



A Rare Case of MOG Antibody Disease

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Abstract

Myelin oligodendrocyte glycoprotein antibody disease (MOGAD) consists of a group of inflammatory demyelinating disorders. Myelin oligodendrocyte glycoprotein antibody-associated disease (MOGAD) covers a wide spectrum of manifestations and is defined by the presence of MOG seropositivity. However, in a proportion of patients, there may be an overlap in some of the clinical and radiological manifestations between MOGAD and multiple sclerosis (MS) or Neuromyelitis optica. Being wary of this entity is critical to ensure appropriate therapy. We present a case report of MOGAD, who along with longitudinally extending transverse myelitis had optic neuritis.

Keywords: case report, myelin oligodendrocyte glycoprotein antibody, multiple sclerosis, transverse myelitis, optic neuritis, NMO.

Introduction

Myelin oligodendrocyte glycoprotein antibody - associated disease (MOGAD) represents a group of inflammatory demyelinating disorders characterized by the presence of Immunoglobulin G (IgG) antibodies to myelin oligodendrocyte glycoprotein (MOG). Over the recent years, it is becoming evident that MOGAD represents a distinct clinical entity separate from acute disseminated encephalomyelitis (ADEM), neuromyelitis optica spectrum disorder (NMOSD), and multiple sclerosis (MS).¹

Here we present a case report of a patient presenting with transverse myelitis and optic neuritis and diagnosed as MOG Antibody disease.

Case Report

A 24 year old female came with complaints of neck pain followed by weakness of all 4 limbs and numbness below the neck for a duration of 3 days. She had H/o difficulty in raising bilateral upper limb above the head, H/O difficulty in mixing food, H/o difficulty in standing, H/O difficulty in rolling over the bed .H/o numbness present below neck and H/o blurring of vision present .She also had h/o difficulty in bladder control for initial 2 days which resolved spontaneously. No history of fever, seizures, trauma. Vitals were stable. On examination visual acuity was reduced to counting fingers in the left eye. Fundus examination showed optic neuritis in the left eye. Power was 3/5 in all four limbs. Bilateral plantar was extensor. Deep tendon reflexes were diminished. Spinothalamic and

posterior column sensations were diminished. Other system examination was found to be normal. Routine blood investigations including total counts was found to be normal. Lumbar puncture was done and sent for analysis. It showed Glucose: 53mg/dl, Protein: 23mg/dl, Wbc:134 cells/cu.mm, ADA:6.8IU/l, No growth with lymphocytic predominance (90%).ANA was negative. NMO [AQ4] Antibodies was Negative.CSF for Oligoclonal bands was negative. Serum ACE levels was 6.1IU/L.MOG Antibody was positive. Contrast MRI of brain showed no significant abnormality in the brain parenchyma (Figure 1.1and 1.2). Contrast MRI of spine showed T2/STIR high signal intensity noted along the cervical cord from cervico medullary junction upto D3 vertebra with cord expansion suggestive of Longitudinally extensive transverse myelitis (Figure 2.1 and 2.2).Visually evoked potential showed amplitude and latency reduced in the left eye suggestive of optic neuritis .Patient was treated with pulse steroids for 5 days followed by oral steroids.

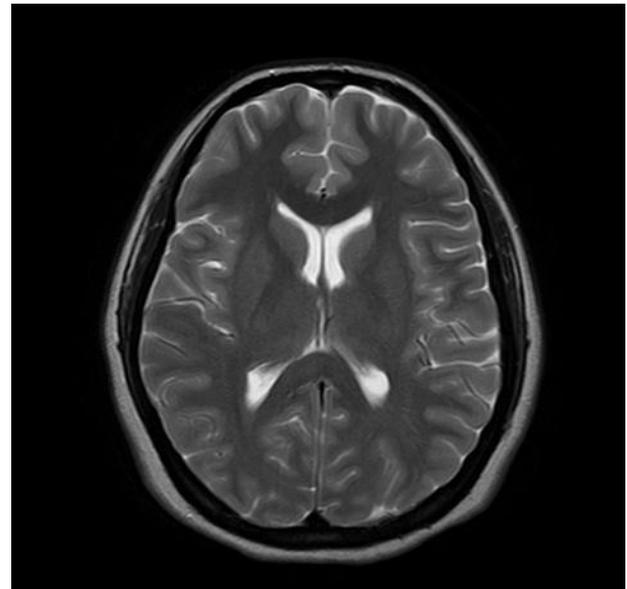
**Figure 1.1****Figure 1.2****Figure 2.1**



