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Charles Bonnet syndrome following head trauma: a case report and literature review

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CASE REPORT

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Background: Charles Bonnet syndrome (CBS) is a condition characterized by vivid, complex visual hallucinations in individuals with visual impairment. Despite its prevalence among the elderly and those with degenerative eye diseases, CBS remains underdiagnosed and undertreated due to a lack of awareness and misconceptions surrounding its etiology and management.
Case Report: A 51-year-old man presented to the emergency room after falling off his bicycle without wearing a helmet. Head imaging revealed a small right frontoparietal traumatic subarachnoid hemorrhage with an associated trace subdural hematoma along the right parietal convexity. Subsequently, he developed non-light perceiving vision loss, after which he began experiencing visual hallucinations.
Conclusion: CBS is frequently overlooked or left untreated. Research on diagnosing and managing CBS following head trauma is limited. Therefore, clear diagnostic criteria for CBS and a better understanding of its underlying mechanisms are needed to improve diagnosis and management strategies.

Keywords: Charles Bonnet syndrome; Traumatic brain injuries; Visual hallucinations; Vision disorders; Eye hemorrhage; Neurocritical care

INTRODUCTION

Charles Bonnet syndrome (CBS) is named after the Swiss naturalist Charles Bonnet, who first described this phenomenon in the 18th century [1]. Despite its long history, CBS remains a relatively obscure condition in clinical practice and is often overshadowed by commonly encountered psychiatric disorders [1]. The hallmark of CBS is the presence of visual hallucinations in individuals with compromised visual function. These hallucinations can manifest in various forms, including patterns, objects, animals, people, and entire scenes [1,2]. The pathophysiology of CBS remains incompletely understood, and multiple hypotheses have been proposed to explain its etiology. Among the prevailing theories, two have garnered significant attention in the literature regarding the genesis of CBS hallucinations. The "release theory" posits that lesions in the visual pathway generate aberrant signals to the visual cortex [3]. Conversely, the "deprivation theory" suggests that a decrease in sensory input, such as sub-threshold visual stimuli, prompts the visual association cortex to generate spontaneous images, resulting in visual hallucinations [4].

Despite being common among the elderly and those with degenerative eye conditions, CBS often remains undiagnosed and

untreated. This disease poses unique challenges to individuals with visual impairment, caregivers, and healthcare providers, with difficulty in the diagnosis and management of these patients. By increasing awareness and understanding of CBS, clinicians can facilitate early recognition, accurate diagnosis, and tailored interventions to improve patient outcomes. Research on the diagnosis and management of CBS after head trauma is limited. The objective of this report is to present an illustrative case of CBS in the setting of Terson Syndrome resulting from head trauma. We also review the existing literature, with a focus on the clinical presentations, fundamental mechanisms, diagnostic benchmarks, and management strategies for CBS. This case underscores the link between traumatic brain injury (TBI) and CBS symptoms, emphasizing the clinical significance of effective diagnosis and management in such cases.

CASE REPORT

A 51-year-old male with no significant medical history was brought to an outside hospital by ambulance after falling off his bicycle without wearing a helmet. His family witnessed the fall and reported that he was unconscious for approximately 10 min. A computed tomography (CT) scan of the head (CTH) at the outside hospital revealed a small right frontoparietal traumatic subarachnoid hemorrhage (tSAH) with a trace subdural hematoma (SDH) along the right parietal convexity (Fig. 1). No associated hydrocephalus or white matter abnormalities were observed. A maxillofacial CT examination revealed a minimally displaced comminuted fracture of the left sphenoid bone and a squamous portion of the left temporal bone that extended cranially through the parietal bone (Fig. 1). On hospital day 0, the patient developed a new left-sided facial droop and progressively became less responsive, and was transferred to our tertiary care hospital for higher level neurological care and neurosurgical evaluation.

On arrival at our institution, the patient was admitted to the neurosurgical intensive care unit (ICU). At that time, the patient had a Glasgow Coma Scale of 7T (intubated), in which he did not open eyes to voice or noxious stimuli. The patient localized the bilateral upper extremities to the stimulation and withdrew from the bilateral lower extremities. The patient's pupils were equally reactive to light. The systolic blood pressure goal was less than 140 mmHg. He was administered levetiracetam 500 mg twice daily for seizure prophylaxis. Urgent repeat CTH was performed on arrival to evaluate blood stability (Fig. 2). Repeat imaging revealed a new 2.7×2.4×2.1 cm right frontoparietal intraparenchymal hematoma (IPH). Additionally, CT revealed a new 2.7×1.2×1.9 cm IPH (intracerebral hemorrhage score of 1) within the right anterior temporal lobe. This was associated with increased sulcal effacement and mass effects on the right lateral ventricle, secondary to vasogenic edema. The patient's trace SDH was stable compared with the original CTH performed at an outside hospital. The patient was administered 3% saline with a sodium goal of 145–155 mEq/L. At this time, urgent neurosurgical intervention was deemed unwarranted, given the minimal midline shift, thus decreasing the necessity of emergent decompression. A repeat CTH was performed the following morning, which demonstrated the stability of the tSAH, frontoparietal IPH, anterior temporal IPH, and trace SDH (Fig. 2). On admission day 3, the patient became hypoxic, and a CT pulmonary angiography was performed, which was negative for pulmonary embolism but

