well. AIND and HC participant serum samples have been collected to obtain NfL, GFAP, UCH-L1, and Tau protein levels using Quanterix SR-XTM SIMOA. We plan to analyze and compare age and sex matched HC samples to AIND patients for each biomarker, ages ranging from 20-80 years old. Concentrations will be log transformed and analyzed with mixed model regression.

Results

Healthy Controls (130), AIND (255), and newly diagnosed/treatment naïve MS samples (681) have already been collected. Nineteen HC and AIND (includes NMDA, LGI1, TRIM46, CRMP5, MOG, DPPX, GABA-A, GAD-65) participants were analyzed. Preliminary results for HC show mean Nfl (10.3 pg/mL), GFAP (78.9 pg/mL), UCH-L1 (5.65 pg/mL), and Tau (0.53 pg/mL) levels, and for AIND patients mean Nfl (186.48 pg/mL), GFAP (434.92 pg/mL), UCH-L1 (71.38 pg/mL), and Tau (51.85 pg/mL) levels. While ongoing biomarker analysis will be completed with HC that are age and sex matched, the significantly higher levels in AIND patients highlights the importance of creating baseline values in HC to understand these same biomarkers in AIND patients.

Conclusions

Preliminary results show NfL and GFAP levels are significantly higher in AIND patients versus HC. Baseline biomarker values are essential for understanding further research in biomarkers related to AIND.

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A Severe Case of Lupus Cerebritis

Christopher Szewczyk, Hemil Gonzalez

Objective

Lupus cerebritis is a rare but potentially fatal complication of SLE. Prompt diagnosis and rapid initiation of therapy can prove lifesaving.

Background

We present the case of a 31-year-old woman who presented to the emergency department with one week of fevers, headaches and confusion. Review of systems was notable for diffuse muscle and joint pains, weakness and a rash. SLE and Sjogren's were both diagnosed six months prior to admission and treatment included prednisone, methotrexate, and hydroxychloroquine. She self-discontinued therapy six weeks prior to seek natural remedies in her native Ecuador. Vitals on admission were 102.2 F, 111 beats/min, 96/67 mmHg, 16 breaths/min and 95% SO2 on 2 L/min via nasal cannula. Physical exam was notable for conjunctival injection, hypopyon (OD), and oral ulcers with visible bleeding. Pelvic exam showed shallow ulcerations of the vaginal mucosa. Neurological exam was significant for diffuse weakness. There were punched-out ulcers on the digits of both hands. Laboratory markers of lupus disease

activity were markedly abnormal. An MRI of the brain showed numerous punctate foci of restricted diffusion in the supra and infratentorial brain parenchyma. Few of the lesions showed subtle rim-enhancement and microhemorrhagic foci. CSF analysis showed 220 WBC/mL (70% PMNs), glucose 13 mg/dL and protein 63.7 mg/dL. No CSF oligoclonal bands were detected. CSF/serum albumin index showed mild impairment of the blood brain barrier. Cultures of CSF, blood, urine and sputum showed no growth.

Design/Methods
NA.

Results
The patient improved significantly upon initiation of pulsed corticosteroids, plasma exchange, and cyclophosphamide. She was transitioned to steroid-sparing agents and is doing well.

Conclusions
Lupus cerebritis can be the dominant syndrome in a patient presenting with uncontrolled SLE. Imaging and CSF findings can be dramatic and evoke infectious syndromes. Once alternative diagnoses have been ruled out lupus cerebritis should be managed aggressively to ensure good outcomes.

Disclosure: Mr. Szewczyk has nothing to disclose. The institution of Hemil Gonzalez has received research support from NIH.

A Rapidly Fatal Case of Anti-GFAP Receptor Encephalitis Due to Acute Brain Edema and Herniation

Roua Kahila, Zafar Kaleem

Objective
Glial fibrillary acidic protein antibody (GFAP) is a newly recognized biomarker for an immunotherapy responsive autoimmune meningoencephalomyelitis with a wide variety of clinical presentations. We report the second GFAP antibody positive case in a young man who died despite appropriate and aggressive immunomodulatory treatment.

