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A Case of Anti-Myelin Oligodendrocyte Glycoprotein Antibody-Positive Late-Onset Acute Disseminated Encephalomyelitis

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Dear Editor,

Myelin oligodendrocyte glycoprotein (MOG) antibody-associated disease (MOGAD) has recently been classified as a distinct inflammatory demyelinating disease of the central nervous system.[1] The clinical manifestations of adult MOGAD resemble those of neuromyelitis optica spectrum disorder (NMOSD), which usually involve the optic nerve and spinal cord. When MOGAD involves the brain, an acute disseminated encephalomyelitis (ADEM)-like phenotype is most commonly observed; however, this is more common in pediatric or young adult patients. Here we report a case of late-onset MOG-immunoglobulin G (IgG)-positive encephalomyelitis with an ADEM-like phenotype.

A 57-year-old female developed gait disturbance with recurrent falls, dysarthria, and cognitive decline 3 months prior to admission to our hospital. She initially experienced recurrent falls and progressive bilateral lower limb weakness. One month later she experienced dysarthria and reduced verbal output, followed by decreased mentality, confusion, and dysphagia. Preceding vaccinations, systemic infections, and other neurologic symptoms were not reported.

On admission she was alert, but she exhibited severe dysarthria and cognitive impairment with decreased orientation to time and place. A neurologic examination revealed paresis of all extremities, which was more severe on the right side (grades 2, 3, 4, and 4 on the Medical Research Council scale in the right arm, right leg, left arm, and left leg, respectively). Bilateral upper and lower extremity deep tendon reflexes were brisk. Ankle clonus and Babinski's sign were observed bilaterally. Her score on the Expanded Disability Status Scale (EDSS) was 9.5.

The results of a cerebrospinal fluid (CSF) examination were unremarkable, with a total nucleated cell count of 3/μL, protein level of 45 mg/dL, and glucose level of 73 mg/dL. CSF cytology, anti-AQP4 antibody, and CSF oligoclonal band tests were negative. Brain magnetic resonance imaging (MRI) fluid-attenuated inversion recovery (FLAIR) images showed multifocal hyperintense subcortical and periventricular white-matter and brainstem lesions (Fig. 1A and B). Gadolinium (Gd)-enhanced T1-weighted images showed multiple open-ring enhancement patterns (Fig. 1C and D). Spinal MRI showed multiple short-segment T2-weighted hyperintense lesions with partial enhancement in the cervical cord (Fig. 1E).

Intravenous steroid pulse therapy was initiated under the clinical diagnosis of ADEM, with oral prednisolone and azathioprine as maintenance therapy. At that time the patient was found to be seropositive for anti-MOG antibodies, as determined by a live-cell fluorescence-activated cell-sorting assay using serum.[2] Follow-up brain MRI conducted 2 weeks after the initial MRI scan showed decreased open-ring enhancements on the Gd-enhanced T1-weighted images without significant changes on the FLAIR images (Fig. 1F, G, and H). Although independent gait was still difficult, her dysarthria and verbal output had improved

