

Floris has nothing to disclose. Dr. Zara has nothing to disclose. Dr. Bhatti has received personal compensation in the range of \$500-\$4,999 for serving as a Consultant for Receptos. Dr. Bhatti has received personal compensation in the range of \$0-\$499 for serving on a Scientific Advisory or Data Safety Monitoring board for NIH LHON gene therapy study. Dr. Sanchez Moreno has nothing to disclose. Matthew Koster has nothing to disclose. Dr. Mariotto has received personal compensation in the range of \$500-\$4,999 for serving on a Speakers Bureau for Biogen. Dr. Pittock has received personal compensation in the range of \$5,000-\$9,999 for serving as a Consultant for Genentech, Inc. Dr. Pittock has received personal compensation in the range of \$500-\$4,999 for serving as a Consultant for Sage Therapeutics, Inc. The institution of Dr. Pittock has received personal compensation in the range of \$500-\$4,999 for serving as a Consultant for Astellas. Dr. Pittock has received personal compensation in the range of \$500-\$4,999 for serving as a Consultant for Prime Therapeutics. Dr. Pittock has received personal compensation in the range of \$500-\$4,999 for serving as a Consultant for UCB. Dr. Pittock has received personal compensation in the range of \$500-\$4,999 for serving as a Consultant for Roche/Genentech. The institution of Dr. Pittock has received personal compensation in the range of \$500-\$4,999 for serving as a Consultant for Alexion. The institution of Dr. Pittock has received personal compensation in the range of \$500-\$4,999 for serving as a Consultant for MedImmune/Viela Bio. Dr. Pittock has received personal compensation in the range of \$500-\$4,999 for serving on a Scientific Advisory or Data Safety Monitoring board for UCB, Inc. Dr. Pittock has received personal compensation in the range of \$5,000-\$9,999 for serving on a Scientific Advisory or Data Safety Monitoring board for Hoffman/LaRoche AG. Dr. Pittock has received personal compensation in the range of \$500-\$4,999 for serving on a Scientific Advisory or Data Safety Monitoring board for Genetech. Dr. Pittock has received personal compensation in the range of \$500-\$4,999 for serving on a Scientific Advisory or Data Safety Monitoring board for F. Hofman/LaRoche. The institution of Dr. Pittock 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. The institution of Dr. Pittock has received research support from Grifols. The institution of Dr. Pittock has received research support from NIH. The institution of Dr. Pittock has received research support from Viela Bio/MedImmune/Horizon. The institution of Dr. Pittock has received research support from Alexion Pharmaceuticals. The institution of Dr. Pittock has received research support from F. Hoffman/LaRoche/Genentech. Dr. Pittock has received intellectual property interests from a discovery or technology relating to health care. Dr. Pittock has received intellectual property interests from a discovery or technology relating to health care. John Chen has nothing to disclose. Alberto Cauli has nothing to disclose. Dr. Solla has nothing to disclose. Dr. Sechi has nothing to disclose.

Neuroinvasive West Nile Virus Disease Presenting as Opsoclonus-Myoclonus-Ataxia Syndrome

Aditi Sharma, Karina Gonzalez Otarula, Lama Abdel Wahed, Adriana Rodriguez, Christine Gill

Objective

Describe a case of probable Neuroinvasive West Nile Virus (WNV) disease presenting as opsoclonus-myoclonus-ataxia syndrome (OMS).

Background

Opsoclonus-myoclonus-ataxia syndrome is a well described condition which is thought to be often of paraneoplastic or autoimmune etiology in adults. Specific pathogenic antibodies have yet to be identified in most cases. It can also be seen in association with CNS infections, although it is unclear if the pathophysiology aligns more with an infectious or a parainfectious process. Here we describe a clinical case where a patient presented with OMS, with CSF findings indicative of a diagnosis of Neuroinvasive West Nile Virus disease.

Design/Methods

Case Report.

Results

A 65-year-old previously healthy man presented with 3 weeks of progressive generalized tremors, oscillopsia and inability to ambulate. Exam revealed opsoclonus, stimulus-induced myoclonus, and generalized ataxia. CSF showed neutrophil-predominant pleocytosis (325 WBCs with 78 neutrophils), which converted 3 days later to lymphocytic

predominance (44 WBCs with 36 lymphocytes). CSF cultures, meningitis/encephalitis multiplex PCR array, brain MRI with and without contrast, body PET-CT and serum autoimmune encephalopathy panel were unrevealing. CSF WNV IgG and IgM were elevated to 2 times and 5 times the upper limit of assay respectively, concerning for Neuroinvasive WNV disease. He received 5 days of intravenous methylprednisolone and immunoglobulins with clinical improvement, and had ultimate resolution of symptoms over the next 6 months.

Conclusions

WNV has been associated with a wide spectrum of movement disorders, and should be considered in the differential diagnosis, especially with CSF pattern as described above. Idiopathic and paraneoplastic opsoclonus-myoclonus-ataxia syndrome remains the most common subtype, however it is important to perform an evaluation for infectious etiologies as well to guide further management and counseling regarding outcome.

