Recurrent posterior reversible encephalopathy syndrome in systemic lupus erythematosus

Melissa Ng, Sadia Saber, Richard Stratton

ABSTRACT

Posterior reversible encephalopathy syndrome (PRES) is an acute encephalopathy that manifests as headache, visual disturbance, altered mental state, and seizures. There are striking characteristic findings on neuroimaging. The PRES is associated with a number of conditions, including autoimmune disease. We describe the case of a 37-year-old female with a history of systemic lupus erythematosus presenting with headache and visual changes. Prompt diagnosis in PRES is important because if it is not recognized and treated early, it may progress to irreversible neurological damage. This patient made a good initial recovery, but suffered a relapse secondary to severe resistant lupus nephritis and refractory hypertension.
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Keywords: Encephalopathy, Headache, Lupus, Posterior reversible encephalopathy syndrome (PRES), Systemic lupus erythematosus (SLE)

INTRODUCTION

Headache with visual changes is a common presentation with a broad differential and it is important not to miss serious causes. Neuroimaging is becoming more readily available, leading to quicker diagnosis of intracranial pathology. Posterior reversible encephalopathy syndrome (PRES) should be suspected in cases presenting with headache, visual disturbance, altered mental state, and seizures. Its associations include toxic agents, hypertension, sepsis, and autoimmune diseases. The PRES, as the name implies, is potentially reversible provided it is recognized and treated early through control of blood pressure and of the underlying cause [1]. However, if it is not adequately treated, it can progress to irreversible neurological damage, hemorrhage, and infarction [2]. Early diagnosis may also be important because treatment of other causes can differ, for example, hypertension as a cause of PRES is controlled as a mainstay of treatment, whereas hypertension in the context of ischemic strokes is more cautiously treated [2].

CASE REPORT

A 37-year-old female presented to the emergency department with a six-day history of worsening headache, severe photophobia, generalized body aches and weakness, and episodes of pyrexia, rigors, and vomiting. She had a background of systemic lupus erythematosus (SLE) complicated by lupus nephritis, pancytopenia, and antiphospholipid syndrome, migraine, and previous complex partial seizures. Previously, she had a left preretinal hemorrhage while on low molecular weight
heparin therapy. She was taking prednisolone 10 mg per
day and mycophenolate mofetil 1 g twice a day, and was
having monthly plasmapheresis. She was a non-smoker
and non-drinker. There was no relevant family history.
She had a heart rate 115 beats/min, respiratory rate
19 mmHg, temperature 37.5°C, blood pressure 124/88
breaths/min, and oxygen saturations 99% on room air.
On examination, heart sounds were normal, chest was
clear, and abdomen was soft and non-tender. She had
peripheral pitting edema. Neurologically, she was very
photophobic and had loss of vision in both eyes. She was
unable to finger count. No ocular pathology was found by
ophthalmology.

Computed tomography scan of head with contrast
showed extensive bilateral low attenuation changes along
the white matter tracts, particularly in the parietal and
occipital lobes. There was no evidence of hemorrhage
and no filling defects identified along the venous sinuses.
Magnetic resonance imaging (MRI) scan showed bilateral
T2/FLAIR hyperintense change involving the parieto-
occipital lobes – features most consistent with PRES.
There was also mild generalized cerebral volume loss.

The patient’s blood pressure increased during her
admission to 170 systolic. She was started on nifedipine
and perindopril to control her blood pressure and limit
progression of PRES. The dose of prednisolone was
increased to 40 mg daily. She was discharged after five
days with rheumatology follow-up.

The patient was re-admitted several weeks later
with a progression of lupus nephritis and refractory
hypertension (blood pressure 190/110 mmHg). During
her admission, she experienced seizures. Repeat neuro-
imaging showed that the previously abnormal areas had
almost completely resolved. There were new areas of
signal change in the frontal, parietal, and occipital lobes,
as well as the cerebellar hemispheres, midbrain and
pons. She then had another episode of severe headache
and visual loss. Imaging once again showed significant
resolution but revealed new lesions in the occipital and
frontal lobes, right corpus callosum, and left caudate. The
development and resolution of lesions are demonstrated
in Figures 1–3. The blood results at the time of each
flare are given in Table 1. Her condition improved
after treatment with intravenous labetalol and nitrate,
steroids, plasma exchange, intravenous immunoglobulin,
and rituximab. She has now made a good recovery and
remains clinically stable.

