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.

Disclosure: Dr. Roy has nothing to disclose. Dr. Mercure-Corriveau has nothing to disclose. Danielle Obando has nothing to disclose. Dr. Wang has nothing to disclose. Dr. Daou has nothing to disclose. Dr. Tobian has nothing to disclose. Dr. Bloch has received personal compensation in the range of $500-$4,999 for serving as a Consultant for Terumo BCT. Dr. Bloch has received personal compensation in the range of $500-$4,999 for serving as a Consultant for Abbott laboratories. Dr. Newsome has received personal compensation in the range of $500-$4,999 for serving as a Consultant for Autobahn. Dr. Newsome has received personal compensation in the range of $500-$4,999 for serving on a Scientific Advisory or Data Safety Monitoring board for Biogen. Dr. Newsome has received personal compensation in the range of $10,000-$49,999 for serving on a Scientific Advisory or Data Safety Monitoring board for Genentech. Dr. Newsome has received personal compensation in the range of $500-$4,999 for serving on a Scientific Advisory or Data Safety Monitoring board for Novartis. Dr. Newsome has received personal compensation in the range of $500-$4,999 for serving on a Scientific Advisory or Data Safety Monitoring board for EMD Serono. Dr. Newsome has received personal compensation in the range of $500-$4,999 for serving on a Scientific Advisory or Data Safety Monitoring board for Greenwich Biosciences. Dr. Newsome has received personal compensation in the range of $500-$4,999 for serving on a Scientific Advisory or Data Safety Monitoring board for Bristol Myers Squibb. Dr. Newsome 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. The institution of Dr. Newsome has received research support from Biogen. The institution of Dr. Newsome has received research support from Genentech/Roche. The institution of Dr. Newsome has received research support from Department of Defense. The institution of Dr. Newsome has received research support from Patient Centered Outcomes Research Institute. The institution of Dr. Newsome has received research support from National MS Society. Dr. Newsome has received personal compensation in the range of $5,000-$9,999 for serving as a Clinical adjudication committee member for clinical trial with medDay Pharmaceuticals. Dr. Newsome has received personal compensation in the range of $10,000-$49,999 for serving as a Lead PI for Clinical Trial with Roche.

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

Cynthia Qi, Tom Hughes, Deborah Gelinas, Yuebing Li, MD, Amit Goyal, Edward Brauer, Arpit Bhuwalka, Mai Sato, Sudhir Jadhav, Glenn Phillips

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

frequency of subsequent IVIg treatment and associated cost during the first 12 months post-IVIg initiation were analyzed. Usage patterns of concomitant gMG treatments during the year preceding and 3 years post-IVIg initiation were compared.

Results

Among 1225 patients with gMG who initiated IVIg treatment, 706 patients (57.6%) received 1 to 5 IVIg treatment courses (intermittent IVIg users), and 519 patients (42.4%) received = 6 IVIg treatment courses (chronic IVIg users) within the subsequent year. Mean annual medical cost per patient was nearly 2.5-fold higher for chronic vs intermittent IVIg users ($161,478 vs $64,888, $p < 0.001$). Usage frequency of other immunotherapy treatments did not decrease over the follow-up period, even for patients who continued annual chronic IVIg for 3 consecutive years post-initiation.

Conclusions

Nearly half of patients with gMG received chronic and multiple IVIg treatment courses within the first year once initiating IVIg treatment, indicating higher usage than expected. For all IVIg initiators, usage frequency of other immunotherapy treatments did not decrease over 3 years despite IVIg initiation.

Disclosure: Ms. Qi has received personal compensation for serving as an employee of argenx. Dr. Hughes has received personal compensation for serving as an employee of Argenx. Dr. Hughes has stock in argenx. Dr. Gelinas has received personal compensation for serving as an employee of argenx. Dr. Gelinas has stock in argenx. Dr. Li MD has nothing to disclose. Mr. Goyal has received personal compensation for serving as an employee of ZS Associates. Edward Brauer has received personal compensation for serving as an employee of argenx. Edward Brauer has stock in argenx. Mr. Bhuwalka has received personal compensation for serving as an employee of ZS Associates India. Mai Sato has nothing to disclose. Mr. Jadhav has received personal compensation for serving as an employee of ZS Associates, India. Dr. Phillips has received personal compensation for serving as an employee of argenx. Dr. Phillips has stock in argenx.

Neurological Immune-Related Adverse Events After COVID-19 Vaccination: A Systematic Review

Ameena Shafiq, Mohammad Salameh, Ibrahim Laswi, Ibrahim Mohammed, Omar Mhaimeed, Nada Mhaimeed, Narjis Mhaimeed, Pradipta Paul, Malik Mushannen, Abdallah Elshafeey, Ahmed Fares, Sean Holroyd, Dalia Zakaria

Objective

The goal of this study is to compile published data reporting neurological immune-related adverse events following COVID-19 vaccination, not including those relating to hematologic abnormalities such as thrombosis or hemorrhage.

