Results

This is a case report of a 32-year-old patient who initially developed meningoencephalitis when he was three. He was stable until age 31, when he developed headaches followed by status epilepticus secondary to cerebral cortical encephalitis with accompanying unilateral right temporal cerebral cortical T2-hyperintensity with leptomeningeal enhancement. Cerebrospinal fluid analysis revealed 505 white cells/μl (normal, 0-5) but oligoclonal bands were negative. Over the subsequent two weeks he developed bilateral severe vision loss (20/400 bilaterally) accompanied by bilateral optic disc edema, and MRI orbits showed bilateral anterior segment optic nerve enhancement extending >50% of the length of each nerve, confirming bilateral optic neuritis. MOG-IgG was tested in serum and was positive at a titer of 1:100. He was treated with intravenous methylprednisolone 1 gram daily for 5 days and a subsequent slow oral prednisone taper along with anti-epileptic treatment with levetiracetam, later transitioned to oxcarbazepine, although he later self-discontinued all anti-epileptics. He received maintenance intravenous immunoglobulins (IVIg) 0.4 g/Kg weekly, that was later weaned to 1 g/Kg every 4 weeks. He then developed recurrent cerebral cortical encephalitis 18 months after his prior episode manifesting with seizures and left occipito-temporal T2-hyperintensity and swelling with leptomeningeal enhancement. He was treated with high dose IV steroids, an oral prednisone taper and his IVIg dose was changed back to 0.4 g/Kg once weekly and anti-seizure medications reinitiated.

Conclusions

Cerebral cortical encephalitis in MOGAD can be recurrent. Close scrutiny of the MRI in patients with MOGAD who develop seizures after prior cerebral cortical encephalitis is important as it can discriminate a recurrent cerebral cortical encephalitis attack from a breakthrough seizure related to prior damage.

Disclosure: Ms. Cacciaguerra has received personal compensation in the range of $500-$4,999 for serving on a Scientific Advisory or Data Safety Monitoring board for Roche. Ms. Cacciaguerra has received personal compensation in the range of $500-$4,999 for serving on a Speakers Bureau for BMS Celgene. Ms. Cacciaguerra has received personal compensation in the range of $500-$4,999 for serving on a Speakers Bureau for Sanofi. John Chen has nothing to disclose. The institution of Dr. Flanagan has received personal compensation in the range of $500-$4,999 for serving on a Scientific Advisory or Data Safety Monitoring board for Alexion. Dr. Flanagan 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. Flanagan has received personal compensation in the range of $500-$4,999 for serving on a Scientific Advisory or Data Safety Monitoring board for Horizon Therapeutics. Dr. Flanagan has received personal compensation in the range of $500-$4,999 for serving on a Speakers Bureau for Pharmacy times. The institution of Dr. Flanagan has received research support from Viela Bio. Dr. Flanagan has a non-compensated relationship as a Member of medical Advisory Board with The MOG Project that is relevant to AAN interests or activities. Dr. Flanagan has a non-compensated relationship as a Editorial board member with Journal of The Neurologic Sciences that is relevant to AAN interests or activities. Dr. Flanagan has a non-compensated relationship as a Editorial board member with Neuroimmunology Reports that is relevant to AAN interests or activities.

Development of New or Enlarging MRI Lesions Outside of Clinical Attacks in MOG-Antibody-Associated Disease

Stephanie Syc-Mazurek, John Chen, Padraig Morris, Elia Sechi, Jayawant Mandrekar, Jan-Mendelt Tillema, Alfonso Lopez, Claudia Lucchinetti, Nicholas Zalewski, Laura Cacciaguerra, Marina Buciuc, Karl Krecke, Steven Messina, M. Tariq Bhatti, Sean Pittock, Eoin Flanagan

Objective

To determine the frequency of new/enlarging T2 or enhancing asymptomatic lesions in myelin-oligodendrocyte-glycoprotein-antibody-associated-disease (MOGAD) and compare to multiple sclerosis (MS) and aquaporin-4 antibody-positive-neuromyelitis-optica-spectrum-disorder (AQP4+NMOSD).

Background

Data on new asymptomatic lesions in MOGAD is limited.

Design/Methods

We retrospectively identified Mayo Clinic MOGAD patients with inclusion criteria of: 1)MOG-IgG positivity by live-cell-based-assay; 2) Fulfilling current MOGAD diagnostic criteria; 3) Baseline and follow-up paired MRIs without interval attacks. Paired MRIs (baseline and follow-up) were categorized as either attack-to-remission or remission-to-remission scans. A neurologist and neuroradiologist reviewed MRIs (T2-FLAIR brain, T2 spine, and T1-post-gadolinium brain and spine) to identify new/enlarging lesions. A subset of MOGAD patients matched for follow-up interval were compared to MS and AQP4+-NMOSD patients.