Background
29 year old previously healthy male with childhood immune disorder presented with 3 week history of acute progressive worsening headaches, bloody emesis, nausea, blurry vision and generalized weakness. Exam was significant for downbeating nystagmus, limb ataxia and tremor and later progressing into inattention, confusion, urinary retention, asymmetric pupils, hyprreflexia and lack of motor or sensory response. Lumbar puncture revealed lymphocytic pleocytosis with elevated protein and opening pressure of 36 cm H20. MRI demonstrated areas of restricted diffusions symmetrically involving white matter of the corpus callosum, middle cerebellar peduncle, cerebellar white matter bilaterally as well as within the pons centrally. Patient was started on intravenous immunogammaglobulin (IVIG) and pulse corticosteroids along with broad spectrum antimicrobial therapy. After an initial apparent response to treatment, repeat head CT showed Diffuse Sulci effacement. Shortly after, He rapidly decompensated with clinical findings indicating brainstem herniation, cardiac arrest and brain death was diagnosed. CSF studies subsequently were reported as positive for GFAP antibodies. An autopsy reported the cause of death as cerebellar tonsillar herniation secondary to diffuse cerebral edema. all sections showed perivascular inflammation and gliosis.

Design/Methods
NA.

Results
NA.

Conclusions
This reported case of anti-GFAP meningo-encephalomyelitis is unusual for the rapid onset of cerebral edema and rapid progression to herniation and brain death occurring only 4 weeks after symptom onset. While this may be a rare complication of Anti GFAP encephalitis, clinicians should be vigilant for acutely increased intracranial pressure in patients with clinical findings of encephalitis in general.

Disclosure: Dr. Kahila has nothing to disclose. Zafar Kaleem has nothing to disclose.

Characterization of Retinal Nerve Fiber Layer Thickness in a Cohort with Glutamic Acid Decarboxylase 65 and Glycine Receptor Autoimmunity

Yoji Hoshina, Ka-Ho Wong, Jonathan Galli, John Greenlee, Julia Klein, M. Paz Soldan, Stacey Clardy, Anette Fjeldstad, John Rose, Robert Kadish

Objective
To describe the retinal nerve fiber layer (RNFL) with the demographic and clinical profile in patients with glutamic acid decarboxylase 65 (GAD65) and glycine receptor (GlyR) neurological autoimmunity.

Background
GAD65 and GlyR autoimmunity can cause a wide range of clinical phenomena, including stiff-person spectrum disorder (SPSD) and epilepsy. Both GAD65, through γ-aminobutyric acid-ergic neurons, and GlyR interact in the retina. Optical coherence tomography (OCT) has previously been used in a variety of neurological disorders to establish baseline characteristics and monitor disease course. This presents a noninvasive opportunity to evaluate for a biomarker that may assist with the treatment of these rare but debilitating disorders.

Design/Methods
OCT measures of RNFL by sectors were studied in patients with GAD65 and GlyR neurological autoimmunity and compared to that of 148 healthy control eyes. Patients' baseline characteristics were also reviewed retrospectively from medical records.

Results
Of the 14 patients included in this study, 12 patients were female, and the mean age was 52.6 ± 16.8 (22-79) years when OCT was performed. Ten had GAD65 autoimmunity and 4 had GlyR autoimmunity. Patients with GAD or GlyR autoimmunity showed lower RNFL thickness in multiple sectors compared to the healthy control group. This result was most apparent in the anti-GAD65 antibody subgroup. Eleven patients had SPSD, one patient had epilepsy, and two had non-specific symptoms.

Conclusions
This study provides insight into baseline RNFL thickness in a group with GAD65 and GlyR autoimmunity, two conditions that may produce varied symptoms. While limited by sample size, RNFL thinning was seen in the GAD65 and GlyR autoimmunity groups, and it was most evident in the anti-GAD65 subgroup. This provides a baseline characterization and suggests that future studies should be conducted to determine the utility of OCT as a biomarker for these conditions.

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A Severe Case of Lupus Cerebritis

Christopher Szewczyk and Hemil Gonzalez

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