Disclosure: Dr. Sharma has nothing to disclose. Dr. Gonzalez Otarula has nothing to disclose. Dr. Abdel Wahed has nothing to disclose. Dr. Rodriguez has nothing to disclose. Dr. Gill has nothing to disclose.

Etiology and Factors Related Outcomes of Longitudinally Extensive Transverse Myelitis in Thailand

Nisa Vorasoot, Pilantana Saichua, Prapassara Sirikarn, Arunnit Boonrod, Narongrit Kasemsap, Kannikar Kongbunkiat, Somsak Tiamkao

Objective

This study aimed to evaluate the clinical features, etiology, and prognosis of longitudinally extensive transverse myelitis (LETM) patients in Thailand.

Background

LETM has various etiology and is different in each region. Proper investigations are essential to reduce misdiagnosis and delay in treatments, which affect clinical recovery and prognosis. In Thailand, there is no clinical study on the etiology of LETM. Therefore, our study aimed to evaluate the clinical features, etiology, and prognosis of LETM patients in Thailand.

Design/Methods

Patients diagnosed with LETM at University Hospital between January 2015 and October 2021 were included. Patient demographics, clinical presentations, Expanded Disability Status Scale (EDSS), imaging, laboratory testing, cerebrospinal fluid profiles, final diagnosis, and treatments were recorded. Factors related to outcomes of LETM were analyzed.

Results

A total of 40 patients, there were 21 females (52.5%), the mean age of onset was 48.4 years (SD = 15.8). NMOSD was the most common etiology of LETM (n = 15), followed by infection (n = 5), SLE (n = 5), idiopathic causes (n = 4), CIS (n = 3), MS (n = 1), spinal dural AVF (n = 2), ADEM (n = 2), either 1 had spinal cord infarction, schwannoma, and vitamin B12 deficiency. Most patients in this study had severe LETM (n = 31). Complete cord had significantly poorer outcome (p-value = 0.003), while dorsolateral and anterior cord had better outcome (p-value = 0.046, 0.046).

Conclusions

NMOSD was the most common etiology of LETM, and a history of prior attacks led to the diagnosis of NMOSD. Complete cord lesion on axial spinal cord MRI was sensitive to NMOSD but not specifically. Factors related to the prognosis of LETM included completed cord lesions on MRI axial view trended to have a poor

outcome, and dorsolateral and anterior cord lesions had a better prognosis.

Disclosure: Dr. Vorasoot has nothing to disclose. Miss Saichua has nothing to disclose. Miss Sirikarn has nothing to disclose. Mrs. Boonrod has nothing to disclose. Narongrit Kasemsap has nothing to disclose. Kannikar Kongbunkiat has nothing to disclose. Somsak Tiamkao has nothing to disclose.

Characterization of Neurosarcoid Myelitis and Assessment of Treatment Response: A Multicenter Retrospective Study

Denis Balaban, Giovanna Manzano, Ahya Ali, Eoin Flanagan, Allen Aksamit, Vyanka Redenbaugh, Stacey Clardy, Rohini Samudralwar, Paunel Agyei, Aram Zabeti, Robert Baughman, Bart Chwalisz, Michael Levy, Shamik Bhattacharyya

Objective

To determine if initial treatment with corticosteroids plus steroid-sparing immunosuppressive therapy (IST) results in faster gadolinium contrast-enhancement resolution in neurosarcoid myelitis (NSM) than corticosteroid monotherapy (CSM).

Background

Neurosarcoidosis is a rare cause of myelitis, treated initially with corticosteroids. Whether combination therapy of corticosteroids plus IST is more effective than CSM at disease onset is unknown.

Design/Methods

We retrospectively reviewed cases of definite or probable NSM, defined by Neurosarcoidosis Consortium Consensus Group, in adults treated at six United States centers. We characterized clinicoradiographic features and treatment outcomes after NSM diagnosis. Treatment groups were defined as CSM, corticosteroids plus intermediate oral IST (methotrexate or mycophenolate mofetil), corticosteroids plus highly effective IST (cyclophosphamide/TNF- α inhibitors), or corticosteroids plus other. We hypothesized that initial treatment with corticosteroids and IST would result in faster gadolinium contrast-enhancement resolution (primary end point).

