Case Report

Anti Myelin Oligodendrocyte Glycoprotein associated Immunoglobulin G (AntiMOG-IgG)-associated Neuromyelitis Optica Spectrum Disorder with Persistent Disease Activity and Residual Cognitive Impairment

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Abstract

Antibodies targeting myelin oligodendrocyte glycoprotein (MOG) have been recently reported in association with idiopathic inflammatory central nervous system disorders. Initially believed to be a benign disorder, anti MOG-IgG was noted to cause steroid responsive recurrent optic neuritis and isolated longitudinally extensive myelitis. However, there is growing evidence that the disease may be predominantly relapsing, often producing severe visual loss and involving regions other than the spinal cord and optic nerve. We report an adolescent male with an aggressive disease course previously undescribed in anti MOG-IgG-associated disease that left him with residual cognitive dysfunction.

Keywords: Anti MOG-IgG, encephalitis, neuromyelitis optica spectrum disorder

Introduction

Idiopathic "non multiple sclerosis" inflammatory demyelinating central nervous system (CNS) diseases constitute a heterogeneous group of disorders. In earlier publications,[1] we have shown that approximately 30% of these patients were seropositive for aquaporin 4-immunoglobulin G (anti AQP4-IgG) closely followed by 20% of patients who were positive for anti MOG-IgG. Recurrent optic neuritis and isolated transverse myelitis were the common disease phenotypes identified.[2,3] Recent reports have indicated that without appropriate intervention, the disease may be more severe than previously thought leaving residual neurological deficits.[4] The patient we describe in this report is one such example and had manifestations previously unreported with anti MOG-IgG related disease.

Case Report

A 17-year-old male developed rapidly progressive quadriparesis with urinary retention, for which he was admitted and evaluated. Magnetic resonance imaging (MRI) showed features [Figure 1a] suggestive of longitudinally extensive transverse myelitis. Brain MRI was unremarkable. Cerebrospinal fluid (CSF) showed mild pleocytosis and marginally elevated protein. He received 5 days of intravenous (IV) methyl prednisolone (1 g daily) followed by an oral taper for 6 weeks and recovered completely. Three months later, he was admitted to our center with headache accompanied by vomiting, altered sensorium, and generalized convulsions. A second MRI was done [Figure 1b-d]. He was given another course of IV steroids followed by IV IgG (0.4 g/kg body weight/day × 5 days) following which he gradually improved. During this admission, a repeat lumbar puncture was done and CSF was evaluated (EUROIMMUN-Autoimmune Panel 1) for anti-N-methyl-D-aspartate receptor antibodies,

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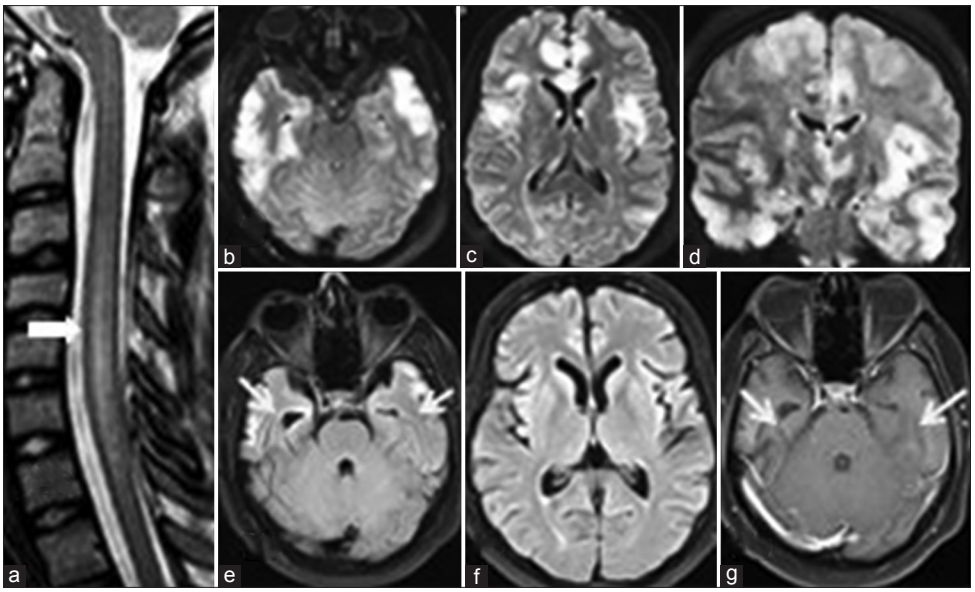

Figure 1: (a) Longitudinally extensive myelitis (sagittal T2W) in cervical cord. (b-d) (Axial and coronal fluid-attenuated inversion recovery) bilateral extensive fluid-attenuated inversion recovery/T2 hyperintense lesions in the cortex of bilateral temporal and paramedian frontal regions with subcortical extension. Review scans after 3 months showing partial resolution of lesions, dilatation of temporal horns (arrows), and sulcal prominence, (e and f) suggesting brain volume loss and persistent gadolinium enhancement (g) of the lesion (arrows)

anti-voltage-gated potassium channel antibodies, anti-alpha-amino-3-hydroxy-5-methyl-4-isoxazole propionic acid receptor antibodies, and anti-γ-aminobutyric acid-B receptor antibodies, all of which were negative. He was also investigated for CNS infections (including herpes simplex virus, dengue, and tuberculosis). Thyroid function was normal and antithyroid antibody and anti AQP4-IgG at Tohoku University at coauthors laboratory) was positive in serum. At discharge, he was dull, inattentive and had dysarthria and gait incoordination. Although he was advised immunosuppressants (azathioprine 150 mg/day and 30 mg of prednisolone), he discontinued after a short period. He was reviewed 3 months later when a detailed cognitive evaluation revealed delayed reaction time, poor attention span, and impaired verbal fluency with perseveration. A repeat MRI [Figure 1e-g] showed partial resolution with subtle evidence of persistent disease activity. He was initiated on injection rituximab with intent to continue every 6 months.

Discussion

Encephalitic illness associated with anti MOG-IgG has been previously described in very young children in the 4–8 years age group[5] and less often in adolescence. Steroid responsive anti MOG-IgG associated encephalitis was recently reported among adults.[6,7] These patients had a monophasic illness with a benign outcome. MRI of the brain shows abnormality in more than one-third of anti-MOG-IgG-associated disease from the onset,[2,4] but symptomatic brain lesions are less common.[4]

Our patient, an adolescent male, presented with fulminant encephalitis as part of a relapsing neuromyelitis optica spectrum disorder[8] and was positive for anti MOG-IgG. He had disease persistence on MRI and residual cognitive impairment 3 months after the second attack. Anti MOG-IgG-associated disorders may be more severe than previously thought. This patient who had an aggressive disease course is such an example. The persistent disease activity and residual brain dysfunction he developed underscore the need for long-term immunosuppression in anti MOG-IgG associated illness with a relapsing disease course.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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