Anti-myelin oligodendrocyte glycoprotein antibody associated neuromyelitis optica spectrum disorder presenting homonymous hemianopia

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Abstract

Anti-aquaporin 4 antibody is the most common cause of neuromyelitis optica spectrum disorder (NMOSD) and anti-myelin oligodendrocyte glycoprotein (MOG) antibody recently has emerged as another cause of NMOSD. Visual field defect can be observed as a manifestation of optic neuritis in patients with multiple sclerosis and NMOSD. However, homonymous hemianopia associated with a visual cortex lesion has been rarely reported in patients with multiple sclerosis and anti-aquaporin 4-positive NMOSD. Recently, we experienced a case of MOG-positive NMOSD who presented with a homonymous hemianopia associated with a visual cortex lesion.

Keywords: Neuromyelitis optica, myelin oligodendrocyte glycoprotein, hemianopia

INTRODUCTION

Neuromyelitis optica spectrum disorder (NMOSD) is an autoimmune disease to present various neurological manifestations including optic neuritis, transverse myelitis, area postrema syndrome, narcolepsy, and other symptoms associated with brainstem or hemispheric lesions.

Anti-aquaporin 4 (AQP4) antibody is the most common cause of NMOSD and anti-myelin oligodendrocyte glycoprotein (MOG) antibody recently has emerged as another cause of NMOSD. Visual field defect can be observed as a manifestation of optic neuritis in patients with multiple sclerosis (MS) and NMOSD. However, homonymous hemianopia associated with a visual cortex lesion has been rarely reported in patients with MS and AQP4-positive NMOSD. Recently, we saw a case of MOG-positive NMOSD who presented with a homonymous hemianopia associated with a visual cortex lesion.

CASE REPORT

A 21 years old man presented with the right visual field defect the day before. Neurologic and ophthalmologic examinations revealed a right homonymous hemianopsia without evidence of papilledema. A generalized tonic-clonic seizure occurred just after arriving at the emergency room. The seizure lasted for 60 sec and ceased after the administration of intravenous lorazepam. Brain magnetic resonance imaging (MRI) exhibited an increased T2-signal intensity and gadolinium-enhancement in the left medial temporal and occipital lobes (Figure 1-A, B). Gadolinium-enhanced lesion was also observed in the right optic nerve and sheath (Figure 1-C). However, he did not complain of the symptom of right optic neuritis. Electroencephalography exhibited an intermittent focal delta slowing in the left temporal region without epileptiform discharges. Full field visual evoked potential (VEP) showed a poor wave formation and delayed P100 latency in both sides (Figure 1-D). Complete blood count and blood chemistry were normal. Serologic tests including anti-AQP4 antibody, anti-nuclear antibody, anti-double stranded DNA antibody, anti-neutrophil cytoplasmic antibody, lupus-anticoagulant, anti-cardiolipin antibody, and anti-phospholipid antibody were negative. HIV and syphilis screening tests were also negative. Cerebrospinal fluid test revealed a pleocytosis (WBC 40 /mm³, lymphocyte 80%) with normal range of sugar and protein.

He had a history of left optic neuritis 4 months ago. Orbit MRI exhibited an enhanced gadolinium enhancement in the left optic nerve (Figure 1-E, F). IgG anti-AQP4 was also negative at that time. Full field VEP exhibited an absence of P100 wave in the left side (Figure 1-H). The visual impairment...
responded well to high dose intravenous steroid and oral steroid tapering therapy.

We referred the patient for an IgG anti-MOG antibody assay in another institute and the result was positive. He was treated with intravenous high dose of steroid and oral levetiracetam. The neurologic deficits responded well to steroid and the lesions in the follow-up brain MRI also resolved (Figure 1-G). He has been on treatment with mycophenolate mofetil for 6 months after discharge and did not have recurrence of epileptic seizures or other symptoms.

DISCUSSION

Several studies have reported that about 25-30% of patients with AQP4-negative NMOSD are positive for anti-MOG antibody. Anti-MOG antibody has also been detected in few patients with MS and in some children with acute disseminated encephalomyelitis. Patients with AQP4-positive NMOSD were mostly negative in anti-MOG antibody test. The histopathologic findings in MOG-positive and AQP4-positive NMOSDs are known to be different. The former exhibit the well-demarcated areas of loss of myelin, relative preservation of axons and astrocytes, numerous lipid-laden macrophages containing myelin debris, and inflammatory infiltrates with predominately perivascular T- and B-cells and terminal complement, while the latter exhibit the astrocyte loss, secondary demyelination, IgG and terminal complement deposition, and inflammatory infiltrate with neutrophils and eosinophils.

Recently, the clinical and radiological distinctions between patients with AQP4-positive and MOG-positive NMOSDs have been elucidated. In a few MOG cohort studies, patients with MOG-positive NMOSD tend to be younger at onset, less likely to develop in women, and more common in Western populations compared to patients with AQP4-positive NMOSD. Bilateral simultaneous optic neuritis, more rapid and complete recovery of optic neuritis, and longer lasting visual field defect are the characteristics of optic neuritis in patients with MOG-positive NMOSD. Involvement of the conus medullaris, a higher frequency of focal myelitis and better
Clinical outcomes are also features of transverse myelitis in patients with MOG-positive NMOSD. Encephalopathy and epileptic seizures have also been reported to occur more frequently in patients with MOG-positive NMOSD than in patients with AQP4-positive NMOSD.\(^1,7\)

In our patient, the left optic neuritis during the first attack was clinically evident, whereas the right optic neuritis during the second attack was clinically silent, though the MRI and VEP have demonstrated the right optic neuritis. Sequential bilateral optic neuritis with different stage can be seen in both AQP4- and MOG-positive NMOSDs. The fact that the patient is a young man, good response to steroid, seizures and encephalopathy are closer to MOG-positive NMOSD than AQP4-positive NMOSD.\(^1,7\) Homonymous hemianopia associated with a lesion in visual cortex is an unusual clinical manifestation in patients with AQP4-positive NMOSD and MS even though visual field defect associated with optic neuritis is common. Central scotoma, altitudinal, quadrant, three quadrant, hemianopia, and bitemporal hemianopia can be observed in patients with NMOSD than MS.\(^1,2,4,5\)

Our patient showed some of the distinctive clinical features indicating MOG-positive NMOSD including male gender, bilateral optic neuritis, acute symptomatic seizure, encephalopathy, and well response to steroid. The homonymous hemianopia associated with a lesion in visual cortex may possibly also be a clinical feature suggestive of MOG-positive NMOSD rather than AQP4-positive NMOSD and MS.

**DISCLOSURE**

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**REFERENCES**