

# Charles Bonnet Syndrome in an elderly blind man with recurrent Pituitary Macroadenoma and optic atrophy: a case report and the review of literature

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## INTRODUCTION

Charles Bonnet Syndrome(CBS) was first described in 1760 by Naturalist and philosopher Charles Bonnet, who first observed lifelike complex visual hallucinations in his grandfather Charles Lullis after bilateral cataract surgery(1); it was a neurologist George de Morsier who coined the term in 1938 after Charles Bonnet himself developed the condition(2). CBS, also known as "phantom image", is characterized by recurrent or persistent complex visual hallucinations in people with a disease of the visual system with intact insight, intellectual function and normal cognitive functioning without primary psychiatric disorders(3,4). With limited knowledge of CBS among physicians, the prevalence of CBS is underreported(4) even though 41-59% of the visually impaired experience elementary visual phenomena, and 11-15% exhibit complex hallucinations, due to a medical condition or artificially produced, such as in preparation for cataract surgery(5,6). The presence of varying inclusion criteria, inconsistent depth of questioning and limited patient disclosure for fear of being ridiculed may all contribute to underreporting(3,5). Visual hallucinations could be a sign of psychiatric disorders, neurological diseases, metabolic abnormalities, and the use or abuse of specific drugs(7). Both elementary forms(3) and complex visual hallucinations are reported in CBS(8); while glaucoma,

cataract and age-related macular degeneration are the most common associated disorders, any ophthalmological conditions can lead to CBS(7), especially in significant visual impairment(9). A pituitary adenoma is rarely associated with CBS (10); however, reduced visual acuity or a visual field deficit can occur from the benign tumour's compressing optic chiasm affecting one or both eyes(11).

There are three main theories associated with the pathogenesis of CBS, and these are; the sensory deprivation or deafferentiation theory related to spontaneous excitation due to a loss of visual input to the brain(12), the release theory associated with excessive excitation and the consequent release of visual hallucinations(13,14), and the "irritative theory" due to distal provocative injuries transmit abnormal input to the visual cortex leading to abnormal excitatory activity to the temporal and occipital lobes(15,16).

Charles Bonnet syndrome is treated multifacetedly with pharmacotherapy, psychosocial therapy, maintaining appropriate eye care, and sensory stimulation. Generally, CBS is treated with antipsychotics and antidepressants(5), and in some cases, antiepileptic medications have also been used with variable benefits (17).

An 81-year-old blind male presented with a three-year history of visual hallucinations after a recurrence of pituitary macroadenoma. His past psychiatric history is uneventful, and he has intact insight with no cognitive impairment. He improved on a low haloperidol dosage, with a few relapses, when he stops the medications.

## CASE PRESENTATION

An 81-year-old blind male in both eyes presented for psychiatric evaluation with a three-year history of the on-and-off visual experience of seeing various images and shapes others could not see.

The visual experience coincided with the recurrence of a pituitary macroadenoma, about which the Trans-Sphenoidal Adenectomy procedure was done fifteen years earlier. The onset of symptoms was preceded by progressive worsening of vision, followed by the sudden appearance and disappearance of various images ranging from elementary simple geometric shapes to well-formed complex “human-like figures.

Within a year, the visual experiences became more apparent, with the human-like figure suddenly appearing, walking alongside in female clothes and shapes before they disappeared abruptly. The humanoid-like creatures manifested in various forms, having distinct faces and extreme sizes of miniature and tall statures, and also displayed “strange behaviour”, such as reading books and offering him money. The visual experience became more diverse involving scenery of both inanimate and animate objects, such as the sudden appearance of food plates and the view of a village full of huts and people of different sizes. Other experiences included sights of river banks with fast-flowing water bodies with swans swimming and playing; he also saw disfigured creatures with “cow-like faces” spontaneously changing their shapes. He reports his typical day begins with “rays of sunlight passing through the curtains”, followed by seeing the scenery of animate or inanimate objects throughout the day until night when he goes to sleep.

Although the experiences sometimes felt intrusive and distressing, the patient held that the phenomena were “not real” and did not interact with the hallucinations. Further interviewing revealed no history of other types of psychotic experiences, depressive or anxiety symptoms.