Figure 2.2

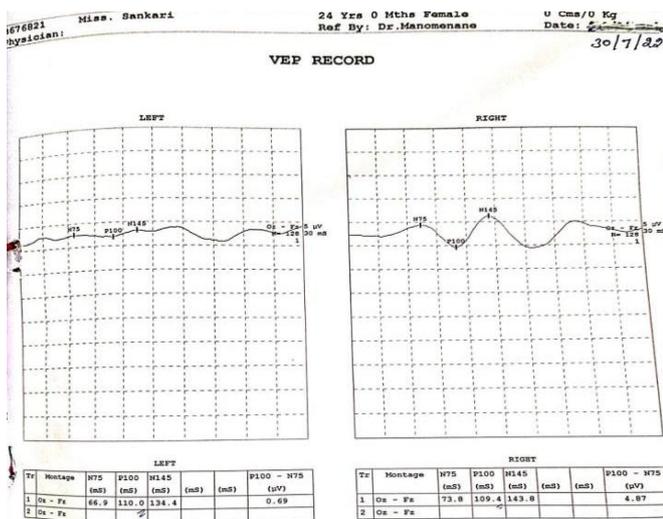


Figure 3.1

Discussion

MOG is a glycoprotein on the myelin surface and present exclusively in the central nervous system. MOGAD is a disease of oligo dendrocytes wherein astrocytes are spared. Median age of onset fourth decade of life, with only a slight predominance in women. MOGAD is an inflammatory demyelinating condition characterised by monophasic or relapsing course of neurological dysfunction which does not meet the criteria for Multiple sclerosis and at lower risk than AQP4 positive patients for further relapses.

Optic neuritis (ON) is the most common initial presentation of MOGAD in adolescence and adulthood, and a frequent presentation in pediatric patients. It is associated with a higher risk of subsequent relapse compared to other clinical presentations. At the onset, vision loss is often severe and up to 80% of patients have bilateral optic nerve involvement, which is highly unusual in MS. Bilateral ON with disc edema can be mistaken for idiopathic intracranial hypertension especially if the patient also complains of headache and has elevated opening pressure on lumbar puncture; however lymphocytic pleocytosis in CSF and enhancement of optic nerve on orbital MRI point toward an inflammatory etiology and should prompt testing for MOGAb. MRI of the orbits during acute MOG-ON typically shows longitudinally extensive optic nerve enhancement with a predilection for the anterior portion of optic nerves, the chiasma and optic tracts are less frequently affected. "Optic perineuritis," characterized by inflammation of the optic nerve sheath and surrounding structures on MRI, is seen in up to 50% of cases of MOGAD-ON. In some cases of MOG-TM, there is an antecedent history of infection or vaccination, but in most patients, no such history can be elicited.

Radiographically, MOG-TM is usually associated with a longitudinally extensive lesion spanning 3-4 vertebral segments. First, cord lesion of MOG-TM during the acute phase are much less likely to demonstrate gadolinium enhancement than in NMOSD. Secondly, spinal cord lesions in MOGAD can be multifocal. MOG-TM affects both gray and white matter of the cord. The involvement of gray matter can manifest as linear hyperintensity of the central spinal canal "pseudo-dilation" or as H-shaped T2-hyperintensity that outlines the anterior and posterior horns (H-sign).

MOGAD can be associated with cases of Acute Disseminated Encephalomyelitis in children and AQP4 seronegative NMO. Brain MRI can be normal or show fluffy areas of increased signal change in white or grey matter. Thalamic and pontine lesions are more common. Conus

involvement is highly specific. More than half of the patients have cervical or thoracic lesions. **Diagnostic Criteria:** (International Panel of Experts -Jarius et al). Monophasic or relapsing acute Optic neuritis, myelitis, brainstem encephalitis or any combination of these symptoms; MRI or electrophysiological findings compatible with CNS demyelination; Seropositivity for MOG -IgG as detected by means of cell based assay employing full length human MOG as target antigen. Treatment includes High dose glucocorticoids followed by low dose prednisone taper, sometimes Plasmapheresis. Brain lesions respond rapidly to treatment. Recurrence following discontinuation of prednisone can occur and become glucocorticoid dependent .Off label empiric treatment- Rituximab, IV Ig, Mycophenolate Mofetil.

Our patient was a young female with first episode of transverse myelitis with unilateral optic neuritis with no typical serological or radiological features suggestive of Multiple sclerosis or Neuromyelitis optica and was diagnosed and treated to be as Myelin Oligodendrocyte Antibody Associated disease. It is vital to differentiate between the three for starting and continuation of appropriate treatment however there can always be an overlap or a change over from one disease to the other in due course of time in the same patient.

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