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(CASE REPORT)

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A case report on seronegative neuromyelitis optica spectrum disorder

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Abstract

An inflammatory, demyelinating, autoimmune disease that primarily affects the central nervous system mainly targeting optic nerve and spinal cord is called neuromyelitis optica (NMO). The term NMOSD refers to a recently revised nomenclature that includes new diagnostic criteria, such as serum aquaporin-4 immunoglobulin G (AQP4-IgG) antibody serological testing.

One of the two common symptoms of NMOSD is optic neuritis or myelitis, which might appear as the initial symptom. And other symptoms include weakness, numbness, paralysis of limbs, nausea, vomiting, imbalance.

It is thought to be related to an autoimmune reaction in which the immune system unintentionally targets healthy cells, particularly targeting proteins in the central nervous system like aquaporin-4. Seronegative NMOSD is treated with methylprednisolone, plasma exchange therapy, and IV immunoglobulin-G therapy.

Keywords: Seronegative neuromyelitis optica spectrum disorder; Autoimmune disease; Serum Aquaporin-4 immunoglobulin G; Methylprednisolone; Optic neuritis; Optic myelitis

1. Introduction

An inflammatory, demyelinating, autoimmune disease that primarily affects the central nervous system mainly targeting optic nerve and spinal cord is called neuromyelitis optica (NMO). The term NMOSD refers to a recently revised nomenclature that includes new diagnostic criteria, such as serum aquaporin-4 immunoglobulin G (AQP4-IgG) antibody serological testing. The patient will be placed in a seronegative subgroup based on negative antibody results [1].

AQP4-antibody serostatus was used to stratify NMOSD into two categories: AQP4-antibody-seronegative NMOSD (or unknown serostatus) and AQP4-antibody-serosepositive NMOSD. When diagnosing AQP4-antibody-seropositive NMOSD, a key clinical characteristic such as optic neuritis, acute myelitis, or brain syndrome is all that is needed if the AQP4-antibody is consistently positive (a cell-based assay is preferable) and other diagnoses are ruled out [2].

One of the two common symptoms of NMOSD is optic neuritis or myelitis, which might appear as the initial symptom. And other symptoms include weakness, numbness, paralysis of limbs, nausea, vomiting, imbalance. Uncertainty surrounds the precise etiology of seronegative neuromyelitis optica spectrum disorder (NMOSD). It is thought to be related to an autoimmune reaction in which the immune system unintentionally targets healthy cells, particularly targeting proteins in the central nervous system like aquaporin-4. Further research is required to fully understand the underlying origins of this illness; however, it is possible that genetic, environmental, and possibly viral variables may contribute to the activation of this immune response [3].

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As exacerbations of NMOSD cause substantial residual disability, acute therapy is vital. Acute therapy includes high dose of IV methylprednisolone (IVMP) 1g/daily for 3-7 days or plasma exchange (PLEX) is done for every other day for 2 weeks (1.5L, 5-7 treatments) when response to IVMP is poor or absent. If response to PLEX is poor IV immunoglobulin-G therapy (IVIgG) can be used [4].

2. Case report

A 45 year old male patient was brought into the hospital with chief complaints of vomiting's 5-6 episodes, no projectile, not blood stained, blurring of vision from past 5 months, progressive, initially he was able to count fingers from a distance, headache. The patient was then taken to the LV prasad eye hospital, where he was treated with steroids i.v.o B/L optic neuritis. The patient was not responding to the treatment and was referred to NIMS for PLEX. He was tapered on wysolone (there was no improvements in vision), and was kept on mycophenolate mofetil (MMF). In NIMS the patient underwent 5 sessions of PLEX and 22 doses of IVIG were given. Then the patient slowly developed to perceive light 4 days after PLEX. Then the patient was discharged and wysolone was prescribed. And after 2 months the patient had developed decrease vision in the right eye and he tried for PLEX then the patient developed hemothorax which was drained by ICP. Then IVIG was started for 5 days which lead to no improvements.

On examination the patient was conscious, coherent, pulse rate was 87 beats per minute, blood pressure was 120/80mmHg, Spo_2 was 97%, CVS was S_1S_2+, respiratory system was BAE+, per abdomen was soft.

The AQP 4 antibody test was negative and MOG Alb was negative, C reactive protein was negative, hemoglobin 12.2 g/dL (decreased), WBC 27.4k/µL (increased), platelets 2.9 lakhs//µL, MCV 86.3 femtoliters, urea 28 mg/dL, AST 20 units/L, ALB 2.9g/dL, Ca^{+2} 8.5 mg/dl , Po_4^- 2.1 mg/dl, Na^+ 127 mEq/L (decreased), K^+ 3.5 mmol/L, Cl^- 87 mmol/L (decreased), CSF fluid was colorless and clear, glucose 85 mmol/L, MRI with orbits with contrast shows enhancement involving immediate retrobulbar portions of B/L optic nerves.

Considering the clinical presentation and by examining the clinical findings the patient was diagnosed with seronegative neuromyelitis optica spectrum disorder with optic neuritis.

The patient was prescribed with the following medications

- Tablet Wysolone 60 mg OD is a glucocorticoid, used for anti-inflammatory and immunosuppressant,
- Tablet Mycophenolate mofetil 1.5 g OD is an inosine monophosphate dehydrogenase inhibitor, used as an immunosuppressant,
- Injection Multivitamin 1 amp in 100ml NS IV OD
- Tablet Pantoprazole 40 mg OD is a proton pump inhibitor, used for erosive gastritis and gastric acid hypersecretion,
- Tablet Chlorpheniramine 4 mg OD is a histamine-H1 receptor antagonist, used for allergic reactions, hay fever, rhinitis.

3. Discussion

The disease's recurrent course, marked by recurrence and partial remissions, results in progressive impairment that significantly lowers a patient's quality of life. Appropriate recognition of the disease has been enhanced by the identification of antibodies against aquaporin 4 (AQP4), their crucial involvement in the etiology of NMO, and the development of diagnostic criteria. Understanding the causes of NMO has advanced quickly in recent years, opening up a wider variety of therapeutic possibilities [5]. For almost all AQP4-IgG-positive individuals, NMOSD has a relapsing course; however, for AQP4-IgG-negative patients, it may be monophasic [6]. The majority of patients received immunosuppressive or immunomodulatory medication at least once. Individuals receiving treatment with a wide range of immunosuppressive or immunomodulatory medications, including interferon beta, glatiramer acetate, azathioprine, rituximab, cyclophosphamide, IVIG, or mitoxantrone [7].

4. Conclusion

The seronegative neuromyelitis optica spectrum disorder is rare, autoimmune disorder. While the symptoms of NMOSD are comparable to those of typical NMOSD, the disorder is not accompanied by the particular antibodies linked to it. Even though the precise etiology is still unknown, it most likely stems from an autoimmune reaction that targets proteins in the central nervous system. Transverse myelitis, region postrema syndrome, optic neuritis, and brainstem

symptoms are among the symptoms. Hemothorax is an uncommon side effect of treatments such as plasma exchange. For those exhibiting possible symptoms of this illness, seeking medical assistance is essential for an accurate diagnosis and effective symptom management.

Compliance with ethical standards

Disclosure of conflict of interest

No conflict of interest to be disclosed.

Statement of ethical approval

'The present research work does not contain any studies performed on animals/human's subjects by any of the authors.

Statement of informed consent

Informed consent was obtained from all individual participants included in the study.

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