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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

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

Elizabeth Matthews, Ide Smets, Ryan Kammeyer, et al.

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