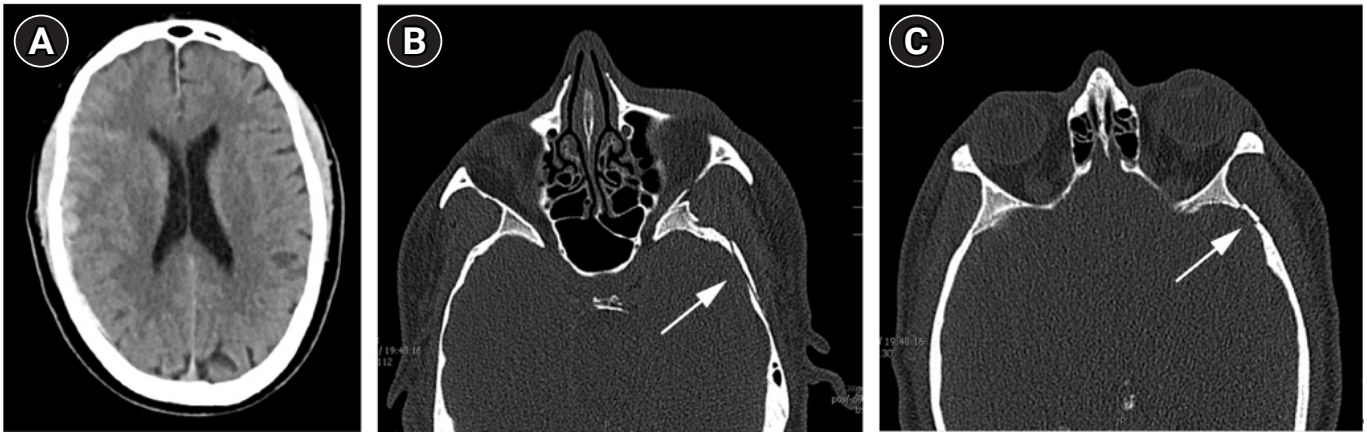

Fig. 1. Computer tomography (CT) without contrast from outside hospital. (A) CT head showing a small right frontoparietal traumatic subarachnoid hemorrhage and subdural hematoma along the right parietal convexity. (B, C) CT maxillofacial thin slices showing minimally displaced comminuted fracture of the left sphenoid bone and squamous portion of the left temporal bone with extension through the parietal bone (white arrows).

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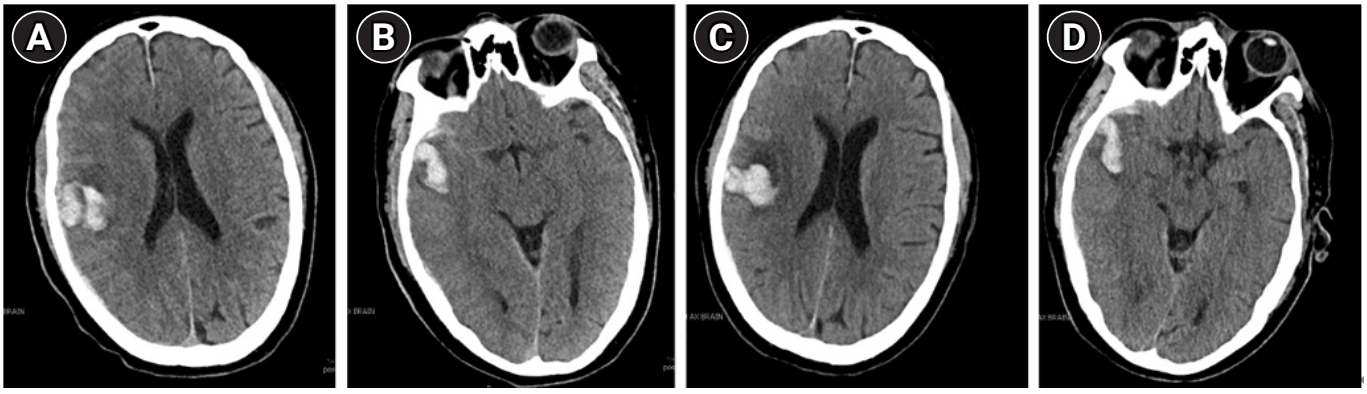

