Rapid Progressive Cerebral Atrophy in Systemic Lupus Erythematosus

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Neuropsychiatric systemic lupus erythematosus (NPSLE) is a serious and potentially life-threatening manifestation of SLE that occurs in 37%–95% of cases and is associated with an increased risk of death¹. Cerebral atrophy (i.e., cortical atrophy and ventricular dilation) determined by magnetic resonance imaging (MRI) was prevalent in 18% of patients with newly diagnosed SLE and was the most frequent abnormal finding by MRI². MRI was more sensitive than brain histopathology for the presence of cerebral atrophy in NPSLE³.

A 30-year-old woman with a 5-year history of SLE had a flare with fever and disturbed consciousness. The initial MRI scan with fluid-attenuated inversion-recovery revealed multiple high-intensity areas in the deep white matter (Figure 1A). Stroke and infection were excluded following investigation. Accordingly, we diagnosed an acute confusional state (delirium) caused by SLE. Despite aggressive treatment, including steroid pulse therapy and pulsed intravenous cyclophosphamide, the patient's consciousness rapidly deteriorated into coma. Four months later, we noted significant cerebral atrophy, characterized by a loss of brain volume, along with mul-

tiple continuous high-intensity areas (Figure 1B). She never recovered from coma and was transferred to another hospital.

Although mild brain atrophy on MRI is a common abnormal finding in NPSLE, rapidly progressing moderate to severe brain atrophy, as in this patient, has seldom been reported⁴. The MRI findings reflect the severe clinical outcome.

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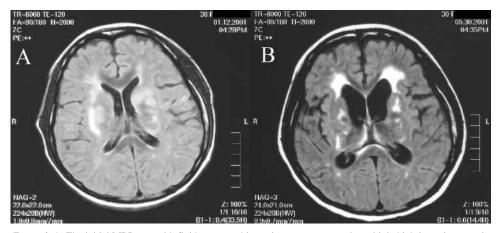


Figure 1. A. The initial MRI scan with fluid-attenuated inversion-recovery reveals multiple high-intensity areas in the deep white matter. B. Four months later, there is significant cerebral atrophy, characterized by a loss of brain volume, along with multiple continuous high-intensity areas.

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