

Repeated Non-enhancing Tumefactive Lesions in a Patient with a Neuromyelitis Optica Spectrum Disorder

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Abstract

A 51-year-old woman had developed fever and consciousness disturbance at 47 years of age. Brain magnetic resonance imaging (MRI) revealed acute disseminating encephalomyelitis (ADEM)-like lesions without gadolinium enhancement (GDE). One year later, she had an episode of bilateral optic neuritis and cerebellar ataxia. Speech deficit and right hand weakness occurred at the age of 51 years. Neurological examination showed motor aphasia, finger agnosia, right-left disorientation, and right hand paresis. Neuromyelitis optica (NMO)-IgG was seropositive. Cerebrospinal fluid examination showed negative results for myelin basic protein and oligoclonal IgG band. The IgG index was normal. Brain MRI revealed a tumefactive lesion in the left temporo-parietal region and conglomerate ovoid lesions in the pericallosal regions. No GDE was found in the brain lesions. Visual evoked potential test showed bilateral prolongation of P100 latencies. She was treated twice with methylprednisolone pulse therapy followed by oral prednisolone, but the motor aphasia did not respond to steroid treatment. She had no prior history of myelitis and was diagnosed as NMO spectrum disorder (NMOSD). Similar to previous studies of NMO-IgG seropositive extensive brain lesions, this patient with NMOSD indicated no GDE in tumefactive lesions at two episodes of encephalopathy. Compared to multiple sclerosis (MS), a high frequency of non-enhancing tumefactive lesions is reported in patients with NMO or NMOSD. The absence of GDE in tumefactive lesions could help to differentiate between NMO and MS.

Key words: neuromyelitis optica, neuromyelitis optica spectrum disorder, NMO-IgG, tumefactive lesion, gadolinium enhancement

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Introduction

Neuromyelitis optica (NMO) is an inflammatory demyelinating disease and anti-aquaporin-4 (AQP4) antibodies (NMO-IgG) play a major role in the diagnosis of NMO or NMO spectrum disorders (NMOSD) (1, 2). Extensive brain lesions resembling brain tumors or acute disseminating encephalomyelitis (ADEM) on magnetic resonance imaging (MRI) are identified in NMO-IgG seropositive patients (3-12). The frequency of cerebral lesions is 6-18% in patients with NMO or multiple sclerosis (MS) who are seropositive for anti-AQP4 antibody (3, 4). Recently, cloud-like gadolinium enhancement (GDE) has been reported as a radiological feature of brain MRI lesions in NMO patients (5).

Several MRI studies have indicated that extensive brain lesions lack GDE in several NMO-IgG seropositive patients (6-11). Little is known whether tumefactive lesions in NMO-IgG seropositive patients are enhanced. We report a unique patient with NMOSD in whom tumefactive lesions without GDE were repeated at two episodes of encephalopathy.

Case Report

A 51-year-old woman had developed fever and disturbance of consciousness at 47 years of age. Brain MRI was performed 3 days later. Extensive ADEM-like lesions were found in the brainstem, middle cerebellar peduncles, subcortical white matter, and corpus callosum. The lesions lacked

