Reversible Posterior Encephalopathy Syndrome in Systemic Lupus Erythematosus and Lupus Nephritis

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Abstract

Reversible posterior encephalopathy syndrome (RPES) is a clinical entity characterized with headache, nausea, vomiting, seizures, consciousness disturbance, and frequently visual disorders associated with neuroradiological findings, predominantly white matter abnormalities of the parieto-occipital lobes. The central nervous system manifestations of systemic lupus erythematosus (SLE) are highly diverse. However, SLE-associated RPES has been seldom reported. Here, we report a case with RPES in SLE and lupus nephritis with exclusive involvement of parietal and occipital cortices. A systematic review of the literature on the pathogenesis and treatment of SLE-associated RPES is included.

Key words: Reversible posterior encephalopathy syndrome (RPES), Systemic lupus erythematosus (SLE), Lupus nephritis, Systematic review

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Introduction

Reversible posterior encephalopathy syndrome (RPES), more commonly called reversible posterior leukoencephalopathy syndrome (RPLS), is a clinical entity characterized by headache, nausea, vomiting, seizures, conscious disturbance, and visual disorder associated with neuroradiological findings, predominantly white matter abnormalities of the parieto-occipital lobes (1, 2). It has been recognized in a wide variety of conditions, including hypertensive encephalopathy, organ transplantation, uremia, preeclampsia/eclampsia, and connective tissue diseases, which is reversible if treated promptly (1-5). Additional risk factors include thrombotic thrombocytopenic purpura, cerebral angiograms, hypertension-inducing treatments such as erythropoietin, blood transfusions, or immunosuppressive therapy particularly cyclosporine A and tacrolimus (FK506), and immunoglobulin as well as various chemotherapeutic agents such as cisplatin (6-11). The central nervous system (CNS) manifestations of systemic lupus erythematosus (SLE) are highly diverse, including cerebrovascular disease, myelopathy, optic neuropathy, meningitis, epileptic seizures, cognitive dysfunction, psychosis, dementia, anxiety, and depression (12). However, few reports have described RPES in SLE. Here, we describe a patient with SLE-associated RPES and systematically reviewed the pathogenesis and treatments of 22 patients with SLE-associated RPES reported in the literature.

Case Report

A 27-year-old woman was admitted for headache, nausea, vomiting, and blurred vision in February 2004. She had had a history of SLE and progressive lupus nephritis for 1 year. The diagnosis was based on American College of Rheumatology (ACR) criteria. Three months before admission the...
dosage of prednisone she had been receiving was reduced to 2.5 mg/d. Then she presented with peripheral edema two weeks before admission. The dosage of prednisone was increased to 50 mg/d. Three days later the edema resolved but she developed the above-mentioned symptoms. She was only able to distinguish between light and dark. After admission, she presented with transient visual inversion, which disappeared two days later. There was no personal or family history of neurological diseases.

On admission her blood pressure was 184/114 mmHg. Her vision was so poor that she only could see the movement of hand near her eyes. Pupils were 3.0 mm in each eye and were reactive to light. Optical fundus examinations were unremarkable and disc borders were sharp. Serum albuminemia was 26.3 g/l, hemoglobin 81 g/l, erythrocyte sedimentation rate 36 mm/1st h, urea nitrogen 21.2 mg/dl, serum creatinine 0.88 mg/dl, C, 0.37 g/l (normal range, 0.75-1.2 g/l), and C4, 46 mg/l (150-350 mg/l). Antinuclear, anti-double-stranded DNA, extractable nuclear antigen, and anti-Ro/SS-A antibodies were positive. Anticardiolipin antibody IgG was negative. Urinalysis revealed proteinuria (+3) and hematuria. Cerebrospinal fluid analysis and cultures were negative. Renal biopsy with light microscopy and immunofluorescence test showed severe and diffuse mesangial proliferative glomerulonephritis, focal endocapillary proliferation and cell crescent (Fig. 1A, B) with basement membrane deposits of IgG, IgM, and C1q, without IgA, C3, C4 or fibrinogen deposition.

