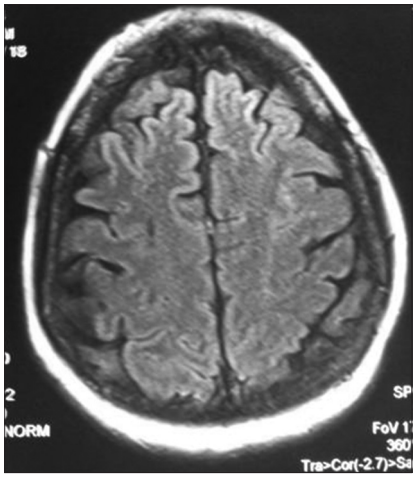

Figure 5: Repeat fluid attenuation inversion recovery image showing resolution of white matter edema in bilateral cerebral hemisphere

cause brain and systemic hypoperfusion, which may be causative factors for PRES in SLE. On the other hand, endothelial cell activation is one of the pathogenic hallmarks of neuropsychiatric SLE (NPSLE). It usually occurs after exposure to interleukin 1 (IL-1) and tissue necrotic factor-α (TNF-α), and may be enhanced by local release of IL-1 and IL-6. SLE patients with high SLE disease activity index have increased serum levels of TNF-α and other pro-inflammatory cytokines that may stimulate endothelial cells of intracranial vessels and astrocytes to produce nitric oxide, causing BBB damage and plasma leakage. In some cases the endothelial dysfunction together with hemodynamic factors may allow the leakage of blood plasma and large amounts of red blood cells resulting in secondary parenchymal hematoma. Histopathology showed the PRES manifestation result from NPSLE were due to focal cerebral edema associated with blood vessel injury and ischemic changes, although in many cases histopathology did not demonstrate specific lesions. SLE patients might develop reversible focal neurological deficits, which responded to steroid therapy.[4]

Even though the classical neurolupus includes seizures and psychosis, a number of other features such as myelopathy, optic neuropathy, meningitis, cognitive dysfunction, and cerebral infarction could be seen in SLE. PRES has been claimed as a particular form of neurological manifestation of SLE with characteristic MRI findings and a usual good outcome. Antihypertensive, antiepileptic, and supportive care are the mainstay of treatment.[5]

In some cases, the diagnosis of PRES remains in doubt. In this situation, regression of the clinical and radiological abnormalities with appropriate treatment supports the diagnosis. Thus, repeated brain imaging is beneficial of diagnosis.[6] Radiographically, PRES is heralded by relatively symmetric, reversible T2 hyperintensities affecting the posterior aspects of the brain, namely the occipital and parietal lobes. It is now known that this description is more of a general rule, and those asymmetric images can be seen, and can involve the deep grey matter as well as the frontal and temporal lobes. The advent of diffusion weighted imaging helped clarify that the MRI changes were not due to ischemia or cytotoxic edema, but due to vasogenic edema.[7]

In our case, as per diagnostic criteria for SLE,[8] more than four well documented features were present, that is, history of arthralgias, photosensitivity, polyserositis, renal impairment and nervous system involvement. Although her vasculitic profile was negative, but her brain imaging was suggestive of diffuse white matter edema, she was treated as seronegative SLE presenting as PRES. She received pulse therapy of i.v. methylprednisolone followed by oral steroids as per body weight. Patient improves clinically and her repeat imaging, done after 6 weeks, was almost normal.

REFERENCES

1. Hinchey J, Chaves C, Appignani B, Breen J, Pao L, Wang A, Pessin MS, Lamy C, Mas JL, Caplan LR. A reversible posterior leukoencephalopathy syndrome. *N Engl J Med* 1996;334:494-500.
2. Legriel S, Pico F, Azoulay E. Understanding posterior reversible encephalopathy syndrome. In: Vincent JL, editor. Annual Update in Intensive Care and Emergency Medicine 2011. Berlin, Germany: Springer; 2011. p. 631-53.
3. Hedna VS, Stead LG, Bidari S, Patel A, Gottipati A, Favilla CG, Salardini A, Khaku A, Mora D, Pandey A, Patel H, Waters MF. Posterior reversible encephalopathy syndrome (PRES) and CT perfusion changes. *Int J Emerg Med* 2012;5:12.
4. Liu B, Zhang X, Zhang FC, Yao Y, Zhou RZ, Xin MM, Wang LQ. Posterior reversible encephalopathy syndrome could be an underestimated variant of "reversible neurological deficits" in systemic lupus erythematosus. *BMC Neurol* 2012;12:152.
5. Marrone L, Streck Ade S, Staub HL, de Freitas CZ, Costa J, Gadonski G, Luiz Staub H. Posterior reversible encephalopathy syndrome (PRES) and systemic lupus erythematosus: report of two cases. *Rev Bras Reumatol* 2012;52:804-10.
6. Kadikoy H, Haque W, Hoang V, Maliakkal J, Nisbet J, Abdellatif A. Posterior reversible encephalopathy syndrome in a patient with lupus nephritis. *Saudi J Kidney Dis Transpl* 2012;23:572-6.
7. Graham BR, Pylypchuk GB. Posterior reversible encephalopathy syndrome in an adult patient undergoing peritoneal dialysis: a case report and literature review. *BMC Nephrol* 2014;15:10.
8. Tan EM, Cohen AS, Fries JF, Masi AT, McShane DJ, Rothfield NF, Schaller JG, Talal N, Winchester RJ. The 1982 revised criteria for the classification of systemic lupus erythematosus. *Arthritis Rheum* 1982;25:1271-7. Updated by: Hochberg MC. Updating the American College of Rheumatology revised criteria for the classification of systemic lupus erythematosus. *Arthritis Rheum* 1997;40:1725.

Cite this article as: Verma S, Yousuf I, Wani MA, Asimi R, Saleem S, Mushtaq M, Shah I, Nawaz S, Daga RA. Posterior reversible encephalopathy syndrome due to seronegative systemic lupus erythematosus. Neuroimmunol Neuroinflammation 2014;1(2):89-91.

Source of Support: Nil. **Conflict of Interest:** No.

Received: *09-02-2014*; **Accepted:** *18-07-2014*