findings, diagnostic studies, treatment response and TPE-related complications were recorded. We also conducted linear regression models to assess for possible predictive factors for TPE response. Literature review of SPSD and TPE was also conducted.

Results

Thirty-six SPSD patients were treated with TPE; mean age was 48 years, 81% female, and average anti-GAD65 antibody titer was 42352 U/mL (range, 0-309,902). Twenty-two patients had classic SPS, 10 had SPS-plus, and 3 had other phenotypes. Thirty-three patients were treated for acute exacerbations, and 3 were on maintenance TPE. There were 4 (11.1%) TPE-related complications (catheter infection, catheter thrombosis, hemorrhage), but no deaths or anaphylaxis. Twenty patients (55.6%) reported improvement in symptoms after TPE, 13 reported no change, and 3 reported worsening of symptoms. Of the 36 total patients who received TPE, 21 received TPE at Johns Hopkins Hospital for an acute exacerbation of their condition, with 12 requiring fewer anti-spasmodic medications 3 months after TPE treatment. There were no predictive factors in a positive treatment response to TPE. Literature review identified 42 more patients; 69% of these patients reported a temporary improvement in their condition.

Conclusions

We describe the safety and tolerability of TPE in patients with SPSD and show that TPE-related complications are uncommon and manageable. Additionally, many patients with SPSD derived improvement with TPE. Further studies could help inform clinicians when to use TPE in SPSD.

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Clinical and Paraclinical Features of Non-Paraneoplastic NIF-Mediated Disease Associated With Concurrent SARS-CoV-2 Infection

Lauren Schmidt, Jon Karel, Stefanie Rodenbeck

Objective

To describe clinical and paraclinical features of non-paraneoplastic NIF-mediated disease associated with concurrent SARS-CoV-2 infection.

Background

Neurologic syndromes associated with neuronal intermediate filament (NIF) immunoglobulin G (IgG) most often are characterized by encephalopathy, cerebellar ataxia, or myelopathy. NIF-IgG has been strongly correlated with the presence of an underlying malignancy, with neuroendocrine tumors being most prevalent. Despite the intracellular target of this antibody, patients with NIF-IgG mediated disease tend to improve clinically with immunotherapy. While some cases have been described in a parainfectious context, this is the first such case in the context of a SARS-CoV-2 infection.

Design/Methods

NA.

Results

We reported a case of non-paraneoplastic NIF-mediated disease in the setting of SARS-CoV-2 infection. The patient presented with first time seizure. He was found to have frequent left temporal lobe spikes then two left temporal lobe seizures on neurotelemetry. Brain MRI displayed abnormal signal throughout the left hippocampus and mesial temporal lobe, without contrast enhancement. LP was subsequently performed. CSF showed elevated protein, 14-3-3, T-tau, interleukin 13, interleukin 2 receptor, and interleukin 6. The meningitis/encephalitis panel, and HSV-1/2 IgG were negative. Serum autoimmune encephalitis panel revealed a high-positive titer for anti-NIF 1:960, with concurrent NIF heavy chain cell-based assay positive. He improved with three days of IV steroids and treatment with levetiracetam and lacosamide. He has since been seizure free.

Conclusions

NIF-mediated diseases usually present with encephalopathy, cerebellar ataxia, or myelopathy and are generally seen in the setting of malignancy. Our case illustrated an example of NIF-mediated disease presenting as seizure in the setting of infection. This highlights the importance of consideration of parainfectious autoimmunity.

Disclosure: Dr. Schmidt has nothing to disclose. Dr. Karel has nothing to disclose. Dr. Rodenbeck has nothing to disclose.

Real-World Utilization Patterns of Intravenous Immunoglobulin in Adults With Generalized Myasthenia Gravis in the United States

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Objective

To evaluate real-world utilization patterns of intravenous immunoglobulin (IVIg) among patients with generalized myasthenia gravis (gMG) over 3 years post-IVIg initiation.

Background

gMG is a rare autoimmune neuromuscular disorder with no known cure. Although IVIg is the most commonly used add-on therapy after standard of care treatments in gMG, it is currently unclear whether it is more commonly used as a "one-off" treatment to manage exacerbations, or as maintenance therapy aimed at reducing or replacing steroid use.

Design/Methods

Patients with gMG who initiated IVIg treatment were identified from a US claims database (Symphony Health, an ICON plc Company, Integrated Dataverse [IDV]®, January 1, 2014-December 31, 2019). The

Therapeutic Plasma Exchange in the Management of Stiff Person Syndrome Spectrum Disorders: A Case Series and Review of the Literature

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