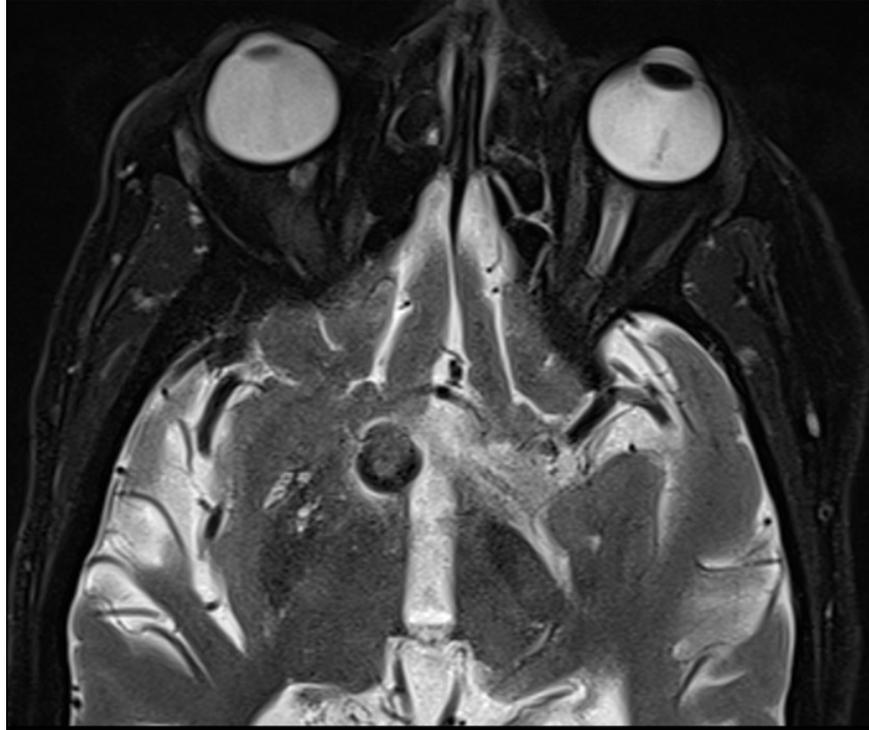
During the interview in the doctor’s office, he reported seeing “a cloudy day, while sat on a chair in an open space on a floor full of white and green leaves” and “small and large artificial plastic trees arranged in a line, some of which were moving.

Past medical history was positive for pituitary macroadenoma fifteen years earlier, which began with a severe headache accompanied by progressive visual impairment. The previous Brain MRI scan showed a large lobulated sellar-suprasellar mass measuring 3.8cm by 3.3cm by 3.1cm in size; the optic chiasm stretched and compressed superiorly, revealing signs of secondary optic atrophy. Hormonal levels (Thyroid Stimulating Hormone, Triiodothyroxine (T3), Tetraiodothyroxine (T4), Prolactin and Growth Hormone) were within normal range.

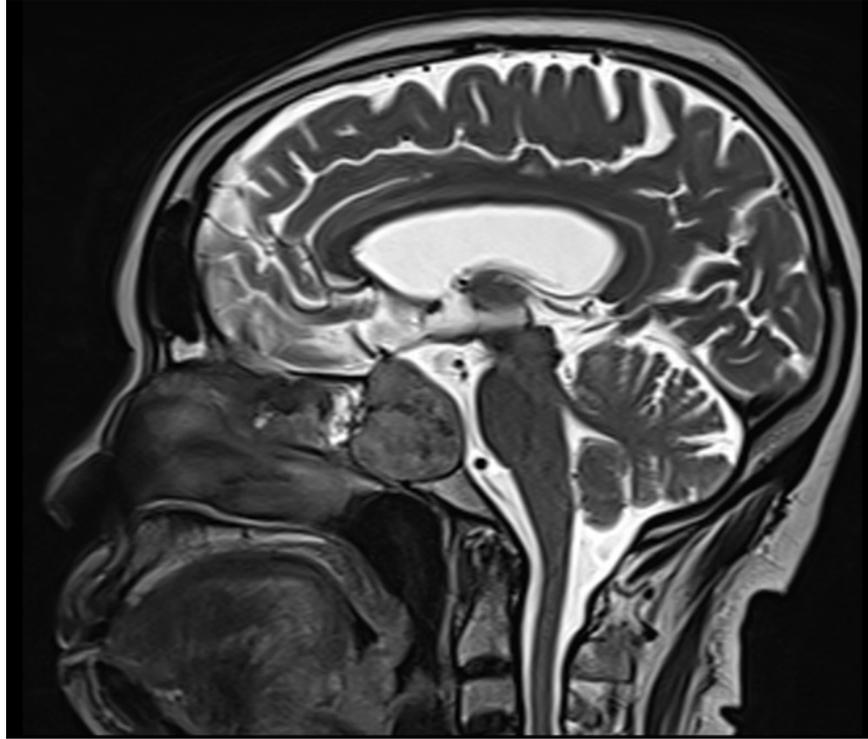
Trans-sphenoidal excision of the pituitary tumour was done: Post-operative CT showed a blood clot with the empty sella with no tumour enhancement. Histopathology results confirmed pituitary adenoma.

There was some improvement in vision during discharge, his left vision was close to normal, and his right eye showed marginal improvement.

During the index assessment, neurological examination showed all cranial nerves were intact except for the optic nerve, where he had a visual acuity of total blindness with no response to light. Motor –bulkiness, tone and power were normal. Deep and superficial reflexes were intact; Babinski sign was negative and had normal sensations for touch, pain, vibration and proprioception. He displayed no gait abnormality, and there were no cerebellar signs.



The mental status evaluation was mostly unremarkable except for visual hallucination. The most recent brain MRI images; Figure 1; Sagittal T2-weighted image showing a solid multilobulated mass measuring 3.4cm by 3.8cm by 2.7cm centred in an expanded sella, with no separate pituitary gland identified. Figure 2; Axial T2-weighted image shows the mass extending into sphenoid and posterior ethmoid sinuses and the suprasellar and interpeduncular cisterns compressing the right aspect optic chiasm and optic nerves atrophy.



