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Neuro-Myelitis Optica Spectrum Disease (NMOSD)

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Abstract:

Neuromyelitis optica spectrum disorder (NMOSD) is a rare autoimmune disease that affects the central nervous system. It is characterized by inflammation of the optic nerve and spinal cord, which can cause vision loss, muscle weakness, and other neurological symptoms. The main objective of this study was to conduct a review study of this disease from different perspectives including an overview of the disease, prevalence, diagnosis, symptoms, and treatment. It can be concluded that NMOSD is a rare autoimmune disorder that primarily affects the optic nerve and spinal cord. Studies on NMOSD have provided important insights into the pathogenesis, diagnosis, and treatment of the condition.

INTRODUCTION

This study discusses one of the rare autoimmune diseases**,** neuromyelitis optica spectrum disorder (NMOSD) in detail. Although this disease is rare (Douedi *et al.,* 2022), but it can be misdiagnosed with multiple sclerosis. The main objective of this study was to conduct a review study of this disease from different perspectives including an overview of the disease, prevalence, diagnosis, symptoms, and treatment in the following sections.

An Overview of NMOSD

Neuromyelitis optica spectrum disorder (NMOSD) is a rare autoimmune disease that affects the central nervous system. It is characterized by inflammation of the optic nerve and spinal cord, which can cause vision loss, muscle weakness, and other neurological symptoms (Wingerchuk *et al.,* 2015).

NMOSD is also known as Devic's disease, named after Eugene Devic, the French neurologist who first described the condition in 1894. NMOSD primarily affects the optic nerves and spinal cord, although it can also affect other parts of the nervous system (my.clevelandclinic.org, 2023).

Symptoms of NMOSD

The primary symptoms of NMOSD include inflammation and damage to the optic nerves, which can lead to vision loss or blindness in one or both eyes. In addition, inflammation and damage to the spinal cord can cause muscle weakness or paralysis, numbness, tingling, and bowel or bladder dysfunction. Other neurological symptoms may include difficulty with speech, memory loss, and seizures (Jarius and Wildemann, 2013).

Causes of NMOSD

The underlying cause of NMOSD is believed to be an autoimmune reaction in which the body's immune system attacks and damages the myelin sheath, the protective covering around nerve fibers in the central nervous system. This damage can lead to inflammation and scarring, which can cause the symptoms of NMOSD (Alkhatib, 2022).

Prevalence of NMOSD

NMOSD is rare, affecting an estimated 1 in 100,000 people worldwide. It is more common in women than in men, and usually first appears between the ages of 20 and 40 years (Kingwell *et al.,* 2013).

Treatment of NMOSD

There is no cure for NMOSD, but there are treatments available that can help manage the symptoms and reduce the risk of relapse. These treatments include immunosuppressive medications, corticosteroids, and plasma exchange therapy (Kim *et al.,* 2018).

The differences between NMOSD and multiple sclerosis

NMOSD is a rare inflammatory illness of the central nervous system that has clinical features with optic neuritis, myelitis, and some disorders of the brain and brainstem. The acronym NMOSD stands for neuromyelitis optica syndrome. The question of whether NMOSD is a severe form of multiple sclerosis (MS) has been debated for a long time. However, the discovery of an NMOSD-specific aquaporin 4 (AQP4) antibody and the subsequent clinical, immunological, and pathological data have demonstrated that NMOSD is, in fact, a separate entity. This is in spite of the fact that the possibility of NMOSD being a severe form of MS has been debated for a long time (Lennon *et al.,* 2004, 2005; Fujihara *et al.,* 2012). Instances that are clinically identified as NMOSD may, at this time, include myelin oligodendrocyte glycoprotein (MOG)-antibody-seropositive inflammatory demyelinating disease, AQP4-antibody- seropositive autoimmune astrocytopathic illness, and double-seronegative disease (Fujihara, 2019).

NMOSD and the expression of Anti-AQP4 antibodies

NMOSD is a demyelinating inflammatory lesion that primarily affects the optic nerves, spinal cord, and brainstem (Nakashima, 2015; Hor *et al.,* 2020). Anti–Aquaporin 4 (a membrane-bound water channel extensively expressed on astrocytic foot processes)–IgG is found in 70–90% of NMO patients. Anti-AQP4 antibodies have been shown to be harmful in vitro and in vivo, and intracerebral injection of AQP4 IgG with complement causes demyelination like NMOSD (Hor *et al.,* 2020).

