CHARLES BONNET SYNDROME (CBS) IN NEUROMYELITIS OPTICA

CIEM-MS Research Center – Federal University of Minas Gerais Medical School – Belo Horizonte, Brazil

Background

Charles Bonnet syndrome (CBS) is a condition characterized by visual hallucinations (VH) in the absence of psychiatric or neurological disorders. Typically patients are aware of the unreal nature of their abnormal perceptions. It most commonly occurs in elderly people with bilateral visual loss due to eye diseases or lesions in the visual pathways. Visual hallucinations are the result of deafferentation. Sensory deprivation produces endogenous activation of the visual cortex which then fires spontaneously. The syndrome has been reported in many diseases, including optic neuritis in multiple sclerosis but never before in NMOSD.

Objective

To report a case of CBS following bilateral optic neuritis as NMOSD.

Case report

A 79-year-old white male, truck driver, was referred with a one-year history of bilateral amaurosis presumably due to arteritic anterior ischemic optic neuropathy. He had presented acute loss of vision associated with pain in the temporal region of the head. A comprehensive examination for giant cell arteritis (GCA) including temporal artery biopsy was negative. Brain MRI was also normal. The patient had been put on prednisone.

Neuro-ophthalmic examination disclosed bilateral NPL and marked optic atrophy (Figure 1). The pupils were dilated and non-reactive to light. He had no other neurological sign. A repeated workup was unrevealing and prednisone was then tapered and stopped.

The patient complained of recurrent episodes of VH which started six months after blindness - perception of lines of trucks inside his bedroom (Figure 2); children playing at his bedside; or faces of people pasted on the walls. Three years after amaurosis he developed an acute attack of transverse myelitis with paraparesis, decreased sensation in both legs and sphincter disturbances. Spinal MRI showed a T6-T8 central spinal cord lesion; (Figure 3). Serum aquaporin-4 IgG was positive. The patient was put on azathioprine. At last visit, one year following the diagnosis of NMO, episodic VH still persisted.

Discussion

Our patient was blind as a result of bilateral simultaneous optic neuritis. He then presented complex visual hallucinations while being fully aware of the unreal nature of his perceptions. He had no symptom of a psychiatric disorder. Three years after optic neuritis he presented acute transverse myelitis associated with a longitudinally extensive spinal cord lesion. Search for serum AQP4-IgG yielded a positive result.

Visual hallucinations have been reported in NMO as a manifestation of PRES and narcolepsy. However, surprisingly, CBS had never before been identified in NMO patients. As severe loss of vision is a common feature of NMOSD clinicians should be aware that VH associated with CBS may not be a rare phenomenon in NMOSD.

References