中文題目：案例報告:系統性紅斑狼瘡之早發性失智症
英文題目：Systemic lupus erythematosus with early onset dementia
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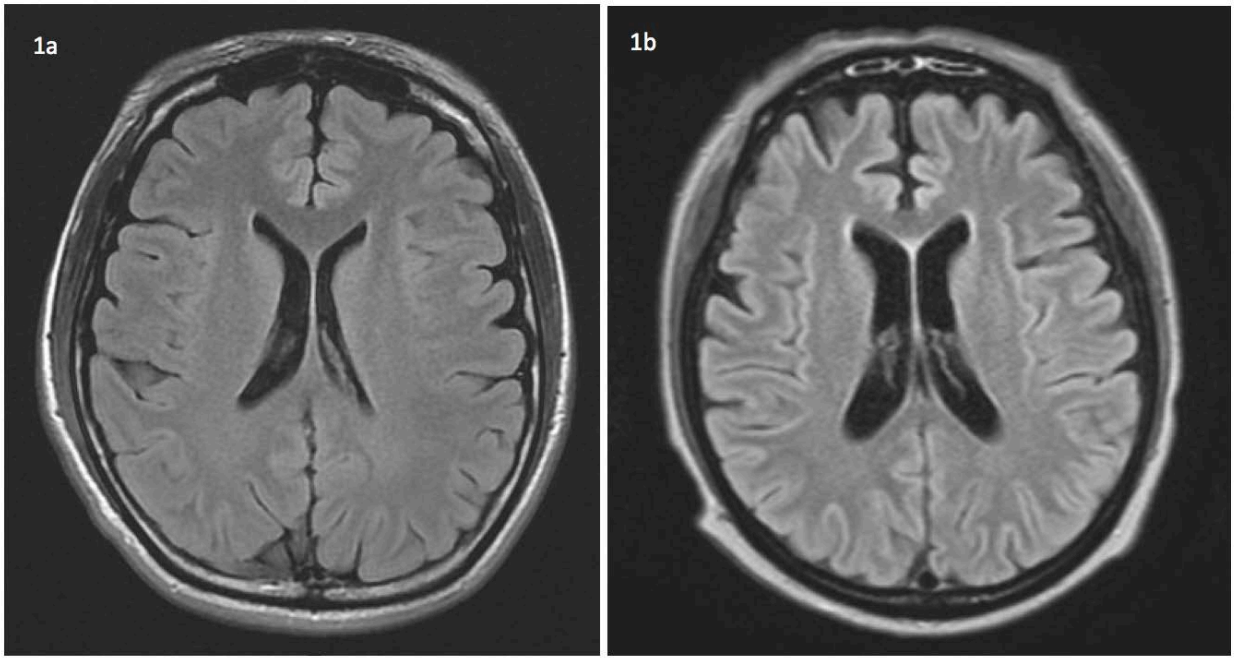
We reported two cases who had experienced neuropsychiatric systemic lupus erythematosus (NPSLE) followed by clinically early-onset dementia and brain atrophy on magnetic resonance imaging (MRI). The first case is a 33-year-old female, educated to master's level, who was diagnosed with systemic lupus erythematosus (SLE) 3 years before manifestations of impaired cognitive function. She had been admitted because of a bad flare characterized by recurrent fever, headaches, proteinuria, mentality changes, incoherent speech and mild dysarthria. The blood analysis showed leukopenia and high serum level of anti-double stranded DNA (anti-ds-DNA) antibodies, anti-cardiolipin IgG, lupus anticoagulant and anti-ribosomal P antibody. Her renal biopsy showed class IV lupus nephritis. Her proteinuria and lupus activity subsided after treatment. Cognitive deficits, mild acalculation and impaired working memory were noticed by her family one year before this admission. Brain MRI showed mild cerebral atrophy compared to age-matched healthy people or lupus patients without NPSLE (figure 1a). The other patient, a 40-year-old female, vocational school graduate, has been diagnosed with SLE for more than 10 years. She has experienced class IV lupus nephritis and received maintenance hemodialysis. The laboratory study revealed low serum complement and high level of anti-ds-DNA antibodies, b2-glycoprotein I and lupus anticoagulant. Recurrent headaches, fluctuated confusion, emotional changes, and dyscalculia developed and progressed one year before this visiting. The brain MRI revealed symmetrical cortical atrophy over frontotemporoparietal lobes, compared to age-matched healthy people (figure 1b).

In our cases, two well-educated young females, carrying with serum antiphospholipid antibodies without thromboembolic events, could not fulfill the diagnostic criteria of antiphospholipid syndrome. By the definition of corticosteroid-induced psychiatric disorders (1), temporary symptoms within 8 weeks of starting or increasing steroids and resolved completely by the reduction in steroid dosage without additional immunosuppressive agents, neither of our patients was under pulse therapy or in a tapering process. Therefore, the cognition dysfunction in our patients was not relevant to steroid therapy. The most common radiographic findings in NPSLE include hyper-intensities in the white or gray matter, parenchymal defect, and focal atrophy (2). Compared the brain MRI of our patients, none of them showed evidence of microinfarction or other structural changes but only mild cerebral atrophy. The complicated manifestation in our patients reminds us that we should be more aware of the risk of cognition dysfunction in SLE patients, especially those patients in young age without active lupus activity and in cases with former NPSLE but without structural damages. Cognition function and brain image check contribute to early diagnosis and treatment.



References:

1. Chau SY, Mok CC. Factors predictive of corticosteroid psychosis in patients with systemic lupus erythematosus. Neurology. 2003;61(1):104-7.

2. Luyendijk J, Steens SC, Ouwendijk WJ, Steup-Beekman GM, Bollen EL, van der Grond J, et al. Neuropsychiatric systemic lupus erythematosus: lessons learned from magnetic resonance imaging. Arthritis Rheum. 2011;63(3):722-32.