

ORIGINAL

Management of neuromyelitis optica spectrum disorder at the Escuela Hospital, Honduras

Manejo del trastorno del espectro de neuromielitis óptica en el Hospital Escuela, Honduras

Dulce Escalante¹ , Gabriel Isaias Lagos¹ , Harlis Estrada Pastrana² , Pablo Carías³ , Alejandro Carías⁴ , Jhossmar Cristians Auza-Santivañez⁵ , Freddy Ednildon Bautista-Vanegas⁶ , Dilma Villca-Villca⁷ 

¹Facultad de Ciencias Médicas. Universidad Nacional Autónoma de Honduras. Tegucigalpa, Honduras.

²Hospital Escuela. Servicio de Neurología. Secretaría de Salud de Honduras. Tegucigalpa, Honduras.

³Facultad de Ciencias Médicas, Departamento de Medicina Interna, Universidad Nacional Autónoma de Honduras. Honduras.

⁴Facultad de Ciencias Médicas, Departamento de Cirugía, Universidad Nacional Autónoma de Honduras. Tegucigalpa, Honduras.

⁵Ministerio de Salud y Deportes, Instituto Académico Científico Quispe-Cornejo. La Paz, Bolivia.

⁶Neurologisches Fachkrankenhaus für Bewegungsstörungen / Parkinson: Beelitz HeilstättenKliniken. Beelitz GmbH, Brandenburg Deutschland, Germany.

⁷Hospital Obrero N° 4, Caja Nacional de Salud - Regional Oruro. Oruro, Bolivia.

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Corresponding author: Alejandro Carías 

ABSTRACT

Introduction: Neuromyelitis optica spectrum disorder is an autoimmune-based nosological entity with inflammatory characteristics that affects the central nervous system: optic nerves, spinal cord and brainstem.

Objective: to present a series of clinical cases to briefly review the management of neuromyelitis optica spectrum disorder.

Method: a retrospective and descriptive analysis was performed on the patients who attended the outpatient clinic of the Teaching Hospital and who have been diagnosed and followed up for neuromyelitis optica spectrum disorder over the last 20 years.

Results: eight patients (100,0 %) were female, ranging in age from 19 to 45 years, with a mean of 32 years. All patients (8) (100,0 %) underwent anti-aquaporin 4 antibodies for diagnosis; four (50,0 %) had unremarkable brain and orbital magnetic resonance imaging (MRI).

Conclusions: Neuromyelitis optica spectrum disorder is a disease that requires an accurate diagnosis based on clinical features, anti-aquaporin 4 antibodies, and imaging studies. Treatment may begin with steroids, although in cases where the disease is not controlled, rituximab may be used.

Keywords: Neuromyelitis Optica; Central Nervous System; Rituximab.

RESUMEN

Introducción: el trastorno del espectro de neuromielitis óptica constituye una entidad nosológica de base autoinmune con características inflamatorias que afecta al sistema nervioso central: los nervios ópticos, médula espinal y tronco encefálico.

Objetivo: presentar una serie de casos clínicos para realizar una breve revisión sobre el manejo del trastorno del espectro de neuromielitis óptica.

Método: se realizó un análisis retrospectivo y descriptivo de las pacientes que asistieron a consulta externa del Hospital Escuela, que han sido diagnosticados y seguidos en los últimos 20 años por trastorno del espectro de neuromielitis óptica.

Resultados: las 8 (100,0 %) fueron pacientes femeninas, en edades entre 19 y 45 años, con una media de 32 años. A todas las pacientes 8 (100,0 %) se les realizó para el diagnóstico los anticuerpos anti-acuaporina 4; 4 (50,0 %) pacientes tenían Imágenes por Resonancia Magnética (IRM) de cerebro y orbitas sin alteración.

Conclusiones: el trastorno del espectro de neuromielitis óptica es una enfermedad que requiere un diagnóstico preciso, que pasa por la clínica, anticuerpos anti-acuaporina 4, y el estudio de imágenes. El tratamiento puede comenzar con el uso de esteroides, aunque en casos en que no tiene control de la enfermedad, se puede usar tratamiento con Rituximab.

Palabras claves: Neuromielitis Óptica; Sistema Nervioso Central; Rituximab.

INTRODUCTION

Described by Eugene Devic over a century ago, neuromyelitis optica spectrum disorder (NMOSD) is an autoimmune-based disease entity with inflammatory features affecting the central nervous system: optic nerves, spinal cord, and brainstem, triggering acute manifestations of optic neuritis, myelitis, and brainstem encephalitis; its immunological nature is associated with immunoglobulin G antibodies against aquaporin-4.^(1,2,3) Pathogenic IgG autoantibodies cause more than ≥ 80 % of NMOSD cases against aquaporin (AQP4-IgG). About 10-40 % of people with NMOSD who lack AQP4-IgG have IgG autoantibodies against myelin oligodendrocyte myelin glycoprotein (MOG-IgG).⁽³⁾

