infection, intracranial hypotension, and vasculitis (p=0.035). Diffuse involvement was more likely to be smooth, and focal (less than 50% of the entire brain circumference) more likely to be nodular (p=0.35).

Conclusions

To our knowledge, this is the largest single-center study of HP with biopsy results. Imaging features can be suggestive of underlying diagnoses, many of which are inflammatory.

Study Supported By: N/A.
Disclosure: Ms. Son has nothing to disclose. Dr. Li has nothing to disclose. Ceren Tozl has nothing to disclose. Sugeidy Ferreira Brito has nothing to disclose. Michael Miller has nothing to disclose. Dr. Wesley has nothing to disclose.

IVIG Associated Hemolysis In NMOSD Patients

Lingxuan Li, Mark Waheed, Ning Zhong

Objective

N/A.

Background

Intravenous immunoglobulin (IVIG) is commonly used to treat neurological conditions, including CNS demyelination disorders. Although rare, IVIG-induced hemolytic anemia is a potentially serious complication, particularly with high-dose IVIG administration. This complication is often overlooked, leading to treatment delays and unfavorable outcomes. Here, we present two cases of hemolytic anemia following IVIG treatment in patients with neuromyelitis optica spectrum disorder (NMOSD).

Design/Methods

N/A.

Results

Case 1: A 39-year-old male with seronegative NMOSD received immunomodulation therapy with a combination of Rituximab and mycophenolate. Despite this treatment, he continued to experience recurrent symptom flares and was found to have new or active lesions on brain and spine MRI. A high dose of IVIG (total dosage 2g/kg) was administered in three infusions. After the second infusion, the patient developed hematuria and mild anemia but did not experience other systemic symptoms. These complications were transient. Case 2: A 51-year-old female with AQP4-positive NMOSD received IVIG (1g/kg split over two days) in addition to Rituximab therapy. Two days after the infusion, she developed chills, followed by intermittent low-grade fever, progressive fatigue, body pain/aches, and difficulty with ambulation two weeks later. Imaging did not reveal active or new lesions. Laboratory findings showed severe anemia, high reticulocyte count (Table 1), elevated ESR, CRP, and LDH, along with reduced haptoglobin. Peripheral smear indicated mild-to-moderate anisopoikilocytosis. The patient symptoms gradually improved, and along with the normalization of laboratory values. Both patients had AB blood type.

Conclusions

The cases we presented demonstrated diverse clinical presentations and outcomes, highlighting the necessity for attentive monitoring and prompt management of such complications. Clinicians must be aware of risk factors like non-O blood types and should consider alternative treatment approaches or dosage modifications when prescribing IVIG to patients with NMOSD or other CNS demyelination disorders to mitigate the risk of hemolytic complications.

Study Supported By: N/A.
Disclosure: Miss Li has nothing to disclose. Dr. Waheed has nothing to disclose. Dr. Zhong has nothing to disclose.

Syncope as a Presenting Symptom of MuSK-Associated Myasthenia Gravis

Danielle Akinsanmi, Steven Pavlakis

Objective

N/A.

Background

Of the different myasthenia gravis (MG) subtypes, dysautonomia has best been characterized in the thymoma group. Cardiac dysfunction and other abnormalities have also been attributed to co-existing autoimmune disorders such as autonomic ganglionopathy. However, further investigation shows qualitative and quantitative evidence of dysautonomia in multiple subgroups without co-existing antibodies. We draw special attention to MuSK-MG where clinical autonomic complaints are often prevalent.

Design/Methods

Cases: We present two cases of recurrent syncope. The first describes a 58-year-old male who develops syncopal episodes and subclinical symptoms of MG following a prostatectomy. He has several hospital visits for syncope before he is hospitalized for a severe orthostatic episode worsened by what is later determined to be a severe myasthenic crisis. He experiences apnea and cardiac arrest. Following stabilization, his MuSK-MG is treated with multiple rounds of steroids and azathioprine before he is able to make a full recovery. The second case is of a 20-year-old female who experiences a year of vasovagal symptoms and tachycardia. She is eventually admitted for respiratory infection triggered respiratory failure in the setting of what is found to be MuSK-MG. She requires rounds of IVIG for stabilization; family declines alternative therapies.

Results

N/A.

Conclusions

In both cases, additional paraneoplastic and autoimmune antibody testing is negative, as well as investigation for a thymoma. These cases underscore the diagnostic complexity of dysautonomia in MG, particularly in MuSK antibody-positive neuromuscular disease. Autonomic instability in these cases may be related to the unique role of MuSK on the formation and maintenance of the neuromuscular junction. There is still much to be elucidated about the role of MUSK and its antibodies. It challenges the conventional diagnostic framework for neuromuscular disease and emphasizes the usefulness of consideration for MuSK in cases of unexplained syncope followed by mild or atypical symptoms of neuromuscular disease.

