contracted a mild COVID-19 infection two months prior and COVID-19 vaccination one month prior to his symptom onset. His exam was remarkable for bilateral facial weakness, diffuse fasciculations and sensory neuropathy on his trunk and extremities. His diagnostic work up including bone marrow biopsy was consistent with a chronic lymphocytic leukemia (CLL)-like immunophenotype. Cerebrospinal fluid (CSF) analysis was remarkable for five WBC (lymph-dominant) and protein of 74 mg/dl. Serum paraneoplastic panel revealed positive CASPR2 antibody with a titer of 1:100. Magnetic Resonance Imaging (MRI) of the brain showed enhancement of bilateral cranial nerve VII. After lack of clinical response to IV methylprednisone (1 gram for 5 days), patient was treated with a single cycle of IV immunoglobulin (IVIG). He had complete recovery of his symptoms except for residual facial weakness. He remains stable at his six months post-treatment follow-up.

Conclusions

Anti-CASPR2 associated autoimmunity following COVID-19 infection or in the setting of CLL has previously been reported. However, cranial neuropathy in association with CASPR2 antibody has never been. A trial of IVIG could be beneficial in patients with viral-spike protein-induced autoimmunity and CLL who do not otherwise meet the criteria for CLL treatment.

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A Woman With Kelch-like Protein-11 Encephalitis and Unmasked Metastatic Carcinoma

Paunel Agyei, Rajesh Gupta

Objective

To illustrate a case of a woman with rhombencephalitis with antibodies to Kelch-like protein-11 (KLHL11) and a metastatic carcinoma.

Background

KLHL11 encephalitis is an autoimmune paraneoplastic syndrome first described in 2019. The clinical presentation consists of a brainstem cerebellar syndrome with symptoms of hearing loss, diplopia, vertigo and ataxia. This entity has been mostly described in male patients with associated testicular seminomas. Few cases have been described in women. This is a case of a woman with a history of hysterectomy and oophorectomy with KLHL11 encephalitis and an associated aortocaval tumor.

Design/Methods

NA.

Results

The patient was a 62-year-old woman that presented to clinic with a 9-month history of vertigo, progressive bilateral sensorineural hearing loss, diplopia, oscillopsia, ataxia and bilateral tremor. Her MRI brain obtained 8 months after symptom onset showed T2 hyperintense lesions in the inferior cerebellar hemispheres and right medial hippocampus with mild contrast enhancement in these areas. Cerebral spinal fluid analysis showed a lymphocytic pleocytosis, elevated protein, and negative infectious work-up. She completed 5 days of intravenous methylprednisolone and continued a steroid taper. She noted mild to moderate improvement in tremors, gait, and diplopia after steroids. Her symptoms, however worsened as she tapered her steroid dose. Serum KLHL11 antibody levels were positive at a titer of 1:7680. Computed tomography of the chest, abdomen and pelvis did not reveal any evidence of malignancy. However, whole body proton emission tomography/computed tomography (PET CT) revealed a large hypermetabolic aortocaval mass soft tissue mass. A biopsy of the mass showed pathology consistent with a metastatic carcinoma of gynecologic origin for which the patient is undergoing chemotherapy with plans for possible tumor debulking.

Conclusions

This case highlights the importance of considering KLHL-11 encephalitis in female patients presenting with rhombencephalitis, and the need for adequate malignancy evaluation in this disorder.

Disclosure: Dr. Agyei has nothing to disclose. Dr. Gupta has nothing to disclose.

Immunotherapy With Subcutaneous Immunoglobulin or Plasmapheresis in Patients With Postural Orthostatic Tachycardia Syndrome (POTS)

Renee Nelson, Katrina Kesterson, Jill Schofield, Svetlana Blitshteyn

Objective

To assess improvement in autonomic symptoms and functional impairment following immunotherapy with subcutaneous immunoglobulin (SCIG) or plasmapheresis (PLEX) in patients with postural orthostatic tachycardia syndrome (POTS).

Background

POTS is a common autonomic disorder defined by an increased heart rate of at least 30 bpm within 10 minutes of standing or a tilt table test, accompanied by orthostatic intolerance, fatigue, dizziness, and headache. Despite pharmacologic and non-pharmacologic therapy, the marked functional impairment associated with POTS reflects great need for improved treatment. Autoimmunity has emerged as a leading etiology of POTS, with case series describing successful treatment with IVIG. To our knowledge, treatment with SCIG has not been described previously.

Design/Methods

Clinical history of seven patients with POTS treated with SCIG or PLEX was reviewed. Response to treatment was assessed using COMPASS-31 and functional ability scale (FAS) completed retrospectively pre- and 3-12 months post-treatment with SCIG or PLEX. Patients with comorbid defined autoimmune disorders or immune deficiency requiring treatment with immunotherapy were excluded from the study.

Results

Of seven patients, all female, ages 28-57, five received SCIG and two received PLEX. Four had comorbid small fiber neuropathy and five had various positive antibodies at low titers. Across all patients, COMPASS-31 and FAS scores improved an average of 50% and 217%, respectively. Six patients were able to discontinue or reduce oral medications and five reported being able to return to work or school. No serious adverse events were reported.

