Conclusions

Central canal dilation is a common radiologic accompaniment of acute MOGAD and AQP4+NMOSD myelitis, but not MS myelitis. The resolution of central canal dilation on follow-up MRI in most patients suggests it is transient and related to the acute inflammatory edema. Central canal dilation may differentiate MOGAD and AQP4+NMOSD from MS. Its recognition could facilitate earlier testing for MOG-IgG and AQP4-IgG, accurate diagnosis, and treatment.

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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.

Exposure to TNF Inhibitors is Rare at MOGAD Diagnosis

Vyanka Redenbaugh, Eoin Flanagan, Valentina Floris, MD, Pietro Zara, M. Tariq Bhatti, Francisco Sanchez Moreno, Matthew Koster, Sara Mariotto, Sean Pittock, John Chen, Alberto Cauli, Paolo Solla, Elia Sechi

Objective

To assess the potential association between TNF-inhibitors and MOGAD

Background

The association of tumor necrosis factor-a (TNF)-inhibitors with MS has previously been suggested, whereas little is known about MOG-IgG-associated disease (MOGAD) in the context of these drugs. We recently encountered two patients who developed MOGAD while receiving TNF-inhibitors, prompting a search for similar cases in the literature and clinical practice.

Design/Methods

The two cases were seen at Mayo Clinic, Rochester (bilateral optic neuritis) and the University-Hospital of Sassari (brainstem syndrome). Three additional cases of MOGAD presenting during treatment with TNF-inhibitors were identified through Pubmed. We searched the medical records of 336 MOGAD patients seen at the Mayo Clinic, to assess if they had been treated with TNF-inhibitors.

Results

A total of 5 patients were identified. The median age at MOGAD presentation was 40 years (range, 36-49); 4/5 were male (80%). The median time from TNF-inhibitor initiation to MOGAD presentation was 6.5 years (range, 2-18). Of 4 patients who discontinued the TNF-inhibitor due to MOGAD onset, two subsequently had a MOGAD relapse. While in another patient, neurological symptoms subsided with corticosteroids despite TNF-inhibitor being maintained. The frequency of MOGAD presenting during TNF-inhibitors treatment at Mayo Clinic was 0.3% (1/336 cases).

Conclusions

We found that MOGAD is unlikely to present during treatment with TNF-inhibitors. The outcomes in these patients seemed not to be influenced by TNF-inhibitor treatment duration or discontinuation. These findings suggest the benefit of TNF-inhibitor withdrawal is not obvious, and the choice of discontinuing vs maintaining treatment with TNF-inhibitors should be weighted based on symptoms severity and activity status of the underlying systemic disorder. When withdrawal is considered, immunosuppression with agents potentially effective for both MOGAD and the immune-mediated disease originally managed by the TNF-inhibitor, could serve as dual purpose treatment.

Disclosure: Dr. Redenbaugh 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.

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 para-infectious 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), imagings, 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

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