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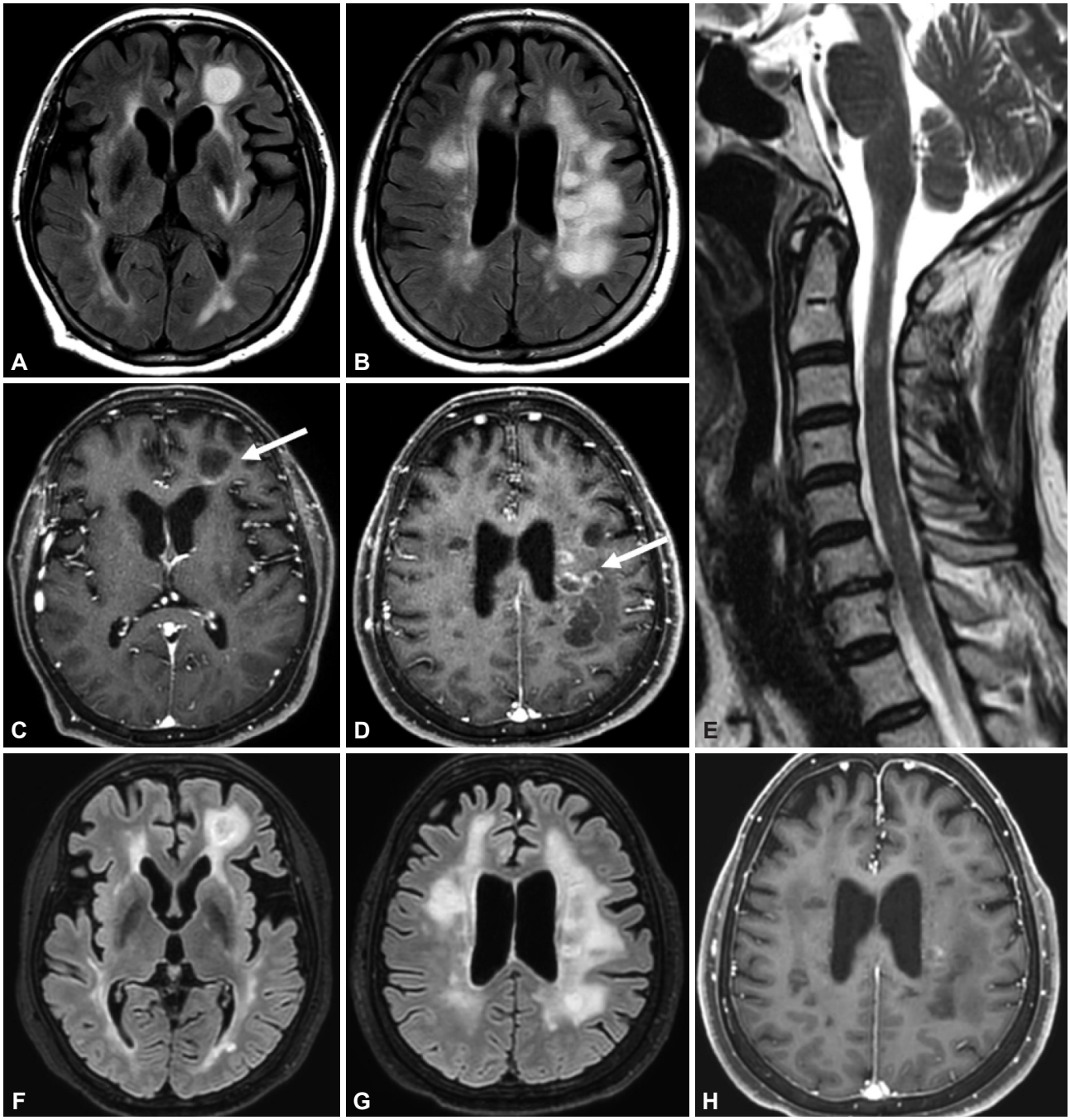

Fig. 1. Brain and spine MRI of the present case. A and B: At admission, FLAIR images showed multiple hyperintense left frontal and bilateral periventricular white-matter lesions. C and D: Gd-enhanced T1-weighted images showed multiple open-ring enhancement patterns (arrows). E: Multiple cervical lesions with short-segment involvement were observed in spinal MRI. F, G, and H: Follow-up brain MRI after 2 weeks showed significantly decreased open-ring enhancement on the Gd-enhanced T1-weighted images, with minimal changes on the FLAIR images. FLAIR: fluid-attenuated inversion recovery, Gd: gadolinium, MRI: magnetic resonance imaging.

significantly after steroid therapy. Her follow-up EDSS score was 9.0. She was transferred to the rehabilitation department 1 month after admission. After intensive rehabilitation treatment for 1 month, she was able to walk with bilateral assistance, with an EDSS score of 6.5.

This patient had monophasic and subacute encephalopathy that could not be explained by fever. Initial brain MRI showed diffuse, large, poorly demarcated lesions predominantly involving the white matter, and there were no newly developed lesions in the follow-up MRI. These clinical and radiologic features fulfill the diagnostic criteria of ADEM.[3]

Recent studies have found that 30–50% of patients with

ADEM are seropositive for anti-MOG antibodies.[4,5] However, most reported cases of MOGAD with the ADEM phenotype were pediatric patients or adult patients younger than 40 years.[6] Adults with MOGAD generally present with optic neuritis or myelitis, and so the clinical presentation of late-onset anti-MOG antibody-positive ADEM has rarely been reported. The case of a 49-year-old male who presented with anti-MOG antibody-positive ADEM was reported recently.[7] To the best of our knowledge, the present case is the first report of anti-MOG antibody-positive ADEM in a patient older than 50 years. Additionally, our patient demonstrated lesions in FLAIR imaging throughout the white matter, brainstem, and spinal cord that were more extensive than those in previously reported cases. The pattern of multiple open-ring enhancements on Gd-enhanced T1-weighted images is also not common in MOGAD. Considering the age and distinct phenotype of the present patient in brain MRI, this report may broaden the understanding of the characteristics of MOGAD.

In conclusion, MOG-IgG-positive encephalomyelitis with an ADEM phenotype can occur in older adults, and serologic testing for anti-MOG antibodies should be considered in patients with late-onset ADEM.

Author Contributions

Conceptualization: Ki Hoon Kim, Seung Woo Kim. Supervision: Ha Young Shin, Seung Woo Kim. Visualization: Ki Hoon Kim, Jinhyuk Cho. Writing—original draft: Ki Hoon Kim. Writing—review & editing: all authors.

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Conflicts of Interest

The authors have no potential conflicts of interest to disclose.

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