On the ninth day after admission, the patient had bilateral parietal cortical lesions with hypointensity on T1-weighted MR imaging (Fig. 2B). The diffusion-weighted imaging (DWI) showed isointensity in the bilateral parietal lobes except for a patchy lesion with hyperintensity in a right parietal gyrus (Fig. 2C). The apparent diffusion coefficient (ADC) maps revealed lesions with hyperintensity in the bilateral parietal cortices, among which there was a patchy lesion with hypointensity in the gyrus corresponding to the lesion on DWI (Fig. 2D). Fluid-attenuated inversion-recovery (FLAIR) MR imaging showed lesions with hyperintensity in the bilateral parietal and occipital cortices (Fig. 2E, F). On the fifteenth day after admission, proton magnetic resonance spectroscopy (MRS) in the lesion showed NAA/ (Cho+Cr) ratio increased and Cho/Cr ratio decreased when compared with the relevant values in the brain region without abnormal signal, which indicated neuronal/axonal dysfunction (Fig. 3). Brain MR venography was normal.

The patient was diagnosed with SLE, lupus nephritis, secondary hypertension, and SLE-associated RPES. She was treated with antihypertensive medication (cilazapril, 5 mg per day). Prednisone and mycophenolate mofetil were used as immunosuppressants to control the activity of SLE and erythropoietin was used to treat anemia. In the subsequent 2 days, the symptoms of headache, vomiting, and visual blurring improved, and visual inversion disappeared. Four days later, her blood pressure was controlled at 147/80 mmHg and all symptoms regressed. One month later, the patient was discharged with no neurological deficit. Those lesions were completely resolved in the follow-up T2-weighted MRI (Fig. 2G), DWI (Fig. 2H), and ADC maps (Fig. 2I) at 70 days after the first MRI scan.

### Systematic Review of the Literature

In total, there were 276 papers in the Medline database from January 1966 to August 2007 when the key words of “leukoencephalopathy and reversible” were used. Among them, 14 papers described 22 patients with RPES in SLE (Table 1) (1, 2, 5, 13-23). Non-English language literature was excluded.

The age of onset ranged from 13 to 40 years old; there was one man and 22 women (including the present case). Symptoms included seizures (91%), headache (70%), visual disturbance (65%), altered mental status (48%), vomiting (26%), and hemiparesis (13%). Except for the patient receiving cyclosporine (case 11), all had blood pressure above normal, ranging from 156/94 to 220/150 mmHg. Lesions on CT or MRI located in posterior parieto-occipital lobes (100%), frontal lobes (39%), temporal lobes (35%), cerebellum (22%), thalamus (13%), brainstem (13%), basal ganglia (9%), or corona radiate (4%). It is noteworthy that, shortly before the syndrome developed, 15 patients (75%) had signs of rapidly progressive renal failure with acute elevation of blood pressure developing against the background of chronic pathology related to SLE [3 patients (case 4, 8, 9) without available data were excluded] and 17 patients (100%) exposed to corticosteroids, azathioprine and/or cyclophos-
Figure 2. A. Cranial T1-weighted magnetic resonance imaging (TR=450.0; TE=10.0) nine days after admission showed lesions with hypointensity in the bilateral posterior parietal cortexes (white arrows). B. Cranial T2-weighted magnetic resonance imaging (TR=3,500.0; TE=95.0) nine days after admission showed symmetric lesions with hyperintensity in the bilaterally posterior parietal cortexes (black arrows). C. Cranial diffusion-weighted imaging (TR=3,000.0; TE=84.0; ep_b1000t/90) showed isointensity in the bilateral parietal lobes except for a patchy lesion with hyperintensity in a right parietal gyrus (thin white arrow). D. Cranial apparent diffusion coefficient maps (TR=3,000.0; TE=84.0; ep_b0_1000/90) revealed lesions with hyperintensity in the bilateral posterior parietal lobes (black arrow), however, a patchy lesion with hypointensity was shown in the gyrus of right parietal lobe (thin black arrow). E, F. Cranial fluid-attenuated inversion-recovery imaging (TR=9,980; TE=108; TI=2,500.0) nine days after admission showed symmetric lesions with hyperintensity in the bilaterally posterior parietal cortexes (E), and occipital cortexes (F). G, H, I. Two months after admission these lesions were completely resolved on follow-up T2-weighted magnetic resonance imaging (TR=4,000.0; TE=120.0/1) (G), diffusion-weighted imaging (TR=3,000.0; TE=84.0; ep_b1000t/90) (H), and apparent diffusion coefficient maps (TR=3,000.0; TE=84.0; ep_b0_1000/90) (I).

Phamide (often high dose) [6 patients (case 2, 3, 4, 8, 9, 13) without available data were excluded]. The neurological signs of all patients, except for case 8, were resolved within 4 days to 1 month after appropriate antihypertensive and anticonvulsant therapy. Case 8 had lesions with a mixture of cytotoxic and vasogenic edema in bilateral parieto-occipital
lobes in the acute stage, and suffered permanent neurological deficit eventually.

