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# A Case of MOGAD Complicated With Cerebral Vasculitis: Case Report and Literature Review

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Dear Editor,

A 39-year-old male experienced a sudden-onset epileptic seizure during strenuous exercise 2 days prior to hospital admission. The seizure, which manifested in twitching of the limbs, foaming at the mouth, and unconsciousness, lasted approximately 5 minutes before it subsided spontaneously. Another seizure occurred within 30 minutes. A physical examination revealed that the muscle strength was grade 5 in both upper extremities and grade 4 in both lower extremities, and that the muscle tone of all four limbs was normal. Tendon reflexes of both upper extremities (++), those of both lower extremities (+), bilateral Babinski sign (−), and neck resistance (−). Laboratory tests (cell-based assay) revealed that the patient was positive for antimyelin oligodendrocyte glycoprotein IgG (anti-MOG-IgG) in the serum (1:100+) and cerebrospinal fluid (CSF, 1:1+). The total cell count in the CSF was $37\times10^6$/L, and the white blood cell count in the CSF was $9\times10^6$/L. The monocyte ratio was 100%, and the IgM level was 0.25 g/L. CSF oligoclonal bands were positive.

The MRI findings (Fig. 1A, C, E, G, H, and I) included abnormal signals in multiple regions of the bilateral cerebral hemispheres, especially white-matter involvement in the right temporal lobe, the right centrum semiovale, and the left frontal lobe. Most lesions were perpendicular to the ventricular wall, and they were distributed parallel to the medullary veins. Irregular patchy enhancement was observed within the lesions of the right temporal and left frontal lobes after gadopentetate injection. A T2-weighted fluid-attenuated inversion recovery (T2-FLAIR) sequence indicated strong signals in parts of the sulci and adjacent cortical areas of the right hemisphere, and contrast-enhanced scans suggested that the leptomeninges that overlaid those areas were also enhanced. Magnetic resonance angiography revealed mild stenosis of the right-side middle cerebral artery (MCA). High-resolution MRI vessel-wall imaging (HR-VWI) revealed segmental wall thickening of the bilateral MCA and basilar artery (BA), with a significant "double-track sign" and circumferential enhancement, which suggested vasculitis.

The patient was treated using glucocorticoids as well as azathioprine and antiepileptic during hospitalization and after discharge, which markedly improved his symptoms. When retested 44 days after hospitalization, his serum MOG-IgG levels were still at an abnormally high titer (1:32). However, the extent of vessel-wall thickening that occurred due to vasculitis was reduced, and the degree of enhancement was diminished (Fig. 1B, D, F, and J).

MOG-IgG-associated disease (MOGAD) is an immune disease of the nervous system that may be caused by MOG-IgG and mediated by T cells. Its manifestations include optic neuritis, meningoencephalitis, brainstem encephalitis, and myelitis.[1] The imaging features in the present case were consistent with demyelination, with the CSF and peripheral blood both testing positive for MOG-IgG antibodies in laboratory examinations. All of the presented evidence supported the clinical diagnosis of MOGAD. In 2019, Budhram et al.[2] presented an exemplar case of a unilateral cortical T2-FLAIR-hyperintense lesion in anti-MOG-associated

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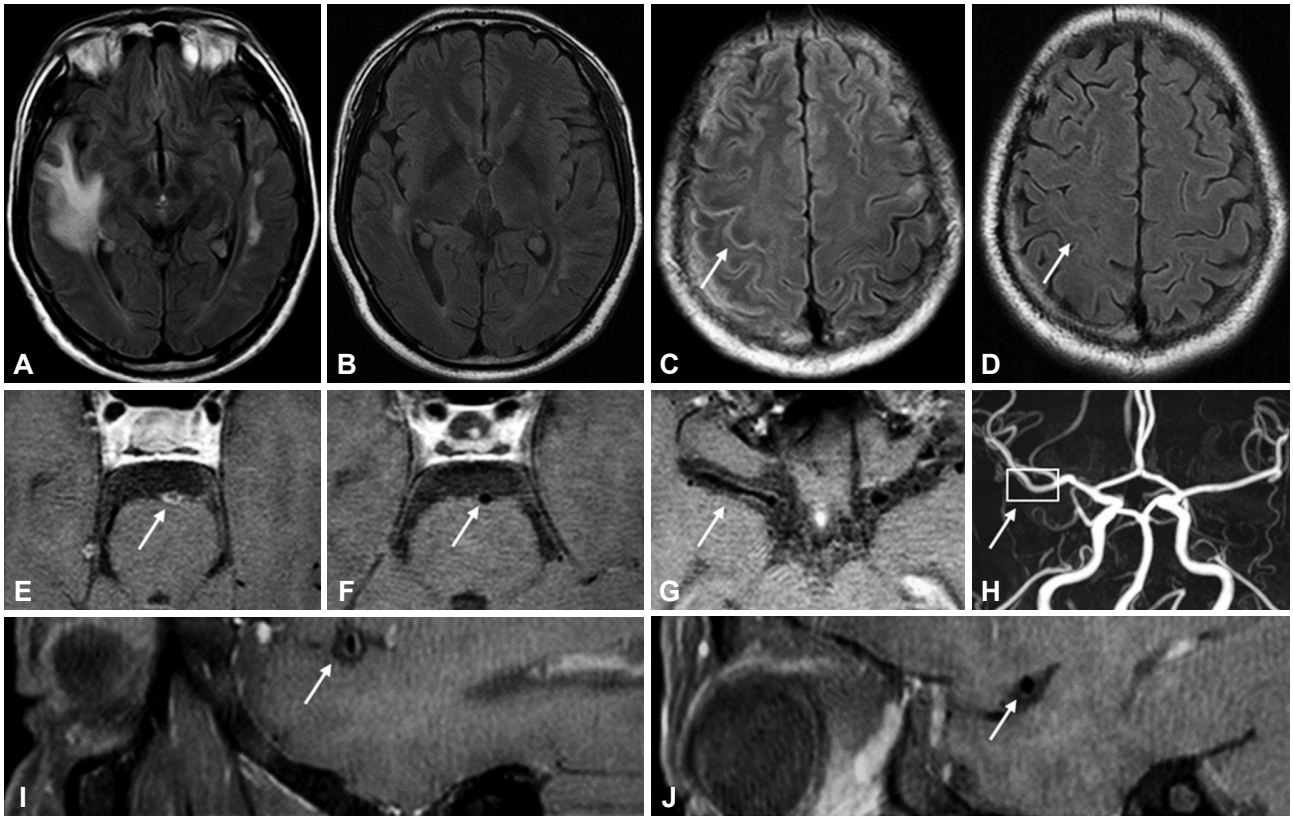


