Finding NMO: A retrospective analysis of the demographics and clinical phenotypes of Neuromyelitis Optica Spectrum Disorder at the University of Kentucky

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BACKGROUND

- Advancements in the field of Neuroimmunology have resulted in a clearer definition and delineation of the major demyelinating processes of Multiple sclerosis, Neuromyelitis Optica Spectrum Disorder (NMOSD) and Myelin oligodendrocyte glycoprotein antibody-associated disease (MOGAD).
- NMOSD is an autoimmune astrocytopathy, distinct from MS and associated with a highly specific serum autoantibody marker (Aquaporin-4 IgG)
- The commercial availability of the Aquaporin-4 IgG antibody in 2007 paved the way for more efficient diagnosis of NMOSD, but it took until 2018 and the arrival of the MOG-IgG live cell-based assay to help discern these diseases further.

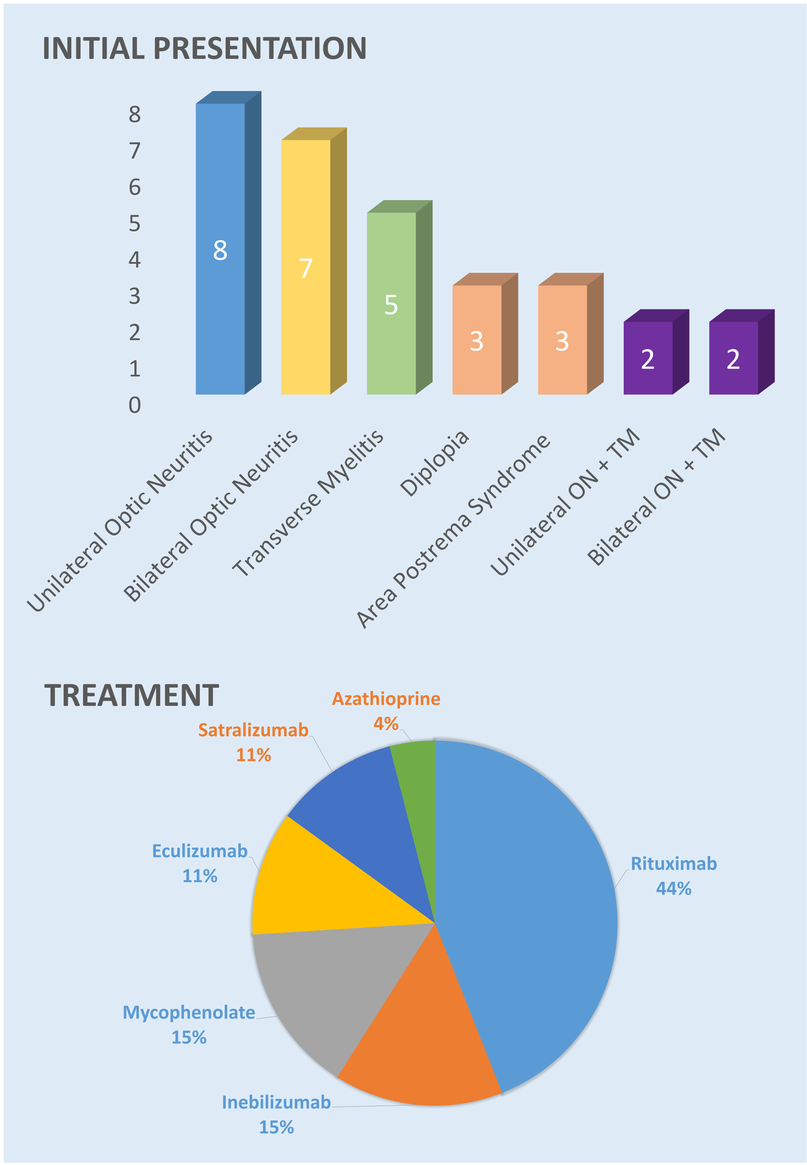
OBJECTIVES

- The aim of this study is to assess the epidemiology, phenotypic variations and overall impact of Neuromyelitis optica spectrum disorder (NMOSD) encountered at the University of Kentucky (UK)

METHODS

- This will be a retrospective study including all patients seen at the University of Kentucky with a tentative or established diagnosis of NMOSD from the year 2018 to present.
- The evaluation will include an analysis of the demographic data, medical history, clinical presentation, neurological exam, diagnostic testing, radiographic evidence, management decisions, disease course and fulfillment of diagnostic criteria.

RESULTS



RESULTS contd.

- 116 patients presented to UK with a high suspicion for NMOSD. 27 patients (23%) were diagnosed as NMOSD, 30 as MOGAD (26%), 8 as Multiple sclerosis (7%), 13 had a separate neurological conditions (11.2%) with the remainder seeking care elsewhere or lost to followup
- Mean age of onset was 38.6 years; youngest at 1 year-3 months and the oldest at 82 years. Mean time to diagnosis was 52.5 months (4.4 years), with the most common alternate diagnosis being MS (31%) followed by Atypical optic neuropathy (23%).
- Initial presentations included Unilateral optic neuritis (30%), Bilateral optic neuritis (26%), Transverse myelitis (19%), Diplopia (11%), Unilateral optic neuritis + Transverse myelitis (7%), Bilateral optic neuritis + transverse myelitis (7%). Area postrema syndrome was seen in 11% of the patients. Two thirds (67%) were AQP-4 positive, with around a third being AQP-4 negative (30%). 3 patients showed concurrent MOG-Ab positivity.
- Rituximab is being used in 44% of patients, Inebilizumab and Mycophenolate mofetil in 15%, Eculizumab and Satralizumab in 11% and a single patient on Azathioprine. No relapses were seen while on FDA approved therapies

CONCLUSIONS

- The results demonstrated the ability of NMOSD to present in the extremes of age, with clinical phenotypes that overlap with other demyelinating conditions. With the majority of cases initially involving the optic nerve and/or spinal cord, it is of utmost importance to reach a prompt diagnosis and initiate an FDA approved therapy, which has showed great effectiveness at preventing relapses in our patients.

REFERENCES

- Dean Wingerchuk, et al. "International consensus diagnostic criteria for neuromyelitis optica spectrum disorders". *Neurology Journals*. 2015 July