**Discussion**

(1) **Neuroimaging and differential diagnosis of SLE-associated RPES**

RPES was firstly coined by Hinchey et al in 1996 (1). Typical lesions are hyperintense on T2-weighted MR images; they are usually hypointense or isointense on DWI, with an increase of the ADC, indicating vasogenic edema (24, 25). The lesions of vasogenic edema in RPES would switch to cytotoxic edema if the precipitating factors were not removed promptly. In the present patient, within the lesions of vasogenic edema (isointensity in DWI and hypointensity in ADC map, Fig. 2C, D) that were exclusively located in parietal and occipital cortices, there was a patchy lesion of cytotoxic edema (hyperintensity in DWI and hypointensity in ADC map, Fig. 2C, D). If our patient had not been treated promptly, those lesions, especially the lesion with cytotoxic edema, might have become more severe and
irreversible. Evolution of initial vasogenic edema into cytotoxic edema, infarction, or hemorrhage has been reported (14, 19, 24), for example, case 8 in Table 1 who suffered permanent neurological deficit had mixed lesions of cytotoxic and vasogenic edema (14).

Proton MRS of the present case showed that the white matter had normal spectroscopy (Fig. 3), which supported the exclusive involvement of the cortex. Casey et al (2) found cortical involvement in 94% of patients with RPES (among them one patient had exclusive involvement of the cortex) and constituted 46% of all lesions, and predominantly cortical involvement tended to be found more often in the mild cases while predominantly subcortical white matter edema was seen more often in the patients with moderate or severe disease. Because cortical involvement is frequent and there is usually not an accompanying destructive process of the white matter, or leukoencephalopathy in those patients, the name of PRES is more suitable than RPLS. In the present case, the determination of lesions on MRI was reliable, because the MRS, done at 6 days after the first MRI scan, confirmed the lesions in the bilateral parieto-occipital cortices and showed abnormal spectroscopy in the lesion (Fig. 3); furthermore, the multiple sequences of MRI during the first scan showed the same lesions (Fig. 2), and clinical manifestation of bilaterally severe visual disturbance corresponded well to the lesion locations.

RPES lesion is very similar to and easily misdiagnosed as ischemic stroke (with cytotoxic edema), which is a common CNS complication of SLE. RPES needs prompt antihypertensive treatment, while ischemic stroke needs mildly elevated blood pressure to improve cerebral perfusion. DWI can help to differentiate an ischemic lesion from RPES lesion since the former (either mild or severe one) had hyperintensity in DWI and hypointensity in ADC maps, while the later had hypo- or isointensity in DWI and hyperintensity in ADC maps (19, 24, 25). Furthermore, infarct lesion always