**Fig. 1.** A and C: Axial T2-FLAIR sequences. Hyperintense lesions were present in the bilateral temporal lobe, with white-matter involvement more prominent in the right than in the left hemisphere. Increased T2-FLAIR signals were observed in the sulci and adjacent cortical areas of the right hemisphere. B and D: The lesions in the brain parenchyma and the sulci of the right hemisphere were improved, and the enhancement was no longer present. E, F, I, and J: The walls of the BA and right-side MCA were thinner than at presentation, and the enhancement was reduced (arrows). G: HR-VWI indicated segmental wall thickening of the right MCA and circumferential enhancement with a significant "double-track sign" (arrows). H: Magnetic resonance angiography indicated mild stenosis of the right MCA (arrows). BA, basilar artery; HR-VWI, high-resolution MRI vessel-wall imaging; MCA, middle cerebral artery; T2-FLAIR, T2-weighted fluid-attenuated inversion recovery.

encephalitis with seizures (FLAMES). In the present case, the bilateral temporal lobes contained obvious cortical T2-FLAIR-hyperintense lesions that were accompanied by epileptic symptoms, which was consistent with the presentation of FLAMES as proposed by Budhram et al.[2] However, the axial T2-FLAIR hyperintensity observed in the bilateral cerebral lobes in our case was more similar to those in the case reported by Ma et al.[3] The same features of meningitis-like leptomeningeal enhancement were observed in both our case and that of Budhram.[4]

Few studies have investigated the association between MOGAD and vasculitis. Patterson et al.[5] performed a biopsy of the lesion area of a patient with anti-MOG encephalitis. Their pathological results indicated that inflammation had infiltrated the gray matter, white matter, and vessel walls. Inflammatory cells were observed in the vessel walls, and lymphocyte infiltration involved the small-vessel walls and perivascular area, which was accompanied by perivascular demyelination. Another researcher found that inflammatory demyelination changes in MOGAD were mostly distributed along small veins, especially around the paraventricular nucleus, which was accompanied by clustered T-lymphocyte infiltration. Debris that predominantly consisted of MOG was observed, with some macrophages phagocytosing the debris in the perivascular space, which suggests a local inflammatory response involving cerebral vessels.[6] Because of the close resemblance between the microscopic changes observed in MOGAD and those in vasculitis, the two are often confused in pathological diagnoses.[5] In patients with MOGAD, the distribution of lesions observed in both unilateral cerebral cortical encephalitis and bilateral medial frontal cerebral cortical encephalitis mostly coincides with the regions supplying the MCAs and anterior cerebral arteries. This observation suggests that the occurrence of MOGAD involves arteries.[7] However, few imaging studies have assessed arterial abnormalities in patients with MOGAD. In contrast to other imaging techniques, HR-VWI provides direct observations of the brain artery walls and can be used to diagnose vasculitis and assess the treatment efficacy.[8,9] In our case, some active demyelinating lesions were distributed in the MCA region, while HR-VWI indicated vessel-wall inflammation in the corresponding MCA trunk and branches. After glu-

cocorticoid treatment, MRI reexamination indicated the synchronous improvement of demyelinated lesions and vascular wall inflammation. All of the above findings indicate that MOGAD may be closely associated with arterial vasculitis, and that it may in essence be an inflammatory process of the central nervous system that involves both large blood vessels and brain parenchyma. In the present case, although the BA and right posterior cerebral artery also exhibited vasculitis, no demyelinating lesions were observed in the infratentorial area. We speculated that this finding was related to the severity or pathological process of vasculitis. The above-mentioned interpretations need to be further tested through imaging and pathological examinations of additional patients.

In conclusion, the clinical data and imaging findings of this case suggest the coexistence of MOGAD and vasculitis; this relationship requires further confirmation in additional studies.

## Ethics Statement

The written informed consent was obtained from participant.

## Availability of Data and Material

Data sharing not applicable to this article as no datasets were generated or analyzed during the study.

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## Conflicts of Interest

The authors have no potential conflicts of interest to disclose.

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