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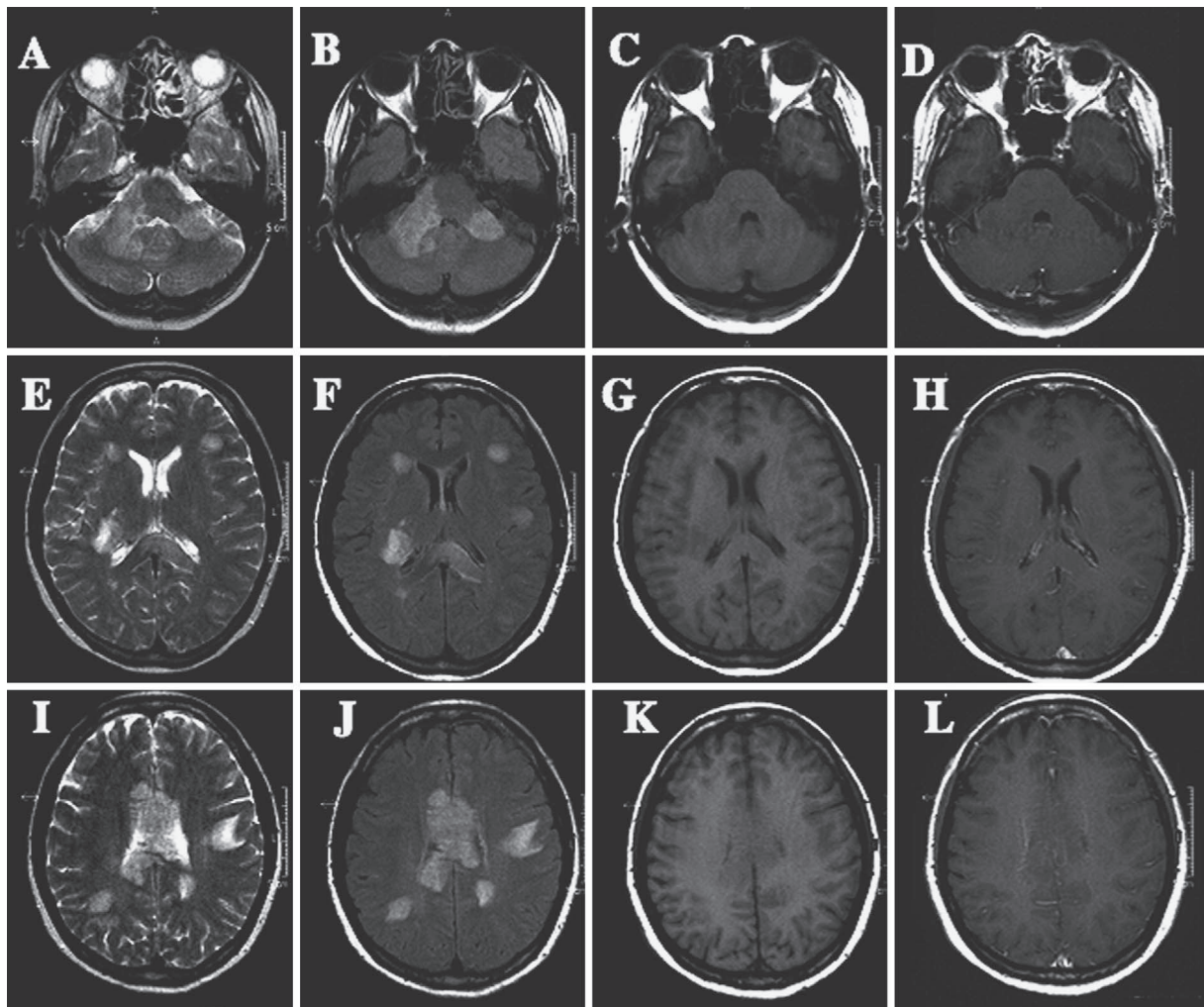


Figure 1. ADEM-like hemispheric lesions at 47 years of age. (A, E, I) T2-weighted imaging. (B, F, J) Fluid-attenuated inversion recovery (FLAIR) imaging. (C, G, K) T1-weighted imaging. (D, H, L) Gadolinium-enhanced T1-weighted imaging. T2-weighted and FLAIR images show extensive hyperintensity lesions, including the infratentorial region, the subcortical white matter and the corpus callosum. All lesions lack GDE.

GDE (Fig. 1). Her consciousness state recovered immediately after methylprednisolone (mPSL) pulse therapy (1 g/day for 3 days). At the age of 48 years, she experienced an episode of bilateral optic neuritis and cerebellar ataxia. Speech deficit and right hand weakness occurred at the age of 51 years. She visited a neighboring hospital and was transferred to our department. Neurological examination showed motor aphasia, finger agnosia, right-left disorientation and right hand paresis. Hematological tests and blood chemistry were normal. Serological NMO-IgG test (Mayo Medical Laboratories, Rochester, MN, USA) was positive. Cerebrospinal fluid (CSF) examination showed a cell count of 20 cells/mm³ and a protein concentration of 18 mg/dL. Myelin basic protein and oligoclonal IgG band were not detected using the isoelectric focusing method. IgG index was 0.54. When brain MRI was performed at admission (3 days from the neurological episode), a tumefactive lesion was found in the left parieto-temporal region (Fig. 2). Fluid-attenuated inversion recovery imaging disclosed widespread

callosal and periventricular hyperintensity, suggesting conglomerate ovoid lesions in the cerebral white matter (Fig. 3). MRI after intravenous administration of gadopentetate dimeglumine (0.2 mL/kg) displayed no GDE in the brain lesions (Fig. 2). Spinal cord MRI revealed no active pathognomonic lesions. P100 latency of visual evoked potential test was 126 msec in the right eye and 124 msec in the left eye. She received mPSL pulse therapy twice followed by tapering administration of prednisolone (50 to 20 mg/day po) for 4 months. Brain single photon emission computed tomography using N-isopropyl-p-[¹²³I]-iodoamphetamine was performed at 6 weeks after steroid treatment. A mild to moderate degree of hypoperfusion was found in the left temporo-parietal region. Four months later, brain MRI showed cystic transformation in the tumefactive lesion (Fig. 4). As she had no prior history of myelitis, we diagnosed her as NMOSD based on the NMOSD diagnostic criteria by Wingerchuk et al (13).