Results

We included 105 MOGAD patients (median age, 31 years[range, 3-80]; 60% female) with 373 paired MRIs (brain, 213, spine 160). In total, 13/373 (3%) scans (10/105 patients) had one or more new/enlarging T2-lesions (brain, 12/213[5.6%]; spine, 1/160[0.6%]) and 8/367 (2%) had enhancing lesions. New spinal lesions were rare across all groups (0-4%). T2 lesions occurred more commonly in attack-remission scans (8/171[4.7%]) then remission-remission scans (5/202[2.4%]). Clinical characteristics did not differ between patients who developed new/enlarging lesions and those who did not. Maintenance immunosuppressants were used in 44/105 (42%) patients. New/enlarging lesions did not predict future clinical relapse. New brain lesions were less in MOGAD (1/25[4%]) than MS (14/26[54%], $p < 0.0001$) but did not differ from AQP4+NMOSD (1/13[8%], $p = 1.0$) in subgroup analysis.

Conclusions

New brain MRI lesions rarely develop outside of attacks in MOGAD which differs from MS. Surveillance MRI in MOGAD may have limited utility as a surrogate biomarker of disease activity in clinical practice and for clinical trials.

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Recurrent Brainstem Lesions as a Manifestation of MOGAD

Rumyar Ardakani, Kyle Blackburn

Objective

NA.

Background

MOG antibody-associated disease (MOGAD) is an inflammatory and demyelinating disease of the central nervous system. Brainstem and cerebellar involvement in MOGAD is relatively common and typically occurs in conjunction with lesions in other parts of the central nervous system such as the optic nerve, supratentorial white matter, or spine. Isolated attacks of the brainstem and cerebellum are rare–especially in a relapsing form.

Design/Methods

A case report.

Results

We present the case of a 60 year-old female who developed new onset weakness and incoordination of her left arm and leg. Her brain MRI revealed restricted diffusion and edema involving the left cerebellar peduncle. She was diagnosed with a stroke and discharged home on antiplatelet therapy. One month later, she developed a left facial droop and was diagnosed with a Bell's palsy. She received oral steroids and had improvement of her facial droop, though it returned a few weeks after completion of her oral steroids. A repeat brain MRI at that time revealed enlargement of the left cerebellar lesion with extension into the left pons. She was again treated with oral steroids and her symptoms improved. She presented again 3 months later with dysarthria and pseudobulbar affect with her brain MRI revealing diffuse T2/FLAIR hyperintensities and edema throughout the bilateral pons and cerebellar peduncles. She received 3 days of high-dose IV methylprednisolone with marked improvement. She ultimately had serum MOG antibody testing performed which returned positive (titer of 1:1000). She was initiated on rituximab, and was clinically stable for over 24 months.

Conclusions

Although brainstem involvement in MOGAD usually occurs in conjunction with other lesions, our case demonstrates that relapsing attacks isolated to the brainstem and cerebellum are within the clinical spectrum of MOGAD.

Disclosure: Dr. Ardakani has nothing to disclose. Dr. Blackburn has received personal compensation in the range of $500-$4,999 for serving on a Scientific Advisory or Data Safety Monitoring board for Genentech.

Spinal Central Canal Dilation in MOG Antibody-Associated Disease Versus Other CNS Demyelinating Disorders

Lauren Webb, Laura Cacciaguerra, John Chen, Elia Sechi, Vyanka Redenbaugh, Divyanshu Dubey, Sean Pittock, Eoin Flanagan

Objective

To assess the frequency of spinal cord central canal dilation on magnetic resonance imaging (MRI) in patients with myelin-oligodendrocyte glycoprotein antibody-associated disease (MOGAD) myelitis compared to myelitis patients with aquaporin-4-positive-neuromyelitis optic spectrum disorder (AQP4+NMOSD) and multiple sclerosis (MS).

Background

In MOGAD myelitis, a sagittal T2-hyperintense line accompanied by axial T2-signal restricted to the spinal cord gray matter forming an H-sign have been recognized and occur more frequently than in AQP4+NMOSD and MS myelitis. Pseudo-dilation of the ependymal canal has also been highlighted in cases of MOGAD myelitis, but detailed studies of this are lacking.

Design/Methods

The spinal cord MRIs of myelitis patients with MOGAD (n = 63), AQP4+NMOSD (n = 37), and MS (n = 26) were evaluated for central canal dilation, defined as T2-hyperintensity with similar consistency to cerebrospinal fluid within a myelitis T2-lesion. Clinical data were collected from the medical record for MOGAD patients. The expanded disability status scale (EDSS) was used to quantify disability at nadir.

Results

The median EDSS score at nadir for the MOGAD myelitis patients was 6 (range: 0-8). MOGAD patients experienced weakness (55/63[87%]), urinary retention/incontinence (50/63[79%]), numbness/paresthesias (48/63[76%]), and stool incontinence/constipation (36/63[57%]). Central canal dilation was more frequent in patients with MOGAD (23/63[37%]) than MS (0/26[0%]); $p < 0.001$) but did not differ from AQP4+NMOSD (14/37[38%]; $p = 0.89$). Spinal canal dilation resolved on follow-up axial MRI for most MOGAD (29/34[85%]) and AQP4+NMOSD (13/14[93%]) patients.

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Stephanie Syc-Mazurek, John Chen, Padraig Morris, et al.

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