

Nephroquiz
(Section Editor: M. G. Zeier)

Transient blindness and seizures in severe lupus nephritis

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Keywords: hypertension; lupus nephritis; posterior reversible encephalopathy syndrome; systemic lupus erythematosus

Case presentation

A 19-year-old woman with severe lupus nephritis was brought to the emergency department for blindness and generalized seizures.

Class IV lupus nephritis had been diagnosed 1 year earlier and treated with corticosteroids and mycophenolate mofetil. Three months before admission, serum creatinine was 1.0 mg/dL. Poor compliance to prescribed therapy 2 months before admission led to worsening nephrotic syndrome (proteinuria 22 g/day) and renal failure (creatinine 5.1 mg/dL), with a systemic lupus erythematosus (SLE) disease activity index of 39. A second kidney biopsy confirmed severe class IV lupus nephritis with crescents in ~50% of the glomeruli and a histological activity index of 18/42. Despite aggressive immunosuppressive therapy including corticosteroids, cyclophosphamide and rituximab, haemodialysis had to be initiated 2 weeks before the current admission.

On the morning of admission, she complained of acute onset blindness and headache, and developed generalized tonic-clonic seizures. Physical examination revealed hypertension (155/100 mmHg), obtundation and visual loss. There were no hydroelectrolytic disturbances, and cerebrospinal fluid examination was normal. Fundoscopic examination showed a grade 2 hypertensive retinopathy. Magnetic resonance imaging showed bilateral parieto-occipital hyperintensities on fluid-attenuated inversion recovery (FLAIR) sequences (Figure 1A), consistent with cerebral oedema.

Question

What is the diagnosis and how should this patient be managed?

Discussion

Clinical and radiological findings in our patient are pathognomonic of posterior reversible encephalopathy syndrome (PRES). This rare syndrome was first described in 1996 by Hinchey in 15 patients with headache, altered mental functioning, seizures and loss of vision, associated with posterior leucoencephalopathy on imaging studies [1]. The term PRES was suggested in 2000 by Casey *et al.* who demonstrated the utility of FLAIR sequences in the detection of bilateral cortical and subcortical hyperintensities of white and grey matter with a predominantly posterior distribution [2]. Conditions associated with this syndrome are hypertensive encephalopathy, eclampsia, renal disease, organ transplantation, immunosuppressive agents and systemic diseases such as SLE [1]. The pathophysiology of PRES is not fully understood. A brain capillary leak syndrome secondary to hypertension and/or cytotoxic effects of drugs or diseases has been proposed. The role of SLE itself in the pathogenesis of PRES remains unclear, but the combination of recent onset hypertension, fluid retention due to renal disease, the use of cytotoxic drugs and possibly endothelial dysfunction related to the disease exposes SLE patients at a particular risk for this complication. The respective roles of SLE flare on the one hand, and corticosteroids, cyclophosphamide and rituximab used in the treatment of severe SLE flare on the other hand, are difficult to delineate. Three retrospective reviews have recently analysed the characteristics of 98 patients with SLE and PRES [3–5]. Although the onset of PRES was related to a SLE flare in >90% of the cases, other factors were often present such as hypertension (82–95%), renal insufficiency (73–84%) and the use of immunosuppressive agents (50%) [4,5].

Prompt recognition of this spectacular clinico-radiological syndrome is crucial because of its potential reversibility with appropriate management. Indeed, intensive treatment of hypertension and other worsening factors, including SLE flare, in combination with anti-convulsant therapy, usually leads to complete resolution of symptoms

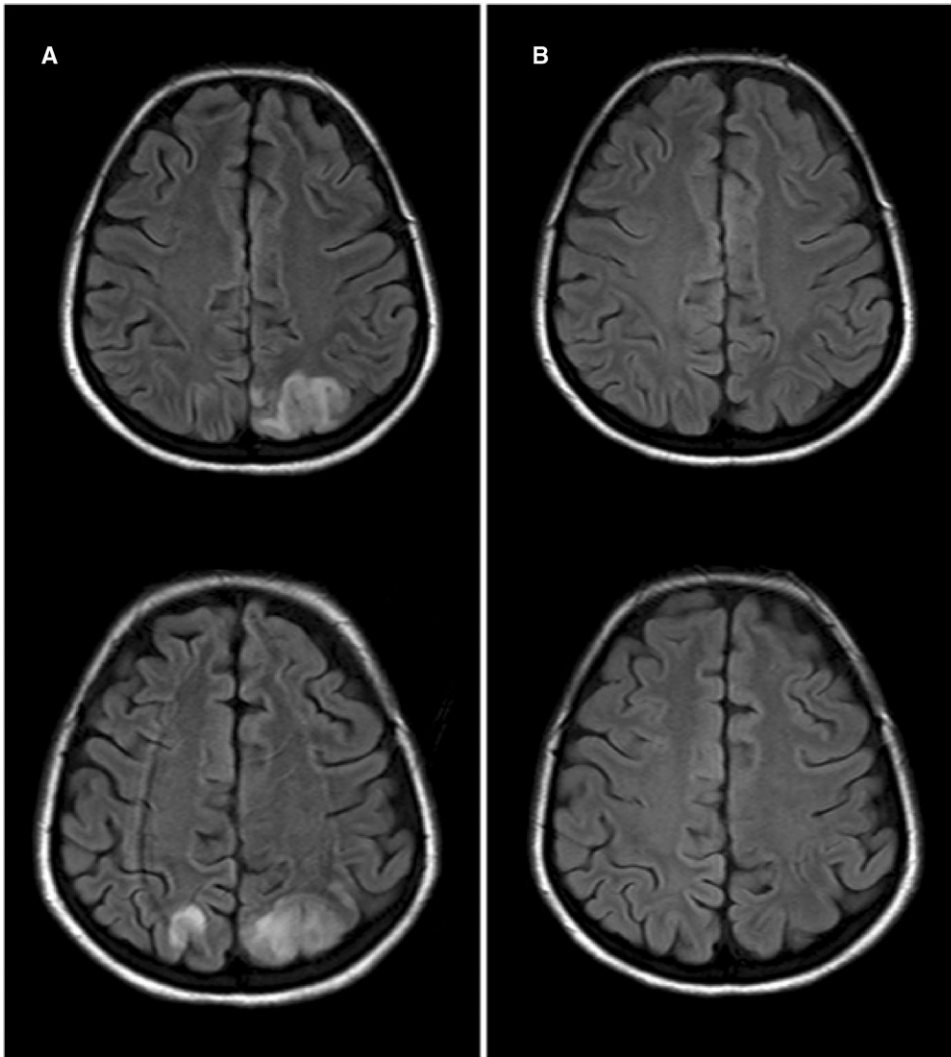


Fig. 1. FLAIR sequences of cerebral magnetic resonance imaging show bilateral occipital cortical hyperintensities on admission (A), and complete resolution of posterior cerebral oedema two months later (B).

and normalization of radiological findings, as in our patient (Figure 1B, 2 months later).

Conflict of interest statement. None declared.

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Received for publication: 25.2.09; Accepted in revised form: 14.5.09