# Tumefactive Demyelination—A Rare Presentation of Anti-MOG Syndrome

Sir,

Anti–myelin oligodendrocyte glycoprotein (MOG) syndrome is an immune-mediated central nervous system demyelinating disorder with a myriad of clinical presentations, most common ones being Acute disseminated encephalomyelitis (ADEM), optic neuritis, and myelitis. Very rarely, they can present with large tumefactive demyelinating lesions that mimic glioma and cause diagnostic challenge to the treating physician. Identifying autoantibodies in these patients is pivotal in taking treatment decisions. We present a case of anti-MOG syndrome presenting as tumefactive demyelination with excellent steroid response.

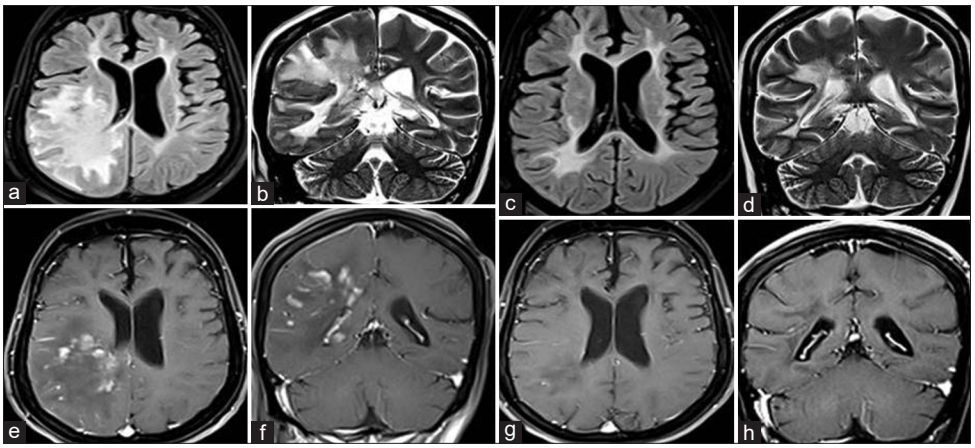


**Figure 1:** (a) MRI Axial FLAIR and (b) Coronal T2W images showing ill-defined heterogeneous hyperintense lesion involving right temporoparietal white matter with adjacent perilesional edema and mass effect. (e) Postcontrast T1 axial and (f) coronal images showing patchy incomplete ring enhancement pattern. MRI brain 6 weeks after treatment, (c) Axial FLAIR and (d) coronal T2W images showing near-complete resolution of the lesion. (g) Postcontrast T1 axial and (h) coronal images showing no enhancement

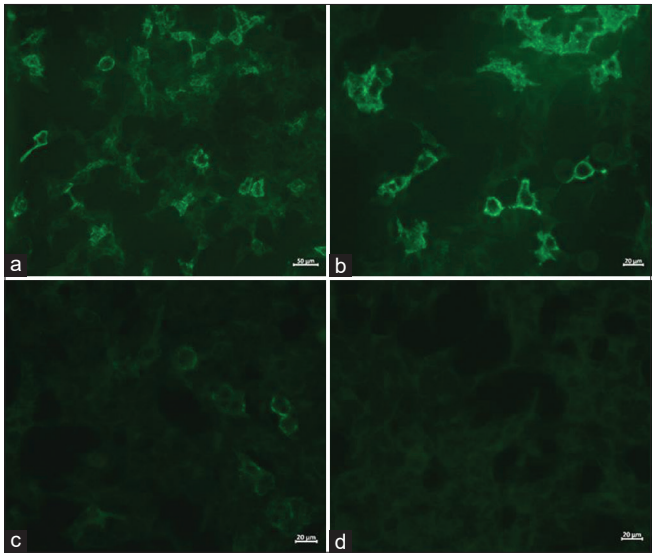


**Figure 2:** Cell-based assay with the test serum showing presence of antibodies against MOG. The intensity of fluorescence was strong positive at (a) and (b) 1:10 dilution, (c) weak positive at 1:100 dilution, and (d) negative at 1:1000 dilution. [magnification = scale bar; a 50 μm, b-d 20 μm]

A 44-year-old lady presenting with 1-week history of progressive left upper and lower limb weakness, without any craniobulbar symptoms or symptoms of raised intracranial pressure. She had previous history of bilateral visual loss 10 years prior to current presentation, which resolved completely in 6 weeks with medical management.

On evaluation, she had visual acuity of 6/9 both eyes, normal pupillary reaction, left hemiparesis, and dysarthria. Clinical differentials considered were stroke, primary demyelinating disorder like multiple sclerosis, and space occupying lesion like glioma. Magnetic resonance imaging (MRI) brain showed an ill-defined T2/FLAIR heterogenously hyperintense lesion involving right temporoparietal white matter with perilesional edema. Patchy areas of plaque like enhancement seen with open ring like pattern around the lesion [Figure 1]. Radiological appearance was most favoring tumefactive demyelination; however, a high grade glioma was also kept as a differential diagnosis.

Routine blood investigations and CSF study were essentially normal. Owing to the previous history of bilateral optic neuritis, there was a high suspicion of demyelinating disorder like neuromyelitis optica or anti-MOG syndrome. Serum was evaluated for antibodies against aquaporin-4 and MOG. Commercially available fixed cell based assay kit employing HEK293 transfected cells (Euroimmun, Lübeck, Germany) was used and the test was performed as per the manufacturer's instructions using appropriate controls. At a starting dilution of 1:10, the test serum was positive for antibodies against MOG and negative for antibodies against aquaporin-4. Strong positive reaction against MOG was noted at 1:10 serum dilution, which became weak positive at 1:100 and negative at 1:1000 dilutions [Figure 2]. Based on the titration, the anti-MOG antibody titre was determined as 1:100.

Following treatment with pulsed intravenous methylprednisolone and maintenance oral steroids, she showed dramatic improvement in neurological status. By 6 weeks, she could walk independently with no neurological deficits and repeat MRI brain showed near-complete resolution of the lesion [Figure 1].

## Discussion

The myriad of clinical presentations of anti-MOG antibody syndrome ranges from ADEM- like presentation in young children to opticospinal presentation in adults.[1] Anti-MOG syndrome presenting as tumefactive demyelination (TDL) is rare with only seven cases in the literature.[2–6] TDL associated with anti-MOG syndrome can be seen as an initial presentation or it can appear during the course of the disease. TDL is defined as demyelinating lesions (2 cm or greater)

with possible mass effect, which are often mistaken for tumour-like space occupying lesions and have a characteristic radiographic appearance.[7] MR imaging features that are found more frequently in patients with TDL than in those with brain tumour are incomplete ring enhancement, mixed T2-weighted iso- and hyperintensity of enhanced regions, absence of cortical involvement, and absence of mass effect.[7] The histopathologic characteristics of patients with anti-MOG syndrome presenting as TDL showed predominantly multiple sclerosis (MS) pathological pattern II (perivascular lymphocyte cuffing with additional complement and antibody deposition) or pattern III (distal oligodendrogliopathy and oligodendrocyte apoptosis).[6]

Our case widens the spectrum of clinical presentations of anti-MOG syndrome and prompts the treating physician to test for anti-MOG antibodies in a case presenting as TDL. Differentiating TDL from glioblastoma is essential as the pathology will dictate the treatment and the long-term prognosis. TDL responds very well to steroids with near-complete recovery within weeks.

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## Conflicts of interest

There are no conflicts of interest.

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