From an epidemiological point of view and taking into consideration the 2015 diagnostic criteria, the prevalence of the disorder in question ranges from 0,7 to 10/100,000 individuals in the populations studied globally, with a preponderance in Asians and Africans; there is a predominance in females with a ratio of 9:1 to male sex, appearing, on average, between 30 and 40 years of age, with the main risk factors being low body mass index, low fruit and vegetable intake, low physical activity, smoking and low vitamin D levels.⁽⁴⁾ In 2015, the International Panel for NMO Diagnosis (IPND) proposed a set of diagnostic methods and criteria covering the entire complex set of patients with or who are AQP4-IG positive, which are now summarised under the umbrella term NMOSD.1 These criteria describe the basic clinical features of NMOSD: Optic neuritis, acute transverse myelitis, area postrema syndrome, acute brainstem syndrome, symptomatic narcolepsy or acute clinical diencephalic syndrome, and symptomatic brain syndrome with brain lesions typical of NMOSD.⁽²⁾

MRI is used to identify and characterize lesions in patients with suspected NMOSD and helps to differentiate between NMOSD and MS. In NMOSD, the radiological findings are usually of greater magnitude compared to those caused by other demyelinating diseases such as Multiple Sclerosis; for example, in Optic Neuritis, there are long segments of inflammation seen on T2 or T1-weighted contrast-enhanced MRI covering more than half the distance from the orbit to the chiasm. In brain parenchyma, peri ependymal contrast-enhancing lesions are typical of AQP4-NMOSD and may occur in the lateral and third or fourth ventricles. MS lesions are usually ovoid or perpendicular (Dawson's Fingers). Subcortical white matter lesions in NMOSD are generally large, confluent, unilateral, or bilateral.⁽⁵⁾ Without proper treatment, patients with NMOSD may develop a significant disability over time due to the recurrence of seizures and insufficient recovery from severe seizures. With a better understanding of the underlying pathogenesis of NMOSD, new monoclonal antibody drugs have been developed targeting three specific components of the immune response, namely B cells, the complement pathway and interleukin-6 (IL-6), resulting in the development of ibalizumab, eculizumab and natalizumab respectively, which after several prospective randomized controlled trials have led to their approval by the FDA for AQP4-IgG-positive NMOSD patients: eculizumab in June 2019, ibalizumab in June 2020 and satralizumab in August 2020.⁽⁶⁾

This case series will provide an overview of neuromyelitis optica spectrum disorder management.

METHODS

A retrospective and descriptive analysis of patients attending the teaching hospital's outpatient clinic who have been diagnosed and followed up in the last 20 years was performed. Data were collected from the patient's clinical records. According to the Helsinki criteria, the privacy of the patients and their data was respected.

Each patient's age and year of the first diagnosed event, sex, presenting symptoms, various complementary diagnostic tests, and treatment were analysed. The tests included four aquaporin antibodies, oligoclonal bands, anti-MOG antibodies, magnetic resonance studies of the cervical, dorsal, and lumbar spine, and the brain and orbits.

RESULTS

All 8 (100,0 %) were female patients aged 19-45, with a mean age of 32. Four antibodies of anti-aquaporin were performed for diagnosis in all 8 (100,0 %) patients; 4 (50,0 %) patients had unaltered magnetic resonance imaging (MRI) of the brain and orbits. Eight (100,0 %) patients received immunosuppressive treatment with methylprednisolone, and three (37,5 %) of the patients also received treatment with rituximab.

Table 1. Clinical description, diagnosis and treatment of patients with neuromyelitis optica nerve (NMO)

No.	NMO syndrome	Anti-aquaporin 4 antibodies	Oligoclonal bands	Anti-MOG antibodies	MRI of the cervical, dorsal and lumbar spine	Brain and orbital MRI	Immunosuppressive treatment with methylprednisolone	Rituximab treatment
1	Bilateral optic neuritis Transverse Myelitis Sensory Level C5-C6. Depression	Positive	Not done	Not done	Hyperintense intramedullary lesion C1-C7	Unchanged	1 cycle	2 dose
2	Bilateral Optic Neuritis (OCT and evoked potentials)	Positive	Negatives	Negatives	Unaltered	Unchanged	3 ciclos	Not applied
3	Transverse myelitis with sensory level T7-T8	Positive	Not done	Not done	Intramedullary Inflammatory Injury C3 -T9	Unchanged	1 cycle	Not applied
4	Transverse myelitis with sensory level T12	Positive	Negatives	Not done	Transverse Myelitis T8-L2	Unchanged	3 cycles	Not applied
5	Right optic neuritis Transverse myelitis at sensory level c6-7 Sind of postrema area Vertiginous syndromes orbitofrontal syndrom Depression and anxiety Daytime sleepiness	Positive	Negatives	Not done	Unaltered	Left frontal subcortical hyperintense lesion	12 cycles	2 doses received
6	Right optic neuritis	Pending result	Negatives	Not done	Not done	Cerebral: hyperintense lesions in hippocampal and left thalamic region. Orbits: myelinating lesion in right optic nerve with acute features.	1 cycle	Not applied
7	Right optic neuritis Transverse myelitis at the L1-L2 level	Positive	Not done	Not done	Demyelinating lesion at the bulbar medullary junction. Chronic lesions with loss of chordal volume.	Cerebral: myelinating lesions at peri-callosal and medulla oblongata level.	3 cycles	Not applied
8	Optic neuritis Transverse myelitis at sensory level T8-T10 Depression and anxiety	Positive	Not done	Not done	Cervical: No alterations Dorsal and Lumbar not performed	Cerebral: Demyelinating lesion in left optic nerve	2 cycles	1 dose

5 (62,5 %) patients were diagnosed in 2024, 5 (62,5 %) patients confirmed in 2024. The time of evolution of the patients was between 5 months and 12 years. 3 (37,5 %) patients started Rituximab treatment in 2024.

