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Case Review of Alternative Treatment in Patient With Immunotherapy Induced Chronic Inflammatory Demyelinating Polyradiculoneuropathy (CIDP)

Melissa Huberman, Oluwagbemiga Larinde

Objective

To emphasize recognition of chronic inflammatory demyelinating polyradiculoneuropathy (CIDP) due to Immune checkpoint inhibitor (ICI) therapy Atezolizumab and efficacy of steroids in treatment.

Background

Atezolizumab is an anti-programmed death-ligand 1 (PD-L1) immune-modulating drug that induces tumor-specific cytotoxic T-cell immunity against the PD-L1 overexpression of many aggressive lung cancers. IVIG was initially thought to have the same benefit in immunotherapy mediated CIDP however recent cases suggest better benefit with plasmapheresis and steroids.

Design/Methods

We report a case of CIDP in a 61-year-old female with high grade neuroendocrine carcinoma of the lung with extensive metastasis. After confirmation of diagnosis, patient began treatment with whole brain radiation therapy and 6 cycles of carboplatin/etoposide followed by atezolizumab maintenance every 21 days for 38 months. Initial treatment reaction included self-resolving loss of balance. Last dose of atezolizumab was given 9 days prior to brain biopsy of left frontal mass. Following biopsy, symptoms of generalized weakness and bilateral lower extremity weakness presented. When symptoms progressed, patient was admitted to hospital, 2-3 weeks after last dose of atezolizumab.

Results

The patient responded to treatment with steroids and cessation of atezolizumab, carboplatin, and etoposide. She recovered to her neurological baseline after a course of IV dexamethasone.

Conclusions

Among ICI therapy, atezolizumab has been less frequently reported with neurologic adverse events than other medications of its class. Increased recognition of the association between ICI and CIDP may improve early detection and improve potentially fatal outcomes. This case and an assessment of the literature indicate ICI induced CIDP may not respond to IVIG and plasmapheresis treatment with consistency. Although IVIG and plasmapheresis maintained efficiency in some reports, adverse outcomes of IVIG treatment may indicate alternative first-line treatment. This case reports successful recovery with the sole use of immunosuppressive therapy which should be considered to optimize future outcomes of ICI associated CIDP.

Disclosure: Miss Huberman has nothing to disclose. Dr. Larinde has nothing to disclose.

Opsoclonus Myoclonus Syndrome and Supraventricular Tachycardia in a Pediatric Patient: A Case Report and Literature Review

Alec Giakas, Sydney Garner, John Korin, Hollie Edwards

Objective

NA.

Background

Opsoclonus myoclonus syndrome (OMS) is a rare movement disorder in children often associated with an underlying neuroblastoma. In other

cases, it is believed that the tumor is occult or there is another immune-stimulating precipitating event. Diagnosis can be difficult, requiring 3 of 4 criteria: opsoclonus or ocular flutter, myoclonus or ataxia, behavioral or sleep disturbances, and neuroblastoma. Prompt treatment of OMS is crucial to preventing permanent neurologic sequelae. In order to better characterize the clinical profile of this syndrome and its associated conditions, we present a case report of a 9 month old male with OMAS without an associated neuroblastoma and with new onset supraventricular tachycardia (SVT) and review the associated literature.

Design/Methods

NA.

Results

A 9-month-old male with a past medical history of macrocephaly, hypotonia, and developmental delay presented with abnormal eye and body movements starting one month prior. Due to concern for OMS, the patient underwent an extensive initial workup including CT neck, chest, abdomen, and pelvis and VHA/HMA levels, which were all within normal limits. The patient's spinal fluid revealed no evidence of paraneoplastic, autoimmune, or infectious processes. Per neurology recommendations, the patient was started on IVIG and dexamethasone. After his first IVIG infusion, the patient's abnormal movements worsened, and his heart rate increased into the 300s. The IVIG was discontinued. However, the patient continued to have recurring bouts of SVT, which was eventually controlled with digoxin. After resuming treatment with IVIG and dexamethasone, the patient's opsoclonus myoclonus symptoms began to improve. All additional metabolic labs resulted normal and the patient was discharged.

Conclusions

Because OMS can cause permanent developmental delay, prompt recognition and treatment of this syndrome is necessary. This is a unique case of OMAS without neuroblastoma, associated with recurrent bouts of SVT. Recognizing rare complications of OMS is crucial to improving medical management of its sequelae.