The diagnosis of Charles Bonnet Syndrome secondary to Pituitary Macroadenoma was made. He was kept on tablets of haloperidol 1.5mg once a day which significantly improved the symptoms with episodes of waxing and waning of hallucinations for the past year when he was not adhering to medications.

## Discussion

This case has several intriguing observations worth noting; first, the visual hallucinatory experiences occurred after the recurrence of the tumour, about which, to the best of our knowledge, this is the first published case of its nature. Furthermore, the patient had visual impairment at the occurrence of the first tumour; his vision improved after adenectomy without experiencing the onset of CBS. Since the start of CBS was also accompanied by optic nerve atrophy, several neuropathological postulates are implicated.

While CBS can appear following Trans-Sphenoidal adenectomy (TSA) without optic atrophy, most pituitary macroadenoma-related cases of CBS accompany optic atrophy. There is only one published case where visual hallucinations occurred in CBS post-TSA without optic atrophy, about which the patient experienced hallucinations only when the eyes were closed. This phenomenon supports sensory deprivation theory (deafferentation theory), where the lack of sensory input, in this case through eye closure, causes a cascade of events that generate visual hallucination. In sensory deprivation theory, the loss of visual input due to either ocular pathology or defect in the visual pathway increases the excitability of the visual association cortex(18), causing spontaneous neuronal discharge and releasing visual hallucinations(12,13,18). The hyperexcitability in visual association cortex is attributed to an increased number of presynaptic neurotransmitters, an increased number of postsynaptic receptors and an increased amount of excitatory glutamatergic N-methyl-D-aspartate(NMDA); conversly, the amount of inhibitory gamma-amino butyric acid (GABA) within the synapse is reduced (20). Given the neuroplasticity capacity of the visual system, these mechanisms cause the sprouting of new axons and reorganizing of receptive fields, leading to hypersensitivity and erratic response of inadequate stimulus from structures within the visual pathways manifesting as visual hallucinations(20).

Based on the overall clinical presentation and optic nerve atrophy, our case aligns more with the release theory. In the release theory, the defect in the visual pathway, in this case, the optic nerve atrophy caused

by pituitary macroadenoma, causes abnormal signal transmission from the sensory end organ to the visual cortex which are coupled with normal visual activity, these signals are then processed and released as hallucinations (9). Both sensory deprivation and release theories are related to cortical disinhibition and spontaneous neuronal excitation of association cortices, resulting from failure to constrain higher cortical function by reduced bottom-up sensory prediction may explain the perception of phantom images. The patient's hallucinatory experiences began with simple geometric shapes and escalated to more complex visual phenomena. Although the progression of hallucinatory symptoms may suggest a direct relationship with the severity of visual impairment, there is no established evidence supporting the relationship between either the frequency of elementary or complex visual hallucinations (VHs) and the degree of visual loss; nevertheless, having both simple and complex VHs is more associated with more visual field loss indicating a problem with serial processing as opposed to a problem with hierarchical processing in the case of a VH appearance(21).

The initial step in managing CBS is treating the underlying ophthalmologic disease, improving visual input and eliminating hallucinations. However, antipsychotic drugs are the mainstay pharmacological treatment of CBS patients. As we observed, several other case reports demonstrated the overall efficacy of haloperidol in CBS as both first or second line option if atypical antipsychotics fail (22). Other effective treatments, including psychoeducation and reassurance, have all been beneficial in the management of CBS (23)

The patient's overall prognosis depends highly on preventing cardiometabolic and cognitive deterioration, as late-life psychosis, regardless of other neurodegenerative symptoms, can be a sign of neurocognitive disorders (24). The fact that the patient has an intact insight of visual hallucinosis, a good response to low-dose antipsychotics, and the absence of a chronic cardiometabolic condition suggests a favourable prognosis of his CBS symptoms. However, since optic nerve atrophy is irreversible, he will remain blind and hallucinations are likely to be lifelong. Furthermore, his advanced age is a relative contraindication for another trans-sphenoidal procedure to remove the tumour because of risks for complications.

## **Conclusion and Recommendation**

Although there is a significant proportion of patients with visual hallucinations in ophthalmology and medical settings, the majority are undiagnosed and do not receive any intervention. There must be a high index of suspicion for surgical and medical aetiology of neuropsychiatric manifestation for early management to improve the patient's outcome and prevent further complications. Furthermore, close follow-up of patients with similar presentations may ensure early intervention should the symptoms recur.

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## **Detailed author's contribution**

GN attended the patient for the first time and referred to AN, SA prepared the first draft, AN supervised, reviewed and edited all the proceeding manuscript drafts. All authors reviewed and agreed with the final version of the manuscript to be submitted.

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## **Conflict of interest**

The author declares there is no conflict of interest.

## **Data availability Statement**

The data supporting this manuscript are available from the corresponding author upon reasonable request.

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