Visual Hallucinations During Visual Recovery After Central Retinal Artery Occlusion

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Background: Charles Bonnet syndrome is characterized by complex, formed visual hallucinations that occur in patients without psychiatric disorders. To the best of our knowledge, it has not been described following central retinal artery occlusion.

Objective: To describe 2 patients who experienced formed visual hallucinations characteristic of Charles Bonnet syndrome after sudden, severe visual loss precipitated by central retinal artery occlusion.

Patients: Two patients, aged 77 and 63 years respectively, experienced sudden deterioration of vision following central retinal artery occlusion. Formed visual hallucinations occurred in patient 1 six days later and in patient 2 two days later.

Results: The hallucinations appeared both within and at the borders of the patients’ residual intact visual fields. They occurred during periods when the patients experienced partial visual recovery associated with enlargement of their visual fields. The visual recovery and hallucinations both ceased at the same time.

Conclusions: We propose that the hallucinations are likely the result of deafferentation and their occurrence during visual recovery suggests that they are a correlate of visual system plasticity.

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We describe 2 patients with partial recovery of visual fields over a period of 10 to 14 weeks following central retinal artery occlusion (CRAO). During these periods of demonstrable visual recovery, the patients experienced formed visual hallucinations (sometimes termed pseudo-hallucinations) characteristic of Charles Bonnet syndrome (CBS) both within and at the borders of the residual visual field. We propose that the hallucinations are likely the result of deafferentation, and their preferred emergence during visual recovery suggests that they are a correlate of visual system plasticity.

REPORT OF CASES

PATIENT 1

A 77-year-old man with significant carotid artery stenosis developed CRAO with complete loss of light perception in the right eye. Two weeks later, CRAO occurred in the left eye, resulting in visual acuity of counting fingers at 1 foot. A small area of the papillomacular bundle remained perfused by the cilio-retinal artery (Figure 1), accounting for a severely constricted island of vision temporal to fixation. Computed tomography and magnetic resonance imaging of the brain did not reveal any vascular or compressive lesions affecting the visual pathways.

After 6 days, the patient experienced complex, repetitive, formed visual hallucinations both within and immediately outside his residual field of vision. He realized that these images were unreal. He reported “seeing” unfamiliar children and medical staff, boxes of supplies stacked in a warehouse environment, workers moving furniture, and dust on his hospital clothes. The hallucinations were solid, colorless, and of normal size and fit naturally into the surroundings. The “images” occurred 6 to 8 times a day at any time and usually lasted several minutes each. There were no specific triggering factors, and the hallucinations disappeared when he closed his eyes. The hallucinations gradually decreased in frequency and ceased completely at week 3.

At week 8, the patient experienced renewed expansion of his left visual field over a 2-week period, which was demonstrated on a Goldmann visual field (Figure 2). His central visual acuity had improved to 20/150 OS but remained at no light perception in the right eye. During this period, similar visual hallucinations recurred within and at the borders of his enlarged visual field, although they...
were less frequent, occurring about 4 times a day. The hallucinations ceased completely by week 10 and his visual field has remained unchanged since that time.

PATIENT 2

A 63-year-old woman developed CRAO with visual acuity of counting fingers at 2 feet in the left eye. Two days later, she reported “seeing” an unfamiliar person walk across her field of vision. The image was colorless, solid, and of normal size; consistently moved from right to left; and disappeared after a few seconds. The hallucinations only appeared when her eyes were open and occurred 2 to 3 times a day. She was fully aware that these hallucinations were unreal.

At week 6, she reported a perceptible enlargement of her visual field, which was demonstrated objectively with Goldmann visual fields (Figure 3A). Coinciding with this, the hallucinations increased in frequency to 5 times a day. By week 14, the hallucinations had ceased completely and repeated visual field examination showed no significant change (Figure 3B).

Charles Bonnet syndrome has been described in association with ocular conditions affecting visual acuity\(^1\)\(^2\) as well as in patients with visual field defects and normal central visual acuity.\(^3\)\(^6\) We are not aware of any previous reports of CBS developing after CRAO. It is likely that sudden, severe visual loss resulting from CRAO precipitated CBS in our patients because the hallucinations began 6 and 2 days respectively after the onset. Although visual impairment is not a prerequisite for the diagnosis of CBS,\(^7\) poor visual acuity has been shown to be a risk factor for its onset.\(^2\) Interestingly, patient 1 experienced hallucinations only after both eyes experienced severe visual loss. In contrast, patient 2, whose hallucinations occurred when vision in the fellow eye was 20/30, demonstrates that CBS may sometimes occur despite good visual acuity in the fellow eye.

In some reports, CBS hallucinations were localized to areas of visual field defect,\(^3\)\(^6\) and it was suggested that the loss of sensory stimuli from the defective region was sufficient to trigger CBS symptoms. However, in 1 report, 30.8% of patients' visual hallucinations occurred outside the vi-
some cases of CBS are manifestations of attempts at visual illusion (Poggel et al, unpublished data, 2005). This is consistent with the time frame during which our patients’ visual hallucinations occurred within 90 days of the onset of the lesion (Poggel et al, unpublished data, 2005). This is consistent with the time frame during which our patients’ visual hallucinations occurred.

We believe that visual hallucinations experienced in some cases of CBS are manifestations of attempts at visual recovery by the surviving neurons. Although the cellular mechanisms of these hallucinations are still unclear, they may be related to excitability changes in deafferented cortical areas. After deafferentation caused by retinal or cortical lesions, neurons become more responsive to neurotransmitter release by increasing the number and/or sensitivity of postsynaptic receptors. This has been well studied in the dopaminergic system.

The receptive field of neurons in the pons/limbic area of the visual cortex may also undergo reorganization and expansion. We believe that such excitability changes provide the neurophysiological/cellular basis of visual hallucinations in some cases of CBS. Because of increased sensitivity of the deafferented tissue, normal levels of intracortical input may trigger visual hallucinations that manifest in areas of the visual field where recovery is occurring because neurons in that region represent specific parts of the visual field.

This hypothesis is illustrated in our patients: the hallucinations corresponded to periods when they experienced improvement in visual field and visual acuity. They occurred with the borders of the residual visual field where there are areas of residual vision in the apparently “blind” region. It is possible that in these areas, surviving neurons might have increased their sensitivity to light stimuli. Hallucinations did not occur in areas of no light perception, where presumably no functional neurons survived. We propose that when visual recovery ceases, the sensitivity of deafferented neurons returns to the normal baseline level and the same level of stimulus is no longer sufficient to trigger hallucinations, hence explaining their termination. The occurrence of visual hallucination only when patients’ eyes were open further supports the hypothesis that some neuronal input from residual visual fields is required to trigger the hallucinations.

This report demonstrates that CRAO may precipitate the onset of CBS. The hallucinations occur both within the normal visual field and in the transition zone (“area of residual vision”) between normal and abnormal vision. We believe that the hallucinations are most likely to occur just before or while the patient experiences improvement in their visual fields. Thus, we propose that hallucinations in some patients with CBS may be due to hypersensitivity of the deafferented cortex and that they are triggered by the activation of surviving neurons during periods of visual recovery. Hallucinations may not always be a cause of concern. Instead, they may be good prognostic sign for neuroplasticity and the patients’ visual field recovery.

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