National Journal of Advanced Research
www.thestudiesjournal.com
ISSN: 2455-216X
Received: 01-04-2025, Accepted: 02-05-2025, Published: 17-05-2025
Volume 11, Issue 1, 2025, Page No. 32-34

A review article: Devic's syndrome

Bansari B Patel[1], Divia Acha Jacob[2], Shalini S[3]

[1] Department of Obstetrics and Gynaecology Nursing, Jubilee Mission College of Nursing, Bangalore, Karnataka, India

[2] Asst.Professor, Department of Medical Surgical Nursing, Jubilee Mission College of Nursing, Bangalore, Karnataka, India

[3] Lecturer, Department of Obstetrics and Gynaecology Nursing, Jubilee Mission College of Nursing, Bangalore, Karnataka, India

Abstract

Neuromyelitis optica (nmo), Or devic's syndrome, is a condition marked by inflammation of the optic nerve (optic neuritis) and spinal cord (myelitis). The discovery of nmo-igg autoantibodies in approximately 80% of patients has sparked significant interest in the scientific and medical communities. These autoantibodies target aquaporin-4 (aqp4), A protein crucial for water regulation in the central nervous system. This article provides a comprehensive overview of nmo's clinical characteristics, Underlying immune mechanisms, And treatment approaches. Key topics include the expanding range of aqp4-related autoimmune disorders, The diagnostic role of mri and optical coherence tomography, And the involvement of, T cells, And granulocytes in the condition's pathology. We also explore promising new therapies for this rare yet often debilitating disease.

Keywords: Neuromyelitis optica, Devic's syndrome, Aquaporin-4 (aqp4), Nmoigg, Myelitis, Optic neuritis

Introduction

Non-formal education is a form of education that occurs outside the formal school system. It encompasses all organized educational activities that take place outside of the formally established system. Non-formal education according to Ihejirika (2000) [3], is any organized or systematic educational activities carried out outside the framework of the formal school system to provide a specific type of learning to a specific sub-group of the population, both adults and children. It includes different types of learning experiences; it is a lifelong process of learning that includes adult and continuing education, the apprenticeship system, in-service programme, on-the-job training programme, personnel and professional development, and workers' and students' industrial training. Short-term learning activities such as conferences, seminars, workshops, and evening classes, as well as specialized purpose programs such as functional and literacy programs, volunteer youth programs, skill acquisition, and liberal education classes, constitute non-formal education, which can be called "living room" or leisure education (Amirize in Ossai & Nwalado, 2014) [8]. The various skills acquisition and apprenticeship programs are examples of non-formal education. Neuromyelitis Optica Spectrum Disorder (NMOSD) is a rare, autoimmune, inflammatory condition that primarily affects the central nervous system, particularly the optic nerves and spinal cord. Previously known as Devic's disease, NMOSD can cause vision loss, muscle weakness, and other neurological symptoms. [1] The disorder is often associated with antibodies against aquaporin-4 (AQP4) or myelin oligodendrocyte glycoprotein (MOG). NMOSD can be relapsing or monophasic, with varying degrees of severity. Accurate diagnosis and treatment are crucial to managing symptoms and preventing long-term damage.

In recent years, the definition of neuromyelitis optica has been expanded as a specific antibody was discovered in the serum of affected patients, and various manifestations have been recognized in a spectrum of diseases. Because of this, the term neuromyelitis optica spectrum disorder is now used to include optic neuritis with spinal cord manifestations and other neurologic disorders associated with the serum aquaporin-4 immunoglobulin G antibodies (AQP4-IgG) [3].

Historical Background

The earliest reports of patients with symptoms of myelitis and vision loss date back to the early 19th century. However, it wasn't until Eugène Devic and Fernand Gault published a comprehensive review in 1894 that neurologists and ophthalmologists began to take sustained interest in this rare condition [1]. Devic and Gault introduced the term "neuro-myélite optique aiguë" to describe the syndrome. Later, in 1907, the Turkish physician Acchioté proposed naming the condition after Devic, in recognition of his contributions [4].

Definition

Devic's disease is an autoimmune disorder that causes severe demyelination and axonal damage, predominantly in the optic nerve and spinal cord, and is distinguished from MS by its association with serum AQP4-IgG antibodies [5].

There are 2 types of NMO

- **Relapsing form:** This type is characterized by periodic relapses, with varying degrees of recovery in between. Women are disproportionately affected, outnumbering men [6].
- **Monophasic form**: This type involves a single episode lasting about a month or two, followed by a prolonged period of remission, sometimes lasting several years. Men and women are equally affected [6].

Causes

- **Autoimmune Factors**

1. Autoantibodies: The detection of NMO-IgG autoantibodies targeting aquaporin-4 (AQP4) is a key diagnostic marker for NMO.

2. Immune system dysregulation: Immune system dysregulation, involving Tcell and B-cell activation, plays a role in NMO's development.

- **Genetic Factors**

1. Genetic predisposition: Genetic predisposition may contribute to NMO risk, but no single gene is known to be the sole cause.

