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Neuromyelitis Optica: Case Report

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Abstract

We present an interesting case of Neuromyelitis optica (NMO), also known as Devic's disease. NMO is an inflammatory and autoimmune central nervous system (CNS) disorder that primarily affects the optic nerve and spinal cord. This case report illustrates the disease's clinical manifestations and highlights the importance of early diagnosis for better treatment outcomes.

Objective: To detail the clinical presentation of NMO and emphasize the role of timely diagnosis and appropriate therapeutic interventions in improving patient prognosis.

Keywords: Neuromyelitis optica (NMO); Devic's disease; Aquaporin-4 (AQP4); Extensive longitudinal transverse myelitis; Optic neuritis; AQP4-IgG antibodies; MOG-IgG antibodies

Introduction

Neuromyelitis optica (NMO), previously known as Devic's disease, is an inflammatory disease characterized by four clinical syndromes: atypical optic neuritis where there is severe vision loss and bilateral involvement, transverse myelitis or longitudinally extensive myelitis, postrema area syndrome, and brainstem syndrome. Unlike multiple sclerosis, this disease affects gray and white matter, and its target of affection is the astrocytes, where aquaporin-4 (AQP4) receptors are found. Part of the diagnosis is the identification of the autoantibody, which is known as anti-AQP4.

The objective of this case is to present the case of a patient who has had multiple clinical syndromes throughout his life and could be consolidated as NMO anti-AQP4 positive.

Clinical Case

We present the case of a 53-year-old male, an independent public accountant, who bravely faced the challenges of NMO. His journey began with left optic neuritis and vision loss in 2017, which was treated with methylprednisolone. A year later, he experienced a relapse with right optic neuritis, again treated with methylprednisolone. In 2024, the patient developed transverse myelitis syndrome, leading to lower extremity weakness and urinary retention. He sought our clinic's help in March 2024 due to gait disturbances, recurrent falls, and numbness, predominantly affecting the right pelvic limb. Neurological examination revealed loss of strength in the right forearm and hand, accompanied by diminished sensitivity to coarse and fine touch. The patient also reported persistent holocranial headache (4/10 intensity), tremors, and a single episode of sphincter incontinence.

MRI findings indicated:

- An affection bilateral of optic manifested nerve signal alterations, with gadolinium enhancement suggesting active bilateral optic neuritis, predominantly affecting the left optic nerve.
- Besides, we find subcortical white matter hyperintensities in the frontal regions without diffusion restriction, consistent with chronic non-specific changes.
- Spinal cord lesions spanning C2-C7 and T2-T7, with gadolinium enhancement indicative of disease activity (Figure 1).

Serology confirmed IgG AQP4 positivity and IgM anti-MOG negativity (<1:1600). These findings, which are consistent with a

diagnosis of NMO, established the diagnosis of neuromyelitis optica spectrum disorder (NMOSD).

The patient's IgG AQP4 positivity indicates a high likelihood of NMO, while the absence of IgM anti-MOG antibodies suggests a lower risk of certain complications and a more favourable prognosis.

The Treatment Plan

We started methylprednisolone in pulses: 1 g daily for five days and Tocilizumab initiation with ongoing biosafety monitoring and clinical evaluations.

Discussion

This clinical case underscores the importance of timely and appropriate management in NMO. Methylprednisolone and tocilizumab are key components of the therapeutic strategy for AQP4-IgG-positive NMOSD. Recent studies support tocilizumab's efficacy in reducing relapse rates and improving the Expanded Disability Status Scale (EDSS) score, particularly in AQP4-IgG-positive patients.

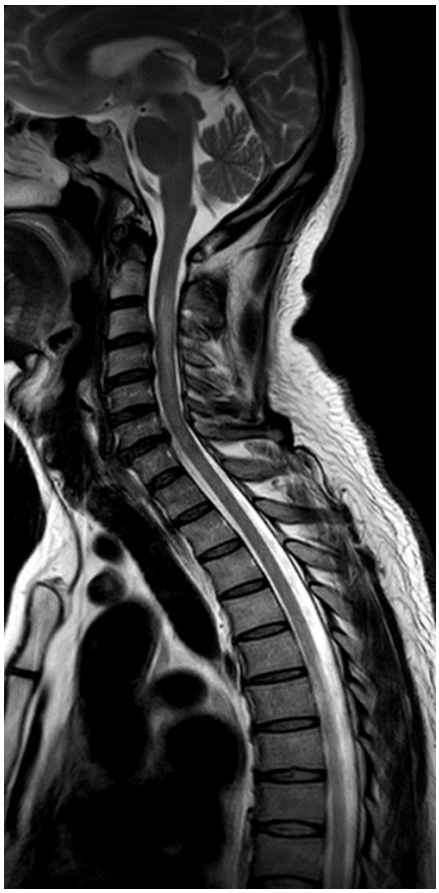

Figure 1: *Magnetic Resonance Imaging showing a longitudinal extensive myelopathy.*

Tocilizumab's ability to block interleukin-6 (IL-6), a pivotal cytokine in autoimmune inflammation, distinguishes it from other immunosuppressant's. Clinical trials and systematic reviews highlight tocilizumab's favourable safety profile, with a poor incidence of severe adverse reactions. While efficacy varies by demographic factors such as ethnicity and gender, the drug consistently improves annualized relapse rate (ARR) and stabilizes Expanded Disability Status Scale (EDSS) scores across patient populations. However, the chronic evolution of NMOSD necessitates long-term management and meticulous monitoring of treatment response, which underscores the gravity of each patient's situation and the need for ongoing support and care. Factors such as age, comorbidities, and treatment adherence may influence treatment and outcome. More studies are needed shortly to address all these variables to optimize individualized and precise care [1-6].

Conclusion

The clinical manifestations in this case presentation demonstrate the principal features and progression of NMOSD and emphasize the benefits of early immunomodulatory therapy. Tocilizumab, in particular, offers promising results for patients with. AQP4-IgG-positive, improving clinical answers, outcomes, and quality of life. Eight months of follow-up revealed no EDSS progression, reinforcing the value of this therapeutic approach.

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