Results

63 patients with NSM (32 female, 30 definite, median age 48) were identified. 86% had spinal cord enhancement on post-gadolinium T1 sequences (8% without enhancement, 6% without data). Time from symptom onset to treatment initiation varied from 11 days to 10 years (median 4 months). All but one patient received corticosteroids initially. 16/63 received corticosteroids alone. 29/63 received corticosteroids and subsequently IST (10 TNF- α inhibitor/cyclophosphamide, 10 mycophenolate/methotrexate, 9 other). Median time to IST initiation was 9.1 months (range 1–132). 16/63 were treated with initial IST (4 TNF- α /cyclophosphamide, 8 mycophenolate/methotrexate, 4 other). There was no significant difference in time to contrast-enhancement resolution between corticosteroids with/without subsequent IST versus corticosteroids + initial IST (Wilcoxon-rank sum test 0.93) or CSM versus corticosteroids + initial IST (Wilcoxon-rank sum test 0.97). Limitations are small sample size, variations in initiation of treatment and doses used, and intervals of clinical/radiographic follow-up.

Conclusions

Standardization of reporting and monitoring of treatment and outcomes is needed for patients with NSM to better assess optimal initial therapy plans.

Disclosure: The institution of Dr. Balaban has received research support from Biogen. Dr. Manzano has nothing to disclose. Dr. Ali has nothing to disclose. The institution of Dr. Flanagan has received personal compensation in the range of \$500-\$4,999 for serving on a Scientific Advisory or Data Safety Monitoring board for Alexion. Dr. Flanagan 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. Flanagan has received personal compensation in the range of \$500-\$4,999 for serving on a Scientific Advisory or Data Safety Monitoring board for Horizon Therapeutics. Dr. Flanagan has received personal compensation in the range of \$500-\$4,999 for serving on a Speakers Bureau for Pharmacy times. The institution of Dr. Flanagan has received research support from Viela Bio. Dr. Flanagan has a non-compensated relationship as a Member of medical Advisory Board with The MOG Project that is relevant to AAN interests or activities. Dr. Flanagan has a non-compensated relationship as a Editorial board member with Journal of The Neurologic Sciences that is relevant to AAN interests or activities. Dr. Flanagan has a non-compensated relationship as a Editorial board member with Neuroimmunology Reports that is relevant to AAN interests or activities. Dr. Aksamit has nothing to disclose. Dr. Redenbaugh has nothing to disclose. Dr. Clardy has received personal compensation for serving as an employee of Veterans Health Administration (VHA). Dr. Clardy has received personal compensation for serving as an employee of University of Utah Health. Dr. Clardy has received personal compensation in the range of \$0-\$499 for serving as a Consultant for Clarion. Dr. Clardy has received personal compensation in the range of \$500-\$4,999 for serving as a Consultant for ExpertConnect. The institution of Dr. Clardy has received personal compensation in the range of \$0-\$499 for serving as a Consultant for VielaBio. The institution of Dr. Clardy has received personal compensation in the range of \$0-\$499 for serving as a Consultant for Genentech. The institution of Dr. Clardy has received personal compensation in the range of \$500-\$4,999 for serving as a Consultant for Alexion. The institution of Dr. Clardy has received personal compensation in the range of \$0-\$499 for serving as a Consultant for GuidePoint. Dr. Clardy has received personal compensation in the range of \$10,000-\$49,999 for serving as an Editor, Associate Editor, or Editorial Advisory Board Member for Neurology/AAN Publications. The institution of Dr. Clardy has received research support from Alexion Pharma. The institution of Dr. Clardy has received research support from Sumaira Foundation for NMO. The institution of Dr. Clardy has received research support from Immune Deficiency Foundation. The institution of Dr. Clardy has received research support from Western Institute for Veteran Research. The institution of Dr. Clardy has received research support from NIH/NINDS. Dr. Clardy has received personal compensation in the range of \$500-\$4,999 for serving as a AAN Summer Meeting CoDirector Travel and Lodging with AAN. Dr. Clardy has received personal compensation in the range of \$500-\$4,999 for serving as a Grand Rounds Travel and Lodging with U of Iowa. Dr. Clardy has received personal compensation in the range of \$500-\$4,999 for serving as a Speaker Honoraria for Grand Rounds with Barrow Neurological Institute. Dr. Samudralwar has received personal compensation in the range of \$500-\$4,999 for serving on a Scientific Advisory or Data Safety Monitoring board for Sanofi Genzyme. Dr. Samudralwar has received personal compensation in the range of \$500-\$4,999 for serving on a Speakers Bureau for Biogen. Dr. Samudralwar has received personal compensation in the range of \$500-\$4,999 for serving on a Speakers Bureau for EMD Serono. Dr. Agyei has nothing to disclose. Dr. Zabeti has received personal compensation in the range of \$10,000-\$49,999 for serving on a Speakers Bureau for Alexion. Dr. Zabeti has received personal compensation in the range of \$5,000-\$9,999 for serving on a Speakers Bureau for Alexion. Dr. Zabeti has received personal compensation in the range of \$10,000-\$49,999 for serving on a Speakers Bureau for Biogen. 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Dr. Levy 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. Levy has received personal compensation in the range of \$5,000-\$9,999 for serving on a Scientific

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Neurology 2022;99;S23-S24

DOI 10.1212/01.wnl.0000903204.16114.2a

This information is current as of December 5, 2022

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