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LETTERS TO THE EDITOR

Charles Bonnet syndrome secondary to hypertensive crisis☆

Síndrome de Charles Bonnet secundario a crisis hipertensiva

Dear Editor:

The increase in such age-related diseases as age-related macular degeneration (ARMD) is causing higher incidence of Charles Bonnet syndrome (CBS), which consists of visual hallucinations in patients with preserved cognitive status.[1] CBS has been associated with systemic diseases[2–4] and topical[5] and systemic[6,7] treatments. We present a case of CBS triggered by a hypertensive crisis, which was self-limiting with monitoring of arterial pressure at the emergency department.

Our patient is an 85-year-old man who attended a private consultation due to a first known episode of hypertensive crisis accompanied by visual alterations. His personal history included atrophic ARMD in both eyes (OU), diagnosed 5 years previously; arterial hypertension (AHT), which had been treated with losartan (50 mg daily) for 10 years, although treatment was not strictly followed; and type 2 diabetes mellitus treated with diet for 3 years. During the examination, the patient presented arterial pressure (AP) values of 210/190 mmHg, then reported seeing several unknown people who did not speak. Hallucinations featured colour and movement. Although the patient knew they were not real, he was alarmed by the fact that he could see them clearly despite his poor vision. He also presented holocranial headache, a sensation of palpitations, and epistaxis as the only accompanying symptoms. No seizures or any other neurological manifestations were observed during the episode. A computed tomography (CT) scan revealed no other alterations. The patient was given 2 oral doses of 50 mg captocapril separated by 30 min to control AP. With the progressive control of AP, the visual hallucinations started to progressively decrease also in number and frequency, disappearing completely 4 hours after symptom onset; AP was 160/80 mmHg at that time (Fig. 1).

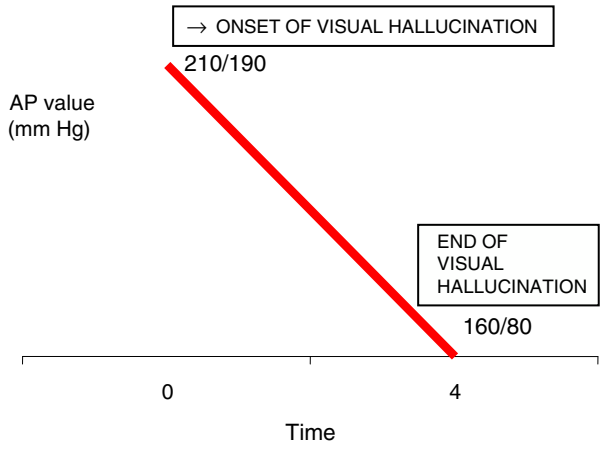

Figure 1 Chronology of the hypertensive crisis and hallucinations. AP: arterial pressure.

He was transferred to our hospital's neuro-ophthalmology department; during the ophthalmological examination, he was able to count fingers at 1 metre with OU. Intraocular pressure was 16 mmHg in OU. Biomicroscopy yielded normal results and eye fundus examination revealed atrophic ARMD. As other causes of hallucinations were ruled out, the patient was diagnosed with CBS secondary to hypertensive crisis.

The origin of the hallucinations in CBS is unknown, although deafferentiation is thought to be a possible trigger factor for these hallucinations.[8–10] According to this theory, since the occipital cortical area receives fewer afferences from the pathological retina, or due to different eye diseases, this compensatory phenomenon would occur in those deafferentiated areas, which would become hyperexcitable before any stimulus.[8–10]

Development of hallucinations has been associated with such trigger factors as flash blindness or dim lighting,[7] systemic[7] and surgical[11] eye treatments,[5,6] and such systemic diseases as anaemia,[2] occipital infarcts,[3] or multiple sclerosis.[4]

In our case, the hypertensive crisis would cause a series of reversible functional changes to the deafferentiated cortical areas, which would trigger visual hallucinations. Sustained high AP would cause haemodynamic, chemical, and histological alterations, which would act as trigger factors for visual hallucinations in the occipital cortex. The mechanism would be alteration of the cortical tissue due to high AP in the blood vessel maintaining haemodynamics in these areas.

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Deafferentiated neurons would be stimulable in this case by hypertension in the cerebral blood vessels corresponding to these cortical areas; with administration of a hypotensive treatment and monitoring of the hypertensive crisis, stimuli and trigger factors disappeared, the episode resolved, and the deafferentiated cortex stabilised.

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Non-motor neurological symptoms in patients with amyotrophic lateral sclerosis☆

Síntomas neurológicos extra-motores en pacientes con esclerosis lateral amiotrófica

Dear Editor:

Amyotrophic lateral sclerosis (ALS) is a neurodegenerative disease traditionally defined as clinical symptoms involving the upper motor neuron, in the motor homunculus, and the lower motor neuron, in the spinal cord.[1,2] However, recent publications have described non-motor signs and symptoms in patients with definite ALS, including impaired higher cerebral functions, signs of dysautonomia, metabolic disorders,[3,4] and cognitive alterations linked to frontotemporal dementia.[5] Post mortem histopathological studies of patients with definite sporadic ALS have identified protein inclusions of transactive response DNA binding protein 43 kDa (TDP-43) in non-motor areas of the central nervous system (CNS), including the nigrostriatal system, cerebellum, forebrain, hypothalamus, and neocortical and allocortical areas.[6,7] ALS and frontotemporal dementia are currently considered to be TDP-43 proteinopathies.

We performed a retrospective study of the medical records of 112 patients diagnosed with definite sporadic ALS according to the El Escorial clinical and neurophysiological criteria.[8] We analysed the non-motor neurological signs and symptoms reported at the time of diagnosis and those manifesting in the first year of clinical follow-up. All patients with diagnosis of definite ALS were assessed with the revised ALS functional rating scale (ALSFRS-r), the Mini-Mental State Examination, a neuropsychological test, a genetic test, and the Hamilton Anxiety and Depression Scale during the baseline consultation, and underwent at least one additional evaluation during the one-year follow-up period. In our sample, we identified 25 non-motor symptoms, with the most prevalent being: depression (in 47% of patients), pain (26%), fatigue (24%), anxiety (18%), pseudobulbar symptoms (13%), and paraesthesias (13%) (Table 1). No diagnosis of frontotemporal dementia was established in any of our patients; curiously, all cases with pseudobul-

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