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 neuroimaging 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.

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
Sadia 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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Table 1: Blood test results

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

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