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.

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Background

Management protocols for pediatric NMDARE increasingly recommend anti-CD-20 agents following first-line treatment with high-dose corticosteroids, intravenous immunoglobulin (IVIg), and/or plasma exchange therapy (PLEX). Even with early, aggressive treatment, some patients exhibit refractory disease or recurrences. Identification of clinical predictors for refractory disease course may allow earlier, targeted treatment escalation in these patients.

Design/Methods

We performed IRB-approved retrospective, descriptive review of patients in our institutional NMDARE database (2011-2021). Refractory disease was defined as lack of neurological improvement within 1-3 months, or recurrent relapse, after standardized treatment protocol (steroids, IVIg or PLEX, and two doses of rituximab 500 mg/m^2). Demographics, clinical information, and diagnostic results from refractory patients were collected.

Results

8/73 (10.9%) patients met criteria for refractory NMDARE (median age 10.0 years, IQR 8.3-14.0. 2 male, 6 female). Median days from symptom onset to first treatment was 12.5 (IQR 8.5-18.8), to detection of +NMDA Ab was 20.5 (17.8-24.0), and to rituximab was 27.5 (26.5-31.0). All were critically ill at disease onset, with seizures, encephalopathy, and respiratory failure. 2 (25%) had associated ovarian teratoma. Oligoclonal bands were tested in 6, with 4 resulting positive (67%). 7 had confirmed B-cell depletion after rituximab. Post-rituximab NMDA titers persisted in serum in 5 (63%) and CSF in 8 (100%). Immune therapy escalation was varied, and included repeat rituximab, mycophenolate, cyclophosphamide, and tocilizumab.

Conclusions

Severe initial presentation is a consistent feature among our refractory patients. The contribution of other factors such as oligoclonal bands and time to diagnosis/treatment are less clear, and warrant inter-group comparison with non-refractory patients. Persistence of serum and CSF titers despite B-cell depletion may suggest utility in targeting CD-20-negative mature plasma cells which may continue to produce disease-causing antibodies.

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Clinical Improvement Following Delayed Initiation of Immunotherapy in a Case of LGI1 Antibody Encephalitis Presenting with Faciobrachial Dystonic Seizures Following COVID-19 Vaccination

Kayla Martin, Matthew Creech, Michael Lane, Jacqueline Bernard

Objective

To demonstrate a case of suspected post-vaccine autoimmune encephalitis associated with leucine-rich glioma-inactivated protein (LGI1) antibodies with significant clinical improvement after initiation of immunotherapy nearly a year after symptom onset.

Background

Although the autoimmune encephalitides have overlap in presentation, some have unique manifestations (such as orofacial dyskinesias seen with NMDA encephalitis). These unique associations can serve as a clinical marker of response to treatment and even allow for earlier initiation of immunotherapy while awaiting results from antibody testing. LGI1 encephalitis characteristically presents with faciobrachial dystonic seizures (FBDS) that are refractory to anti-seizure medications (ASMs) but responsive to immunotherapy.

Design/Methods

Case report

Results

A previously healthy and highly independent 89-year-old woman developed what she described as abnormal posturing and spasms of the right shoulder two to three weeks after receiving the J&J COVID-19 vaccine. The abnormal movements progressed to involve the right side of her face and were refractory to multiple ASMs. EEG captured multiple events without epileptiform correlate. Several months later she developed paranoia, delusions, and hallucinations. Autoimmune encephalopathy panel returned positive for the LGI1-antibody around nine months after the onset of FBDS. Upon our initial exam, she had a fluctuating level of arousal, impaired recall of recent events, and was tangential in conversation. There were frequent, brief, repetitive, dystonic movements of the right side of the face consistent with FBDS. Admission was arranged for immunotherapy (intravenous methylprednisolone and intravenous immunoglobulin). Upon follow-up four weeks later, there was significant improvement in arousal and concentration with resolution of FBDS and delusions.

Conclusions

This case highlights a classic case of LGI1 encephalitis after vaccination presenting with FBDS and progressive cognitive changes. Despite immunotherapy being delayed, there was marked clinical improvement. It is important to recognize this entity and that it typically has a favorable outcome.

Disclosure: Dr. Martin has nothing to disclose. Dr. Creech has nothing to disclose. Dr. Lane has nothing to disclose. Dr. Bernard has received personal compensation in the range of $500-$4,999 for serving as a Consultant for Genentech. Dr. Bernard has received personal compensation in the range of $500-$4,999 for serving as a Consultant for Alexion. Dr. Bernard has received personal compensation in the range of $500-$4,999 for serving as a Consultant for Biogen. Dr. Bernard has received publishing royalties from a publication relating to health care.

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

Shuvro Roy, Nicolas Mercure-Corriveau, Danielle Obando, Yujie Wang, Laetitia Daou, Aaron Tobian, Evan Bloch, Scott Newsome

Objective

To describe the safety, tolerability, and response to TPE in patients with SPSD.

Background

Stiff person syndrome spectrum disorders (SPSD) are a rare group of disabling neuroimmunological disorders. SPSD often require immune therapies especially in the setting of inadequate response to symptomatic treatments. The safety and efficacy of therapeutic plasma exchange (TPE) in SPSD is unclear.

Design/Methods

A retrospective review of medical records for patients with SPSD seen at Johns Hopkins Hospital was performed. Patient characteristics, exam

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