the range of $5,000-$9,999 for serving as a Consultant for Alexion. The institution of Dr. Piquet has received personal compensation in the range of $0-$499 for serving as a Consultant for UCB. The institution of Dr. Piquet has received personal compensation in the range of $0-$499 for serving as a Consultant for EMD Serono. Dr. Piquet has received personal compensation in the range of $500-$4,999 for serving on a Scientific Advisory or Data Safety Monitoring board for Genentech. Dr. Piquet has received personal compensation in the range of $500-$4,999 for serving on a Scientific Advisory or Data Safety Monitoring board for Alexion. The institution of Dr. Piquet has received personal compensation in the range of $500-$4,999 for serving on a Scientific Advisory or Data Safety Monitoring board for Genentech. Dr. Piquet has received personal compensation in the range of $10,000-$49,999 for serving as an Expert Witness for Sands Anderson PC. Dr. Piquet has received personal compensation in the range of $5,000-$9,999 for serving as an Expert Witness for Joe Jones Law Firm. The institution of Dr. Piquet has received research support from Rocky Mountain MS Center. The institution of Dr. Piquet has received research support from Novartis. The institution of Dr. Piquet has received research support from Abbvie. The institution of Dr. Piquet has received research support from Roche/Genentech. The institution of Dr. Piquet has received research support from NYU. The institution of Dr. Piquet has received research support from Anokion. The institution of Dr. Piquet has received research support from UCB. The institution of Dr. Piquet has received research support from Foundation for Sarcoidosis. Dr. Piquet has received publishing royalties from a publication relating to health care. Dr. Piquet has received publishing royalties from a publication relating to health care. Dr. Piquet has received personal compensation in the range of $10,000-$49,999 for serving as a Litigative Consultant with US-Dept HHS/DICP. Dr. Piquet has a non-compensated relationship as a Medical Advisory Board Member with Autoimmune Encephalitis Alliance (AEA) that is relevant to AAN interests or activities. Dr. Piquet has a non-compensated relationship as a Medical Advisory Board Member with Stiff Person Syndrome Research Foundation (SPSRF) that is relevant to AAN interests or activities.

Early Relapse in Anti-GAD65 and Anti-NMDA Co-expression Associated Autoimmune Encephalitis: A Case Report

Irene Gomez Oropeza, Irving Fuentes, Salvador Martinez Medina

Objective

N/A.

Background

Autoimmune encephalitis prevalence has risen notably in recent years, driven by advancements in neuronal antibody research. AE is the most common form of encephalitis of non-infectious origin, and it has a wide spectrum of clinical phenotypes. However, it remains a rare autoimmune disease frequently underdiagnosed. This report elucidates the diagnostic complexities, therapeutic challenges, and the possibility of relapse regarding the rare phenomenon of co-expression of Anti-GAD65 and Anti-NMDA autoimmune encephalitis.

Design/Methods

N/A.

Results

We present a case report of an 18-year-old female who presented to the hospital with a biparietal subacute headache that progressed with paresthesias and behavioral changes like mutism, inattention, hypersomnia, and myoclonus. Her physical examination revealed mixed dysarthric speech (flaccid and spelled), dyscalculia, and altered abstraction. She underwent an electroencephalogram that showed the presence of mild generalized dysfunction, without epileptiform activity. Imaging studies and a lumbar puncture did not support the diagnosis of infectious pathology. The Bush Francis score was rated 1 and a MOCA test 17, revealing cognitive impairment. Anti-NMDA and Anti-GAD65 antibodies were reported in the cerebrospinal fluid autoantibodies panel. She started treatment with methylprednisolone plus sessions of plasmapheresis, which finally showed improvement and she was discharged, but within less than two months she had a relapse of neuropsychiatric symptoms and was admitted again. Thus rituximab treatment was initiated as a second-line option that resulted in good clinical evolution and no signs of relapse in the following 2 months.

Conclusions

This case underlines the complexities of management in AE, particularly when confronted with co-expression antibody positivity. Clinicians should maintain a high index of suspicion, particularly in underrepresented populations, and consider the need for long-term assessment alongside optimal immunotherapy, keeping in mind its therapeutic nuances. Future research directions are needed, to enhance our comprehension of AE pathogenesis, optimize treatment strategies, and address disparities in diagnosis across diverse demographics.

