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Anti-Myelin Oligodendrocyte Glycoprotein Antibody Positivity in Setting of Aqp4-Seronegative Encephalomyelitis

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Abstract

Myelin oligodendrocyte glycoprotein (MOG) is a protein found on the outer surface of oligodendrocytes. Although not clearly understood, it is thought to play a role in myelination of the central nervous system. Antibodies against MOG result in an inflammatory demyelinating process. Clinically, this manifests as an array of symptoms including optic neuritis (unilateral or bilateral), seizures, encephalopathy and myelitis which extends longitudinally over three or more vertebral segments. The lumbar cord and conus medullaris are predominantly involved, which sets Anti-MOG apart from other demyelinating disorders. To date, only a handful of cases of positive anti-MOG antibody with viral prodromal, and aquaporin 4 seronegative related encephalomyelitis has been reported. We have reviewed the data regarding Anti-MOG in the context of demyelinating spectrum disorders. Our review focuses on the potential role of anti-MOG being a separate entity that overlaps clinically and phenotypically with acute disseminated encephalomyelitis (ADEM) and neuromyelitis optica spectrum disorder (NMOSD).

Keywords: Anti-MOG; Encephalomyelitis; Demyelinating disorder

Introduction

Anti-MOG is a fairly new entity with extensive overlap in clinical presentation and pathophysiology with other demyelinating disorders such as acute disseminated encephalomyelitis (ADEM) and neuromyelitis optica spectrum disorders (NMOSD). We present a case of a 16-year-old female who developed an extensive demyelinating encephalomyelitis. Through this case, we seek to highlight both similarities and differences between anti-MOG and other disorders.

Case Report

A 16-year-old female who presented with a chief complaint of altered mental status and fever. A week prior to presentation, she was seen at urgent care for peri-orbital edema of the right eye, fever and URI symptoms. She had a negative strep test and was discharged home with topical antibiotics [1]. This was followed by a severe headache with nausea. She became obtunded and incoherent. On presentation, she was combative, twitching, and incontinent of bladder and bowels. A lumbar puncture (LP) was performed which showed an opening pressure of 43cm H2O, protein 69mg/mL, glucose of 60, 168 WBCs and 12 RBCs. She was started on broad spectrum antibiotics. Her initial brain MRI showed diffusion restriction within the left temporoparietal subcortical white matter in the periventricular regions (Figure 1A). EEG showed intermittent rhythmic theta discharges but no electrographic seizures. Due to persistent encephalopathy, a LP was repeated; protein 65, glucose 63, RBC 6, and WBC 136 (25 neutrophils, 75 mononuclear) [2]. CSF samples was sent for EBV panel, CMV, West Nile, bartonella antibodies, Lyme antibodies, anti-NMDA, paraneoplastic panel, and limbic encephalitic panel. Three days later, her MRI brain was repeated, which showed T2 hyperintensity in the medulla/pons and edema in the basal ganglia (Figure 1B).

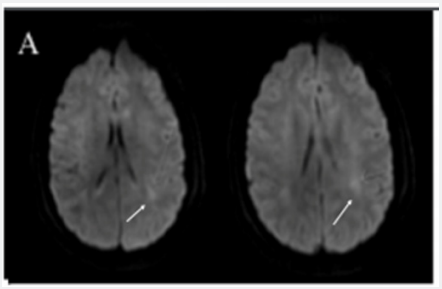

Figure 1A: Initial MRI showing DWI restriction within the subcortical white matter in the periventricular regions.

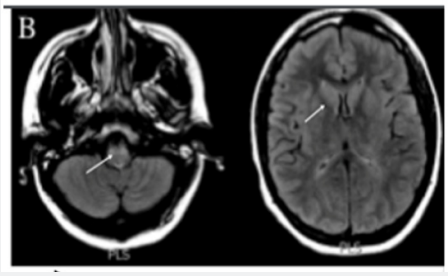

Figure 1B: Day 3 MRI showing T2 hyperintensity in the medulla/pons and edema in the basal ganglia.

