Design/Methods

A 23-year-old female was diagnosed with NMDAR encephalitis. She was treated with ovarian teratoma removal, corticosteroids, intravenous immunoglobulin therapy, rituximab, and tocilizumab. She continued to experience severe, self-mutilating orofacial dyskinesias. Tetrabenazine, haloperidol, and diazepam did not yield any sustained improvement. Tramadol was started based on prior case reports suggesting its efficacy as well as clonazepam.3

Results

Tramadol 50 mg po q6h led to immediate improvement in symptoms. Over the next 5 days, tramadol was increased to 150 mg NG q6h and further reduced movements. When tramadol was held for one day, the movements significantly worsened and improved when it was restarted. Clonazepam 1 mg NG QID also led to further improvement.

Conclusions

Tramadol and clonazepam effectively treated severe orofacial dyskinesias in a patient with NMDAR encephalitis and refractory symptoms despite aggressive management. We propose early use of tramadol and clonazepam be considered for severe orofacial dyskinesias secondary to NMDAR encephalitis.

Disclosure: Dr. Fernandes has nothing to disclose. Dr. Clift has received personal compensation in the range of $5,000-$9,999 for serving as a Consultant for Biogen. Dr. Clift has received personal compensation in the range of $5,000-$9,999 for serving as a Consultant for EMD Serono. Dr. Clift has received personal compensation in the range of $5,000-$9,999 for serving as a Consultant for Novartis. Dr. Clift has received personal compensation in the range of $5,000-$9,999 for serving as a Consultant for Sanofi. Dr. Clift has received personal compensation in the range of $5,000-$9,999 for serving as a Consultant for Roche. Dr. Clift 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. Clift has received personal compensation in the range of $5,000-$9,999 for serving on a Scientific Advisory or Data Safety Monitoring board for Biogen. Dr. Clift has received personal compensation in the range of $5,000-$9,999 for serving on a Scientific Advisory or Data Safety Monitoring board for EMD Serono. Dr. Clift has received personal compensation in the range of $5,000-$9,999 for serving on a Scientific Advisory or Data Safety Monitoring board for Novartis. Dr. Clift has received personal compensation in the range of $5,000-$9,999 for serving on a Scientific Advisory or Data Safety Monitoring board for Roche. Dr. Clift has received personal compensation in the range of $5,000-$9,999 for serving on a Scientific Advisory or Data Safety Monitoring board for Sanofi. Dr. Chu has nothing to disclose.

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Predictors for the Development of Neurological Immune-Related Adverse Events of Immune Checkpoint Inhibitors and Impact on Mortality

Chen Yan, Merry Huang, Carol Swetlik, Karlo Toljan, James Bena, Pauline Funchain, Marisa McGinley

Objective

To report the incidence, predictors for development, impact on mortality, and impact on pre-existing neurological conditions of neurological immune-related adverse events (irAEs) in a large clinical cohort.

Background

Immune checkpoint inhibitors (ICI) are associated with irAEs. Although neurological complications have been described, little is known about risk factors for their development and their impact on mortality. The impact of ICIs on pre-existing neurological conditions is also not well understood.

Design/Methods

Patients who received ICI between January 2013 and December 2018 were identified using a tertiary cancer center registry. Patient demographics, cancer characteristics, treatment type, and concurrent oncologic therapy were summarized using descriptive statistics. Patients with neuro-irAE were compared to those without neuro-irAE during the study timeframe. Odds ratios from univariable and penalized multivariable logistic regression models were calculated to identify potential predictors for developing a neuro-irAE. The impact of a neuro-irAE on overall survival was estimated by Kaplan-Meier and multivariable Cox proportional-hazard models.

Results

Overall frequency of neurological irAEs was 2.3% (28/1228). Peripheral nervous system complications such as myasthenia gravis, myositis, and neuropathies were the most frequent (53.6%). Melanoma, younger age, prior chemotherapy, prior resection, CTLA-4 ICI exposure, and combination ICI exposure had significantly higher odds for developing a neuro-irAE ($p < 0.05$), but these findings were not statistically significant in the multivariable models. Those with a neuro-irAE had greater survival at 3 years compared to those without a neuro-irAE (69% vs 55%, $p = 0.004$), but after adjusting for patient and cancer characteristics, this effect was no longer statistically significant. Relapse rate of pre-existing neurological condition after exposure to ICI was 15.4% (2/13).