DISCUSSION

Posterior reversible encephalopathy syndrome is an
under-recognized clinical and radiological syndrome
which usually presents with headache, visual changes
including cortical blindness, nausea and vomiting,
altered mental state, and seizure activity [3]. Acute
hypertension is strongly associated with PRES, though it
does not correlate with severity [2]. Posterior reversible
encephalopathy syndrome has been associated with toxic
agents such as immunosuppressive therapies, sepsis, pre-
eclampsia and eclampsia, and autoimmune conditions
such as systemic lupus erythematosus, systemic sclerosis,
and polyarteritis nodosa and other vasculitides.

Magnetic resonance imaging scan remains the
gold standard of diagnosis in PRES. Previously, the
radiological findings were classically reported as cerebral
edema along the white matter tracts in the posterior
parietal and occipital lobes [4], but other patterns on
neuroimaging are increasingly recognized [2]. The
pathophysiology is not well understood but the current
theory postulates that impaired cerebral autoregulation
results in vasogenic edema secondary to capillary leakage
and endothelial disruption [4].
A number of cases of PRES have been described in the context of SLE [1, 5]. Given the multisystem nature of SLE, it is likely that there are several contributing factors to the development of PRES, such as hypertension, renal disease, and use of immunosuppressive agents. Furthermore, given the wide range of neuropsychiatric manifestations in SLE, the diagnosis of PRES can be difficult if neuroimaging is not readily available. The fact that our patient was not hypertensive at the time of presentation clouded the picture given the strong association between hypertension and PRES.

The mainstay of treatment of PRES is early blood pressure control and removal of the causative agent to prevent progression to permanent neurological damage. Prompt treatment has a good prognosis, and patients often make full neurological recovery, but the risk of neurological impairment and up to 15% risk of mortality need to be noted [2]. Recurrent PRES is uncommon. This patient had acute flare-ups of severe resistant lupus nephritis and refractory hypertension which contributed to recurrent PRES.

**CONCLUSION**

In conclusion, posterior reversible encephalopathy syndrome presents as headache, visual changes, altered mental state, and seizures. It is associated with toxins, hypertension, sepsis, and autoimmune conditions. Typical magnetic resonance imaging scan show cerebral edema in the white matter tracts with predominance towards the posterior parietal and occipital lobes. This case brings to light a few key take-home lessons: firstly, early diagnosis of posterior reversible encephalopathy syndrome (PRES) is crucial so blood pressure and any underlying cause can be aggressively treated. Secondly, flares of associated conditions can lead to PRES recurrence. Finally, it is important to bear in mind that PRES if often, but not always, reversible and can result in permanent neurological damage.

**Table 1: Blood test results**

<table>
<thead>
<tr>
<th></th>
<th>Admission</th>
<th>5 weeks</th>
<th>7 weeks</th>
<th>6 months</th>
</tr>
</thead>
<tbody>
<tr>
<td>Hemoglobin</td>
<td>89</td>
<td>82</td>
<td>84</td>
<td>82</td>
</tr>
<tr>
<td>White blood cells count</td>
<td>2.64</td>
<td>6.67</td>
<td>5.91</td>
<td>6.61</td>
</tr>
<tr>
<td>Platelets</td>
<td>69</td>
<td>178</td>
<td>43</td>
<td>353</td>
</tr>
<tr>
<td>Urea</td>
<td>2.9</td>
<td>16.4</td>
<td>15.1</td>
<td>15.1</td>
</tr>
<tr>
<td>Creatinine</td>
<td>60</td>
<td>304</td>
<td>346</td>
<td>158</td>
</tr>
<tr>
<td>Erythrocyte sedimentation rate</td>
<td>23</td>
<td>95</td>
<td>19</td>
<td>53</td>
</tr>
<tr>
<td>uPCR</td>
<td>1592</td>
<td>1525</td>
<td>1399</td>
<td>364</td>
</tr>
<tr>
<td>dsDNA</td>
<td>200</td>
<td>99</td>
<td>15</td>
<td>19</td>
</tr>
<tr>
<td>Lactate dehydrogenase</td>
<td>266</td>
<td>278</td>
<td>346</td>
<td>188</td>
</tr>
</tbody>
</table>

Abbreviations: uPCR: Urinary protein creatinine ratio, dsDNA: Double stranded DNA

**Author Contributions**

Melissa Ng – Substantial contributions to conception and design, Acquisition of data, Analysis and interpretation of data, Drafting the article, Revising it critically for important intellectual content, Final approval of the version to be published

Sadie Saber – Substantial contributions to conception and design, Acquisition of data, Revising it critically for important intellectual content, Final approval of the version to be published

Richard Stratton – Substantial contributions to conception and design, Acquisition of data, Analysis and interpretation of data, Revising it critically for important intellectual content, Final approval of the version to be published

**Guarantor**
The corresponding author is the guarantor of submission.

**Conflict of Interest**
Authors declare no conflict of interest.

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