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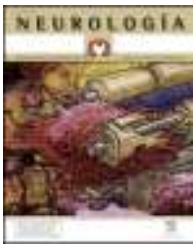

LETTERS TO THE EDITOR

Charles Bonnet syndrome in patient with impaired visual field and good visual acuity☆

Síndrome de Charles Bonnet en paciente con alteración campimétrica y buena agudeza visual

Dear Editor:

Charles Bonnet syndrome (CBS) is a condition characterised by the appearance of simple or complex visual hallucinations in patients with impaired vision and conserved cognitive status.[1—3] Increased life expectancy and quality of life in these patients, many of whom have age-related eye disease, such as age-related macular degeneration, is causing an increased incidence of CBS, which can affect up to 60% of the patients with low visual acuity (VA).[1—4] However, CBS can also manifest in patients with good VA but with visual field alterations secondary to such neurological disorders as those caused by certain cerebrovascular accidents (CVA).

The patient was a 75-year old woman assessed in the neuro-ophthalmology unit of our hospital due to stable visual field alterations consisting of right homonymous hemianopsia secondary to CVA. She had suffered an atherothrombotic infarct of the left posterior cerebral artery a year before. She reported sudden onset of hallucinations one month before. Hallucinations consisted of distorted faces in colour and movement which lasted for approximately 30 minutes and predominantly occurring when waking up in the morning. Although hallucinations affected the whole visual field, the patient reported that they manifested predominantly on her left visual hemifield. She reported no other types of hallucinations. Personal history included medically treated arterial hypertension and diabetes mellitus. The patient's personal and family history did not reveal any other relevant finding or known allergy.

Examination revealed a VA of 0.8 in both eyes (OU) but anterior pole, intraocular pressure, and eye fundus were normal in OU. Campimetry study (OCTOPUS 1-2-3, programme G1X) showed stable left homonymous hemianopsia since the CVA. The patient was assessed in our neuro-opthalmology multidisciplinary unit, which included ophthalmology, neurology, and psychiatry consultations. Imaging studies showed that cerebrovascular involvement was stable and trigger factors for hallucinations were ruled out. Our patient was diagnosed with CBS secondary to visual field alterations despite having good VA.

CBS is characterised by the presence of complex visual hallucinations which are usually repetitive, persistent, and appear suddenly. The patient did not exhibit cognitive deficit or any other sensory hallucinations. They typically manifest as images of people, animals, or trees, and they do not make sounds. Images can also be in black or white or in colour, and they may be static or appear to move.[5] In most cases, these hallucinations last less than 10 minutes and are usually repetitive. The course of the disease can be episodic or cyclical with a duration generally lasting less than 18 months, although progression had lasted several years in some published cases.[3,4]

The cause originating these hallucinations is still unknown, but neuronal deafferentation is thought to be responsible for CBS development. According to this theory, loss of stimulation of retinal nerve cells caused by any eye disease decreases stimulation of the occipital cortex, but unlike in amaurosis, stimulation does not disappear completely. Residual afferent signals are thought to trigger the deafferentation phenomenon by causing histological, biochemical, and anatomical changes to synapses in an attempt to compensate for the limited stimulation. This transforms neurons into hyperexcitable cells.[3,5] These changes were evidenced both in the presynaptic and postsynaptic terminal, as well as in the dysfunction in primary and secondary areas of visual cortex. Although the cause of CBS is still unknown, there are some trigger factors such as fatigue, stress, dim lighting, and flash blindness.[3,5,6]

Our patient did not show any eye disease that could justify development of CBS. However, cerebral impairment with lesion of the optic pathway and associated visual field alteration would be responsible for neuronal deafferentation, which, as a compensatory phenomenon, would trigger the development of visual hallucinations. This has happened in cases of visual field alterations[7,8] and visual field alterations after surgery of the central nervous system.[9—12] Deficit of afferent nerve fibres typical of left homonymous hemianopsia would lead to the development of hallucinations in the whole visual field but predominantly on the left side.

We do not know the cause of CBS in our patient, who was ophthalmologically stable and showed no decreases in VA. However, progressive neuronal deafferentation caused

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by decreased afferent fibres to the occipital cortex would be responsible for the development of hallucinations, also favoured by the trigger factor of morning flash blindness.

As conclusion, CBS can develop in patients with good VA; therefore, ophthalmologists, neurologists, and psychiatrists should be aware of this process to avoid incorrect diagnoses and treatments.[13,14] Progressive development of neuro-ophthalmology multidisciplinary units by the 3 specialties would promote a better knowledge of CBS, which will surely improve quality of life of our patients.[13,14]

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Acute disseminated encephalomyelitis as a complication of systemic lupus erythematosus☆,☆☆

Encefalomielitis aguda diseminada como complicación del lupus eritematoso sistémico

Dear Editor:

Around 50% of patients with systemic lupus erythematosus (SLE) experience central nervous system alterations at some stage of the disease.[1] Common manifestations of neurological lupus include headache, seizures, visual alterations, stroke, myelitis, movement disorders, memory impairment, personality changes, and depression.[2]

We describe the case of a 33-year-old man who presented a haemorrhagic variant of acute disseminated encephalomyelitis (ADEM), which led to a diagnosis of LES.

The patient arrived at the hospital with nausea, vomiting, diarrhoea, and fever. His medical history included an isolated psychotic episode 10 years previously and several consultations with his primary care doctor due to episodes of pleuritic pain and painful joint inflammation. He was being treated with risperidone, biperiden, and citalopram.

On the fourth day of hospitalisation, he presented weakness in the lower limbs and sphincter dysfunction. Physical examination revealed flaccid areflexic paraparesis and hypoaesthesia. Brain and spinal CT scans showed no relevant findings. Cerebrospinal fluid (CSF) analysis revealed lymphocytic pleocytosis with 90 cells/μL (96% mononuclear cells), elevated protein levels (328 mg/dL), and normal glucose levels (57 mg/dL). The patient started corticosteroid treatment: 10 mg dexamethasone followed by 4 mg every 6 hours. Ten hours later, he presented dysarthria, diplopia, and progressive dyspnoea. A neurological examination revealed bilateral horizontal ophthalmoplegia, bilateral facial paralysis, and upper limb weakness. Oxygen saturation was 88% at an oxygen flow rate of 15 L/min. The patient was intubated orotracheally to maintain adequate ventilation, and transferred to the intensive care unit.

The brain and spinal MRI scan showed an extensive hyperintense lesion on FLAIR; the T2 sequence showed a lesion extending from the brain to the medullary cone

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