Conclusions

Neuro-irAEs are rare and are not associated with an increase in mortality. Potential predictors for the development of neuro irAEs are younger age, melanoma, prior chemotherapy and resection, CTLA-4, or combination ICI exposure. Relapse of a pre-existing neurological condition was uncommon.

Disclosure: Dr. Yan has nothing to disclose. Dr. Huang has nothing to disclose. An immediate family member of Dr. Swetlik has received personal compensation for serving as an employee of Global Blood Therapeutics. An immediate family member of Dr. Swetlik has stock in Global Blood Therapeutics. Dr. Toljan has nothing to disclose. James Bena has nothing to disclose. Pauline Funchain has received personal compensation in the range of $500-$4,999 for serving on a Scientific Advisory or Data Safety Monitoring board for Eisai. The institution of Pauline Funchain has received research support from Pfizer. The institution of Pauline Funchain has received research support from Bristol Myers Squibb. Dr. McGinley has received personal compensation in the range of $500-$4,999 for serving on a Scientific Advisory or Data Safety Monitoring board for Genentech. The institution of Dr. McGinley has received research support from Novartis. The institution of Dr. McGinley has received research support from Biogen.

Do Aspirin and Other Non-Steroidal Anti-Inflammatory Drugs (NSAIDs) Have a Protective Effect Against Neuro-Autoimmune Disease and Comorbidity?

Mohsen Ahmed, Afaaq Ahmed, Ronak Trivedi, Muhammed Ors, Nabeel Ahmed, Nizar Souayah

Objective

To investigate the neuroprotective potential of Aspirin (AS) and other non-steroidal anti-inflammatory drugs (NSAIDs) against neuro-autoimmune diseases (NAD) and additional comorbidity.

Background

AS is an NSAID used for the treatment and prevention of cardiovascular and neurologic disease. However, the extent of Aspirin and other NSAIDs neuroprotective benefits and their prevention of additional comorbidity in patients with NAD has not been completely characterized.

Design/Methods

A retrospective analysis on 34464 patients hospitalized at a tertiary care center in a major metropolitan area was conducted. 6168 and 8228 patients were taking either AS or NSAIDs on admission, respectively. 18707 patients were not taking aspirin or NSAIDs (nMED). Patients with NAD were further categorized as taking Aspirin alone (aAS), taking NSAIDS alone (aNSAID), or no medications (anMED). The outcomes compared included the prevalence of NAD and risk of comorbidities.

Results

Patients taking AS or NSAID had no significant difference in NAD compared to those in nMED (0.45%, 0.6%, 0.56%, $p > 0.05$). Patients in aAS had a mortality rate at 10.7% compared to 1.9% in those in anMED ($p < 0.05$). 57% of aAS compared to 14.4% of anMED had high risk comorbidities ($p < 0.0001$). The prevalence of encephalopathy or overall neurological complications was significantly higher in aAS compared to anMED (17.8%, 6.7%, $p > 0.05$; 28.6%, 13.%, $p > 0.05$). Patients in aNSAID had no significant increase in mortality, high risk comorbidity, or overall neurological complications when compared with anMED (4%, 6.7%, $p > 0.05$; 10%, 6.7%, $p > 0.05$; 6.1%, 13.5%, $p > 0.05$).

Conclusions

These results suggest that AS may provide a strong protective benefit against neurological complications among those with NAD despite a significantly higher associated mortality and high risk comorbidity.

Disclosure: Mr. Ahmed has nothing to disclose. Mr. Ahmed has nothing to disclose. Mr. Trivedi has nothing to disclose. Mr. Ors has nothing to disclose. Mr. Ahmed has nothing to disclose. Dr. Souayah has received publishing royalties from a publication relating to health care.

Therapeutic Plasma Exchange in a Patient With Acute Motor Axonal Neuropathy Subtype of Guillain-Barre Syndrome and Systemic Lupus Erythematosus

Melissa Huberman, Yamac Akgun, Jake Langlie, YanYun Wu

Objective

To highlight treatment resistance of concurrent Guillain-Barre syndrome (GBS) and active systemic lupus erythematosus (SLE).

Background

Coincidence of SLE and GBS is an uncommon and complicated course of autoimmune disease. Treatment is further complicated by pregnancy exacerbation of SLE.