### Table 1. Clinical Characteristics and Neuroimaging Findings of 23 Patients with SLE and RPES

<table>
<thead>
<tr>
<th>No</th>
<th>Age/sex</th>
<th>Bp (mmHg)</th>
<th>Serum creatinine (mg/dl)</th>
<th>precipitating factors</th>
<th>Symptoms and signs</th>
<th>Lesions on CT or MRI</th>
<th>Complete resolution of neurologic signs</th>
</tr>
</thead>
<tbody>
<tr>
<td>1*</td>
<td>27/F</td>
<td>184/114</td>
<td>0.88</td>
<td>HP, MP</td>
<td>headache, vomiting, visual blurring, visual inversion</td>
<td>pet (B), occ (B)</td>
<td>within 4 days</td>
</tr>
<tr>
<td>2[1]</td>
<td>50/M</td>
<td>200/110</td>
<td>3.3</td>
<td>HP</td>
<td>headache, lethargy, vomiting, cortical blindness</td>
<td>pet (B), occ (B), temp (B), fr (L), thalamus (L)</td>
<td>within 2 weeks</td>
</tr>
<tr>
<td>3[1]</td>
<td>40/M</td>
<td>200/150</td>
<td>3.1</td>
<td>HP</td>
<td>headache, seizures, vomiting, confusion, hemiparesis</td>
<td>occ (L), pet (B), temp (L), fr (L), occipital (L)</td>
<td>within 2 weeks</td>
</tr>
<tr>
<td>4[2]</td>
<td>15/F</td>
<td>100</td>
<td>NaH</td>
<td>HP</td>
<td>status epilepticus</td>
<td>pet (B), occ (B), temp (B), fr (B), thalamus (L), corona radiata (L)</td>
<td>Complete resolution</td>
</tr>
<tr>
<td>5[3]</td>
<td>22/F</td>
<td>200/130</td>
<td>4.4</td>
<td>HP, RF, CYC, MP, CYC</td>
<td>headache, seizures, visual blurring, diaphoria, confusion, hemiparesis</td>
<td>ftr (B), occ (B), temp (B), chl (B)</td>
<td>within 4 days</td>
</tr>
<tr>
<td>6[3]</td>
<td>22/F</td>
<td>170/110</td>
<td>9.4</td>
<td>HP, RF, MP, CYC, CYC</td>
<td>headache, vomiting, seizures, confusion, visual blurring</td>
<td>occ (B), pet (B), temp (B)</td>
<td>within 10 days</td>
</tr>
<tr>
<td>7[3]</td>
<td>30/M</td>
<td>210/125</td>
<td>6.2</td>
<td>HP, MP</td>
<td>seizures, confusion, stupor, visual blurring</td>
<td>occ (B), ftr (B)</td>
<td>within 2 weeks</td>
</tr>
<tr>
<td>8[4]</td>
<td>22/F</td>
<td>200/144</td>
<td>NaH</td>
<td>HP</td>
<td>seizures</td>
<td>pet (B), occ (B), ftr (B)</td>
<td>permanent neurologic deficits</td>
</tr>
<tr>
<td>9[4]</td>
<td>27/F</td>
<td>174/105</td>
<td>NaH</td>
<td>HP</td>
<td>seizures, visual loss, confusion</td>
<td>pet (B)</td>
<td>within 2 weeks</td>
</tr>
<tr>
<td>10[5]</td>
<td>30/F</td>
<td>170/100</td>
<td>RF, CYC</td>
<td>cortical blindness, status epilepticus</td>
<td>chl (L)</td>
<td>within 2 weeks</td>
<td></td>
</tr>
<tr>
<td>11[6]</td>
<td>24/F</td>
<td>130/80</td>
<td>Normal CYC</td>
<td>Pemphigus, headache, GTCS</td>
<td>ftr (B), pet (B), occ (B)</td>
<td>within 1 month</td>
<td></td>
</tr>
<tr>
<td>12[7]</td>
<td>13/F</td>
<td>215/100</td>
<td>Normal HP, CSF</td>
<td>headache, vomiting, confusion, GTCS</td>
<td>occ (B), pet (B)</td>
<td>within 6 days</td>
<td></td>
</tr>
<tr>
<td>13[8]</td>
<td>20/F</td>
<td>220/150</td>
<td>2.2</td>
<td>HP</td>
<td>seizures, headache, visual blurring</td>
<td>occ (B), pet (B), ftr (B), temp (B), chl (B)</td>
<td>after 4 separate seizures</td>
</tr>
<tr>
<td>14[9]</td>
<td>22/F</td>
<td>200/110</td>
<td>3.8</td>
<td>HP, MP, CYC</td>
<td>headache, diaphoria, confusion, seizure, sensory, blindness</td>
<td>pet (B), temp (B), chl, basal ganglia (B), brainstem</td>
<td>within 4 days</td>
</tr>
<tr>
<td>15[9]</td>
<td>37/M</td>
<td>175/97</td>
<td>3.7</td>
<td>MP, PC</td>
<td>cortical blurring, GTCS</td>
<td>occ (B), temp (L), basal ganglia (L)</td>
<td>within a week</td>
</tr>
<tr>
<td>16[9]</td>
<td>24/F</td>
<td>210/100</td>
<td>5.2</td>
<td>HP, RF, MP</td>
<td>headache, diaphoria, GTCS</td>
<td>occ (B), thalamus (B), brainstem</td>
<td>Within 24 hours</td>
</tr>
<tr>
<td>17[9]</td>
<td>32/F</td>
<td>150/94</td>
<td>1.8</td>
<td>MP, CYC</td>
<td>headache, GTCS</td>
<td>occ (B), pet (B), chl</td>
<td>Complete resolution</td>
</tr>
<tr>
<td>18[9]</td>
<td>30/F</td>
<td>130/110</td>
<td>3.1</td>
<td>RF, MP, CYC</td>
<td>headache, GTCS</td>
<td>occ (B)</td>
<td>within 2 days</td>
</tr>
<tr>
<td>19[5]</td>
<td>40/F</td>
<td>180/100</td>
<td>3.0</td>
<td>HP, RF, MP</td>
<td>headache, GTCS</td>
<td>pet (B), ftr (L), thalamus (L)</td>
<td>within 1 month</td>
</tr>
<tr>
<td>20[20]</td>
<td>25/F</td>
<td>190/127</td>
<td>Normal HP, Aza, Pred</td>
<td>headaches, dizziness</td>
<td>occ (B)</td>
<td>2 weeks later</td>
<td></td>
</tr>
<tr>
<td>21[21]</td>
<td>27/F</td>
<td>165/105</td>
<td>Normal HP, Pred, Aza</td>
<td>cortical blindness, confusion, hemiparesis, gait preference, GTCS</td>
<td>occ (B)</td>
<td>within a week</td>
<td></td>
</tr>
<tr>
<td>22[20]</td>
<td>30/F</td>
<td>100</td>
<td>HP, RF, MP</td>
<td>stupor, seizures, paresis</td>
<td>occ (B)</td>
<td>after 10 days</td>
<td></td>
</tr>
<tr>
<td>23[20]</td>
<td>20/F</td>
<td>190/110</td>
<td>4.5</td>
<td>HP, RF</td>
<td>headache, seizures, cortical blindness</td>
<td>occ (B)</td>
<td>within 2 days</td>
</tr>
</tbody>
</table>

