

Neuromyelitis Optica: A Mini Review



Taru Dewan* and Nidhi Sharma

Department of Ophthalmology, Dr. Ram Manohar Lohia Hospital, India

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***Corresponding author:** Taru Dewan, Department of Ophthalmology, Post Graduate Institute of Medical Education and Research, Dr. Ram Manohar Lohia Hospital, New Delhi-110001, India, Tel: 9810673180; Email: tarudewan@hotmail.com

Abstract

Neuromyelitis optica (NMO) is an idiopathic inflammatory disorder affecting the central nervous system. Previously thought to be related to Multiple sclerosis NMO recently came out to be a distinct clinical identity with new diagnostic criteria and physical tests. Broader understanding and confirmatory assay for anti-aquaporin-4 antibodies have changed the diagnostic and treatment protocol for the disease. It carries a significant mortality and morbidity. Newer modalities of treatments for acute exacerbations and for relapses have come up with better understanding. This review describes some of the new findings, clinical manifestations, biomarkers associated with the disease and magnetic resonance imaging characteristics.

Keywords: NMO; MRI; NMOSD; Anti-aquaporin 4 IgG; TM

Abbreviations: NMO: Neuromyelitis Optica; NMOSD: Neuromyelitis Optica Spectrum Syndrome; MRI: Magnetic Resonance Imaging; Anti-AQ4 IgG: Anti Aquaporin-4 Immunoglobulin; SLE: Systemic Lupus Erythemaotosis; ANCA: Anti-Neutrophilic Cytoplasmic Antibody; TM: Transverse Myelitis; CSF: Cerebrospinal Fluid; AION: Anterior Ischaemic Optic Neuropathy; PION: Posterior Ischaemic Optic Neuropathy

Introduction

Neuromyelitis optica (NMO) is a severe idiopathic immune-mediated necrotizing and demyelinating disease that mainly involves the spinal cord and optic nerve [1]. The target antigen is aquaporin-4, the dominant water channel in central nervous system, found in the astrocyte foot processes at the blood-brain barrier [2]. A number of conditions were discovered with anti-AQ4 IgG necessitating coining of the term Neuromyelitis Optica Spectrum of disorders [3]. Antibodies against aquaporin-4 differentiate it from Multiple sclerosis [4].

As NMO carries significant morbidity and mortality, early diagnosis with quick initiation of therapy in the form of corticosteroids, plasma exchange helps in proper management.

Discussion

In India, there is scarcity of epidemiological data for demyelinating disorders. Predominantly females are involved with a mean age of onset 32.6- 45.7 years and median time to first relapse is 8- 12 months [5].

In Indian population an association of HLA-DRB1*03 [6] with NMO has been found which is supported with evidence from Brazil [7], the Carribean Islands [8] and France [9]. It

may be associated with other systemic autoimmune disorders like SLE, Sjogren's syndrome or p-ANCA associated vasculitis. Sometimes onset may be associated with acute infection like varicella zoster or HIV.

Clinical Manifestations

Longitudinally Extensive Transverse Myelitis (LETM) defined as longitudinal cord lesions extending in more than three vertebral segments and acute severe optic neuritis are classical features of NMO [3]. The core features of NMO include optic neuritis, acute myelitis, area postrema syndrome, acute brainstem syndrome, symptomatic narcolepsy or acute diencephalic clinical syndrome and symptomatic cerebral syndrome [1]. In 15-40% of cases there is concomitant occurrence of both optic neuritis and transverse myelitis [3]. Severe visual impairment, bilateral simultaneous or sequential optic neuritis in rapid succession with poor recovery with visual acuity of 20/200 or worse [10,11] and non central scotoma deficits such as altitudinal field defects should raise suspicion for NMOSD [12]. Classical tumefactive and longitudinal lesions in MRI may be missed in early course of disease and during resolution phase [13]. MRI findings of posterior optic nerve

involvement particularly chiasm is highly suggestive of NMO. Other common findings are neuropathic pain, Lhermitte's sign and painful tonic spasms [14].

Laboratory Testing

Suspected patients should be evaluated for anti-aquaporin-4 antibodies. Cell based assay is preferred over tissue based assay or ELISA because of higher sensitivity and specificity [15]. CSF analysis is not mandatory and only indicated in doubtful cases. CSF findings are dependent on activity status of the disease. In acute stages WBC count (>380cells/ul) [16], glial fibrillary acidic protein, serum proteins mainly albumin [16] and IL-6 [17,18] are raised.

Differential Diagnosis

It includes infectious causes like bacterial and viral, reactive causes like post infectious, vascular like AION, PION, nutritional causes like vitamin deficiency, tobacco consumption, compressive lesions like ocular tumors, familial and hereditary causes. Differentials of TM are spinal infarction, fistulas, compressive lesions, B12 deficiency, paraneoplastic syndromes, multiple sclerosis and infections [1].

Treatment

Corticosteroids and plasma exchange are most commonly employed therapies for acute exacerbations. IV immunoglobulins may also be used [1].

Corticosteroids

Acute exacerbations are treated with IV methylprednisolone 1g/day for 5 days followed by prolonged prednisolone taper for 2 to 6 months [1].

Plasma exchange

Considered as first line therapy in patients not responding to steroid therapy or in patients who were successfully treated with plasma exchange previously. Typical protocol is an exchange of 1 to 1.5 volumes of plasma every other day for a total of five treatments [1].

IV immunoglobulins

Suggested in patients with no response or contraindication to corticosteroids or plasma exchange [1].

Conclusion

NMO is an inflammatory disorder with high mortality and morbidity so early recognition of disease offers a window of opportunities for treatment and induction of remission.

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