Conclusions

Patients with POTS experienced significant functional improvement with reduction in autonomic symptoms following immunotherapy with SCIG or PLEX. This case series suggests that SCIG and PLEX may be safe and effective treatments for patients with severe POTS refractory to standard therapies. Randomized controlled trials are needed to determine the efficacy and safety of these long-term therapies.

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Role of Immunotherapy in Down Syndrome Disintegrative Disorder (DSDD)

Nidhiben Anadani, Deepti Chrusciel

Objective

To describe case series of patients with DSDD, successfully treated with immunotherapy including Intravenous Immunoglobulin (IVIG) at a single academic center.

Background

Down syndrome is the most common chromosomal disorder, and in most cases, is due to trisomy of chromosome 21. DSDD is under-recognized, rapidly progressive neuropsychiatric syndrome with various postulated etiology including psychological stress, primary psychiatric disorder and autoimmunity.

Design/Methods

Case-1: A 20-year-old fun loving female with trisomy-21 and infantile spasms started having complex partial seizures, hallucinations, speech regression, tics, abnormal head movement and obsessive-compulsive behavior. Case-2: A 20-year-old female cheerleader with trisomy-21, started having rapid regression in language, cognition, social skills and agitation over one year. Case-3: A 22-year-old female dancer with trisomy-21, started having subacute onset depression, hallucinations, sleep changes, anorexia and speech regression over one year.

Results

Case-1: MRI brain and cerebrospinal fluid (CSF) studies were normal including negative autoimmune encephalitis panel. Serum thyroglobulin and thyroid peroxidase antibody were high. Prolonged oral steroid therapy helped but caused adverse effects. She was able to return to her premorbid baseline with chronic IVIG therapy every 10 weeks. Case-2: MRI brain and CSF were normal. Serum autoimmune encephalitis panel, thyroglobulin antibody and thyroid peroxidase antibody were negative. Pulse IV steroids improved symptoms, however she regressed after stopping steroids. IVIG every 6 weeks along with electroconvulsive therapy improved neurological symptoms. Case-3: MRI brain and EEG were normal. CSF showed elevated white blood cell count. Serum Thyroid antimicrosomal and thyroglobulin antibody were high. One dose of IVIG caused significant improvement in neurological symptoms for 6 weeks.

Conclusions

DSDD should be considered in patients with down syndrome with rapid regression. It is often associated with positive thyroid peroxidase antibody suggesting immune mediated etiology. Various immunotherapy treatments have been reported in literature including steroid, IVIG, mycophenolate and rituximab with significant improvement in selected patient with autoimmunity.

Disclosure: Dr. Anadani has nothing to disclose. Dr. Chrusciel has nothing to disclose.

EEG Characteristics in Hospitalized Patients With Acute COVID-19 Symptoms

Ganesh Murthy, Daniel Fayard, Ryan Chung, Steve Chung

Objective

Our objective was to evaluate the incidence of seizures, pattern of EEG abnormalities, and localization of abnormal discharges in hospitalized patients with COVID-19.

Background

The COVID-19 epidemic has revealed significant neurological manifestations including de novo seizures in patients who do not have a prior history of epilepsy or clear epilepsy risk factors. Our center is located in Arizona, which in the early part of January 2021 had more cases per capita than any other place in the world.

Design/Methods

We performed a retrospective review to observe the electroencephalogram (EEG) patterns of hospitalized adult patients with COVID-19 between March 2020 and February 2021.

Results

We identified 99 patients who were COVID-19 positive and had EEG testing during the same hospitalization. The most common EEG abnormality was diffuse background slowing, which was seen in 63.6% of patients (n = 63/99), compare to 15.1% of focal background slowing. Epileptiform discharges were seen in 11.1% of patients and seizures were found in 5.1% of patients, as newly diagnosed seizures. When combining all focal abnormalities, the most common location for these abnormalities was in the frontal regions 36.4% (n = 8/22). Even though 21 patients had acute focal neuroradiologic findings, only 5 had correlated EEG abnormalities within the same region. When EEG was obtained with suspected seizures (n = 33), 4 cases (12.1%, n = 4/33) indeed showed ictal pattern compared to 1.6% when seizures was not suspected (p = 0.087).

Conclusions

Abnormal EEG findings are most commonly found in the frontal lobe among hospitalized patients with acute COVID-19 symptoms. De novo seizures may be seen with COVID-19 infection. Suspicion of seizures should be raised in patients with COVID-19 encephalopathy. The utility of an EEG may help allow us better insight into how and where the COVID infection affects our central nervous system.

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Progressive Multifocal Leukoencephalopathy Associated With Sarcoidosis: A Multi-Center Case Series

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Immunotherapy With Subcutaneous Immunoglobulin or Plasmapheresis in Patients With Postural Orthostatic Tachycardia Syndrome (POTS)

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