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.

Disclosure: Dr. Sanghi has nothing to disclose. The institution of Dr. Gombolay has received research support from CDC. The institution of Dr. Gombolay has received research support from NIH. Tuba Khan has nothing to disclose.

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.

Disclosure: Dr. Wright has nothing to disclose. The institution of an immediate family member of Dr. Liu has received research support from NIH. The institution of Mr. Wong has received research support from Biogen Idec. Mr. Mizenko has nothing to disclose. The institution of Dr. Kammeyer has received personal compensation in the range of $500-$4,999 for serving as a Consultant for Genentech. The institution of Dr. Schreiner has received personal compensation in the range of $500-$4,999 for serving as a Consultant for CDC. Dr. Schreiner has received personal compensation in the range of $500-$4,999 for serving on a Scientific Advisory or Data Safety Monitoring board for Biogen. Dr. Schreiner has received personal compensation in the range of $500-$4,999 for serving as an Expert Witness for Mary C Bush, LLC. The institution of Dr. Schreiner has received research support from Biogen. The institution of Dr. Schreiner has received research support from Roche Genentech. Dr. Kadish has received personal compensation in the range of $500-$4,999 for serving on a Scientific Advisory or Data Safety Monitoring board for Genentech. The institution of Dr. Kadish has received research support from Alexion Pharmaceuticals. Dr. Smith has nothing to disclose. Dr. Galli has nothing to disclose. An immediate family member of Ms. Klein has received personal compensation for serving as an employee of Amgen. An immediate family member of Ms. Klein has received personal compensation in the range of $500-$4,999 for serving on a Speakers Bureau for Amgen. Dr. Greenlee has received personal compensation in the range of $500-$4,999 for serving as an Editor, Associate Editor, or Editorial Advisory Board Member for Medlink. Dr. Greenlee has received publishing royalties from a publication relating to health care. Dr. Greenlee has received publishing royalties from a publication relating to health care. The institution of Dr. Rose has received research support from National Multiple Sclerosis Society. The institution of Dr. Rose has received research support from Guthy Jackson Charitable Foundation. The institution of Dr. Rose has received research support from NIH. The institution of Dr. Rose has received research support from Friend's of MS. The institution of Dr. Rose has received research support from Biogen. Dr. Rose has received intellectual property interests from a discovery or technology relating to health care. Dr. Paz Soldan has received personal compensation in the range of $500-$4,999 for serving as a Consultant for TG Therapeutics. The institution of Dr. Paz Soldan has received research support from National Institutes of Health. The institution of Dr. Paz Soldan has received research support from National Multiple Sclerosis Society. The institution of Dr. Paz Soldan has received research support from Western Institute for Biomedical Research. The institution of Dr. Paz Soldan has received research support from Biogen. The institution of Dr. Paz Soldan has received research support from Novartis. The institution of Dr. Paz Soldan has received research support from Clene Nanomedicine. Dr. Bennett has received personal compensation in the range of $10,000-$49,999 for serving as a Consultant for Horizon Therapeutics. Dr. Bennett has received personal compensation in the range of $10,000-$49,999 for serving as a Consultant for Alexion. Dr. Bennett has received personal compensation in the range of $10,000-$49,999 for serving as a Consultant for Genentech-Roche. Dr. Bennett has received personal compensation in the range of $500-$4,999 for serving as a Consultant for TG Therapeutics. Dr. Bennett has received personal compensation in the range of $500-$4,999 for serving as a Consultant for Reistone Bio. Dr. Bennett has received personal compensation in the range of $5,000-$9,999 for serving as a Consultant for Abbvie. Dr. Bennett has received personal compensation in the range of $0-$499 for serving as a Consultant for Novartis. Dr. Bennett has received personal compensation in the range of $500-$4,999 for serving as a Consultant for Chugai. Dr. Bennett has received personal compensation in the range of $500-$4,999 for serving as a Consultant for Mitsubishi Tanabe. Dr. Bennett has received personal compensation in the range of $500-$4,999 for serving on a Scientific Advisory or Data Safety Monitoring board for Clene Nanomedicine. Dr. Bennett has received personal compensation in the range of $500-$4,999 for serving on a Scientific Advisory or Data Safety Monitoring board for Roche. Dr. Bennett has received personal compensation in the range of $500-$4,999 for serving as an Expert Witness for Parker,Whiteford Taylor. Dr. Bennett has received personal compensation in