Study Supported By: N/A.

Disclosure: Ms. Gomez Oropeza has nothing to disclose. Mr. Fuentes has nothing to disclose. Dr. Medina has nothing to disclose.

Patient Preferences for Generalized Myasthenia Gravis Treatment Profiles: Results of a Web-based Survey

Karen Yee, Christine Poulos, Cooper Bussberg, Kelley Myers

Objective

To better understand treatment preferences of patients with anti-acetylcholine receptor antibody-positive (AChR-Ab+) generalized myasthenia gravis (gMG) and to estimate the relative importance of preferred treatment attributes.

Background

gMG is a rare, chronic autoimmune disorder. Key therapeutic approaches differ among the available treatments for AChR-Ab+ gMG, but no studies on patient treatment preferences have been published to date.

Design/Methods

US adults with a self-reported physician diagnosis of AChR-Ab+ gMG completed a web-based survey. Two object-case, best-worst scaling (BWS) exercises were analyzed. The first BWS exercise obtained preferences for a treatment profile similar to ravulizumab compared with 4 other treatment profiles (eg, similar to eculizumab, efgartigimod intravenous and subcutaneous, and zilucoplan). The second BWS exercise obtained preferences for the individual attributes used to define the treatment profiles. Profile scenarios were defined by mode of administration and dosing frequency only, followed by the addition of consistent disease control and meningococcal vaccination requirements. Additionally, the most important gMG treatment attribute was identified.

Results

Of 153 respondents, mean age was 49 years, 77% female, 84% white, 54% with college degree or higher, 41% employed, and 27% had been diagnosed for <3 years. Mean MG-Activities of Daily Living score was 8.0 (min–max: 0–17). Respondents preferred the ravulizumab-like profile vs all 3 other profile-based scenarios: 35% vs 10%–22% when considering mode and dosing frequency only, 44% vs 3%–31% when considering addition of consistent disease control, and 39% vs 5%–29% when considering all 4 attributes. Consistent disease control was selected as the most important attribute when choosing a gMG treatment (82%), followed by mode of administration (10%), dosing frequency (6%), and meningococcal vaccination requirements (3%).

Conclusions

Patients with gMG preferred treatments with less frequent dosing schedules and consistent disease control, and consistent disease control was considered the most important attribute when choosing a therapy.

Study Supported By: Alexion, AstraZenica Rare Disease.

Disclosure: Dr. Yee has received personal compensation for serving as an employee of Alexion. Dr. Yee has stock in Alexion. Dr. Yee has stock in Takeda. Ms.

Poulos has nothing to disclose. Cooper Bussberg has received personal compensation for serving as an employee of RTI Health Solutions, a not for profit research institute which conducts research for the pharmaceutical industry. His salary is unconnected to the projects on which he works and the details of the financial arrangements between RTI Health Solutions and its clients are confidential. Dr. Myers has nothing to disclose.

Impact of Clinical Progression on Brain Volume Changes in Patients with Anti-NMDAR Autoimmune Encephalitis

Enrique Gomez Figueroa, Veronica Rivas-Alonso, Mariana Espinola-Nadurille, Jesus Ramirez-Bermudez, Sandra Orozco-Suarez, Antonio Arauz-Gongora, Jose Flores-Rivera

Objective

To assess the impact of clinical symptoms and complications on volumetric brain parameters in individuals with anti-NMDA receptor-associated autoimmune encephalitis (NMDARE).

Background

NMDARE is a disorder characterized by psychiatric symptoms, altered consciousness, dysautonomia, hypoventilation, and seizures, with pathophysiology involving NMDA receptor dysfunction. Brain atrophy, although reported inconsistently, remains poorly understood regarding prognostic implications. This study aims to evaluate cerebral atrophy in Mexican patients with autoimmune encephalitis and its correlation with clinical phenotypes.

Design/Methods

We conducted a longitudinal retrospective study including patients admitted to National Institute of Neurology and Neurosurgery with autoimmune encephalitis and positive cerebrospinal fluid tests for anti-NMDA receptor antibodies. Patients with initial and follow-up magnetic resonance imaging were analyzed. Automated segmentation and comparative analysis were performed using Volbrain and SIENA, respectively. Parametric and non-parametric statistics, including linear models, were employed to assess volume changes.

