Letters to the Editor

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.
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 heterogeneously hyperintense lesion involving right temporoparietal white matter with adjacent 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.[5-6] 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)
Letters to the Editor

Dear Editor

Brain histopathology and clinical course of MOG-antibody-associated idiopathic epilepsies do not show any apparent cognitive clinician. and does not change the management decisions of the in most of these cases, it does not require any medication subclinical impairment in neuropsychological tests, but epilepsy or electroclinical syndromes also demonstrate some Not only the patients of JME, but patients with other idiopathic patients for their comprehensive management. We wish to Hence, detailed higher mental function tests supplemented for anti-MOG antibodies in a case presenting as TDL. 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.

Financial support and sponsorship
Nil.

Conflicts of interest
There are no conflicts of interest.

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Submitted: 05-Mar-2021 Accepted: 06-Mar-2021 Published: 21-Apr-2021

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DOI: 10.4103/aiian.AIAN_185_21