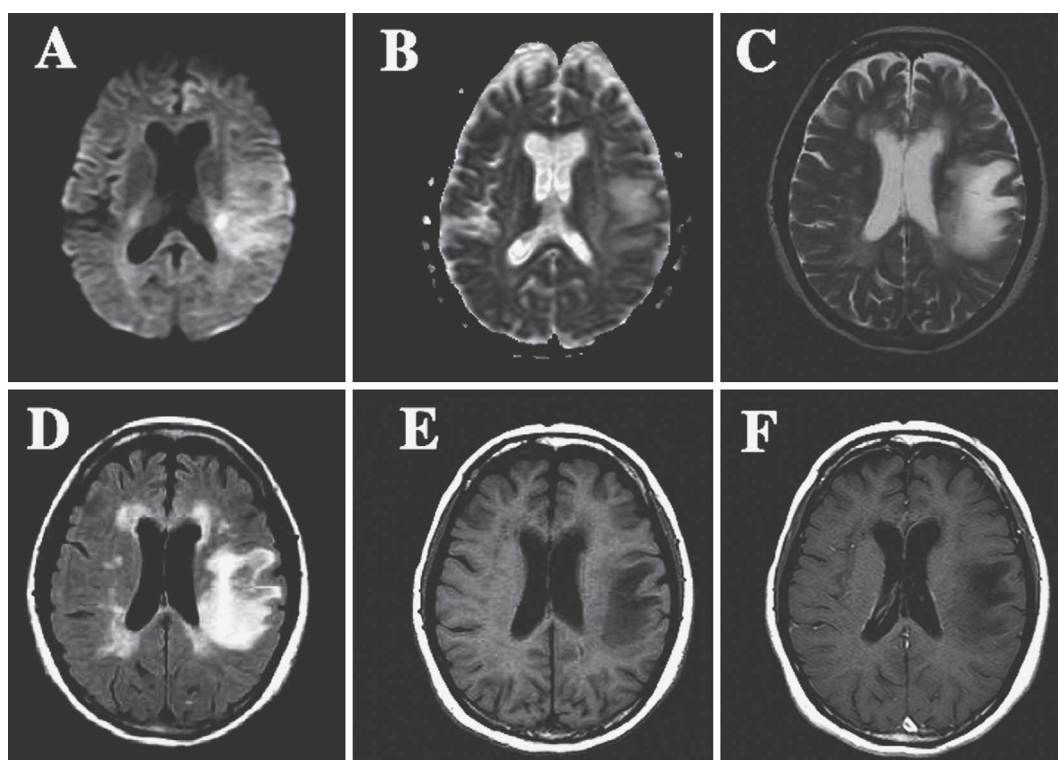


Figure 2. A non-enhancing tumefactive lesion. (A) Diffusion-weighted imaging (DWI). (B) Apparent diffusion coefficient (ADC) map. (C) T2-weighted imaging. (D) FLAIR imaging. (E) T1-weighted imaging. (F) Gadolinium-enhanced T1-weighted imaging. DWI, ADC map and FLAIR images show a huge hyperintensity lesion in the left parieto-temporal region. GDE is absent in the tumefactive lesion.

