

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 meningo-encephalomyelitis 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, hyperreflexia and lack of motor or sensory response. Lumbar puncture revealed lymphocytic pleocytosis with elevated protein and opening pressure of 36 cm H₂O. 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 immunoglobulin (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 PSD, 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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Differential Diagnosis in the Management of CPI Immunotoxicity: Case Series of Etiologies not to Miss

Timothy Gregory, Sudhakar Tummala

Objective

To present on treatable conditions arising with exposure to checkpoint inhibiting immunotherapy for malignancy. Each case was diagnostically obscured by presumed immunotoxicity.

Background

Neurological immune-related adverse events (n-irAEs) are rising in incidence with adoption of checkpoint inhibitors (CPIs) for many cancers. 1-3% of patients treated with CPIs experience severe n-irAEs with potential for persistent functional disability or mortality. Diagnosis can be challenging for immunologically vulnerable patients with frequently multifactorial problems from their cancer and potential infectious, metabolic, and iatrogenic complications.

Design/Methods

Three informative cases from a single institution were analyzed.

Results

1. An 80-year old woman with metastatic melanoma and recent treatment with ipilimumab+ pembrolizumab developed acute leg weakness.

Given her EMG and CSF findings, she began treatment for suspected CPI-induced atypical GBS and myositis. Concomitantly she was found to have B12 and folate deficiencies, then gradually improved to baseline with vitamin repletion, steroids, and plasma exchange. 2. A 27-year old woman with metastatic melanoma and recent treatment with ipilimumab+nivolumab developed autoimmune hepatitis and intractable vomiting. Three weeks after she began dabrafenib and trametinib, she developed confusion, diplopia, and ataxia along with weakness and areflexia. She was treated for possible GBS, but was concurrently found to have thiamine deficiency with sequela of Wernicke's encephalopathy on MRI Brain. Her confusion improved with thiamine supplementation but had persistent weakness. 3. A 57-year old woman with lung adenocarcinoma who had progressed on durvalumab began pembrolizumab. Two weeks later, she developed fevers, rash, and lethargy. She was treated supportively but continued to worsen until neurological workup revealed limbic hyperintensities on MRI Brain and CSF pleocytosis with +HSV1. She had minor clinical improvement with acyclovir but remained cognitively debilitated.

Conclusions

Given frequently complex clinical circumstances when working up n-irAEs, a systematic approach and a broad differential must be utilized for this important intersection of cancer neurology and immunology.

Disclosure: Dr. Gregory has nothing to disclose. Dr. Tummala has received personal compensation in the range of \$500-\$4,999 for serving as a Consultant for REVHEALTH LLC.

Giant Cell Arteritis of the Superior Mesenteric Artery Presenting With Wernicke Encephalopathy From Thiamine Deficiency

Sarah Shapiro, David Renner, Ludovica Farese

Objective

N/A.

Background

Giant cell arteritis (GCA) is one of the most common systemic vasculitides in adults over the age of 50 with incidence ranging from 15 to 35 per 100,000 individuals. The disorder is often included in the differential diagnosis of maladies producing atypical facial pain, headache, visual loss, amaurosis fugax, jaw pain, elevated inflammatory markers, and anemia. GCA is typically known to affect cranial arteries with physical exam findings of tenderness to palpation of the temporal arteries and cranial neuropathies. Clinical diagnosis is further supported by new headache, temporal artery abnormality, elevated ESR (= 50 mm/h), and abnormal artery biopsy.

Design/Methods

N/A.

Results

A 68-year-old female with history of primary generalized seizures presented to clinic with a 6-week history of paroxysms of acute confusional episodes, the inability to arise from a seated position due to lower extremity weakness bilaterally, alterations of consciousness without loss of consciousness, severe anorexia, and weight loss. MRI with contrast including Axial FLAIR/T2/Diffusion revealed bilateral pan-lobar cortical and subcortical atrophy with ex-vacuo ventriculomegaly and mild leukoaraiosis in the subcortical white matter tracts. PET-CT body revealed linear uptake involving the aortic root, extending into subclavian arteries bilaterally with segmental involvement of proximal common carotids, and extending inferiorly to the level of the common iliac arteries and the mesenteric arteries. Temporal artery biopsy revealed presence of granulomas with multinucleated giant cells.

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Yoji Hoshina, Ka-Ho Wong, Jonathan Galli, et al.

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