

中文題目：可逆性後腦病變症候群——紅斑性狼瘡影響神經中樞以可逆性後腦病變症候群表現

英文題目：Posterior Reversible Encephalopathy Syndrome: A Case of Lupus CNS Involvement Presented with PRESS

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Background:

Posterior reversible encephalopathy syndrome (PRES) is a clinico-radiological syndrome characterized by headache, seizures, altered mental status or visual loss, while image findings include white matter vasogenic edema affecting mainly the posterior occipital and parietal lobes of the brain.

Case presentation:

This 23-year-old kindergarten teacher presented with the chief complaint of intermittent fever for one week. She reported exertional dyspnea, dorsal hand skin rashes, skin yellowing and dark urine recently. Laboratory examination revealed anemia (Hb=4.7g/dL) with positive direct and indirect Coomb's test, leukocytosis of 14800/uL, thrombocytopenia at 105×10^3 /uL, acute kidney injury (creatinine=2.19 g/dL) with proteinuria, total bilirubin was 2.3mg/dL and high LDH levels at 880/uL. Immunological profiles revealed ANA with speckle 1:160 and anti-dsDNA antibody 614 IU/ml, and low C3 (20.5 mg/dL) and C4 (2.1 mg/dL). Right pleural effusion and mild pericardial effusion were observed in image studies. She was admitted with systemic lupus erythematosus (SLE).

On the second day of admission, she complained of blurry vision, which followed by a tonic-clonic seizure lasting for one minute. The CSF examination was not remarkable. Brain MRI T2W FLAIR image demonstrated hyperintense lesion over posterior brain regions (bilateral parietal, occipital, left cerebellum, right thalamus, bilateral posterior midbrain, and bilateral posterior pons). EEG indicated diffuse cerebral dysfunction. She was diagnosed with SLE-related posterior reversible encephalopathy syndrome (PRES) and was prescribed dexamethasone 4 mg Q6H, levetiracetam 1200mg Q12H and Mycophenolate 2 tabs BID for seizure control. A week later, follow up MRI demonstrated regressive change of hyperintensity of bilateral occipital and parietal lobes with multifocal stenosis of intracranial arteries.

Conclusion:

PRES can be a clinical presentation of lupus with CNS involvement. Seizures, headache, altered mental status or visual loss were the most common clinical symptoms. In our patient presented with seizure and blurry vision. In view of good prognosis when supportive treatment was given early, prompt recognition of PRES in SLE patients is crucial to allow administering appropriate management. If promptly recognized and treated, the clinical syndrome usually resolves within a week, and the changes seen in magnetic resonance imaging (MRI) resolve over days to weeks.