On day 8, she began moving her upper extremities spontaneously, however, was plegic in her lower extremities. MRI of her spine showed signal hyperintensity involving the gray matter, particularly anterior horn cells extending from C6 through T8 without evidence of associated abnormal enhancement (Figure 1C). She was started on methylprednisolone 500 mg BID for 5 days followed by 1-2 mg/kg/day taper dose with oral prednisolone taper [3]. Her serum was sent for thyroid studies, myeloperoxidase, ACE, dsDNA, ANCA panel, HTLV I/II neg, HIV, quantiferon gold and vitamin B12 and E. Given no improvement, patient was started on IVIG treatment, and subsequently plasmapheresis. On Day 20, she was found to be anti-MOG positive; titers 1:40. On day 28, patient was discharged to rehab facility to complete a 4-week steroid taper. At her 3 month follow up she had improved remarkably, and she ambulated using a walker. Her mentation was normal and she had regained bowel and bladder control (Figure 1D).

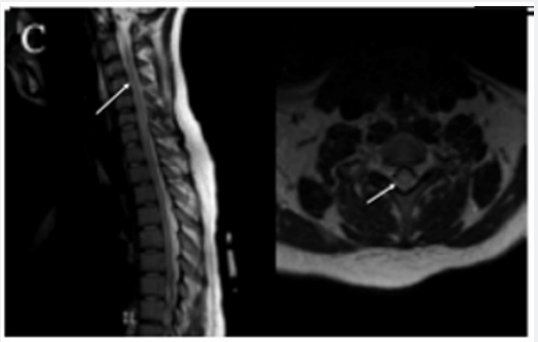

Figure 1C: MRI of cervical/thoracic spine showing hyperintensities involving the gray matter and anterior horn cells.

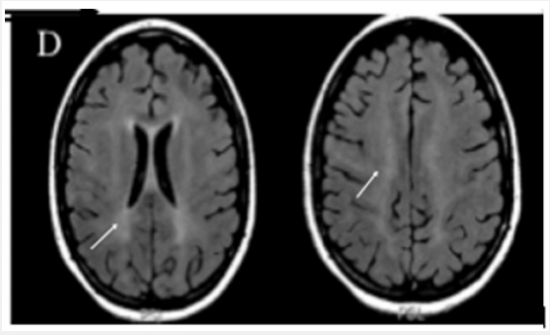

Figure 1D: Repeat MRI on day 16 showing confluent subcortical hyperintensities.

Discussion

Pediatric demyelinating disorders constitutes an umbrella of disease processes that overlap in presentation, symptoms and clinical course. Examples include ADEM, multiple sclerosis (MS), neuromyelitis optica (NMO), and optic neuritis. In this population, demyelination typically follows an acute viral illness and or vaccination, and can be either monophasic or have a relapsing course [4]. ADEM, like MS, is a clinical diagnosis with no associated seromarker. Myelin oligodendrocyte glycoprotein antibodies has been shown to be present in approximately 40% of those diagnosed with ADEM, and about 47% of anti-MOG cases are preceded by a viral illness. Additionally, anti-MOG antibodies has been detected in those who clinically satisfy the criteria for NMOSD, however, are negative for the aquaporin-4 antibody. This constitutes roughly 25% of NMOSD cases. Perhaps, these cases represent a separate disease entity given antibodies specific to anti-MOG. Notably, the presence of anti-MOG antibody is associated with a good clinical recovery. These antibodies are more readily detected in serum compared to CSF. In comparison, NMOSD is associated with antibodies targeting aquaporin-4 (AQP4) which is present on astrocytes, not oligodendrocytes [5].

Anti-MOG and MS have very similar histopathology. Given this overlap with other entities, testing for MOG antibodies is essential as its treatment and clinical course differ [6]. Treatment of demyelinating diseases associated with MOG antibodies are IVIG or PLEX followed by an oral prednisone taper over 4 to 6 months. Relapses are more common in females, those older than 10 years at onset, and those with a baseline titer greater than 1:1,280 [1]. Follow up is warranted, and serum/CSF studies should be repeated to assess for the presence of anti-MOG antibodies. If present, immunosuppressive agents such as azathioprine, rituximab or mycophenolate mofetil should be considered. Despite significant overlap with other demyelinating disorders, due to its distinctive features, antibody target, clinical course and prognosis, anti-MOG should be set apart as its own clinical entity [7]. Therefore, testing for this antibody should be considered on a more routine basis if there is compelling evidence of an underlying inflammatory or demyelinating disorder.

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