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Glial Fibrillary Acidic Protein (GFAP) Antibody-Associated Astrocytopathy in Systemic Sarcoidosis

Elizabeth Matthews, Ide Smets, Ryan Kammeyer, Maarten Titulaer, Amanda Piquet

Objective

To report two cases of glial fibrillary acidic protein (GFAP) antibody-associated meningoencephalitis in patients with biopsy-proven systemic sarcoidosis.

Background

GFAP astrocytopathy is an autoimmune neurologic disease first defined in 2016. To our knowledge, no association with systemic sarcoidosis has been previously reported.

Design/Methods

Case Series

Results

Patient 1 is a 47-year-old woman with pre-existing pulmonary sarcoidosis treated with steroids and methotrexate with remission 6 years prior. She subsequently developed new-onset epilepsy, progressive ataxia and vertical diplopia. GFAP antibodies were positive in the cerebrospinal fluid (CSF) by cell-based assay (CBA). Body PET scan showed diffuse FDG avidity in her lungs, spleen, and lymph nodes, suggesting simultaneous reactivation of her systemic sarcoidosis. She was treated with steroids followed by infliximab with resolution of her symptoms. Patient 2 is a 58-year-old man with known pulmonary sarcoidosis, who was off immunosuppression at the time of his presentation but had received steroids 17 years prior. He presented with progressive apathy, memory disturbance, dysarthria, and gait instability. MRI revealed widespread T2 hyperintensities. GFAP antibodies were positive in CSF on CBA and confirmed by tissue-based immunofluorescence assay. He received steroids with initial response but relapsed after steroid discontinuation. He improved after restarting steroids and was subsequently transitioned to infliximab with sustained neurologic recovery.

Conclusions

Sarcoidosis is a poorly understood multi-system disorder that is presumably an immune-mediated response to yet unidentified antigen(s). It is known to co-exist with other autoimmune diseases, with autoimmune thyroiditis being most common. GFAP astrocytopathy is also poorly understood. GFAP is found intracellularly and similar to other antibody-mediated diseases against intracellular epitopes, the antibodies are believed to be a biomarker of underlying autoimmunity but not directly pathogenic. We report these cases to highlight a potential association between production of intrathecal GFAP antibodies and systemic sarcoidosis, which may provide insights into the pathogenesis of these two diseases.

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Case of Anti-NMDA Receptor Encephalitis Presenting in a Toddler With Hemorrhagic Cavernomas

Kayla Jacques, Lydia Marcus

Objective

N/A.

Background

N/A.

Design/Methods

Introduction: Anti-N-methyl-D-aspartate (anti-NMDA) receptor encephalitis signifies an autoimmune antibody-mediated neuropsychiatric

disease that often presents with a set of well-described clinical characteristics and other times manifests with more rare features. The heterogeneity of patient presentation can propose a diagnostic challenge to even the best clinical neurologist. Anti-NMDA receptor encephalitis should be considered for patients who possess an alternative existing diagnosis that shows atypical progression because early recognition and treatment of the disease can help reduce long-term complications.

Results

Case Report: We illustrate a 14 month-old previously healthy boy with anti-NMDA receptor encephalitis who first presented with focal seizures. Initial neurologic imaging revealed intracranial hemorrhage with underlying cavernous malformations. He responded well to a single antiseizure agent, but re-presented one week later with transient weakness that was ultimately attributed to worsening intracranial hemorrhage with surrounding edema. Upon his third presentation, he developed dyskinesias, sleep dysfunction, autonomic instability, cognitive changes, and motor regression, prompting further work-up with lumbar puncture. Cerebrospinal fluid analysis showed a positive NMDA antibody titer of 1:40. Treatment with intravenous steroids, plasma exchange (PLEX), and intravenous immune globulin (IVIg), followed by infusions of Rituximab and Cyclophosphamide resulted in gradual, marked clinical improvement.

Conclusions

This case study and literature review explores the relationship between cavernous malformations, intracranial hemorrhage, and anti-NMDA receptor encephalitis, and how these diagnoses respond to escalating immunomodulation therapies. Consideration of this entity should be made when the neurologic examination does not follow an expected course of a previously established diagnosis. With timely recognition and aggressive treatment approaches, patients can achieve substantial clinical improvement.