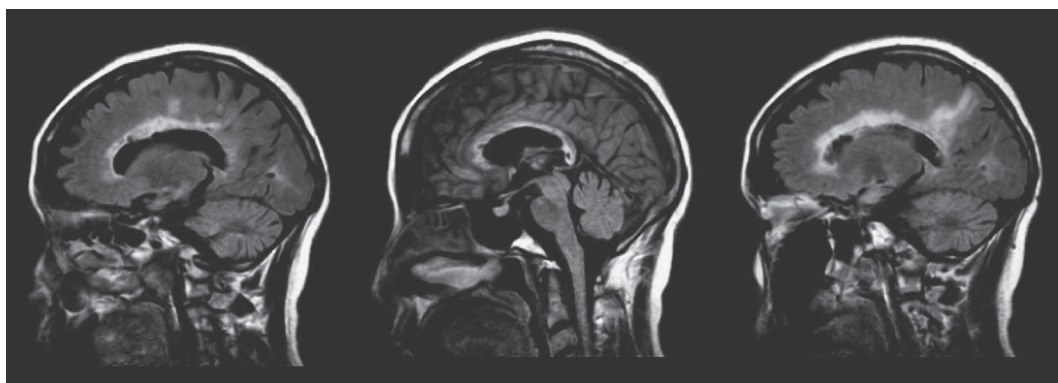


Figure 3. Conglomerate ovoid lesions in the callosal and periventricular regions. Sagittal FLAIR imaging shows widespread pericallosal and periventricular hyperintensity lesions.

Discussion

Neuroradiological features of the present patient with NMOSD suggested non-enhancing ADEM-like and tumefactive lesions at two episodes of encephalopathy. Moreover, MRI revealed conglomerate ovoid lesions in the corpus callosum and the periventricular regions. These ovoid lesions were undistinguishable from those in MS.

Brain MRI abnormalities are detected in 60% of patients with NMO (7). The frequency of extensive brain lesions is 18.5% in Japanese patients with NMO or MS who are sero-

positive for anti-AQP4 antibody (3). A study using brain gadolinium-enhanced MRI showed cloud-like GDE in 50% of MNO patients. This blurred-margin patchy enhancement has been highlighted as a specific MRI finding in NMO patients (5). However, the above study did not state whether GDE existed in the extensive brain lesions and whether the GDE patterns differed between patients with NMO who were seropositive and those who were negative for anti-AQP4 antibody (5). GDE of extensive brain lesions has been depicted in three patients with NMOSD and posterior reversible encephalopathy syndrome (PRES). Blood pressure fluctuations and rapid fluid shifts following several therapies for

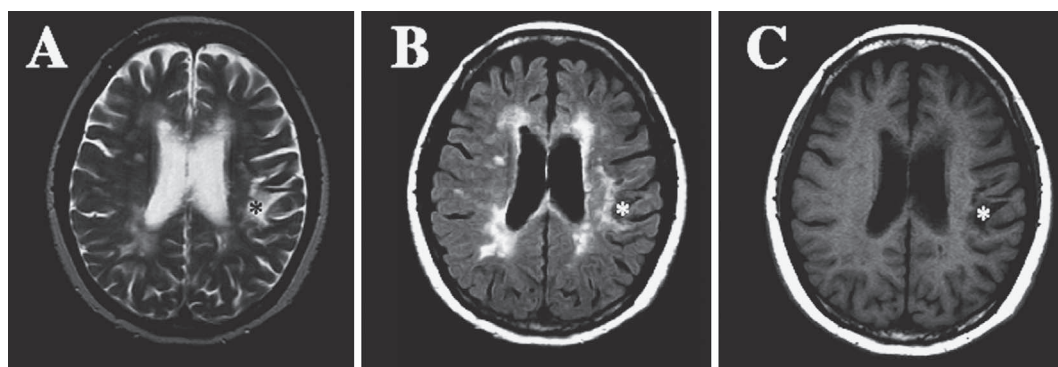


Figure 4. MRI at 4 months after admission. (A) T2-weighted imaging. (B) FLAIR imaging. (C) T1-weighted imaging. T2-weighted images show a hyperintensity lesion. FLAIR and T1-weighted images show a hypointensity lesion. These signal intensities suggest cystic transformation (asterisks).

NMO seemed to occur in NMO-IgG seropositive patients with PRES (12). GDE-lacking tumefactive lesions have been reported in eight cases with NMO-IgG seropositive NMO or MS, in addition to the present patient (6-11). Of five patients with anti-AQP4 antibody seropositive NMOSD, GDE was absent in four patients. Spotty enhancing lesions of the basal ganglia were found only in one patient with systemic lupus erythematosus (11). In contrast, recent studies of tumefactive demyelinating lesions have suggested a high frequency of GDE (93 or 95%) in patients with clinically proven MS (14, 15). As compared to MS, non-enhancing tumefactive lesions seem to occur commonly in NMO-IgG seropositive patients with NMO or NMOSD. The neuroradiological changes of the present patient indicated that the absence of GDE might be another MRI profile in patients with NMO or NMOSD. Thus, GDE patterns of tumefactive demyelination lesions could facilitate the differential diagnosis between NMO and MS.

The authors state that they have no Conflict of Interest (COI).

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