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Anti-Myelin Oligodendrocyte Glycoprotein Syndrome with Findings Resembling "Snake-Eye Appearance": a Case Report

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Case Report

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Anti-myelin oligodendrocyte glycoprotein (anti-MOG) syndrome is an immune-mediated inflammatory condition of the central nervous system, which usually involves spinal cord and optic nerves. Herein, we studied the case of a 57-year-old female patient who presented with acute/subacute symptoms of sphincter dysfunction, paraparesis, and ocular pain. The patient was diagnosed with anti-MOG syndrome with findings resembling snake-eye appearance (SEA), characterized by nearly symmetrical round high signal intensity lesions located at anterior horns (gray matter) on T2-weighted image.

Keywords: Myelin-oligodendrocyte glycoprotein; Spinal cord; Snake-eye appearance; Magnetic resonance imaging

INTRODUCTION

Snake-eye (or owl-eye) appearance (SEA) is characterized as nearly symmetrical round high signal intensity lesions of anterior horns (gray matter) of the spinal cord on T2-weigthted images. It is a well-known and common magnetic resonance imaging finding in acute anterior spinal artery ischemia. In addition, SEA has been reported in chronic compressive myelopathy, amyotrophic lateral sclerosis, but rarely in neuromyelitis optica spectrum disorder (NMOSD). However, there are no published studies of SEA in anti-MOG syndrome as far as we know (1, 2).

Anti-myelin oligodendrocyte glycoprotein (anti-MOG) syndrome is an immune-mediated inflammatory condition of the central nervous system (CNS) that damages the surfaces of oligodendrocytes and myelin sheaths (3, 4). Previous investigation reported that the spinal involvement of anti-MOG syndrome typically involves both gray and white matters, covering more than 50% of the axial section of the medullary cord (4). Here, we describe a case of a female patient with anti-MOG syndrome with MRI findings of the spinal cord resembling SEA.

CASE REPORT

A 57-year-old female patient was referred to our hospital after experiencing two days of increasing lower spinal cord weakness and six days of sphincter dysfunction. She also had a 1-month history of ocular pain. The patient was a non-smoker, with no remarkable past medical history, such as diabetes, hypertension, or dyslipidemia. Neurologic examination revealed paraparesis (motor Grade 4), and hypoesthesia below the thoracic dermatome, patellar hyperreflexia.

The patient underwent spine MRI using a 3-Tesla MRI scanner. Axial T2-weighted image (Fig. 1a) showed bilateral round high signal intensity lesions at anterior horn cells of T2-6 level without expansion of spinal cord, intervertebral disc herniation, and spinal canal stenosis. It was expected that there was enhancement on the left side of the corresponding area on T1-weighted fat suppressed contrast enhanced image (Fig. 1b). Sagittal T2-weighted image showed suspicious "pencil-thin T2 high signal intensity lesions" involving the T2-T6 spinal cord level (Fig. 1c). Brain and orbit MRI for the evaluation of ocular pain revealed multiple cortical gray matter and juxtacortical white matter T2 high signal intensity lesions (Fig. 1d) without contrast enhancement (Fig. 1e) and contrast enhancement of the anterior segment of the right optic nerve (Fig. 1f).

Despite cerebrospinal fluid (CSF) protein was elevated (52.3 mg/dL), the cell count was normal and the oligoclonal bands were negative. Serum anti-aquaporin 4 antibody was negative and anti-MOG IgG antibody was positive in the serum and cerebrospinal fluid, and the patient was diagnosed as anti-MOG syndrome. It was observed that the patient had a rapid recovery after being treated with intravenous steroids for six days and was discharged 12 days later admission. A year has passed and the patient is currently on surveillance without significant sequelae, or recurrence.

DISCUSSION

Anti-MOG syndrome is an immune-mediated inflammatory condition of the CNS, usually involving the spinal cord and optic nerves (3). Earlier, anti-MOG syndrome was not clearly distinguished from NMOSD due to the overlapping of clinical manifestation and repetitive relapsing course. Although both diseases were treated with steroids and plasma exchange, anti-MOG syndrome was thought to be at low risk of relapses, with relatively more promising long-term visual and motor outcomes (4). Therefore, in order to provide appropriate treatment to patients and to accurately predict the prognosis, the need for rapid identification and confirmation of MOG-IgG is emerging. Due to the recent developments over the last decades, anti-MOG syndrome was shown to have an independent pathophysiological mechanism from NMOSD. Anti-MOG syndrome is caused by damage to MOG, a membrane protein expressed on the outermost surface of myelin sheaths of oligodendrocytes (3, 4). Although the exact role remains unclear, it is believed to act as a mediator of the complement activation cascade (4). When the blood-brain barrier (BBB) is destroyed by traumas such as infectious conditions, the peripherally generated anti-MOG antibodies invade the CNS, activating the complement system, causing demyelination (3, 4). With the introduction of the cell-based assay (CBA) and immunoprecipitation techniques for the detection of MOG-IgG, the diagnosis has become clear, due to the rapidly accumulating knowledge regarding this disease entity (3). However, because CBA test results takes a long period of time to obtain, radiologic findings therefore play an important role in the initial evaluation and management of anti-MOG syndrome.