- **Environmental Factors**

1. Triggers: Infections or other autoimmune diseases might trigger NMO onset in people with a predisposition.

- **Other Factors**

1. Inflammation: Inflammation in the central nervous system (CNS) plays a key role in the development of NMO lesions.

Symptoms

Symptoms of neuromyelitis optica (NMO) vary between people and can be mild or severe.

The symptoms can include [5]

- Eye pain, diminished colour vision in one or both eyes
- Sharp, burning, or shooting sensations (nerve pain) may be felt in the back, neck, arms, or legs.
- Muscle weakness, numbness, or paralysis can occur in the arms or legs.
- Muscles may contract suddenly, causing spasms.
- Bowel and bladder dysfunction can occur, leading to symptoms like incontinence, constipation, or difficulty emptying the bladder.
- Sexual function can be affected, leading to issues like erectile problems or impaired orgasmic function.
- Vomiting and hiccups

Diagnostic evaluation

- Physical and neurological examinations. A neurological exam plays a crucial role in detecting issues with senses, reflexes, muscle function, balance, and facial movements.
- Blood tests: AQP4 or MOG antibodies.
- MRI scan: to detect the brain and spinal cord abnormalities.
- Lumbar puncture: to assess take a sample of fluid from spinal cord.
- zzOphthalmoscopic examination: to check the eyes and vision abnormalities [7].

Management

NMO is a manageable condition, and treatment focuses on two main areas: acute care to address relapses and long-term management to prevent future attacks.

- Acute treatment targets the immediate consequences of an NMO relapse, primarily inflammation, with corticosteroids or other anti-inflammatory medications being the primary options. Prompt treatment is crucial to minimize lasting damage**.**
- Long-term management of NMO involves immunosuppression or immunomodulation to prevent the immune system from attacking the nervous system, thereby reducing the frequency, severity, and duration of attacks.

There are several drugs and treatments that can help treat NMO, including:

- **Anti-inflammatory drugs:** These drugs reduce the inflammation of your nervous system. The most common drugs that do this are corticosteroids, such as prednisone.

- **Plasma exchange (plasmapheresis):** Plasma exchange is an option when steroids aren't effective. It involves exchanging a patient's plasma with donor plasma to remove immune cells and chemical markers that drive inflammation**.**

- **Intravenous immunoglobulin (IVIG):** This treatment involves IV infusion of immunoglobulin, a plasma product containing donor antibodies that won't trigger an immune response.

- **Immune-suppressing drugs (immunosuppressants)** [8]

Complication

There are several complications of NMO including:

- Visual impairment or blindness
- Paralysis or weakness of 1 or more limb
- Stiffness or muscle spasms
- Loss of bowel or bladder control
- Depression
- Fatigue [10]

Conclusion

Devic's syndrome, or Neuromyelitis Optica (NMO), is a rare and complex autoimmune disorder that requires prompt diagnosis and treatment. While there is no cure, advances in research and treatment options have significantly improved outcomes for individuals affected by this condition. With ongoing management and care, people with NMO can lead fulfilling lives, and continued research holds promise for even better treatments and quality of life in the future.

References

1. Jarius S, Wildemann B. The history of neuromyelitis optica. J Neuroinflammation,2013:10:8.
2. Guier CP, Kaur K, Stokkermans TJ. StatPearls [Internet]. StatPearls Publishing; Treasure Island (FL): Jan 20, 2025. Optic Neuritis.
3. Wingerchuk DM, Banwell B, Bennett JL, Cabre P, *et al*, International Panel for NMO Diagnosis. International consensus diagnostic criteria for neuromyelitis optica spectrum disorders. Neurology,2015:85(2):177-89.
4. Jarius S, Wildemann B. 'Spinal amaurosis' (1841). On the early contribution of Edward Hocken to the concept of neuromyelitis optica. J Neurol,2014:261:400–404.
5. https://www.mayoclinic.org/diseases-conditions/neuromyelitis-optica/symptomscauses/syc,20375652.
6. https://www.hopkinsmedicine.org/health/conditions-and-diseases/neuromyelitisoptica.
7. Wingerchuk DM, Banwell B, Bennett JL, *et al*. international consensus diagnostic criteria for neuromyelitis optica spectrum disorders

https://www.ncbi.nlm.nih.gov/pmc/articles/PMC4515040/). Neurology,2015:85(2):177-189.
8. Jarius S, Wildemann B, Paul F. Neuromyelitis optica: clinical features, immunopathogenesis and treatment.
9. {https://www.ncbi.nlm.nih.gov/pmc/articles/PMC3992027/). Clin Exp Immunol,2014:176(2):149-164.
10. Douglas VC, Aminoff MJ. Neuromyelitis Optica. In: Papadakis MA, McPhee SJ, Rabow MW, McQuaid KR, eds. Current Medical Diagnosis & Treatment 2023. McGraw Hill, 2023.
11. Wingerchuk DM, Pittock SJ, Lucchinetti CF, Lennon VA, Weinshenker BG. A secondary progressive clinical course is uncommon in neuromyelitis optica. Neurology,2007:68:603–605.