Background

Anti-myelin oligodendrocyte glycoprotein (MOG) antibodies have been commonly associated with optic neuritis, myelitis, and acute disseminated encephalomyelitis but rarely with tumefactive lesions, especially in children. We report a young child with MOGAD presenting with a tumefactive cerebellar demyelinating lesion.

Design/Methods

A retrospective chart review

Results

A 3-year-old developmentally appropriate boy with fever for five days prior presented for gait changes and a self-resolved seizure lasting less than 5 minutes. Neurologic examination showed abnormal finger to nose on the left side, weakness of the left lower extremity and an ataxic gait. The differential at the time was Todd's paralysis versus an intracranial process. MRI showed a non-enhancing, ill-defined T2 hyperintense tumefactive lesion with mass effect within the left cerebellum concerning for tumor, abscess, or demyelination. On EEG, a lack of a well sustained and modulated posterior dominant rhythm and lack of a well-developed anterior to posterior gradient, with moderate background slowing was seen. Cerebrospinal fluid showed 8 white blood cells, 0 red blood cells, 55 mg/dl glucose, 31 mg/dl protein, 0.61 IgG index, and 0 oligoclonal bands. The Mayo Clinic cell-based assay detected anti-MOG IgG antibody in the serum with titer of 1:100. Neurological symptoms gradually improved after steroid pulse therapy.

Conclusions

This report highlights the novel spectrum of radiologic manifestations associated with MOGAD in pediatric patients and MOGAD should always be considered in the differential diagnosis of tumefactive lesions.

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Myelin oligodendrocyte glycoprotein antibody-associated disease (MOGAD) as a novel presentation of CNS autoimmunity in a pediatric patient with Wiskott-Aldrich syndrome (WAS)

Vivien Xie, Alexandra Kornbluh

Objective

Report a novel case of myelin oligodendrocyte glycoprotein antibody-associated disease (MOGAD) presenting as relapsing bilateral optic neuritis in a pediatric patient with Wiskott-Aldrich syndrome (WAS).

Background

WAS is a rare X-linked primary immunodeficiency caused by mutations in the WAS gene that leads to increased susceptibility to infections, thrombocytopenia, eczema, malignancies, and autoimmunity. Known CNS autoimmune manifestations include cerebral vasculitis, but optic neuritis, CNS demyelination, and MOGAD have not been previously reported.

Design/Methods

Chart review

Results

A 5-year-old boy with a history of chronic immune thrombocytopenia, hypogammaglobulinemia, anemia, and focal epilepsy developed binocular vision loss. MRI of the brain demonstrated enlargement of bilateral optic nerves with marked enhancement of the nerve sheaths consistent with optic neuritis, as well as multiple small enhancing supratentorial lesions. He was treated with pulse methylprednisolone followed by oral steroid taper, and he returned to baseline with no reported residual visual deficits. Five months later, he experienced a relapse of bilateral vision loss, and repeat MRI re-demonstrated bilateral optic neuritis as well as resolution of prior brain lesions. He was treated with repeat course of steroids and experienced moderate improvement in his vision. Rituximab was then initiated to prevent further relapses of optic neuritis while treating his chronic suspected immune-mediated thrombocytopenia. Myelin oligodendrocyte glycoprotein antibody (MOG-IgG) via serum fluorescence-activated cell sorting assay was positive (titer 1:100), confirming a diagnosis of MOGAD. At age six, molecular panel testing for genes associated with primary immunodeficiency identified a missense WAS gene variant. He was subsequently found to have decreased WAS protein expression consistent with a diagnosis of WAS.

Conclusions

We describe a case of pediatric MOGAD presenting with multiphasic bilateral optic neuritis in a patient with WAS. This case expands the reported spectrum of CNS autoimmunity associated with WAS and may help to inform indications for therapeutic options such as bone marrow transplant.

Disclosure: Dr. Xie has nothing to disclose. Dr. Kornbluh has nothing to disclose.

MOGAD in the Mountain West: Epidemiology and Outcomes in Pediatric and Adult Patients at Two Large Academic Referral Centers

Melissa Wright, Suzanne Liu, Ka-Ho Wong, Christopher Mizenko, Ryan Kammeyer, Teri Schreiner, Robert Kadish, Tammy Smith, Jonathan Galli, Julia Klein, John Greenlee, John Rose, M. Paz Soldan, Jeffrey Bennett, Joshua Bonkowsky, Lisa Peterson, Amanda Piquet, Stacey Clardy

Objective

To describe the characteristics and outcomes in adult and pediatric patients diagnosed with myelin oligodendrocyte glycoprotein antibody-associated disease (MOGAD) at the two major referral centers in the Mountain West of the United States, a geographic area encompassing roughly 15% of the land mass of the continental US.

Background

Since the development of commercial assays, MOGAD has become increasingly recognized as an etiologic diagnosis for several CNS demyelinating phenotypes, yet the epidemiological characteristics, relapse rates and outcomes of large populations are not well-described

Design/Methods

A retrospective chart review for patients within the health systems at the University of Utah and the University of Colorado, and affiliated children's hospitals, was conducted. To identify MOGAD patients, we queried the ICD10 codes corresponding to demyelinating disease of CNS, neuromyelitis optic spectrum disease, optic neuritis, transverse myelitis, and acute disseminated encephalomyelitis. These patients were then cross matched against antibody testing results and existing research databases at each institution. Search dates included 1/1/2016-12/1/2021 to encompass the period of commercially available MOG IgG testing. Patients were cross referenced with a list of positive MOG IgG assays at each institution.

Results

We describe the characteristics of over 50 patients (adults and children) with MOGAD, including age of onset, gender, symptoms at onset, associated autoimmunity, antibody titers, response to therapies and relapse rates.

Conclusions

This is a comprehensive characterization of a diverse population of pediatric and adult MOGAD patients seen at the two major referral

hospitals in the Mountain West. The treatment regimens and outcomes in this population may inform approaches to current management and future clinical trials.

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A Case of Recurrent Cerebral Cortical Encephalitis in MOG Antibody-Associated Disease

Laura Cacciaguerra, John J. Chen, Eoin P. Flanagan

Objective

NA.

Background

Cerebral cortical encephalitis is a recently recognized syndrome of myelin-oligodendrocyte-glycoprotein-antibody-associated disease (MOGAD), yet most descriptions report single episodes without recurrence.

Design/Methods

Case report of recurrent cerebral cortical encephalitis in MOGAD.

MOGAD in the Mountain West: Epidemiology and Outcomes in Pediatric and Adult Patients at Two Large Academic Referral Centers

Melissa Wright, Suzanne Liu, Ka-Ho Wong, et al.

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