v. 2, n. 8, 2022

International Journal of Health Science

DOUBLE-SERONEGATIVE NMOSD PRESENTING AS ALIEN HAND SYNDROME: CASE REPORT

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Abstract: Seronegative neuromyelitis optica spectrum syndrome (NMOSD) can be diagnosed in the presence of key clinical features associated with MRI criteria. We present a clinical case of a male patient who initially manifested a longitudinally extensive transverse myelitis, with a negative autoantibody test; later evolving with corpus callosum lesion and, clinically, with alien hand syndrome (AMS), in association with cognitive deficit. The patient met diagnostic criteria for seronegative NMOSD and showed clinical improvement with immunotherapy.
Keywords: Hemineglect, corpus callosum, alien hand, neuropsychology, neurology.

INTRODUCTION

Neuromyelitis Optica Spectrum Syndrome (NMOSD) is an uncommon autoimmune disease of the Central Nervous System, mediated by autoantibodies. The presence of major clinical features (longitudinally extensive myelitis, optic neuritis, area postrema syndrome, trunk syndrome, diencephalic syndrome, or cerebral syndrome) with anti-aquaporin-4 antibody is sufficient to make a diagnosis of NMOSD. In seronegative cases, the criteria are more stringent and MRI requirements must also be met. The term neuromyelitis optica may suggest inflammation unique to the spinal cord and optic nerve. However, in about 60% of patients there may be brain involvement, although most of the changes observed are nonspecific (Kim et al, 2019). In this sense, in the study by Zeng (2021), patients with NMOSD presented the following aspects: decreased cognitive performance in attention, short-term memory and verbal memory; which were associated with significantly decreased degree centrality, nodal efficiency, and increased nodal shortest pathway of various brain regions.

Hemineglect (HMN) is the difficulty that the individual has to orient himself, act and respond to stimuli and/or actions that occur on the contralateral side of a hemispheric lesion; they are not due to elementary sensorimotor disorders. Typically, HMN is described by lesions that affect the parietal lobe - mainly the right one - which contains the body schema and mediates spatial perception (Arguelles et al, 2012). In this disorder, there is a loss of configuration of sensory input to the brain, especially the neocortex. Including several cortical and subcortical regions in the right hemisphere, especially in the parietal and frontal lobe, in addition to the temporal lobe and the thalamus; they are part of neural networks critically involved in spatial and attentional functions. Damage to different regions within these networks can cause hemispatial neglect (Cojan, Saj, Vuilleumier, 2021)

SMA is defined as an involuntary and apparently purposeful autonomous motor activity, added to the patient's erratic perception of the affected limb (Ortega-Albás, 2003); generally associated with difficulty in recognizing this limb in the absence of visual clues (visual hemineglect), resulting from lesions in the frontal lobe or lesions in the anterior portion of the corpus callosum and, in rarer cases, lesions in the parietal lobe.

GOAL

The present article aims to present the clinical case of a patient with diagnostic criteria for NMOSD, presenting with apraxia and left hemineglect.

METHODOLOGY

The present study consists of a case report, submitted to neurological and neuropsychological evaluation at a reference Federal University Hospital. Procedures included patient and child caregiver interviews, laboratory tests (including anti-MOG antibody, anti-aquaporin-4 and CSF

analysis), brain MRI, and neuropsychological tests (Wechsler Abbrev. Neuropsychological (NEUROPSIC-R), Clock Drawing Test, Digit Span, Mini-Mental State Exam).

ETHICAL ASPECTS

The present case is included in the research sample approved by CEPSH/UFSC (CAAE: 00783512.2.0000.0121), with signed informed consent.

CASE REPORT

A 56-year-old patient, right-handed, graduated in Physical Education with a Master's degree and active as a capoeira master.

Five years ago, he presented with subacute paraparesis. The MRI performed at the time showed longitudinally extensive transverse myelitis (D3-D9) of probable demyelinating etiology. He performed diagnostic investigation exams: anti-MOG and anti Aquaporin-4 antibody research, serology for HIV, hepatitis and syphilis; all negative. CSF analysis without changes. Pulse therapy with intravenous methylprednisolone was administered, with improvement of the paraparesis. Initiated azathioprine 50 mg bid. The patient remained clinically stable for 4 years.

About 1 year ago, he was admitted to our hospital again due to cognitive impairment and difficulty walking. Neurological and neuropsychological examinations demonstrated ideational apraxia, hemineglect (image 1, SMA). Despite showing satisfactory verbal skills, he had significant cognitive impairment in executive functions, especially in terms of working memory, cognitive flexibility, visuoconstruction, visuopraxia, visual information processing and abstract reasoning (Total IQ: 75 - Borderline, 5th percentile, IQ verbal: 109 - Average, IQ performance: 45 - Extremely low). Qualitatively, when the patient was asked to perform gestures referring to ideomotor praxis (sign of the cross and sending a kiss) and to ideational praxis (toothbrushing), it was observed that the execution was performed with the body side preserved. Associated with mood swings and manipulative behavioral traits in the verbal sphere, sertraline was indicated.