Typical spinal cord manifestation of anti-MOG is a longitudinally extensive transverse myelitis (LETM) pattern, with no or mild patchy contrast enhancement. However, a pattern of multiple short lesion is not infrequent (~30%) (3-6). Preferential gray matter involvement is also an important finding for differential diagnosis. The gray matter involvement in anti-MOG syndrome on MRI is represented by the presence of a sagittal line hyperintensity (a.k.a pseudo-dilatation of the ependymal canal) and H-shaped gray matter hyperintensity can be observed in the axial view, also known as the axial H sign (4).

The etiology of spinal cord gray matter T2 hyperintensity in anti-MOG syndrome is unclear. This finding is supported by the fact that oligodendrocytes containing MOG are found in spinal cord gray matter, and experimental autoimmune encephalomyelitis animal models immunized with MOG 35-55 peptide showed lesions in these areas (7).

The SEA, and another common finding in spinal cord infarction, the anterior pencil like hyperintensity (2) observed in the sagittal plane (which was similar to the sagittal line hyperintensities observed in our case), are considered consequences of the fact that neurons in the gray matter are more susceptible to hypoxia or ischemic damage than axons in the surrounding white matter (8).

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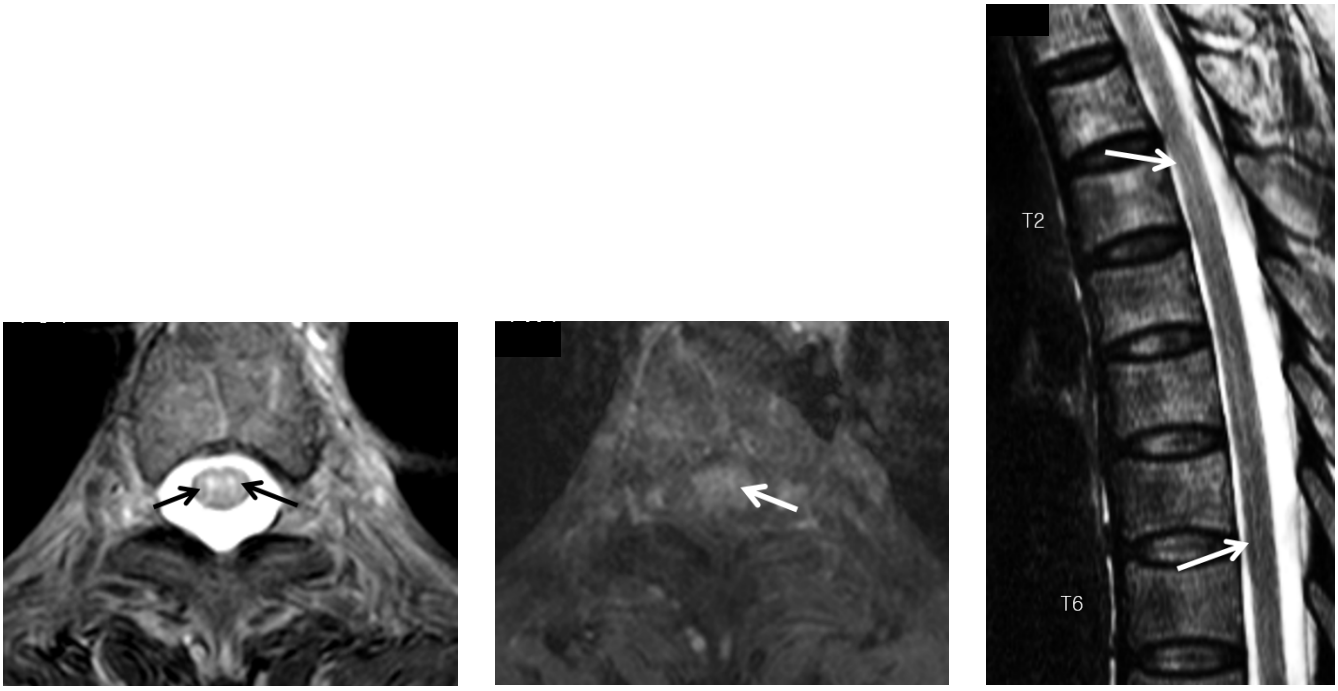