Fig. 2. Computer tomography (CT) without contrast. (A, B) CT head on arrival revealing new intraparenchymal hematomas in the frontoparietal and anterior temporal lobes, sulcal effacement, and mass effect of the right lateral ventricle. (C, D) Repeat CT showing stability of the subarachnoid hemorrhage, frontoparietal hematoma, and anterior temporal hematoma.

raised concerns about multifocal pneumonia. The patient's sputum cultures showed gram-positive cocci; thus, he was transferred to the medical ICU for management. On admission day 8, he was noted to move his upper extremities and withdraw his lower extremities from noxious stimuli. Magnetic resonance imaging of the brain on day 14 of admission showed the evolution of his IPHs and improvement in edema and midline shift (Fig. 3). Given his improved mental status and the passing of spontaneous breathing trials, the patient was extubated.

Following extubation, the patient was slightly delirious but voiced concerns regarding decreased visual acuity. An ophthalmologist was consulted, and bedside examinations were performed. Complete loss of visual acuity was observed bilaterally in all visual fields, with loss of light perception and no response to confrontational testing. Fundoscopic examination revealed bilateral intraretinal hemorrhages of the superior and inferior arcades, chalky white and pale optic nerves with obscuration of the optic disc margins, and prominent flame hemorrhages superiorly and inferiorly. His examination was consistent with bilateral non-light perceiving blindness and most concerning Terson Syndrome, caused by a sudden increase in intracranial pressure. Ophthalmologists stated that they did not believe that vitrectomy would benefit this patient because an examination showed no vitreous hemorrhage, but only scattered intraretinal hemorrhage. The patient underwent electroencephalogram for three days which demonstrated a focal right hemispheric seizure. The neurology service was consulted, and the patient was administered valproic acid 500 mg tid (three times daily) with no further recommendations.

The patient began to experience vivid visual hallucinations 31 days after the accident. He reported experiencing new-onset bilateral visual hallucinations that could be distinguished from reality. For example, he visualized "three fighter jets that were laughing at him"; the patient works with fighter jets which give him insight into the hallucinations. He also stated that he could visualize holes and staircases while trying to walk, which made him feel unsteady. Additionally, when he was pushed into his wheelchair, he experienced a sensation of falling. A psychiatrist was consulted, and it was determined that there was no psychological basis for these hallucinations. Through interdisciplinary discussions between psychiatry and ophthalmology services, it was determined that visual hallucinations were most likely due to CBS or visual release hallucinations, in which the occipital lobe spontaneously fires due to a lack of stimuli. The patient had been taking valproic acid 500 mg bid (twice daily) since being diagnosed with CBS, and since then, has experienced fewer visual hallucinations, though they have not fully resolved.

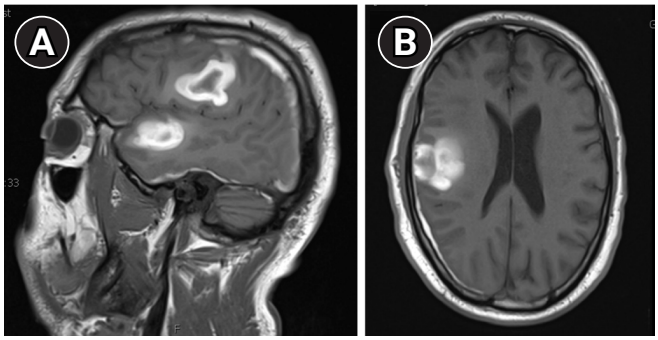

Fig. 3. Magnetic resonance imaging without contrast obtained on admission day 14. T1 weighted (A) sagittal image showing the evolution of intraparenchymal hematomas and (B) axial image showing improvement of edema and midline shift.

The patient was discharged on day 34 with continued outpatient rehabilitation. He was advised to continue taking valproic acid (500 mg tid) to prevent seizures during the TBI. Trazodone

and melatonin were also prescribed. Follow-up with neurology and neurosurgery was recommended. He was also connected with resources to help with new-onset blindness and adjust to his new lifestyle.

DISCUSSION

CBS after trauma

There is a scarcity of research on TBI, which causes CBS. Tobe [5] discussed a patient who developed CBS after a motor vehicle accident and experienced visual hallucinations involving people, animals, and objects.