Background

COVID-19 vaccination has been repeatedly shown to reduce the incidence and severity of COVID-19 infection. The expedited timeline of these vaccines has given rise to many discussions pertaining to their safety. Many neurological and non-neurological adverse events have been linked to COVID-19 vaccination including acute MI, anaphylaxis, appendicitis, Bell's palsy, deep vein thrombosis, disseminated intravascular coagulation, encephalomyelitis, transverse myelitis, and many others.

Design/Methods

The following databases were searched in April 2021 using different keywords: PubMed, Medline, Embase, Scopus, Web of Science, Science, Direct, MedRxiv, and Lens.org. Studies were included if they reported any adverse immune-related neurological events secondary to COVID-19 vaccination. Studies were excluded if they were not in English, included self-reported events only, or did not report primary data. Screening and extraction were conducted by 2 different reviewers using Covidence.

Results

The search strategy yielded 18 studies which reported a total of 61 patients who had received a COVID-19 vaccination and experienced = 1 neurological adverse events. Most reported adverse events were facial nerve palsy (52.5%), reactivation of herpes zoster (11.5%), Guillian-Barré syndrome (6.6%), demyelinating disease (6.6%), and neuropathy (11.5%). Other reported adverse effects were delirium, periauricular vesicular rash, bilateral sensorineural hearing loss, visual disturbance, gait disturbance, serotonin syndrome, and vestibular ataxia (16.4%).

Conclusions

The symptoms were time-limited and self-resolving in nature. In addition, the incidence of the reported events following COVID-19 vaccination compared to the general population is similar. Hence, there is little to no evidence suggesting a causal relationship between COVID-19 vaccination and neurological adverse events.

Disclosure: Ms. Shafiq has nothing to disclose. Mr. Salameh has nothing to disclose. Mr. Laswi has nothing to disclose. Mr. Mohammed has nothing to disclose. Dr. Mhaimeed has nothing to disclose. Miss Mhaimeed has nothing to disclose. Ms. Mhaimeed has nothing to disclose. Mr. Paul has nothing to disclose. Mr. Mushannen has nothing to disclose. Dr. Elshafeey has nothing to disclose. Dr. Fares has nothing to disclose. Dr. Holroyd has nothing to disclose. Dr. Zakaria has nothing to disclose.

Post COVID-19 Vaccination Associated Acquired Neuromyotonia

Shivangi Shivangi, Amit Shankar Singh, Jeenendra Prakash Singhvi, Namit Bansal, Rajat Gupta

Objective

Acquired neuromyotonia or Isaacs' syndrome is an immune mediated inflammatory disorder characterized by involuntary continuous muscle fiber activity manifesting as twitching and stiffness along with autonomic dysfunctions like hyperhidrosis and/or tachycardia. Here we report a young male who developed acquired neuromyotonia following COVID-19 vaccination.

Background

A 20-year-old male presented in our clinic with gradually progressive pain and numbness in bilateral lower limbs, tremors in both hands, shivering while walking, excessive sweating and difficulty in micturition for last 15 days. He also noticed twitching of muscles in calf and thigh muscles along with these symptoms. According to patient, these symptoms started after he took his first dose of COVID-19 vaccination (Covishield- Oxford-AstraZeneca viral vector vaccine) 10 days back. There was no history of fever or backache. He had no chronic illness and was not on any medications. Examination revealed hyperhidrosis, mild proximal muscle weakness in both lower limbs with twitching in muscles suggestive of myokymia. There were quivering and rippling movements of intrinsic muscles of both hands resembling polyminimyoclonus. In view of the above findings, possibility of acquired neuromyotonia possibly following COVID-19 vaccination was kept and further evaluation was done.

Design/Methods

Routine blood investigations, thyroid function test, anti-thyroid peroxidase antibodies and anti-nuclear antibodies were normal. Cerebrospinal fluid analysis was normal. Anti-VGKC antibodies were detected in serum with strongly positive anti-CASPR and weakly positive anti-LGI1 antibodies confirming diagnosis of acquired neuromyotonia.

Results

Pulse dose of intravenous methylprednisolone for 5 days was given which resulted in visible improvement in pain, twitching, hyperhidrosis and urinary symptoms. He was continued on oral steroids and complete resolution of his symptoms was noted over a period of 2 months.

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

Cynthia Qi, Tom Hughes, Deborah Gelinas, et al.

Neurology 2022;99;S29-S30
DOI 10.1212/01.wnl.0000903256.02960.8f