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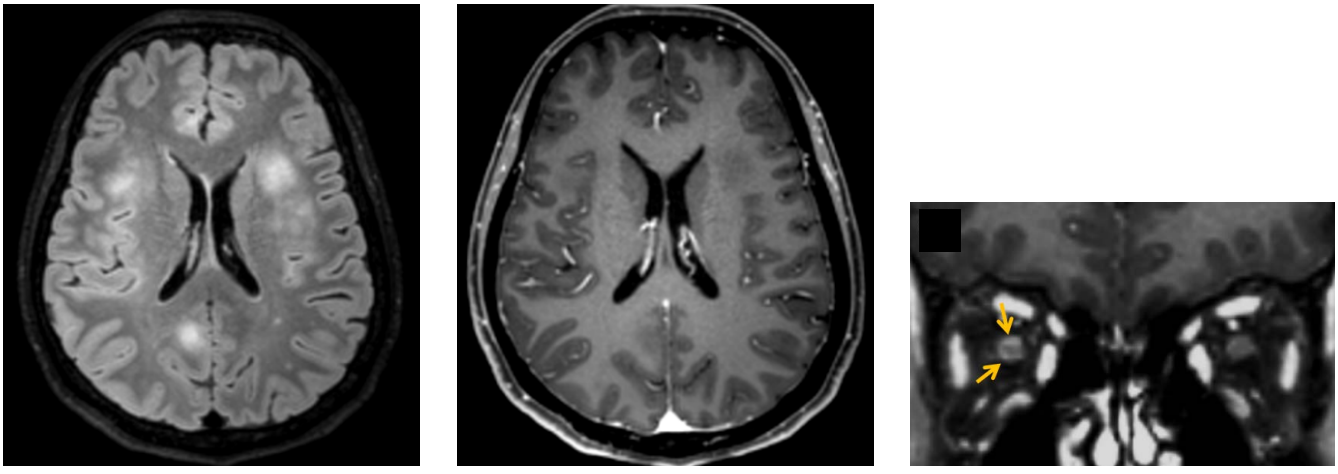

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Fig. 1. A 57-year-old female with anti-MOG syndrome showing "snake-eye appearance". (a, b) Axial T2-weighted images showed bilateral round high signal intensity lesions (black arrows) at anterior horns of T2-T6 spinal cord (a) without expansion of spinal cord, intervertebral disc herniation, and spinal canal stenosis. Contrast enhancement on the left side of the corresponding area (white arrow) on T1-weighted fat suppressed contrast enhanced image was expected (b). (c) Sagittal T2-weighted MRI of the spinal cord demonstrates longitudinally extensive "pencil thin T2 high signal intensity lesion" (white arrows) involving T2-T6 level. (d, e) Axial FLAIR image (flip angle/TR/TE = 90°/4800 ms/314.8 ms) of brain shows multiple T2 high signal intensity lesions in cortical and subcortical area of bilateral cerebral white matter (d) without definite enhancement on axial T1-weighted fat suppressed contrast enhanced image (e). (f) Coronal T1-weighted fat-suppressed contrast-enhanced MRI shows contrast enhancement (yellow arrows) in right optic nerve, mainly anterior aspect, suggestive of the optic neuritis.

Previous investigations reported that spinal cord infarcts are usually located in the central area of the anterior spinal artery, and in the anterior spinal artery infarction, it occurs most frequently in the thoracolumbar area, followed by the cervical area, and rarely in the mid-thoracic spinal cord (9, 10). Also, it can be noted that in early finding of neuromyelitis optica (NMO), there is an increase in metabolic activity and reduced collateral supply of the anterior horns. According to Deneve et al. (4), central thin and linear T2 hyperintensity with gadolinium enhancement along the ependymal canal, "pseudo-dilatation of the ependymal canal," was more commonly observed in anti-MOG syndrome cases and the "bright spot" sign, an intramedullary lesion with a higher T2 signal intensity than CSF, indicated NMOSD rather than anti-MOG syndrome.

Given the pattern of gray matter predilection in both spinal cord infarction and anti-MOG syndrome, we believe that faint high signal around the central canal of the spinal cord with relative prominence of high signal in the anterior horn cell may mimic SEA, as in our presented case.

In conclusion, we report a case of anti-MOG syndrome with findings resembling SEA. We suggest that the diagnosis of anti-MOG syndrome should be considered in patients with this imaging appearance. When there are related symptoms, such as optic neuritis and specific patterns of brain lesion on MRI, and that clinical situations are not consistent with spinal cord infarction, suspicion for anti-MOG syndrome should be raised.

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