

Epidemiology and Burden of NMOSD, MS, and MOGAD in Thailand: a Population-Based Study

Nanthaya Tisavipat, Pornpong Jitpratoom, Sasitorn Siritho, Naraporn Prayoonwiwat, Metha Apiwattanakul, Natthapon Rattanathamsakul, Jiraporn Jitprapaikulsan

Objective

To determine cumulative incidence and point prevalence of neuromyelitis optica spectrum disorder (NMOSD), multiple sclerosis (MS), and myelin oligodendrocyte glycoprotein antibody-associated disease (MOGAD) in Thailand using population-based data of Chumphon province.

Background

CNS inflammatory demyelinating diseases (CNSIDDs) have a great interracial heterogeneity. The epidemiology of CNSIDDs in Thailand, a Mainland Southeast Asian country, is unknown.

Design/Methods

Searching for CNSIDD patients at a public secondary care hospital in Chumphon from January 2016 to December 2021 was performed using relevant ICD-10-CM codes. All neurology patients were systematically referred to this hospital as it was the only hospital in the province with a neurologist. Diagnoses were individually ascertained by retrospective chart review. Cumulative incidence over 2016-2021, point prevalence on December 31st, 2021, attack rate, mortality rate, and disability-adjusted life years (DALYs) were calculated. Population data were obtained from the National Statistical Office of Thailand. As of December 31st, 2021, the population census of Chumphon was 509,479.

Results

NMOSD was the most prevalent CNSIDD in adult Thai population at 3.33 per 100,000 persons (crude prevalence 2.55). The age-adjusted prevalence of aquaporin-4 antibody-positive NMOSD alone was 3.08 per 100,000 persons. Age-adjusted incidence rate of NMOSD was 1.65 per 100,000 persons/year (crude incidence rate 0.20). Age-adjusted prevalence of MS followed at 0.77 and MOGAD at 0.51 per 100,000 persons (crude prevalence 0.59 and 0.39, respectively). Although most had a fair recovery, disability was worst among NMOSD with a DALY of 3.47 years per 100,000 persons. Mortality and attack rates were highest in NMOSD as well. No increase in incidence or attack rate were observed during the COVID-19 pandemic.

Conclusions

CNSIDDs are rare diseases in Thailand. The prevalence is comparable to that of East Asian countries. NMOSD caused the highest DALYs among CNSIDDs.

Disclosure: Miss Tisavipat has nothing to disclose. Dr. Jitpratoom has nothing to disclose. Dr. Siritho has received personal compensation in the range of \$0-\$499 for serving on a Speakers Bureau for Merck. Dr. Siritho has received personal compensation in the range of \$0-\$499 for serving on a Speakers Bureau for Teva. Dr. Siritho has received personal compensation in the range of \$0-\$499 for serving on a Speakers Bureau for Novartis. Dr. Siritho has received personal compensation in the range of \$0-\$499 for serving on a Speakers Bureau for Eisai. Dr. Prayoonwiwat has nothing to disclose. Dr. Apiwattanakul has received personal compensation in the range of \$0-\$499 for serving on a Speakers Bureau for Merck. Dr. Apiwattanakul has received personal compensation in the range of \$0-\$499 for serving on a Speakers Bureau for Teva. Dr. Apiwattanakul has received personal compensation in the range of \$0-\$499 for serving on a Speakers Bureau for Novartis. Dr. Rattanathamsakul has nothing to disclose. Dr. Jitprapaikulsan has nothing to disclose.

Acute Central Nervous System Demyelination Following COVID-19 Vaccination

Sydney Lee, Alexandra Muccilli, Raphael Schneider, Daniel Selchen, Kristen Krysko

Objective

To describe features of central nervous system (CNS) demyelinating events following vaccination against coronavirus disease 19 (COVID-19).

Background

Several reports suggest a potential association between COVID-19 vaccines and acute CNS inflammation.

Design/Methods

A case series was performed at the BARLO MS Centre in Toronto, Ontario, Canada. Clinicians reported patients who experienced an acute CNS demyelinating event within 60 days after receiving at least one COVID-19 vaccination from March 2021 to January 2022. Clinical characteristics were evaluated.

Results

Twenty patients were identified (median age 39 years (range 25-82); 13 (65.0%) female). Two had pre-existing multiple sclerosis (MS). Individuals received the Pfizer (n = 14), Moderna (n = 5) or Astrazeneca (n = 1) COVID-19 vaccines. Within 1-53 days (median 12) of the first (n = 8) or second (n = 12) vaccine dose, patients developed transverse myelitis (TM) (n = 15), optic neuritis (n = 4) or brain demyelination (n = 4). Diagnoses at last follow up (median 114 days (range 39-255)) were relapsing remitting MS (n = 8), post-vaccine TM (n = 5), clinically isolated syndrome (n = 3), myelin oligodendrocyte glycoprotein antibody disease (n = 2), MS relapse (n = 1) and neuromyelitis optica spectrum disorder (n = 1). Thirteen patients received pulse corticosteroids, and of these, 4 received plasma exchange. Seven did not receive acute treatment. 20.0% returned to baseline (n = 4), 75.0% partially recovered (n = 15) and 5.0% worsened (n = 1). At last follow up, 11 were on disease modifying therapy and 9 were not. Nine patients received a subsequent COVID-19 vaccine. Of these, one experienced symptom recrudescence without radiologic evidence of a new demyelinating attack.