the range of $5,000-$9,999 for serving as an Expert Witness for Podoll. Dr. Bennett has received personal compensation in the range of $5,000-$9,999 for serving as an Expert Witness for Hogan Lovells. The institution of Dr. Bennett has received research support from Novartis. The institution of Dr. Bennett has received research support from Alexion. Dr. Bennett has received intellectual property interests from a discovery or technology relating to health care. Dr. Bennett has received publishing royalties from a publication relating to health care. Dr. Bonkowsky has received personal compensation in the range of $500-$4,999 for serving as a Consultant for Bluebird bio. Dr. Bonkowsky has received personal compensation in the range of $500-$4,999 for serving as a Consultant for Neurogene. Dr. Bonkowsky has received personal compensation in the range of $500-$4,999 for serving as a Consultant for Passage Bio. Dr. Bonkowsky has received personal compensation in the range of $0-$499 for serving as a Consultant for Takeda. Dr. Bonkowsky has received personal compensation in the range of $500-$4,999 for serving on a Scientific Advisory or Data Safety Monitoring board for Autobahn. Dr. Bonkowsky has received stock or an ownership interest from Orchard. The institution of Dr. Bonkowsky has received research support from NIH. An immediate family member of Dr. Bonkowsky has received intellectual property interests from a discovery or technology relating to health care. Dr. Bonkowsky has received publishing royalties from a publication relating to health care. The institution of Dr. Peterson has received research support from Kronus. The institution of Dr. Piquet has received personal compensation in the range of $500-$4,999 for serving as a Consultant for Genentech. Dr. Piquet has received personal compensation in the range of $500-$4,999 for serving on a Scientific Advisory or Data Safety Monitoring board for Genentech. Dr. Piquet has received personal compensation in the range of $500-$4,999 for serving on a Scientific Advisory or Data Safety Monitoring board for Alexion. The institution of Dr. Piquet has received research support from Rocky Mountain MS Center. The institution of Dr. Piquet has received research support from Novartis. The institution of Dr. Piquet has received research support from Abbvie. The institution of Dr. Piquet has received research support from Roche/Genentech. The institution of Dr. Piquet has received research support from NYU. Dr. Piquet has received publishing royalties from a publication relating to health care. Dr. Piquet has received publishing royalties from a publication relating to health care. Dr. Piquet has received personal compensation in the range of $5,000-$9,999 for serving as a Litigative Consultant with US-Dept HHS/DICP. Dr. Clardy has received personal compensation for serving as an employee of Veterans Health Administration (VHA). Dr. Clardy has received personal compensation for serving as an employee of University of Utah Health. Dr. Clardy has received personal compensation in the range of $0-$499 for serving as a Consultant for Clarion. Dr. Clardy has received personal compensation in the range of $500-$4,999 for serving as a Consultant for ExpertConnect. The institution of Dr. Clardy has received personal compensation in the range of $0-$499 for serving as a Consultant for VielaBio. The institution of Dr. Clardy has received personal compensation in the range of $0-$499 for serving as a Consultant for Genentech. The institution of Dr. Clardy has received personal compensation in the range of $500-$4,999 for serving as a Consultant for Alexion. The institution of Dr. Clardy has received personal compensation in the range of $0-$499 for serving as a Consultant for GuidePoint. Dr. Clardy has received personal compensation in the range of $10,000-$49,999 for serving as an Editor, Associate Editor, or Editorial Advisory Board Member for Neurology/AAN Publications. The institution of Dr. Clardy has received research support from Alexion Pharma. The institution of Dr. Clardy has received research support from Sumaira Foundation for NMO. The institution of Dr. Clardy has received research support from Immune Deficiency Foundation. The institution of Dr. Clardy has received research support from Western Institute for Veteran Research. The institution of Dr. Clardy has received research support from NIH/NINDS. Dr. Clardy has received personal compensation in the range of $500-$4,999 for serving as a AAN Summer Meeting CoDirector Travel and Lodging with AAN. Dr. Clardy has received personal compensation in the range of $500-$4,999 for serving as a Grand Rounds Travel and Lodging with U of Iowa. Dr. Clardy has received personal compensation in the range of $500-$4,999 for serving as a Speaker Honoraria for Grand Rounds with Barrow Neurological Institute.

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.

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 and Alexandra Kornbluh

Neurology 2022;99;S18
DOI 10.1212/01.wnl.0000903172.52024.08