developed a variety of hallucinations ranging from simple to complex forms.

In the 1930s', the medical community decided to honour Charles Bonnet with the naming of Charles Bonnet Syndrome; to reflect that Bonnet had been the first person ever to realise that visual hallucinations in eye disease were something rather different to the visual hallucinations encountered in mental illness and Alzheimer's disease.[6]

Different views:

There have been various prevalences reported for CBS. A 1989 review identified just 46 cases in the literature.[2] More recent prevalence estimates range from less than 1% to 40% of patients with impaired vision.[4] Several reasons account for this variation in prevalence: differences in history taking, the definitions used and patients' willingness to report their hallucinations.[1]

Some CBS definitions do not include visual loss.[1]

Furthermore, researchers do not agree whether certain images like bright lights, diffuse colours or auditory hallucinations are part of CBS.[1]

Explanations for this mysterious disease:

An early psychoanalytical interpretation suggested that 'the ego defends itself by creating a substitute world full of entertaining scenes to compensate for the visual loss.[2] However, far from being entertained, the hallucinations leave some people profoundly distressed.

Awidely supported theory suggests that the hallucinations are a mental equivalent of a phantom limb.[1]Half of the afferent neuronal fibres projecting to the brain begins in the eyes[7] and so, the brain expects a considerable perceptual input. According to the differentiation theory, lack of stimuli to the visual association areas of the cerebral cortex causes a release phenomenon similar to phantom limb symptoms.[1,4]As Draaisma points out, the images often appear when vision begins to decline and then disappear when blindness occurs. According to the release theory, the vision should then be at their clearest as he notes.

Sensory deprivation offers another possible explanation. CBS hallucinations develop when some sufferers shut their eyes.[8] In one study, 10 of 13 normally sighted patients report hallucinations while being blindfolded for 5 days.[1]This is however contradictory to Draaisma's view that the images disappear when blindness occurs.

Another theory proposes a dysfunction in the brain's censors. A psychoanalytic theory published in 1962 suggested that the failing eyesight decreases the perception of reality, which makes room for the products of the imagination.[2]

A neurophysiologic version suggests that the brain's censorship mechanism continually removes irrelevant sensory impulses from conscious perception. However, this depends on normal sensory input . If the sensory input falls below a certain threshold, the brain allows subconscious perceptions to surface producing visual hallucinations.[8]

Charle's Bonnet Syndrome: An early marker for Dementia?

The results of a study indicated that patients diagnosed with CBS evidence neuropsychological changes commonly associated with the early stages of dementia. Therefore, in patients with impaired vision, the appearance of cognitive deficits, albeit subtle, occurs with the onset of visual hallucinations. From this study, it has been proposed that isolated visual hallucinations in the older adults may be an indication of the early stages of dementia.[9]

Treatment:

There is no specific treatment for CBS although antidepressants and anticonvulsants have been used for CBS with good effect in previous reports.[10,11] The efficacy of these drugs in the treatment is somewhat questionable and should be reserved for those who exhibit high levels of distress and have not responded to conventional intervention.[12]The treatment approach

would also depend on other comorbid physical or mental disorders.[13,14]

Reassurance about the nature of the illness (i.e not being a mental illness) could be highly comforting and all that may be needed. Combination of this and programmed blinking , intermittent closing and opening of the eyes have been reported as helpful.[15]

The best solution is finding effective treatment to the eye disorder because Charles Bonnet Syndrome has been reported to regress with effective treatmentof the cause of visual loss and improvement in visual activity.[16,17]

Conclusion

In conclusion, doctors are advised to seek for hallucinatory experiences by asking the visually impaired about it. Doing this would go a long way at ameliorating the suffering of the visually impaired from this syndrome that could possibly be passed off as mental disorder. It would also strengthen the consultation liaison between psychiatry and ophthalmology.

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