Disclosure: Dr. Jacques has nothing to disclose. Dr. Marcus has received personal compensation in the range of $500-$4,999 for serving as an Expert Witness for Ragsdale LLC. The institution of Dr. Marcus has received research support from NIAID.

Anti- DPPX Antibody Encephalitis With a Pan-Positive Review of Systems

Mustafa Donmez, Maria Mazzola, Wissam Deeb, Carolina Ionete

Objective

NA.

Background

Subacute encephalitides like anti-DDPX encephalitis are challenging diagnoses due to their unusual presentations. Anti-DPPX encephalitis usually involves gastrointestinal, nervous, and respiratory systems. Common symptoms include diarrhea, weight loss, cognitive dysfunction, amnesia, myoclonus, tremor, and exaggerated startle response.

Design/Methods

NA.

Results

A 48-year-old man with no significant past medical history was referred to the neurology clinic to evaluate his tremor. The patient's first symptoms included alternating diarrhea and constipation, weight loss, fatigue, generalized pain, chest pain, dyspnea, and leg cramps. Several emergency departments, primary care physicians, and specialty clinic visits did not yield a diagnosis. During the first neurology clinic visit, he reported new urinary symptoms, chills, insomnia, tremor, anxiety, and depression. His physical exam revealed exaggerated physiological tremor only. During the second neurology clinic visit and resultant urgent admission, he reported new symptoms of forgetfulness, inattention, muscle jerks, numbness in the extremities, and seizure-like episodes. His exam was remarkable for mild left-sided rigidity and MOCA of 23/30 with impairment in delayed recall. The patient's MRI brain with and without contrast and spot EEG were unremarkable. CSF studies were negative except for protein elevation with lymphocytic pleocytosis. The patient's autoimmune CSF panel was positive for anti-DPPX antibodies, though available only after discharge. The patient was urgently seen in the neuroimmunology clinic and received a steroid course and monthly rituximab. The patient is improved with the treatments. His repeat MOCA 28/30 and returned to working full-time.

Conclusions

Anti-DPPX antibody encephalitis has been reported in a handful of cases in the literature. It has a subacute presentation with often a delay to diagnosis–an average diagnosis time of 8 months after the onset of symptoms. We present the case to raise awareness about anti-DPPX encephalitis, especially its typical clinical triad of GI dysmotility, cognitive decline, and myoclonus/tremor.

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Acute Autoimmune Encephalitis With Features of Bickerstaff Brainstem Encephalitis (BBE) and Two Abnormal Autoantibodies Presenting With Prominent Cerebellar Abnormality on MRI–A Case Report

Osman Ozel, Eugene Lai, Maya Ramy, Joseph Masdeu, Christine Rizk, Meryim Pourshеykhi, Abdulmunaim Eid, Sanaa Karim, Sara Benitez, Belen Pascual, Timea Hodics

Objective

To present an unusual cerebellar imaging finding of a patient with clinical features of BBE

Background

BBE is characterized by progressive ataxia, ophthalmoplegia and impaired consciousness. Magnetic resonance imaging (MRI) of the brain is usually normal. However, rare T2 Flair changes have been reported. Scarcity of cerebellar findings on imaging led to the controversy of peripheral vs central etiology for the ataxia. Despite other modalities including positron emission tomography, magnetic resonance spectroscopy and molecular level evidence pointing towards involvement of the cerebellum, MRI is usually unrevealing.

Design/Methods

A 62-year-old woman presented with acute onset ataxia with multiple falls, dysarthria, diplopia, and blurred vision that started 3 days prior to presentation. She had left face angioedema couple days after her flu inoculation 6 weeks prior to presentation. Her exam revealed normal mental status, scanning speech, bilateral dysmetria and ataxia as well as left-sided facial palsy, square wave jerks and hyperreflexia. CSF showed 30 RBC, 17 WBC, normal glucose, and elevated protein of 68 mg/dl. No infectious etiologies were identified. MRI brain showed infratentorial leptomeningeal enhancement with T2 hyperintensities in the both cerebellar hemispheres. Anti-GQ1b antibodies were 51 IV (negative < 30 IV) and anti-GAD65 antibodies were also weakly positive only in serum, 0.12 nmol/L (negative < 0.02 nmol/L).

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Kayla Jacques and Lydia Marcus

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