Neuropsychological testing revealed left-sided brain impairment and significant cognitive function deficits. A neurological examination later showed symptoms indicative of Lewy Body Dementia and Alzheimer disease, which were likely exacerbated by a combination of family history, age, and TBI. Girgis et al. [6] presented the case of an 84-year-old woman who experienced vision loss in her right eye after falling from a standing height. By day 3 of admission, she began experiencing hallucinations in her right eye, describing seeing children playing in the garden while in her hospital bed. The patient was diagnosed with indirect traumatic optic neuropathy and CBS. The patient underwent conservative treatment, and within 2 weeks, vision in the right eye improved to the point where she could perceive hand motions. To date, only one case of CBS following Terson syndrome due to SAH has been reported. Cebulla et al. [7] reported the case of a 57-year-old patient with visual loss due to a ruptured cerebral aneurysm. The aneurysm was managed with ventriculostomy and aneurysm clipping. On postoperative day 31, the patient experienced visual hallucinations involving familiar objects and people. Her visual acuity was limited to light perception, and bilateral dense vitreous hemorrhages were noted on examination. The patient subsequently underwent a vitrectomy, and her vision improved to 20/30 in the right eye and 20/20 in the left eye.

Diagnosing CBS

Diagnosis of CBS can be challenging because of its overlap with other psychiatric and neurological conditions. Clinical assessments should include comprehensive ophthalmic evaluations, neuroimaging studies to rule out organic pathology, and psychiatric assessments to exclude primary psychotic disorders. Although the fundamental characteristics of CBS are widely acknowledged, there are controversies surrounding the specific diagnostic criteria and their implications in clinical practice and research [2,8]. Bonnet's initial description highlighted vision loss as a primary feature. However, the precise degree of vision loss required to trigger hallucinations has not been thoroughly investigated. Some studies have suggested that a visual acuity of 20/50 or worse is associated with hallucinations; however, patients with better visual acuity still experience hallucinations [2,9]. It is equally crucial to differentiate CBS from other causes of visual hallucinations, especially in cases of minimal vision loss where treatable conditions may be overlooked. An insight into the unreal nature of hallucinations is often considered a hallmark of normal cognition in CBS; however, some patients with CBS also lack complete insight [2]. For example, the definitions of normal cognition in CBS require clarification. For example, it is unclear whether the diagnostic criteria for CBS require a normal neuropsychiatric examination and the absence of dementia. Additionally, there is uncertainty as to whether there are potential contributions of subclinical Alzheimer's or Lewy Body pathology to CBS. Several case reports have found associations between CBS and later diagnosis of Alzheimer's or Lewy body dementia [5,8,10,11].

Management strategies for CBS

The management of CBS requires a multidisciplinary approach that addresses both the underlying visual impairment and associated hallucinatory symptoms. For many patients with CBS, hallucinations are not overly troublesome, and reassurance about their harmless nature is typically adequate, as they are not indicative of an underlying neurological or psychiatric condition [1,2]. However, some patients find the hallucinations bothersome when seeking treatment. In cases where vision loss causing CBS is reversible, such as cataracts, resolution of hallucinations has been reported after vision restoration alone; however, low vision aids can be beneficial for improving visual acuity and function [12]. Pharmacological management options, though limited in evidence, include atypical antipsychotics, cholinesterase inhibitors, selective serotonin reuptake inhibitors, antiepileptic medications like valproic acid, and others [13-15]. In this case, valproic acid effectively reduced the frequency of visual hallucinations.

To the best of our knowledge, this is the first report of CBS in a patient with TBI without a vitreous hemorrhage. CBS presents several challenges in diagnosis and management, which are exacerbated by TBI. The permanent blindness experienced by our patient was likely due to multiple factors related to the complex hospital course. Intraretinal hemorrhage secondary to trauma is an obvious culprit, but fluctuating respiratory status and hypoxia likely also influenced the poor visual prognosis. A limitation of this case is that occipital lobe seizures cannot be ruled out, as repeat electroencephalogram was not performed after the onset of CBS symptoms. Although vision loss is a key feature of CBS, the threshold of vision loss required to trigger hallucinations has not

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been clearly defined. Distinguishing CBS from other conditions such as psychiatric disorders or neurodegenerative diseases can be difficult. The criteria for normal cognition in CBS remain unclear, and the potential role of subclinical Alzheimer's or Lewy Body pathology is uncertain. Further research is needed to establish diagnostic criteria and understand the underlying mechanisms of CBS, as well as optimal management strategies for complex cases, such as ours.

ARTICLE INFORMATION

Ethics statement

This study did not meet institutional review board (IRB) review requirements and did not require IRB approval. Written informed consent was obtained from the patient, and the CARE guidelines were followed to enhance the quality and standardization of the reported cases.

Conflict of interest

No potential conflict of interest relevant to this article.

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Conceptualization: JDW, UO. Data curation: GW, JDW, UO. Visualization: UO, JPB. Writing–original draft: GW, JDW. Writing–review & editing: all authors.

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