BRIEF COMMUNICATION

Charles Bonnet Syndrome Associated with a First Attack of Multiple Sclerosis

Keiichi Komeima^{1,2}, Takashi Kameyama², and Yozo Miyake¹

¹Department of Ophthalmology, Nagoya University Graduate School of Medicine, Nagoya, Japan; ²Department of Neurology, Gifu Prefectural Tajimi Hospital, Tajimi, Japan

Abstract

Background: We treated a rare case of Charles Bonnet syndrome (CBS) manifested during temporary blindness in both eyes caused by optic neuritis associated with a first attack of multiple sclerosis (MS).

Case: A 66-year-old Japanese woman became completely blind for 3 months due to optic neuritis after a first attack of MS. During the blind period, she experienced vivid visual hallucinations for about 2 weeks.

Observations: The patient had no psychiatric disorders or cognitive impairments; therefore, the visual hallucinations during the period of blindness were indicative of CBS. Unexpectedly, the hallucinations disappeared without treatment following her recovery of vision.

Conclusions: Although rare, visual impairment during a first attack of MS can be associated with visual hallucinations indicative of CBS. The hallucinations can disappear spontaneously with the recovery of vision without treatment. **Jpn J Ophthalmol** 2005;49:533–534 © Japanese Ophthalmological Society 2005

Key Words: Charles Bonnet syndome, multiple sclerosis, visual hallucinations

Introduction

Charles Bonnet syndrome (CBS) is characterized by complex visual hallucinations in the presence of normal cognition and insight, and it usually occurs in elderly patients with visual impairments.¹ Numerous cases of CBS have been reported to be associated with different ocular disorders; however, only a few cases of CBS have been reported to be associated with multiple sclerosis (MS).^{2,3} The ophthalmological characteristics of these patients are not well defined. We report on a woman who was diagnosed with CBS during the period of her blindness caused by optic neuritis associated with a first attack of MS.

Case Report

A 66-year-old Japanese woman who had diabetes mellitus visited our hospital for a periodic fundus examination. At the initial visit (day 1), her corrected visual acuity was 0.8 OD and 1.0 OS. She was noted to have mild cataracts, but the fundus and optic discs were unremarkable. There was no evidence of diabetic retinopathy in either eye. The next day (day 2), she returned complaining of blurred vision. Her corrected visual acuity was reduced to 0.3 OD and 0.8 OS. Her pupillary reflexes were depressed, and Goldmann perimetry showed that the peripheral visual fields were significantly constricted in both eyes.

On day 4, her visual loss had progressed to no light perception, with dilated pupils and absence of pupillary reflexes indicating complete blindness in both eyes. In addition, she developed motor weakness involving her left arm and left leg, and she rapidly became wheelchair-bound.

Magnetic resonance imaging (MRI) of the orbit and brain demonstrated no particular findings that were related to her blindness. Funduscopic examination revealed hyperemia of the optic discs with blurred disc margins.

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Correspondence and reprint requests to: Keiichi Komeima, Department of Ophthalmology, Nagoya University Graduate School of Medicine, 65 Tsuruma-cho, Showa-ku, Nagoya 466-8550, Japan e-mail: kkome@med.nagoya-u.ac.jp

She was then treated for 3 days with intravenous methylprednisolone (1g/day) followed by 11 days of oral prednisolone (1mg/kg per day). However, her ocular and systemic conditions did not improve. About 1 month later, she lost voluntary action, and she finally became bedridden. MRI of the brain was repeated and demonstrated multiple high-intensity signal lesions in the cerebral white matter, bilaterally but predominantly on the right side and in the left middle cerebellar peduncle. A cerebrospinal fluid (CSF) test revealed mild lymphatic pleocytosis and slight elevation of protein content and IgG. An oligoclonal band was not present in CSF. Thus, the diagnosis of clinically probable MS was established.⁴ Therefore, her visual impairment was thought to be caused by optic neuritis associated with a first attack of MS. At this point, another course of treatment with intravenous methylprednisolone and oral prednisolone was carried out. One week after initiation of the second steroid treatment, the patient rapidly recovered her voluntary action and even began to walk with assistance.

About 1 month after she was deprived of vision, despite her recovery from the severe paralysis, she remained blind and began to experience vivid visual hallucinations, although she had no behavioral or cognitive symptoms. These complicated images included beautiful sunsets, flowers of the Japanese apricot tree, a scene of the Pacific Ocean, and blurred sun, which occurred whether she had her eyes open or closed. The duration of the hallucinations was usually a few to 10 minutes. She said that she enjoyed the beautiful hallucinations and was happy to see them.

The hallucinations continued for about 2 weeks, and then they gradually disappeared as she recovered light perception in both eyes. Six months after she lost her vision during the first attack of MS, her corrected visual acuity recovered to 0.03 OD and 0.01 OS, but severe atrophy of the optic discs and central scotomas remained in both eyes.

Discussion

We have described the case of a 66-year-old woman who experienced vivid visual hallucinations while she was completely blind as a result of optic neuritis associated with a attack of MS. In cases of CBS associated with MS reported by psychiatrists, the patients had suffered from MS for about 20 years and were distressed by the hallucinations. In these cases, the visual hallucinations were significantly reduced by treatments with carbamazepine or olanzapine.^{2,3} However, our case was different in several respects. Firstly, CBS was observed during a first attack of MS. Secondly, the visual hallucinations disappeared spontaneously without specific treatment as the patient recovered her vision. And lastly, the patient enjoyed the hallucinatory images rather than being troubled by them.

Although the exact cause of CBS is not known, Cogan⁵ suggested that deprivation of visual input can cause a release of memory traces, which are experienced as visual hallucinations in patients with CBS.

Visual hallucinations in patients with MS, to the best of our knowledge, have not been described by ophthalmologists. Visual hallucinations rarely occur without cognitive impairment in MS patients. On the other hand, patients with CBS seldom complain about their hallucinations for fear of being considered mentally unstable.

We recommend that ophthalmologists pay more attention to patients with visual impairment associated with MS so that the possible diagnosis of CBS is not overlooked. Some MS patients might be troubled by unreported visual hallucinations.

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