*Present case


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follows a vascular territory that usually is not symmetric; the calcarine fissure is often involved in posterior cerebral infarction but it is normally spared in RPES (1, 19), for example, Fig. 2F of the present patient showed no lesion in the calcarine fissure but distinct lesions in symmetric cortical cortices. The above-mentioned small patchy portion of the lesion of the present case had cytotoxic edema (with hypointensity in DWI and hyperintensity in ADC map, Fig. 2C, D), which was reversible and could be mild ischemia; however, the rest of the lesion had vasogenic edema (isointensity in DWI and hypointensity in ADC map, Fig. 2C, D) and was not an ischemic change. All of the lesions of the present case still existed on the localizing slices for MRS at 6 days after the first MRI scan (Fig. 3), and disappeared at 70 days after the first MRI scan, which indicated the lesions had lasted for more than 6 days (because the first MRI scan was done at 9 days after the appearance of the visual disturbance) and were not considered as ischemic change, because an ischemic lesion lasting for more than 6 days would leave a permanent lesion of encephalomalacia and would not disappear totally on MRI after several months. The distinction between RPES and CNS vasculitis is also important in patients with SLE. Management of RPES may require withdrawal of immunosuppressants, whereas CNS vasculitis requires immunosuppressive therapy. CNS vasculitis lacks the typical MRI findings of RPES, and diagnosis relies on corroborative data from cerebrospinal fluid analysis, angiography, and perfusion studies, and is made definitively by brain biopsy (19).

(2) Systematic review

In the present series, it was noted that SLE-associated RPES has been increasingly reported from 1996 to 2007 (Fig. 4) in part due to the fact that MR imaging has been widely used and much attention has been paid to this syndrome. It favored women mostly because SLE always happens in women, and the lesion locations, symptoms, and outcome were similar to those of patients with other causative factors. Just before the syndrome developed, most of these patients had signs of rapidly progressive renal failure with acute elevation of blood pressure. It seems that the occurrence of SLE-associated RPES was basically caused by hypertension induced by fluid retention secondary to acute renal failure and the high dose of corticosteroids. Anti-hypertension treatment was the most important to reverse the RPES lesion.

(3) Pathophysiology of SLE-associated RPES and immunosuppressive treatment

The mechanism behind the development of PRES is yet unproved. Two broad theories have been considered. Severe hypertension with autoregulatory failure and hyperperfusion, resulting in vasodilation and subsequent vasogenic edema, is often cited as the underlying mechanism (26-29). Alternatively, vasospasm has been demonstrated (catheter angiography, MR angiography), which could result in decreased cerebral blood flow [shown on MR perfusion, single photon emission CT (SPECT)], ischemia, and cytotoxic edema (30-33). The main difference between those two hypotheses is
with reference to the change of cerebral perfusion.

There have been some reports on the existence of hyperperfusion in PRES lesions. Schwartz et al (27) found increased vascular perfusion adjacent to lesions in 2 patients with reversible hypertensive encephalopathy (in accordance with the diagnosis of PRES). Schwartz (28) reviewed the clinical and radiographic signs of hypertensive encephalopathy and found that, all patients had lesions predominantly in occipital and/or parietal lobes, and the lesion had increased cerebral perfusion on SPECT and perfusion MR imaging, did not show restricted diffusion. The syndrome resolved completely in most cases after the administration of antihypertensive agents. Wartenberg and Parra (29) found increased cerebral blood volume and blood flow in both parietal-occipital lesions on perfusion computed tomography imaging in a patient with PRES caused by triple-H therapy for symptomatic subarachnoid hemorrhage.