Conclusions

To our knowledge, this is the largest series to date describing acute CNS demyelination after vaccination against COVID-19. The rate of vaccination in the eligible general population was high during the time of the cases and we could not determine whether the number of demyelinating events was higher than expected. Repeat vaccination was not associated with recurrent adverse events in this small observational series.

Disclosure: Dr. Lee has nothing to disclose. Dr. Muccilli has received personal compensation in the range of \$500-\$4,999 for serving on a Scientific Advisory or Data Safety Monitoring board for Biogen. Dr. Muccilli has received personal compensation in the range of \$500-\$4,999 for serving on a Scientific Advisory or Data Safety Monitoring board for Novartis. Dr. Muccilli has received personal compensation in the range of \$500-\$4,999 for serving on a Scientific Advisory or Data Safety Monitoring board for EMD Serono. Dr. Schneider has received personal compensation in the range of \$500-\$4,999 for serving as a Consultant for Bristol-Myers Squibb. Dr. Schneider has received personal compensation in the range of \$500-\$4,999 for serving as a Consultant for Novartis. Dr. Selchen has received personal compensation in the range of \$10,000-\$49,999 for serving on a Speakers Bureau for EMD Serono. Dr. Selchen has received personal compensation in the range of \$5,000-\$9,999 for serving on a Speakers Bureau for Roche. Dr. Selchen has received personal compensation in the range of \$5,000-\$9,999 for serving on a Speakers Bureau for Novartis. Dr. Selchen has received personal compensation in the range of \$10,000-\$49,999 for serving as an Expert Witness for Teva. Dr. Krysko has received personal compensation in the range of \$500-\$4,999 for serving as a Consultant for Normative Inc. Dr. Krysko has received personal compensation in the range of \$5,000-\$9,999 for serving as a Consultant for Biogen. Dr. Krysko has received personal compensation in the range of \$500-\$4,999 for serving as a Consultant for Roche. Dr. Krysko has received personal compensation in the range of \$500-\$4,999 for serving as a Consultant for EMD Serono. Dr. Krysko has received personal compensation in the range of \$500-\$4,999 for serving as a Consultant for Novartis. Dr. Krysko has received personal

compensation in the range of \$500-\$4,999 for serving on a Scientific Advisory or Data Safety Monitoring board for Novartis. The institution of Dr. Krysko has received research support from Biogen Idec. The institution of Dr. Krysko has received research support from National MS Society.

Spinal Cord Neurosarcoidosis: A Clinical-Radiological Correlation of 39 Cases

Rami Al-Hader, Lonni Schultz, Justin Nofar, Vivek Rai, Mirela Cerghet

Objective

Present radiological and clinical data of spinal cord neurosarcoidosis and response to treatment.

Background

The diagnosis of neurosarcoidosis is challenging. Stern et al. have used histopathological data, clinical scenarios, and response to treatment to propose diagnosis criteria for definite, probable, or possible neurosarcoidosis. There is no definitive confirmatory test except sample biopsy, which is not a preferred test for the central nervous system due to potential complications. MRI studies can help detect nervous system involvement; however, it is neither sensitive nor specific.

Design/Methods

Retrospective analysis with descriptive statistics.

Results

Our cohort consisted of 39 patients with spinal neurosarcoidosis. On MRI, 62% of the patients had a longitudinally extensive intramedullary lesion, 21% had one or multiple patchy intramedullary lesions, 31% had leptomeningeal involvement, and 18% had nerve roots enhancement. The cervical spine was most commonly affected (85%), followed by the thoracic (38%) and lumbar (15%). Thirty-seven patients were treated with oral or IV corticosteroids at first presentation, followed by maintenance with oral steroids and maintenance immunosuppressive agents. The three most used agents were Methotrexate (49%), Azathioprine (31%), and Mycophenolate mofetil (18%). Thirty-four patients had MRIs during follow-up, and twenty-nine patients had documented improvement during follow-up, with a median improvement time on MRI of 10.8 months (95% CI = 6.1 to 17 months). Thirty-one patients had enhancement on MRI at presentation, and 18 (58%) had complete enhancement resolution during follow-up, with a median time for resolution of enhancement of 51.8 months (95% CI = 24.9 to 83.4 months).

Conclusions

The diagnosis of spinal neurosarcoidosis can be challenging; however, we found that resolution of MRI enhancement can require a few years of immunosuppression, which is longer compared to other spinal neuroimmunological pathologies. The current knowledge about the treatment and prognosis of neurosarcoidosis is limited, and there is no FDA medication approved nor clinical trials data regarding the treatment of neurosarcoidosis.

