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CASE REPORT

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Keywords: Encephalopathy, Headache, Lupus, Posterior reversible encephalopathy syndrome (PRES), Systemic lupus erythematosus (SLE)

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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 pre-retinal 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

Figure 1: Magnetic resonance imaging scan of head, axial section, T2-weighted images, (A) (On admission): hyperintense changes in parieto-occipital lobes, (B) (Five weeks): resolution of changes. New patchy areas of high signal change in frontal and parietal regions, and (C) (Seven weeks): significant resolution with new area at right posterior temporal-occipital junction.

Figure 2: Magnetic resonance imaging scan head, axial section, T2-weighted images, (A) (On admission): bilateral hyperintense changes in parieto-occipital lobes, (B) (Five weeks): resolution of changes, (C) (Seven weeks): new lesions in occipital lobes, posterior temporal-occipital junction, splenium of corpus callosum.

Figure 3: Magnetic resonance imaging scan head, axial section, T2-weighted images, (A) (On admission): no significant areas of hyperintense change, (B) (Five weeks): extensive signal change of pons, right middle cerebellar peduncle, cerebellar hemispheres, and inferior vermis, and (C) (Seven weeks): resolution of previously abnormal areas.

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].

Table 1: Blood test results

	Admission	5 weeks	7 weeks	6 months
Hemoglobin	89	82	84	82
White blood cells count	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 sedimentation rate	23	95	19	53
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

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 is 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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