Short communication

Paradoxical cessation in a case of Charles Bonnet syndrome


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ABSTRACT

Case report: A 78-year-old male patient diagnosed with Charles Bonnet syndrome (CBS) showing secondary visual deficit toward end-stage glaucoma. He progressed to amaurosis, with an abrupt disappearance of hallucinations in parallel to the loss of residual vision.

Discussion: The paradoxical cessation of CBS occurs when the patient loses residual vision and progresses to amaurosis. The lack of stimulation, both in the corresponding retina and the cortex, leads to the disappearance of hallucinations because the deafferented and hyper-excited neurons lose the necessary stimulus that triggers CBS.

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Caso clínico: Paciente varón de 78 años diagnosticado de síndrome de Charles Bonnet (SCB) por déficit visual secundario a glaucoma terminal. Evolucionó a amaurosis, presentando desaparición brusca de las alucinaciones de forma paralela a la pérdida del resto visual.

Discusión: El caso paradójico del SCB se produce cuando el paciente pierde el resto visual, evolucionando a amaurosis. La falta de estimulación en la retina y el córtex correspondiente hacen desaparecer las alucinaciones debido a que las neuronas desaferentadas e hiperexcitadas pierden el estímulo que desencadena el SCB.

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Introduction

The Charles Bonnet syndrome (CBS) is a clinical condition characterized by the appearance of simple or complex visual hallucinations in patients with significant visual impairment and preserved cognitive condition.1–3 The estimated prevalence of simple visual hallucinations in patients with significant vision impairment is 45–59%, and 11–15% for complex visual hallucinations. However, case studies demonstrate a prevalence of 1.84–3.15% due to lack of knowledge by the physician and the fear of the patients to be labeled as psychiatric cases.2,4

The cause of CBS is not known. The most accepted theory is neuronal deafferentation.4 Paradoxical CBS cessation occurs in patients who lose the remaining vision and evolve to amaurosis. The absence of stimuli in the retina and corresponding cortex causes the disappearance of said hallucinations because deafferented and hyper-excited neurons lose the stimuli which gives rise to CBS.

Clinic case

A male patient, 78 years, who visited the Emergency Service due to sudden amaurosis after complete loss of remaining vision in the right eye (RE) secondary to bilateral terminal glaucoma with several years’ evolution, intervened with trabeculectomy and in treatment with topical betablockers. The patient was in follow-up by the Neuroophthalmology Unit for CBS with 10 months evolution, consisting in visions of deformed people and faces, in color and in movement, mainly during the morning and lasting 5 min. In the last 2 months the patient was in treatment with 10 mg per day of donezepil hydrochloride (Aricept®, Pfizer S.A., Alcobendas, Madrid, Spain), with good response as he referred lower frequency and intensity of said hallucinations. In addition, the patient referred that when losing the remaining vision the visual hallucinations also suddenly disappeared. Personal history of the patient included arterial hypertension in medical treatment. The patient did not refer allergy to any drug and the personal or familial history was not relevant.

The examination evidenced amaurosis in both eyes (BE) and the anterior pole exhibited pupils in medium non-reactive midriasis, stable pseudophakia in BE, superior trabeculectomy in BE, with intraocular pressure at 14 mmHg in BE and terminal cup in ocular fundus of BE. The most recent campimetry (OCTOPUS TOP G1), taken at the practice 3 months before, evidenced total campimetric restriction in RE and campimetric abolition in LE (Fig. 1). After being examined in the Glaucoma Unit, the patient was referred to the Neuroophthalmology Multidisciplinary Unit comprising Ophthalmology, Neurology and Psychiatry, where he was diagnosed with paradoxical CBS cessation produced by total loss of remaining vision in BE and evolution to bilateral amaurosis.

Discussion

CBS is characterized by the presence of complex, stereotyped and elaborate visual hallucinations which are usually repetitive, persistent and of sudden appearance. The patient did not exhibit cognitive impairment or other type of sensory hallucination. Accordingly, CBS is a neuro-ophthalmological and not a psychiatric disease.
Hallucinations can be simple, such as basic geometric lines and figures with photopsia, or multicolored, or complex and structured, generally involving people, faces, animals or trees which, in the absence of other types of hallucinations, typically do not speak or make sounds. They can also be in black or white or in color, static or moving. In the vast majority of cases, these hallucinations last less than 10 min and are usually repetitive in the form of a persistent fixed stereotype. The course can be in episodes, cycles or chronic, with duration generally under 18 months, although some described cases experienced an evolution comprising several years.

The cause of the hallucinations is unknown although there are trigger factors such as fatigue, stress, poor lighting and glare. The deafferentiation theory could account for the development of CBS, as it states that the loss of retina nerve cell stimuli for any ocular disease produces obvious occipital cortex stimuli reduction but without disappearing entirely as in amaurosis. Residual afference would give rise to deafferentiation with histological, biochemical and anatomic changes in the synapses in an attempt to compensate for the lack of stimuli, thus becoming hyper-excitability. These changes are evidenced in the presynaptic as well as postsynaptic terminal as well as in the dysfunction of the primary and secondary visual cortex areas.

Paradoxical cessation of CBS occurs in patients exhibiting visual loss with evolution to amaurosis. The total absence of light stimuli in the retina, together with the disappearance of afference with the cerebral cortex, produces the disappearance of the stimuli to deafferentiated and hyper-excited neurons which give rise to the hallucinations.

To conclude, paradoxical cessation of CBS is a complex phenomenon which can give reasons for doubt in diagnosis because the disappearance of hallucinations could be due to the natural evolution of the syndrome or to the treatment (in the present case, with donepezil hydrochloride). However, the sudden disappearance of symptoms, which does not occur in the previous assumptions, together with the evolution toward total amaurosis, points to the paradoxical cessation of the process. It is necessary to study CBS in greater depth to facilitate the knowledge of all the associated etiopathogenic factors.

Conflict of interests

No conflict of interests has been declared by the authors.

REFERENCES