Results

Sixty-five patients were included (mean age 30.52 years, 49.2% male). Thalamic reduced volume was associated with psychosis (ß= -26.39±12.87, p=0.047), limbic cortex changes with status epilepticus (ß=137±64.89, p=0.04), and hippocampal loss with dysautonomia (ß=-16.66±8.42, p=0.05). Reduced volume was observed in cerebral white matter, occipital lobe, and limbic cortex with EEG abnormalities, while thalamic enlargement correlated with neoplasm presence (all p<0.05). Rituximab treatment was linked to brainstem volume gain, and cyclophosphamide with global white and gray matter changes.

Conclusions

Our findings indicate that cerebral volume changes in NMDARE are heterogeneous and influenced by clinical presentation and treatment. Further longitudinal studies are warranted to ascertain the reversibility of these changes and their impact on long-term outcomes.

Study Supported By: N/A.

Disclosure: Dr. Gomez Figueroa has received personal compensation in the range of $500-$4,999 for serving as a Consultant for Biogen Mexico. Dr. Gomez Figueroa has received personal compensation in the range of $500-$4,999 for serving on a Scientific Advisory or Data Safety Monitoring board for Astra Zeneca Mexico. Dr. Gomez Figueroa has received personal compensation in the range of $500-$4,999 for serving on a Scientific Advisory or Data Safety Monitoring board for Johnson & Johnson LATAM. Veronica Rivas has nothing to disclose. Mariana Espinola-Nadurille has nothing to disclose. Jesus Ramirez-Bermudez has nothing to disclose. Dr. Orozco-Suarez has nothing to disclose. Dr. Arauz has nothing to disclose. Dr. Flores-Rivera has nothing to disclose.

Risk Factors Influencing Relapse Rate in Double Seronegative Neuromyelitis Optica Spectrum Disorder (DSNMOSD): A Multicenter Study

Gerome Vallejos, Takahisa Mikami, Joao Vitor Mahler Ferreira Oliveira, Guilherme Silva, Samira Apostolos-Pereira, Dagoberto Callegaro, Michael Levy

Objective

To identify the risk factors associated with the relapse rate in patients with DSNMOSD.

Background

Double Seronegative Neuromyelitis Optica Spectrum Disorder (DSNMOSD) is a rare subset of NMOSD that is characterized by a negative test against Aquaporin 4 (AQP4) IgG and Myelin Oligodendrocyte Glycoprotein (MOG) IgG.

Design/Methods

This is a multicenter retrospective study of cases of DSNMOSD at the MassGeneralBrigham and the Hospital University of Sao Paulo. Patients were included if they (1) met the diagnostic criteria for Seronegative NMOSD according to the 2015 International Panel for NMO Diagnosis (IPND) and (2) tested negative for AQP4-IgG and MOG-IgG at least once. Patients were excluded if they had an alternative diagnosis. To examine the association of the different clinical and paraclinical factors on relapses, we calculated the incidence rate ratio (IRR) using a Poisson regression analysis.

Results

A total of 37 relapsing DSNMOSD patients were analyzed. In a univariate Poisson regression analysis, being female and of non-Caucasian race was predicted to have a higher rate of relapse (IRR 1.63, 95%CI 1.1-2.6; p= 0.034) and (IRR: 1.60, 95% CI 1.10-2.35; p=0.017), respectively. Simultaneous optic neuritis (ON) and transverse myelitis (TM) at onset resulted in a lower IRR of 0.04 (95%CI 0.01-0.13; p <0.001). After adjusting for sex, race, laterality of ON, and use of disease-modifying therapy, age at onset at ≥40 years old was associated with higher rate of relapses (IRR 2.10, 95% CI 1.23-3.51; p = 0.006), while initial clinical manifestations of TM only (IRR 0.47, 95%CI 0.22-0.97; p=0.043) and simultaneous ON and TM (IRR 0.05, 95% CI 0.10-0.21; p <0.001) were linked to low relapse rates when compared to ON only.

Conclusions

Later age onset (≥40 years old) and patients presenting with TM only and simultaneous ON and TM at onset significantly influence the relapse rate observed in DSNMOSD.

Study Supported By: N/A.

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