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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.

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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

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

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

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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-alpha 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-alpha 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-alpha/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.

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