AntiAQP4 antibody titers in serum are 1,000-fold higher than in CSF, and CSF oligoclonal bands, which are found in only 15–30% of NMO patients, frequently fade away with disease progression, showing that B cell activation and the origin of the humoral immune response are located outside the CNS. Following BBB breach, activated B lymphocytes may penetrate the CNS and produce disease, as previously stated. Tolerance is maintained during normal early B cell growth by the elimination of most self-reactive B cells. AntiAQP4 IgG-producing B cells appear to emerge from early B cell tolerance checkpoint abnormalities, both centrally and peripherally in the bone marrow, resulting in an increased number of autoreactive B cells in the mature naive B cell population (Cotzomi *et al.,* 2019). After somatic hypermutations, this reservoir of auto-reactive B cells can provide B cell clones that produce pathogenic anti-AQP4 antibodies. The fact that unmutated precursors of B cells secreting anti-AQP4 antibodies do not bind to autoantigen supports the conclusion that anti-AQP4 specificity and the generation of pathogenic autoantibodies require affinity maturation and the acquisition of somatic hypermutations, both of which are regulated by T cells (Cotzomi *et al.,* 2019). Additionally, B cell clones may be activated by an antigen other than AQP4, such as a peptide from *Clostridium perfringens*, an intestinal bacterium native to the United States (which has homology to an immunodominant epitope of AQP4) (Hor *et al.,* 2020).

Variations between NMO and neuromyelitis optica

The comparative clinical and demographic aspects of neuromyelitis optica are not as well-known as they might be (NMO). In this study, we investigated publications that were evaluated by specialists to identify the incidence and prevalence of NMO, as well as its clinical phenotypes and demographic factors. We also looked at the papers' abstracts to see how well they described the disease. According to research that was carried out on populations in Europe, South East and Southern Asia, the Caribbean, and Cuba, the incidence and prevalence of non-communicable diseases like NMO range from 0.05–0.4 and 0.52–4.4 per 100,000, respectively. This research was conducted in Europe, South East and Southern Asia, the Caribbean, and Cuba. The language used for the research was English. The same age range was revealed for both the mean age at onset (32.6–45.7) and the median time to first relapse. The mean age at onset was determined to be the age at which symptoms first appeared (8–12 months). According to the majority of studies, females are more likely to be afflicted by the disorder, and it appears to have a relapsing pattern, particularly in people who have tested positive for an anti-aquaporin 4 antibody (anti AQP4-IgG). It is probable that ethnicity has an effect, not only on the clinical outcome of the disease, but also on the phenotypic of the disease itself. In spite of the limitations that are inherent to the review process, common patterns that have been identified in the clinical and demographic aspects of NMO in a variety of populations contribute to a greater global understanding of NMO and the strategies that may be used to combat it. These patterns can be found in the clinical and demographic aspects of NMO (Pandit *et al.,* 2015).

Neuromyelitis optica, often known as NMO, is an autoimmune disease that causes inflammation of the central nervous system and mostly affects the optic nerve and spinal cord. Up until quite recently, it was believed that NMO was a clinical variation of multiple sclerosis (MS). On the other

hand, clinical characteristics, neuroimaging characteristics, immunological characteristics, and histological characteristics have now been established that differentiate NMO from MS. In 1894, Clifford and Gault provided a summary of the disease's most important characteristics, despite the fact that descriptions of the sickness had been published much earlier. The initial Wingerchuk diagnostic criteria for NMO were established in 1999 and included the generally lengthy spinal cord segmental involvement, longitudinally extensive transverse myelitis (LETM), and neutrophilic pleocytosis in the cerebrospinal fluid (CSF) (Wingerchuk *et al.,* 1999). After that, the subsequent finding of anti-AQP4-IgG (Lennon *et al.,* 2004) led to the revision of the Wingerchuk criterion in the year 2006 (Wingerchuk *et al.,* 2006). The various types of NMO that do not meet the criteria set in 2006 are grouped together under the umbrella term "NMO spectrum disorders" (NMOSD). Examples of this condition include isolated unilateral or simultaneous bilateral or recurrent optic neuritis (ON), isolated or recurrent transverse myelitis (TM), and typical NMO brain lesions (hypothalamus, corpus callosum, brainstem, periventricular) with or without detectable anti AQP4-IgG autoantibody. In light of this, the diagnostic criteria have been improved, and the range of clinical symptoms associated with NMO has broadened during the past few years (Wingerchuk *et al.,* 2007).

CONCLUSION

Neuromyelitis optica spectrum disorder (NMOSD) is a rare autoimmune disorder that primarily affects the optic nerve and spinal cord. Studies on NMOSD have provided important insights into the pathogenesis, diagnosis, and treatment of the condition.

NMOSD studies pointed out that:

1. The discovery of aquaporin-4 (AQP4) antibodies as a biomarker for NMOSD: AQP4 is a water channel protein that is expressed on the surface of astrocytes, and the presence of AQP4 antibodies is highly specific for NMOSD. The discovery of AQP4 antibodies has greatly improved the accuracy of NMOSD diagnosis and has enabled the development of targeted therapies.

2. The identification of disease triggers: Studies have identified various factors that can trigger NMOSD, including infections, vaccinations, and pregnancy. Understanding these triggers can help to prevent or manage NMOSD flares.

3. The development of effective treatments: There is currently no cure for NMOSD, but studies have led to the development of several effective treatments. These include immunosuppressive therapies, plasma exchange, and monoclonal antibody therapies that target AQP4.

4. The importance of early diagnosis and treatment: Early diagnosis and treatment are crucial for improving outcomes in NMOSD. Studies have shown that delaying treatment can lead to irreversible neurological damage and disability.

CONFLICT OF INTEREST

The author declares that this article content has no conflict of interest.

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