Table 2. Year of diagnosis, confirmation, time of evolution and treatment with Rituximab of the patients.

Patient	Year of clinical diagnosis	Year of confirmation of diagnosis by Anti-Aquaporin 4	Time of disease evolution	Start of Rituximab	Rituximab dose received
1	2024	2024	1 year	March 2024	2 dose
2	2021	2021	4 years		
3	2024	2024	4 months		
4	2024	2024	1 years		
5	2017	2023	6 years	May 2024	2 dosis
6	2024	Pending result	5 months		
7	2012	2024	12 years	October 2024	1 dose
8	2024	2024	Died in November 2024		

DISCUSSION

Neuromyelitis optica is a nosological entity whose pathophysiological substrate is antibody-mediated inflammation of the central nervous system, the understanding of which has been developed over the last century. The estimated prevalence is 0,34 to 10 patients per 100,000 people in the adult population; in the present study, the entire sample belongs to the female sex, consistent with estimates for white and black populations in which it was 2,3 to 7. In the present investigation, the patients in this study present an age range of 19 to 45 years, which contrasts with studies in Indian and Spanish populations in which the ages with the highest prevalence correspond to the ranges of 30 to 39 years and 40 to 59 years, respectively.⁽⁷⁾

Neuromyelitis optica spectrum disorder, being linked to antibodies against aquaporin-4, is considered, as mentioned, an autoimmune disease. In this study, the patients studied had positive anti-aquaporin-4 antibodies, except for one of them, whose result was yet to be confirmed, with an age range between 19 and 45 years; these results are similar to those reported by Arnett et al., who found a mean age at diagnosis of 38,3 years with the aforementioned positive antibodies.⁽⁸⁾

Magnetic resonance imaging is key in evaluating neuromyelitis optica, as it provides information about a possible lesion of the optic pathway. In the present investigation, 50 % of the cases did not present lesions in the orbit and brain by this diagnostic method; in contrast, Wang et al., in their investigation, determined the presence of lesions in up to 87 % of patients studied, with the optic nerve being the most frequently affected.⁽⁹⁾

Finally, Rituximab, a chimeric monoclonal antibody, has been used in the treatment of neuromyelitis optica spectrum disorder and other immunosuppressive therapies, such as methylprednisolone and monoclonal antibodies. The drug targets antibody-producing B cells. In this study, 37,5 % received combined treatment with steroids and Rituximab, with the latter having a lower annual relapse rate and, therefore, a better prognosis.⁽¹⁰⁾

CONCLUSIONS

Neuromyelitis optica spectrum disorder is a disease that requires an accurate diagnosis based on clinical findings, detection of anti-quaporin-4 antibodies, and imaging studies. It remains a challenge in neurology today, and its management requires a comprehensive approach encompassing investigation, early diagnosis, and personalization of treatment.

Current evidence underlines the importance of early identification of serological markers and the use of targeted immunomodulatory therapies to improve patients' prognosis and quality of life. The ongoing evolution in our understanding of this disease invites further exploration of new therapeutic strategies that will lead to better disease control and less burden on sufferers.

CONSENT

Consent was obtained from the patient for this work to be carried out.

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CONFLICT OF INTEREST

The authors declare that there is no conflict of interest.

AUTHORSHIP CONTRIBUTION

Conceptualisation: Dulce Escalante, Alejandro Carías.

Formal analysis: Dulce Escalante, Gabriel Isaías Lagos.

Research: Alejandro Carías, Jhossmar Cristians Auza-Santivañez.

Methodology: Pablo Carías, Alejandro Carías, Jhossmar Cristians Auza-Santivañez.

Project management: Dulce Escalante.

Supervision: Freddy Ednildon Bautista-Vanegas.

Visualisation: Dulce Escalante, Gabriel Isaías Lagos.

Writing - original draft: Dulce Escalante, Gabriel Isaías Lagos, Harlis Estrada Pastrana, Pablo Carías, Alejandro Carías, Jhossmar Cristians Auza-Santivañez, Freddy Ednildon Bautista-Vanegas, Dilma Villca-Villca.

Writing - proofreading and editing: Dulce Escalante, Gabriel Isaías Lagos, Harlis Estrada Pastrana, Pablo Carías, Alejandro Carías, Jhossmar Cristians Auza-Santivañez, Freddy Ednildon Bautista-Vanegas, Dilma Villca-Villca.