Advisory or Data Safety Monitoring board for Horizon. 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 Genentech. Dr. Levy has received personal compensation in the range of $500-$4,999 for serving as an Editor, Associate Editor, or Editorial Advisory Board Member for Elsevier. Dr. Levy has received personal compensation in the range of $10,000-$49,999 for serving as an Expert Witness for Various law firms. The institution of Dr. Levy has received research support from National Institutes Health. Dr. Bhattacharyya has received personal compensation in the range of $500-$4,999 for serving on a Scientific Advisory or Data Safety Monitoring board for Alexion Pharmaceuticals. The institution of Dr. Bhattacharyya has received research support from Alexion Pharmaceuticals. Dr. Bhattacharyya has received publishing royalties from a publication relating to health care. Dr. Bhattacharyya has received publishing royalties from a publication relating to health care. Dr. Bhattacharyya has received publishing royalties from a publication relating to health care.

Resolution of Recurrent VZV Myelitis With the Use of Intravenous (IV) Immunoglobulin (IG)

Danelvis Paredes, Elijah Lackey, Christopher Eckstein

Objective

To present a case of a patient with recurrent VZV myelitis successfully treated with Intravenous Immunoglobulin (IVIG).

Background

VZV myelitis is a rare complication of VZV reactivation that tends to be monophasic. There is no solid evidence for a particular treatment regimen for VZV myelitis. No prior reports or studies have looked at using intravenous immunoglobulin for this condition, particularly for refractory cases.

Design/Methods

75 year old female with history of hypertension presented with paresthesia on lateral side of right lower extremity, followed by vesicular rash T2 dermatomal distribution. Over a period of 2 weeks patient started experiencing bilateral lower extremity weakness R>L. CNS imaging revealed C3-C6 enhancing lesion and T2 hyperintense lesion. VZV confirmed by skin biopsy. Lumbar puncture (LP) was remarkable only for mildly elevated protein (67 mg/dL), although this LP was 4 weeks after initial symptoms. Patient was treated initially with 5 days of 1 g IV Solumedrol and valacyclovir 1 g TID with improvement of weakness and rash. However, over the next two years, the patient continued to get occasional vesicles with a flare of her myelitis shortly after valacyclovir down-titration trial. She was started on monthly IVIG (1 g/kg IVIG over 2 days) given her refractory and recurrent myelitis. Since starting monthly IVIG, the patient has not had any more zoster outbreaks or further episodes of myelitis. She has gradually improved her balance and gait as well.

Results

N/A.

Conclusions

We present an unusual case of recurrent VZV myelitis successfully treated with monthly IVIG. The successful treatment of this patient with IVIG should prompt consideration for its use in similar cases of recurrent VZV myelitis and may provide insight for future studies on how to treat VZV-related diseases. Rapid initiation of this treatment when the condition is recognized early could significantly improve outcomes and patients' quality of life.

Disclosure: Dr. Paredes has nothing to disclose. Dr. Lackey has nothing to disclose. The institution of Dr. Eckstein has received research support from Biogen. The institution of Dr. Eckstein has received research support from Genzyme.

Sjogren's Sensory Neuropathy: A Potentially Treatable Condition with Early Intervention

Ryan Naum, Kelly Gwathmey

Objective

To describe 2 cases of Sjögren's syndrome sensory neuronopathy (SSSN) in which early intervention resulted in excellent clinical outcomes.

Background

Acquired sensory neuronopathies (i.e. dorsal root ganglionopathies) are rare sensory neuropathies most commonly associated with Sjögren's syndrome (SS) and paraneoplastic syndromes.

Design/Methods

We describe 2 patients who presented with painful sensory symptoms, sicca symptoms and with positive SSA and antinuclear antibodies. The first patient presented with perceived symmetric hand weakness (despite no motor nerve conduction abnormalities), total body numbness and paresthesia, as well as sensory ataxia, while the other presented with asymmetric numbness and pain in the left foot and hand. The electrophysiological profiles varied considerably between the 2 patients. The first patient demonstrated complete absence of sensory nerve action potentials (SNAPs) whereas the second patient had only asymmetrical superficial peroneal and sural SNAPs, corresponding with his clinical deficit. The first patient was diagnosed with SSSN, while the other was diagnosed with asymmetrical sensory neuropathy in SS. The first patient was treated with mycophenolate mofetil (MMF) with dramatic symptomatic improvement and near recovery of clinical deficits. The second patient was treated with prednisone, methotrexate, and ultimately switched to MMF and rituximab with significant improvement in symptoms.

Results

NA.

Conclusions

SS is commonly associated with sensory neuropathies including sensory neuronopathies. Both patients met diagnostic criteria for SSSN, though with differing severities. To date, there has been no randomized controlled trial evaluating treatments of SSSN. However, our findings suggest that early use of MMF could result in considerable benefit in a disease that is often functionally devastating.

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NMDA Receptor Encephalitis With Severe Orofacial Dyskinesias Treated With Tramadol and Clonazepam

Falen Fernandes, Fraser Clift, Laura Chu

Objective

N/A.

Background

Anti-N-methyl-D-aspartate receptor (NMDAR) encephalitis is a neuroinflammatory disease mediated by antibodies targeting the GluN1 subunit of the NMDAR. It presents with well-defined neuropsychiatric symptoms, including psychosis, agitation, seizures, and memory disturbances.1 Movement disorders including orofacial dyskinesias are common, but often difficult to manage, with no specific published guidelines.[1,2,3]

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.

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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.

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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.

Sjogren's Sensory Neuropathy: A Potentially Treatable Condition with Early Intervention

Ryan Naum and Kelly Gwathmey

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