MULTIPLE SCLEROSIS AND RELATED DISORDERS

Long-term outcome and prognosis in patients with neuromyelitis optica spectrum disorder from Serbia

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Highlights

- 2015 NMOSD criteria increase the rate of diagnosis.
- Higher EDSS at onset is an independent predictor of reaching severe disability.
- Severe visual deficit is reached earlier after optic neuritis or opticospinal onset.
- Longer time from NMOSD onset to maintenance treatment predicts severe disability.
- NMOSD studies should utilize OSIS as an outcome disability measure.

Abstract

Background

Neuromyelitis optica spectrum disorder (NMOSD) most commonly, although not exclusively, targets optic nerves and spinal cord. Untreated, early and severe disability is common. We evaluated the long-term outcome in NMOSD patients diagnosed according to the 2015 criteria.

Methods

We retrospectively analyzed 74 patients from the hospital-based NMOSD cohort at the Clinic of Neurology, Belgrade, Serbia, who fulfilled the 2015 NMOSD criteria. We identified patients based on 2015 criteria; 51.4% of whom would not have fulfilled 2006 criteria. Median follow-up was 6.9 years. Aquaporin-4 (AQP4) IgG was tested in all patients using a cell-based indirect immunofluorescence assay. The level of neurological disability was assessed by the Expanded Disability Status Scale (EDSS) score, and by Opticospinal Impairment Scale (OSIS), visual acuity (VA) and motor function subscores.

Results

The disease course was monophasic in 17.6% patients and relapsing in the remainder; none developed progressive disease. AQP4-IgG was detected in 89.2% of patients. 45 of 74 patients were treated with immunosuppressants, 40 with azathioprine, 3 with mycophenolate mofetil, 1 with cyclophosphamide, 1 with mitoxantrone, and 2 patients with rituximab. The median intervals from onset to EDSS 4.0, 6.0 and 7.0 were 6.5 years, 11.9, and 22.0 years, respectively. Higher baseline EDSS was associated with risk of attaining EDSS 4.0, 6.0 and 7.0; a shorter first inter-attack interval for reaching EDSS 4.0 and 6.0; longer time to the start of treatment for reaching EDSS 7.0. Worse visual acuity at the disease onset predicted faster assignment of OSIS VA = 6 and VA = 8. Severe visual deficit (OSIS V_A 6) was reached earlier after optic neuritis (median time, 10.0 years) or combined opticospinal onset (median time, 11.4 years) than after myelitis onset (median time, 18.0 years) ($p = 0.002$).

Conclusion

Our results support the benefits of early diagnosis and treatment of NMOSD, especially in persons with severe optic and spinal disability at onset.

Keywords:
Neuromyelitis optica spectrum disorder (NMOSD), Long-term outcome, Opticospinal Impairment Scale (OSIS), NMOSD diagnostic criteria