Complex Visual Hallucinations After Occipital Cortical Resection in a Patient With Epilepsy Due to Cortical Dysplasia

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Background: Charles Bonnet syndrome is a rare disorder characterized by complex and recurrent visual hallucinations in elderly patients with visual pathway pathologic defects. To date, to our knowledge, it has not been described in patients undergoing surgical resection for occipital lobe epilepsy due to cortical dysplasia.

Objective: To describe a patient who experienced complex visual hallucinations following resection of cortical dysplasia on the right occipital lobe and who was diagnosed as having Charles Bonnet syndrome.

Patient: A 35-year-old woman underwent surgical resection for medically intractable epilepsy caused by cortical dysplasia involving the right occipital lobe.

Results: Two months after resection of the epileptogenic zone, complex visual hallucinations in the left visual field not associated with loss of consciousness or delusion developed in the patient. Hallucinations persisted for more than 12 months despite treatment with antiepileptic medications. During hallucination, no electrographic seizures were recorded through long-term video-electroencephalographic monitoring.

Conclusions: Charles Bonnet syndrome may occur in a patient with occipital lobe epilepsy following resection of the diseased brain with a developmental malformation. Charles Bonnet syndrome associated with surgical treatment of occipital lobe epilepsy may have been overlooked.

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Charles Bonnet syndrome (CBS) is a rare disorder characterized by complex visual hallucinations not accompanied by psychotic symptoms in elderly patients with impaired vision, and it is most commonly associated with ocular pathologic features. Charles Bonnet syndrome has been described in association with variable pathologic conditions of the eyes and central visual pathways, including macular degeneration, occipital lobe infarction, multiple sclerosis with optic neuritis, temporal arteritis, and venous congestion of occipital cortex caused by arteriovenous malformation.

Cortical dysplasia is an important cause in refractory occipital lobe epilepsy (OLE) and surgical treatment is increasingly considered in patients with OLE. To our knowledge, CBS has never been identified in a patient who had undergone surgery for OLE arising from cortical dysplasia. We describe herein a patient who experienced complex visual hallucinations following occipital cortical resection.

A 35-year-old, left-handed woman was admitted for presurgical evaluation of medically intractable epilepsy. She had experienced seizures since the age of 4 years, and her current seizure frequency was 1 to 2 per month with adequate antiepileptic therapy. She was mentally challenged, with a full-scale IQ of 53. She felt green lights as a visual aura, and her seizures, which were characterized by vocalization and flexed posture of both upper extremities with mild dystonia of the left arm, were always followed by head deviation to the left side and secondary generalization. Brain magnetic resonance imaging showed flattening or obliteration of the gyri on the right occipital lobe consistent with cortical dysplasia (Figure 1). Ictal and ictal-interictal subtracted single-photon emission computed tomography (SPECT) showed increased perfusion in the right parieto-occipital area (Figure 2A and B). An intracarotid amobarbital test demonstrated language dominance in the
right hemisphere. An interictal electroencephalogram (EEG) showed frequent regional spikes with most occurring in the right parieto-occipital area (Figure 3A). An ictal EEG showed that low-voltage fast activities arose from the same location. We performed intracranial monitoring with a subdural grid on the right occipital lobe and strips on the medial and inferior occipital areas. Results showed that ictal discharges occurred independently from the medial and inferior occipital areas, suggesting multiple foci or variable spread from a single nearby locus. These findings led us to perform a broad right-sided occipital cortical resection.

Two months after the operation, the patient developed complex visual hallucinations in the left visual field, a symptom she had never before experienced. These hallucinations, which included images of a strange child, geometrical shapes, and a human face–like moon with black slanted eyes and a mouth, intensified over time despite continuous antiepileptic therapy. During these hallucinations, the patient experienced fear. The images seen by the patient were in color, with varied degrees of clarity depending on the stimuli, which included blinking of the eyes, intensity of lights, and sights of moving objects. When she closed her eyes, the images were diminished and the colors faded. Because she feared reappearance of the images, the patient was reluctant to open her eyes. The images, however, did not disappear completely but persisted in the left visual field. The hallucinations occurred when the patient was fully awake, and they were not accompanied by loss of consciousness or abnormal behavior. No auditory components were present.

