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Nmo Spectrum of Disorders – Clinical, Radiologic and Serologic Profile in Indian Patients

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Dr.SREERAMULU DIGUVINTI
ASSOCIATE PROFESSOR OF MEDICINE, GOVERNMENT GENERAL HOSPITAL, KURNOOL MEDICAL COLLEGE, KURNOOL, ANDHR PRADESH

Dr. CHENNAKESAVULU DARA
SENIOR RESIDENT IN GENERAL MEDICINE, GOVERNMENT GENERAL HOSPITAL, KURNOOL MEDICAL COLLEGE, KURNOOL,ANDHRA PRADESH

ABSTRACT *Neuromyelitis optica (also known as Devic's disease) is an idiopathic, severe, demyelinating disease of the central nervous system that preferentially affects the optic nerve and spinal cord. Neuromyelitis optica has a worldwide distribution, poor prognosis, and has long been thought of as a variant of multiple sclerosis; however, clinical, laboratory, immunological, and pathological characteristics that distinguish it from multiple sclerosis are now recognised. The presence of a highly specific serum autoantibody marker (NMO-IgG) further differentiates neuromyelitis optica from multiple sclerosis and has helped to define a neuromyelitis optica spectrum of disorders.We correlate the severity of disease and response to treatment.*

INTRODUCTION:
Neuromyelitis optica (also known as Devic's disease) is an idiopathic, severe, demyelinating disease of the central nervous system that preferentially affects the optic nerve and spinal cord. Neuromyelitis optica has a worldwide distribution, poor prognosis, and has long been thought of as a variant of multiple sclerosis; however, clinical, laboratory, immunological, and pathological characteristics that distinguish it from multiple sclerosis are now recognised. The presence of a highly specific serum autoantibody marker (NMO-IgG) further differentiates neuromyelitis optica from multiple sclerosis and has helped to define a neuromyelitis optica spectrum of disorders[1]. Optico-spinal phenotype of multiple sclerosis dominates demyelinating disorders in Asia. This has been speculated to be, due to, over representation of NMO and its variants among the Indian patients. As, NMO IgG antibody is a reliable tool, with high sensitivity and specificity, in distinguishing NMO spectrum of disorders from other demyelinating illnesses, we undertake a prospective observational study**.**

METHODS:
Clinical and demographic details and results of investigations were collected and compiled in all the patients. Sera were collected for NMO-IgG estimation during the acute phase of illness and send to neuro-ophthalmology laboratory, Sankara Nethralaya, Chennai for testing.

RESULTS:
Of the 40 pts (29F, 11M), 17 had ATM, 9 had ON and 14 had ATM+ON**.** The overall NMO-IgG sero-positivity was 42.5% (17/40), with higher sero-positivity (57%) in ATM+ON group when compared to the other two groups. A higher abnormal Brain MRI (35% vs 21%) and higher relapse rates (59% vs 35%) were found among the sero-positive group compared to sero-negative group.

DISCUSSION:
The term *neuromyelitis optica* was first used in 1894 by Devic, although the first reported cases were described in 1870[2].NMO-IgG reacts with the water channel aquaporin 4. Data suggest that autoantibodies to aquaporin 4 derived from peripheral B cells cause the activation of complement, inflammatory demyelination, and necrosis that is seen in neuromyelitis optica. The knowledge gained from further assessment of the exact role of NMO-IgG in the pathogenesis of neuromyelitis optica will provide a foundation for rational therapeutic trials for this rapidly disabling disease. The relation of neuromyelitis optica to optic-spinal multiple sclerosis in Asia is uncertain. We assessed the capacity of a putative marker for neuromyelitis optica (NMO-IgG) to distinguish neuromyelitis optica and related disorders from multiple sclerosis[3]. Although originally considered a variation of multiple sclerosis (MS), clinical, radiological, pathological, and especially immunological data (detection of NMO IgG antibody) have led to a novel definition of this clinical entity [4]. Despite the use of sensitive assays, aquaporin-4 (AQP4)-specific antibodies are not detected in 10%–40% of patients diagnosed with neuromyelitis optica (NMO) or NMO spectrum disorder [5]. It is also recognized that AQP4 immunoglobulin (Ig) G^+ NMO patients frequently produce other autoantibodies, including antibodies that target nuclear and cytoplasmic antigens identified in certain systemic rheumatologic diseases, including systemic lupus erythematosus and Sjögren syndrome[6].

CONCLUSION:
NMO represents a distinctive subset of CNS demyelinating disorders. The interim data of 40 pts in our study showed high NMO-IgG sero-positivity (42.5%) & its correlation with severity of the disease, poor response to treatment and higher relapse rates.

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