There have also been reports on the existence of cerebral hypoperfusion and vasoconstriction in PRES. Tajima et al (30) found temporary hypoperfusion in the PRES-lesions on Xe-SPECT and reversible abnormal irregular narrowing of the posterior cerebral artery on angiography. By using stable xenon CT perfusion imaging, Casey et al (31) found hypoperfusion within a parietal occipital lesion in a PRES patient with a renal transplant who presented with hypertension and seizures; after the high blood pressure was controlled, the hypoperfusion changes were resolved. Using serial MR imaging techniques, Brubaker et al (32) found there was a significant increase in ADC values in the lesions of all patients with PRES, and a significant decrease in cerebral blood volume (CBV) and cerebral blood flow (average 30% of control). Bartynski and Boardman (33) found that vasculopathy (ie, vasoconstriction or vasodilatation) was a common finding on catheter angiography and MR angiography in the patients with PRES, and a MR perfusion study demonstrated reduced cortical CBV in PRES lesions.

Whether having cerebral hypoperfusion or hyperperfusion, the vast majority of the reported cases of PRES had lesions of vasogenic edema, which had been demonstrated by DWI and ADC mapping (24, 25, 28, 29, 32, 33); only a few reported cases had cytotoxic edema which were always mixed with vasogenic edema (9, 14, 32, 34, 35). That indicated that, generally, this kind of hypoperfusion would not cause brain ischemia if treated promptly. We suppose that vasculopathy (either vasoconstriction or vasodilatation), abnormal cerebral perfusion (hypo- or hyper-perfusion), and brain edema (mostly vasogenic) are parts of pathophysiological change caused by various agents, such as hypertension, organ transplantation, uremia, eclampsia, immunosuppressive agent, and connective tissue disease. Removing those precipitates promptly is the most important for recovery. Both of the above hypotheses are one-sided and not comprehensive and the pathogenesis of PRES needs further clarification.

In our series of SLE-associated RPES, we found that most of the common precipitants of RPES (1, 3, 5), including acute elevations of blood pressure, renal decompensation, fluid retention, and treatment with immunosuppressants, are frequently found in SLE patients, especially in the period of acute exacerbation when an extended course of intravenous cyclophosphamide and high-dose methylprednisolone are usually suggested. It is difficult to ascertain the exact contribution of cyclophosphamide as compared with hypertension and fluid overload (resulting from renal failure and high-dose methylprednisolone). We postulate that, all the mechanisms, together with the possible role of autoimmune-mediated vascular injury secondary to the activity of SLE, might contribute to the development of SLE-associated RPES. SLE patients, especially those with CNS involvement, always have significant cerebral hypoperfusion (36, 37). Furthermore, there was a patchy ischemic portion within the PRES lesion in our patient, which may have been due to the severe cerebral hypoperfusion around the lesions, although angiography and CBF imaging were not done. Involvement of the CNS in SLE is sometimes caused by cerebral embolism, which has been confirmed by several studies on detectability of microembolic Doppler signals in the intracranial circulation. Therefore, the patchy ischemic lesion in the present case was also probably caused by cerebral embolism (38, 39). However, normally the embolic infarct would leave a permanent lesion of encephalomalacia since it was not a temporary vasoconstriction.

In the present patient, mycophenolate mofetil and erythropoietin were unlikely the precipitating factor of RPES because RPES occurred before they were used. To date, we found no report on an association between mycophenolate mofetil and RPES. After admission she was treated with antihypertensive drugs, while immunosuppressants (methylprednisolone and mycophenolate mofetil) were continued. Her RPES was completely reversed. Although sudden discontinuation of immunosuppressants may be helpful to RPES it might risk the subsequent rapid deterioration of renal function in the phase of activity. In the last decade, sequential regimens of short-term cyclophosphamide induction followed by mycophenolate mofetil maintenance have been shown to be more efficacious and safer than the long-term exposure to cyclophosphamide (40, 41). As suggested, it is safer to change the immunosuppressive therapy to mycophenolate mofetil and high-dose corticosteroids need not be withdrawn in treating SLE-related RPES, as long as the blood pressure and fluid retention are well controlled (18, 19).

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