Disclosure: Mr. Giakas has nothing to disclose. Ms. Garner has nothing to disclose. Mr. Korin has nothing to disclose. Dr. Edwards has nothing to disclose.

GABABR IgG Associated Encephalitis: Clinical Presentations and Measures to Improve Diagnostic Assay Specificity

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Objective

To review the clinical/oncological presentations of gamma aminobutyric acid-B receptor (GABABR)-IgG and evaluate the clinical specificity of antibody testing methodologies.

Background

GABABR-IgG is an intermediate-risk paraneoplastic autoantibody commonly associated with encephalitis and/or seizures.

Design/Methods

GABABR-IgG positive patients tested at Mayo Clinic Neuroimmunology Laboratory were identified. Available archived sera were retested by cell-based assay (CBA) at 1:10 and 1:100 dilutions.

Results

105 GABABR-IgG seropositive patients with clinical details were identified (females, n = 56; median age 63 [range 8-82]). Most patients had one of three anti-GABABR encephalitis/seizure presentations: focal-onset seizures with altered mental status (n = 39), new-onset

refractory status epilepticus (NORSE [n = 23]) or rapidly progressive dementia (n = 14). Fifty-three of 72 patients (74%) with available data had associated malignancy, primarily small cell lung cancer (SCLC). Two P/Q type VGCC-IgG positive patients had isolated Lambert-Eaton Myasthenic Syndrome. Additionally, 27 patients presented with non-specific symptoms or alternate diagnoses (atypical presentation); none had underlying SCLC (13 with malignancy data). Most patients with anti-GABABR encephalitis/seizure presentations had GABABR-IgG detected in CSF (95%, 52/55; of 3 who were negative, 1 had NMDA-IgG and another AMPA-IgG and CRMP5-IgG) and/or GABABR-IgG detected by CBA in serum at a dilution of 1:100 (76%, 31/41). Five encephalitis/seizure patients with GABABR-IgG detected in CSF were CBA positive only at 1:10 dilution in serum. Forty-eight of 76 patients had samples positive on tissue IFA (serum = 17/42, CSF = 40/49). Among 27 patients with atypical presentations, most did not have GABABR-IgG CSF CBA positivity (91%, 21/23) and/or GABABR-IgG 1:100 serum titer by CBA (0/17). None of 27 cases were tissue IFA positive (serum = 0/27, CSF = 0/21).

Conclusions

GABABR-IgG is associated with autoimmune encephalitis/seizure presentations. Our study highlights the importance of evaluating CSF and testing serum at both 1:10 and 1:100 dilutions, as seropositivity at only the 1:10 dilution by CBA without CSF GABABR-IgG positivity has poor clinical specificity.

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A Case of Paraneoplastic NMO With Sarcoma and Unusual Cauda Equina Enhancement

Mohammed Hussein, Danny Samkutty, Sarah Durica, Nidhiben Anadani

Objective

To describe a case of sarcoma associated with NMO presenting with myeloradiculitis.

Background

Neuromyelitis optica is a demyelinating disease of the CNS that predominantly affects the spinal cord and optic nerves. Reports of NMO occurring in the setting of cancer suggest that aquaporin-4 autoimmunity may have a paraneoplastic basis. Here, we describe a patient with metastatic sarcoma who tested positive for aquaporin-4 IgG consistent with NMO.

Design/Methods

66-year-old man with left thigh soft tissue sarcoma with metastasis to the lungs admitted for 2-day history of urinary retention. Neurological exam showed bilateral lower extremity weakness, with decreased patellar reflexes. MRI spine showed cord swelling with enhancement from C6 downwards into the thoracic spine with associated syringomyelia and diffuse enhancement of the cauda equina. MRI brain was normal without abnormal enhancement in the optic nerves. CSF analysis demonstrated elevated protein and 0 oligoclonal bands. Was started on methylprednisolone 1 gram daily for 5 days empirically for transverse myelitis. Extensive serum and CSF workup for other causes of myelopathy including rheumatologic, infectious, and nutritional etiologies were unremarkable until NMO IgG antibody came back positive at 1:10000 titer. He had minimal clinical improvement after methylprednisolone course and so was started on plasmapheresis for five days. His weakness improved slightly but continued to have urinary retention. Discharged to a long-term acute care facility on prednisone 60 mg daily with intermittent bladder catheterization and plan for follow-up with Neuroimmunology.

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