Study Supported By: N/A.
Disclosure: Dr. Akinsanmi has nothing to disclose. Dr. Pavlakis has nothing to disclose.

The Patient Journey in Myelin Oligodendrocyte Glycoprotein Antibody Disease (MOGAD): Results From Cross-sectional Patient and Physician Surveys

Michael Levy, Sofia Zappacosta, Silvia Narduzzi, Meheni Khellaf, Mia Unsworth, Emily Trenholm, Friedemann Paul

Objective

Describe the myelin oligodendrocyte glycoprotein antibody disease (MOGAD) patient journey to diagnosis.

Background

MOGAD is an autoimmune rare disease of the central nervous system, with no approved treatment. Patient journey insights are limited.

Design/Methods

The Adelphi MOGAD Disease Specific Program collected real world data from neurologists and their patients through cross-sectional surveys (June to November 2022) across France, Germany, Spain, Italy, the UK and the USA. Neurologists managing ≥1 patients with MOGAD completed a patient record form (PRF) for each patient (≤10 patients). Following informed consent, patients with completed PRFs were invited to fill in a patient self-completion form (PSC). Patients ≥18 years diagnosed with MOGAD were included.

Results

Overall, 74 physicians completed 268 PRFs, with 66 patients completing PSCs. The PRF data showed patients had a median age of 36 years; most were White (85%) and female (65%). Roughly half (136/268, 51%) visited a neurologist as first point of contact because of initial MOGAD symptoms, which were most often optic (182/268, 68%) and/or myelitic (175/268, 65%). Subsequently, 64% (172/268) of patients initially received an alternative diagnosis that was later attributed to MOGAD, most commonly optic neuritis, transverse myelitis or multiple sclerosis. Physician-reported median time from alternative diagnosis to MOGAD diagnosis was 31 days and from first consultation to MOGAD diagnosis was 42 days. Physicians also reported that all patients underwent a MOG antibody test to aid MOGAD diagnosis. Negative serology for AQP4 antibodies was the most common prompt (165/268, 62%) for a MOGAD antibody test. Nearly all patients were ultimately diagnosed by a neurologist (262/268, 98%).

Conclusions

Almost 2/3 of patients initially received an alternative diagnosis that was later attributed to MOGAD, suggesting suboptimal early disease management. Better MOGAD symptom awareness may help to improve the diagnosis pathway and support patient outcomes. Analysis funded by UCB Pharma.

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MRI Characteristics of LGI1- and CASPR2-antibody Encephalitis: Independent Discovery and Validation Cohorts

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Objective

To identify simple MRI characteristics which accurately distinguish two common forms of autoimmune encephalitis (AE); LGI1- and CASPR2-antibody encephalitis (LGI1/CASPR2-Ab-E), from two major differential diagnoses, viral encephalitis (VE) and Creutzfeldt-Jakob disease (CJD).

Background

Rapid and accurate diagnosis of AE encourages prompt immunotherapy initiation, towards improved patient outcomes. However, clinical features alone may not sufficiently narrow the differential diagnosis. Further, awaiting autoantibody results can delay immunotherapy administration.

Design/Methods

Retrospective, cross-sectional, blinded analysis of first available brain MRIs from 192 patients across two tertiary referral centres with LGI1/CASPR2-Ab-E, VE or CJD evaluated for predetermined MRI features by two neuroradiologists (discovery cohort; n=87) and validated in an independent cohort by three neurologists (n=105). Groups were statistically compared with contingency tables.

Results

MRIs from 192 participants were reviewed (n = 71 female, 37%; median age = 66). By comparison to VE and CJD, in LGI1/CASPR2-Ab-E T2/FLAIR-hyperintensities were less likely to extend outside the temporal lobe (7% vs 94% in VE; P<0.001 and 75% in CJD; P<0.01), less frequently exhibited swelling (22% of LGI1/CASPR2-Ab-E vs 59% of VE; P<0.001), and showed no diffusion restriction (0%, vs 73% in VE and 80% in CJD; both P<0.001) and rare contrast-enhancement (5% vs 41% in VE, P=0.013). These findings were validated in an independent cohort and generated an area under the curve of 0.97, sensitivity of 90% and specificity 95% amongst cases with T2/FLAIRhyperintensity in the hippocampus and/or amygdala.

Conclusions

T2/FLAIR-hyperintensities confined to the temporal lobes, without diffusion restriction or contrast-enhancement, robustly distinguish LGI1/CASPR2-Ab-E from key differentials. These observations should assist clinical decision making towards expediting immunotherapy. Their generalisability to other forms of AE/VE should be examined in future studies.

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