It is also mentioned about the lesion, that it is directly related to the patient's worse performance in parietal lobe skills bilaterally and with worsening of attention and movement ability, mood swings and apathy (more related to the anterior part of the body). corpus callosum).

Magnetic resonance imaging performed after 1 month of symptoms showed a confluent lesion with T2/FLAIR hyperintensity and diffusion restriction in its periphery, extensively affecting the corpus callosum, with an incomplete enhancement pattern of the ring in its anterior portion (image 2 A and B). He was submitted to a new pulse therapy with methylprednisolone, azathioprine was suspended and cyclophosphamide was started. The patient improved clinically after 1 month of treatment.

CONCLUSION

The patient in this case was diagnosed with double seronegative neuromyelitis optica and presented with hemineglect and alien hand syndrome, manifesting significant executive dysfunction, in addition to mood swings. The contrast enhancement pattern on cranial magnetic resonance imaging is typical of a lesion of a demyelinating nature, suggesting activity. Immunotherapy treatment showed improvement in the condition. In short, it is a rare neurological disorder that impacts the daily life of the patient and his family.

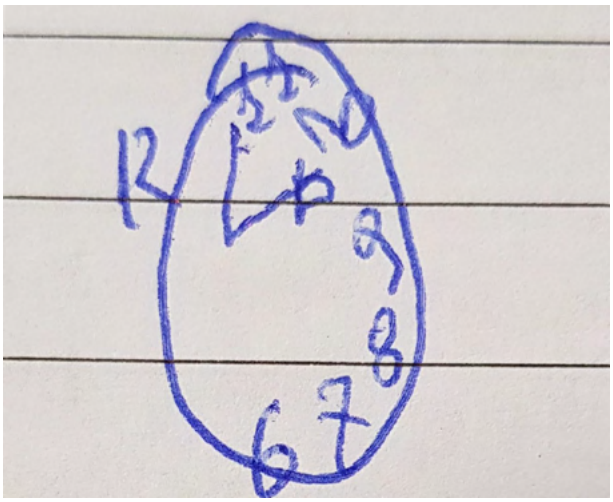

Image 1. Clock drawing demonstrating left hemineglect.

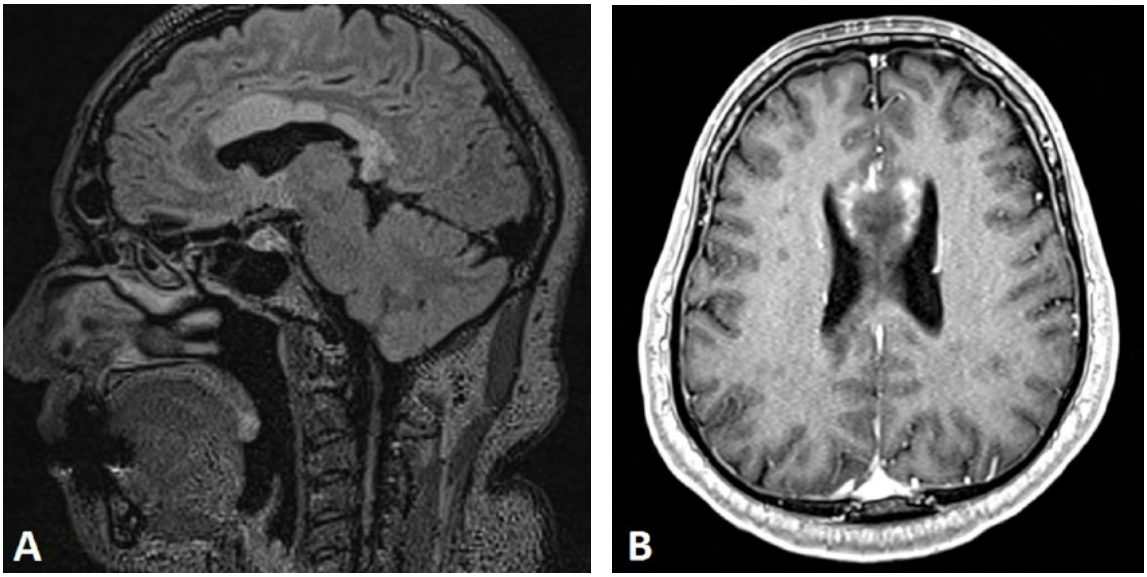

Image 2. MRI. (A) Confluent lesion with FLAIR hyperintensity affecting the corpus callosum. (B) Incomplete annular enhancement pattern in the anterior portion of the corpus callosum, typical pattern of a demyelinating lesion.

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