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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