Disclosure: Dr. Al-Hader has nothing to disclose. Dr. Schultz has nothing to disclose. Dr. Nofar has nothing to disclose. Dr. Rai has nothing to disclose. Dr. Cerghet has nothing to disclose.

Serum Autoantibody Lowering by the Anti-FcRn Monoclonal Antibody, Nipocalimab, Correlates With Clinical Improvement in Generalized Myasthenia Gravis Patients

Sindhu Ramchandren, Jeff Guptill, Carlo Antozzi, Vera Bril, Josep Gamez, Sven Meuth, Richard Nowak, Dianna Quan, Maria Teresa Sevilla Mantecon, Leona Ling, Yaowei Zhu, Keith Karcher, Hong Sun

Objective

To evaluate the relationship between clinical improvement in Myasthenia Gravis-Activities of Daily Living (MG-ADL) scores and the pharmacodynamic effects of IgG autoantibody lowering induced by nipocalimab in the Vivacity MG Phase 2 study.

Background

Nipocalimab is a fully human, aglycosylated, effectorless IgG1 anti-FcRn monoclonal antibody that targets the neonatal Fc receptor (FcRn) with high affinity, thereby lowering IgG pathogenic antibodies in autoimmune disease.

Design/Methods

The relationship between the reduction in acetylcholine-receptor (AChR)- and Muscle-Specific-Tyrosine-Kinase (MuSK)- autoantibodies with improvement in MG-ADL scores were explored across the four nipocalimab dose arms in the Vivacity MG Phase 2 Study in generalized myasthenia gravis (gMG) patients.

Results

Of the 68 patients enrolled, 54 were randomized to one of the four nipocalimab dosing arms. 51 (94%) were seropositive for anti-AChR, 3 (6%) for anti-MuSK. Nipocalimab was well-tolerated and achieved substantial, dose-dependent and rapid reductions in serum total IgG, including all IgG subtypes and anti-AChR autoantibodies. These reductions were associated with dose-dependent, durable and rapid MG-ADL responses in all nipocalimab-treated groups. A similar trend in IgG4 reduction was noted, though the sample size of MuSK positive patients was small.

Conclusions

The results support the rapid, dose-dependent and predictable effect of nipocalimab in lowering pathogenic autoantibodies and inducing clinical improvement in patients with gMG. In addition, the close correlation between serum IgG, anti-AChR and clinical response suggest the potential of using serial serum IgG levels as a biomarker in management of gMG patients treated with nipocalimab; this will be tested in the ongoing Phase 3 gMG trial.

Disclosure: Dr. Ramchandren has received personal compensation for serving as an employee of Janssen Pharmaceutical Companies of Johnson & Johnson. Dr. Ramchandren has a non-compensated relationship as a Scientific Advisory Board Member with CMT Research Foundation (CMTRF) that is relevant to AAN interests or activities. Dr. Guptill has received personal compensation in the range of \$500-\$4,999 for serving as a Consultant for Grifols. Dr. Guptill has received personal compensation in the range of \$5,000-\$9,999 for serving as a Consultant for Jacobus. Dr. Guptill has received personal compensation in the range of \$500-\$4,999 for serving as a Consultant for Cabaletta. Dr. Guptill has received personal compensation in the range of \$500-\$4,999 for serving as a Consultant for Regeneron. Dr. Guptill has received personal compensation in the range of \$500-\$4,999 for serving as a Consultant for BD. Dr. Guptill has received personal compensation in the range of \$5,000-\$9,999 for serving as a Consultant for Piedmont Pharmaceuticals. Dr. Guptill has received personal compensation in the range of \$500-\$4,999 for serving as a Consultant for Immunovant. Dr. Guptill has received personal compensation in the range of \$500-\$4,999 for serving as a Consultant for Argenx. Dr. Guptill has received personal compensation in the range of \$500-\$4,999 for serving as a Consultant for Momenta. Dr. Guptill has received personal compensation in the range of \$5,000-\$9,999 for serving as a Consultant for Alexion. Dr. Guptill has received personal compensation in the range of \$500-\$4,999 for serving as a Consultant for Signant. Dr. Guptill has received personal compensation in the range of \$5,000-\$9,999 for serving on a Scientific Advisory or Data Safety Monitoring board for Immunovant. Dr. Guptill has received personal compensation in the range of \$5,000-\$9,999 for serving on a Scientific Advisory or Data Safety Monitoring board for Alexion. Dr. Guptill has received personal compensation in the range of \$500-\$4,999 for serving on a Scientific Advisory or Data Safety Monitoring board for Argenx. Dr. Guptill has received personal compensation in the range of \$500-\$4,999 for serving on a Scientific Advisory or Data Safety Monitoring board for Momenta. Dr. Guptill has received personal

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Neurology 2022;99;S34-S35

DOI 10.1212/01.wnl.0000903296.89024.e9

This information is current as of December 5, 2022

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