Design/Methods

We report a case of acute motor axonal neuropathy (AMAN) subtype of GBS in a young pregnant woman in her early 20s presenting with SLE. Patient was 10 weeks pregnant at presentation. Lumbar puncture study and electrical muscle stimulation (EMS) were consistent with acute motor axonal neuropathy subtype of GBS. Patient also had increased proteinuria and renal biopsy performed that was consistent with lupus nephritis. Despite treatment with IVIG and pulse dose corticosteroids, the patient had minimal neurological improvement with respiratory decline requiring intubation. Her pregnancy was terminated at this point and a course of therapeutic plasma exchange (TPE) was started followed by cyclophosphamide.

Results

The patient responded to the combination of therapy and had slow but gradual neurologic recovery as well as improvement of proteinuria.

Conclusions

Concurrent GBS and active SLE in the setting of pregnancy may be more treatment resistant, and combination therapy including TPE, immunosuppression, and termination of pregnancy may be indicated. For patients with concurrent GBS and SLE, especially axonal subtype of GBS and during pregnancy, TPE should be considered as a primary treatment option with respect to both efficacy and safety.

Disclosure: Miss Huberman has nothing to disclose. Dr. Akgun has nothing to disclose. Mr. Langlie has nothing to disclose. YanYun Wu has nothing to disclose.

Neurochondrin Autoimmunity With Slow Saccades: A Case Report

Laurel Tanke, Eric Eggenberger, Misha Pless

Objective

N/A.

Background

Neurochondrin is a cytoplasmic neuronal antigen that can be targeted by specific antibodies, resulting in an antigen-specific T-cell response and autoimmune cerebellar and brain stem degeneration.1 Previously reported cases of neurochondrin antibody positivity have been predominantly associated with rapidly-progressive rhombencephalitis, but various other neurological symptoms and signs may be present.1,2

Design/Methods

N/A.

Results

34-year-old woman presented with 2 years of constant numbness in her toes, gradually worsening to involve the entire lower and upper extremities. She also reported dysarthria, imbalance, and leg spasticity. Neurological examination showed a slow, wide-based gait, brisk reflexes bilaterally, and bilateral extensor plantar responses. MRI brain was normal and CSF analysis showed 7 oligoclonal bands. The patient was diagnosed with probable multiple sclerosis and steroids were initiated but resulted in no improvement of symptoms. Plasmapheresis was also ineffective. 3 months later, MRI showed subtly increased T2 signal of the central cord mainly involving the gray matter at the level of C5 and C6. CSF analysis showed 15 oligoclonal bands and an elevated IgG index of 1.39. VDRL was nonreactive. HTLV, ANCA, AQP4, CSF VZ PCR, HSV PCR, and HHV6 PCR were all negative. EMG showed mild length-dependent axonal sensory motor peripheral neuropathy. On neuro-ophthalmic examination, the patient showed slowed saccades with some preservation of optokinetic nystagmus (OKN) fast phases. Repeat MR showed abnormal T2 hyperintensity in the anterior horns of the gray matter from C1-2 to T1. Serum testing at that time showed a positive neurochondrin antibody. She was then started on cyclophosphamide and mycophenolate mofetil. 6 months after initiation of cyclophosphamide, dysarthria and upper limb mobility improved, and she had no further neurological deterioration at that time.

Conclusions

Neurochondrin IgG-related autoimmunity is rare and can result in a wide variety of neurological signs and symptoms. Ocular dysmotility, including slowed saccades, may be present.

Disclosure: Ms. Tanke has nothing to disclose. Dr. Eggenberger has nothing to disclose. Dr. Pless has nothing to disclose.

Refractory Pediatric NMDA Receptor Encephalitis: A Case Series

Varun Kannan, Delia Rospigliosi, Victoria Adeseye, Yi-Chen Lai, Timothy Lotze, Eyal Muscal, Nikita Shukla

Objective

To characterize clinical features of our institution's refractory pediatric NMDA receptor encephalitis (NMDARE) patients, in the hopes of identifying predictive risk factors and specific treatment escalation targets.

Do Aspirin and Other Non-Steroidal Anti-Inflammatory Drugs (NSAIDs) Have a Protective Effect Against Neuro-Autoimmune Disease and Comorbidity?

Mohsen Ahmed, Afaaq Ahmed, Ronak Trivedi, et al.

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