Figure 1. Brain magnetic resonance image and postoperative computed tomographic scan of a 35-year-old patient with epilepsy caused by cortical dysplasia after occipital cortical resection. A, Brain magnetic resonance image shows obliteration of gyri and increased signal intensities on the right occipital lobe. B, Postoperative computed tomographic scan of the brain shows a tissue defect in the same area.
Eight months after the operation, the patient experienced her first episode of habitual seizure. On admission to the hospital, video-EEG monitoring was conducted for 2 days to determine the cause of her visual hallucinations, as well as whether recurrent electrical ictal discharges accompanied these hallucinations. The symptoms persisted throughout the whole day, although they waxed and waned. The EEG showed intermittent regional spikes and waves over the right centrotemporal region, which were maximal at T8 and C4, although the skull defect should be considered for its impact spike localization (Figure 3B). Electrographic seizures were not recorded when she was experiencing visual hallucinations. Brain SPECT (Figure 2C and D), conducted during visual hallucinations with maximal intensity, revealed a drop in perfusion in the right parieto-occipital area, unlike the finding of ictal manifestation. Despite continued therapy with antiepileptic medications, this patient has experienced visual hallucinations for more than 12 months, persisting until the present time.

We have described a patient who experienced complex visual hallucinations after resection of cortical dysplasia involving the right occipital lobe. The patient had typical CBS according to the diagnostic criteria of Gold and Rabins. The visual hallucinations experienced by this patient were different from her previous visual auras and have persisted for more than 12 months without changes of characteristics. In addition, the results of long-term video-EEG monitoring and SPECT could rule out the possibility of ictal phenomena.

Charles Bonnet syndrome has been reported primarily in elderly patients with impaired vision. The crucial characteristics of CBS include formed and complex visual hallucinations, full or partial retention of insight, and the absence of delusion. Following impairment or loss of vision, hallucinations usually develop after a latency period and disappear within several weeks or months.

While the pathophysiology of CBS is generally interpreted as a release phenomenon, it may be caused by sensory deprivation or "phantom vision." Its preponderance in elderly patients has also suggested that CBS is an early manifestation of dementia. This possibility is considered less likely, however, owing to the occurrence of CBS in pediatric patients, as well as cases of CBS associated with structural damage, such as infarction or vascular malformation.

Several reports using SPECT have suggested the association of CBS with abnormalities of cerebral perfusion. Patients suffering from eye disease showed asymmetric hyperperfusion in the lateral temporal cortex, striatum, and thalamus, with visual hallucinations precipitated by excessive cortical compensation. Occipital hypoperfusion has also been suggested as a cause of CBS.

In our patient, CBS developed after resection of a longstanding abnormal cortex with developmental malformation. In contrast, most previous cases of CBS were reported to have developed after a new insult to the occipital lobe. Visual functions in the premorbid state of these patients may have been normal, and an acute insult to the normal visual system may have led to disruptions caus-
ing visual hallucinations. Occipital lobe dysplasia is a lesion formed during the first trimester of intrauterine life. This leads to reorganization of visual function, such that few deficits are present despite extensive developmental malformations.\textsuperscript{10,19} Thus, in these individuals, the functional deficit after resection is expected to be low.

Although surgical treatments of OLE are common, to our knowledge, there have been no reports on hallucinations associated with CBS. It is possible, however, that visual hallucinations of CBS may occur episodically and transiently, disappearing before patients report the symptoms. It is also possible that clinicians may interpret visual hallucinations as part of the remaining visual aura, that the incidence of CBS may be low owing to reorganization of visual function, or that OLE is relatively uncommon compared with ocular pathology or other forms of epilepsy.

We could not fully exclude the possibility that these hallucinations were caused by ictal phenomena. Seizure symptoms could be modified by resection, and small seizure foci could be missed during scalp-recorded EEG or ictal SPECT.\textsuperscript{20} Moreover, intracranial recordings were not performed on this patient. Since the characteristics of this patient’s hallucinations are identical to those of CBS and since they have persisted for more than 1 year, we consider it unlikely that they are caused by an ictal phenomenon.

This case demonstrated that CBS could develop after a patient went through resection of the occipital lobe. Following surgery, it is important to determine if the remaining visual aura is caused by persisting occipital lobe seizures or to newly developed CBS. Any possibility of misdiagnosis can be prevented through a detailed medical history of the nature of the visual hallucinations and through video-EEG monitoring.

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