Neuromyelitis optica with very late onset

Neuromielite óptica de início tardio

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Neuromyelitis optica (NMO or Devic’s disease) is a relative rare relapsing inflammatory disorder of the central nervous system that closely resembles multiple sclerosis¹. Strong evidence indicates that aquaporin 4 antibodies (NMO-IgG) have a pathogenic role in NMO and serve as a useful diagnostic marker. The well-established diagnostic criteria for definitive NMO, which are accepted worldwide, include optic neuritis, myelitis and, at least, two of the following three supportive criteria: magnetic resonance image (MRI) evidence of a contiguous spinal cord lesion that is three or more segments in length, brain MRI that does not fulfill the diagnosis criteria for multiple sclerosis, or NMO-IgG seropositivity².

Several studies have shown that the onset of NMO is predominant in the second and third decades of life, and this result have been confirmed in three classical Brazilian studies on the subject ³–⁵. Only five cases of NMO developing after the age of 70 years have been reported⁶–¹¹. The present study describes another case of NMO with onset at the age of 75 years.

A female patient aged 75 years presenting with blurred vision in the right eye and weakness in both legs was seen at the Emergency department. These symptoms became established over a 24-hour period. She presented diabetes mellitus type II, high blood pressure and visual loss in the left eye, secondary to toxoplasmosis that had started two years earlier. The patient’s condition progressed, with urinary retention and worsening of her paraparesis, such that her maximum sensory level became T4. Her brain MRI showed leukoaraiosis, and the spinal fluid analysis showed 58 cells (22% neutrophils, 78% lymphocytes), glucose 56 mg%, proteins 117 mg% and was negative for syphilis, bacteria, fungi and oligoclonal bands. Her blood analysis was IgM positive for toxoplasmosis and CMV, and negative for HIV, HTLV I and II, rheumatoid factor and anti-nucleus factor. Carotid ultrasound, blood cell counts and serum biochemical analysis were unremarkable. Electrophysiological studies showed no peripheral neuropathy or myopathy. NMO-IgG was positive, detectable at concentrations of 1:160. Her spinal cord MRI showed an extensive lesion, enhanced by gadolinium (Figure).

The patient was treated with intravenous corticosteroids (pulse therapy for five days) followed by intravenous human immunoglobulin for five days. Her visual deficit showed some improvement and she was released from the hospital with oral corticosteroids prescribed. Four months later, the patient was seen to be continuing to show progressive but slow improvement. Toxoplasmosis was considered to be an additional hazard for immunosuppression and, therefore, treatment has been mainly based on rehabilitation, since immunosuppression is not an option for this patient.

The present case shows that NMO must be considered as a differential diagnosis in any patient presenting optical neuritis and/or myelitis, irrespective of the patient’s age.

Figure. Extensive spinal cord lesion in a 75-years-old patient (sagittal and transversal images).
References