

# Neuromyelitis Optica Spectrum Disorders with Autoimmune Diseases

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Neuromyelitis optica (NMO) spectrum disorders (NMOSD) represent an evolving spectrum of inflammatory demyelinating central nervous system-based auto-immune diseases extending beyond the optic nerves and spinal cord to include the brain stem, cerebrum, skeletal muscles, the circumventricular organs and the diencephalon, from monophasic to a polyphasic illness with multiple recurrences [1-3]. NMOSD is a kind of multi-faceted and complex disease. Besides host genetic and environmental factors, the major immunopathological mechanisms of the NMOSD is the presence of aquaporin-4 (AQP4) antibodies (also known as AQP4-IgG or NMO-IgG). The discovery of highly specific AQP4-IgG which is predominantly located on astrocyte foot processes contributing to the formation of blood-brain barrier (BBB), opened a new era in the classification and understanding of NMOSD pathogenesis and holded promise for greater diagnostic accuracy and individualized care in NMOSD [4-6].

The growing attention is now focusing on the associations between NMOSD and the variety of autoimmune diseases, including non organ specific autoimmune diseases (rheumatoid arthritis (RA), systemic lupus erythematosus (SLE), Sjogren syndrome (SS), antiphospholipid syndrome (APLS), undifferentiated connective tissue disease (UCTD)), organ-specific autoimmune diseases (thyroid diseases, myasthenia gravis, ulcerative colitis), and co-existent autoantibodies without diseases [3,7-10]. However, the pathophysiological mechanism of this association remains unclear and it is unknown if these diseases are an indication, the progressive forms, the primary cause of NMOSD or a concomitant autoimmune disease. Some studies suggest immune pathologic mechanism and humoral immune response may be involved in the pathogenesis of NMOSD co-occurring with autoimmune diseases [1-13]. We extend the pathogenic mechanism of coexistence of NMOSD and autoimmune diseases from previous hypothesis and provide additional evidences, all of these suggest that it is etiopathogenetically heterogeneous, there is disorder in immune regulation or changes of the multiple targets with autoantibodies, have a dysimmune state or show higher susceptibility to multiple autoimmune conditions, share certain autoimmune aetiology similarities with autoimmune disorders which trigger a cross immune response [14]. NMOSD diagnosis is primarily clinical manifestations, MRI and pathology. The molecular biomarkers, including Anti-AQP4, Anti-myelin oligodendrocyte glycoprotein (MOG), glial fibrillary acidic protein (GFAP, a key indicator of astrocytic injuries) and Th1, Th2, Th17, Treg related cytokines [15,16]. Especially, Anti-AQP4, play an important role in the diagnosis, evaluation of disease activity, and therapeutic strategies for NMOSD. As for NMOSD co-occurring with autoimmune diseases, it is not only help for the nosologic classification, but also makes a paradigm shift in diagnosis and treatment of these. acute attacks aims at minimizing residual disability, such as high doses of intravenous corticosteroids and plasma exchange (PE), whereas The maintenance treatment are mainly focused on reducing relapse, low doses of corticosteroids and immunosuppressive therapy, azathioprine, mycophenolatemofetil, and rituximab tend to be the most-recommended first-line therapies, methotrexate, mitoxantrone, cyclophosphamide, cyclosporine and immunoglobulin are the second-line therapies for NMOSD. However, there are many open questions

regarding the treatment of NMOSD, such as drug comparisons, duration of treatment and toxicities in children and pregnancy [17-21].

Overall, clinical and immunopathological studies of the NMOSD associated with autoimmune diseases available in the literature is limited. Furthermore, there is no consensus about these. We should continue to explore the clinical utility and cost-effectiveness of the detection of these autoimmune diseases and autoantibodies in NMOSD. Moreover, further research is necessary to elucidate the relationship between autoimmune diseases and NMOSD, especially neurologists and rheumatologists, more importantly, help to choose suitable and effective therapies and improve the prognosis.

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