

# Posterior reversible encephalopathy syndrome: a rare case with delayed reverse

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### Abstract

*Posterior reversible encephalopathy syndrome (PRES) is a reversible phenomenon diagnosed with both clinical and radiological criteria. We present an atypical case of PRES with unusual delayed reversal. A 29-year-old female presented with seizure and altered consciousness during post-partum periods. The initial computed tomography scan of brain was normal. But subsequent magnetic resonance imaging of brain showed hyperintense lesion in right parieto-occipital region. After 7 weeks of multidisciplinary team effort, the patient recovered both clinically and radiologically. High index of suspicion with long term follow up is crucial for this curable and reversible syndrome. Because of rarity in delayed recovery, this case of PRES is reported.*

**Keywords:** Posterior reversible encephalopathy syndrome, vasogenic edema, delayed recovery.

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### Introduction

Posterior reversible encephalopathy syndrome (PRES) is a clinical-radiological diagnosis which is often under diagnosed or misdiagnosed due to rarity of the disease. PRES manifests clinically with headache, visual disturbance, seizure or decreased consciousness.<sup>1</sup> Radiologically, cerebral edema at parietal and occipital lobes is diagnostic for PRES.<sup>1</sup> Vasogenic edema in the posterior cerebral circulation region is the hallmark for the diagnosis of PRES.<sup>1</sup> The commonest etiology for PRES is eclampsia.<sup>2,3</sup> Post-partum eclampsia is also a common cause for PRES and the average duration from delivery to hospital admission due to PRES is 6.9 days.<sup>3</sup> Subjective improvement occurs within few weeks and improvement of imaging may require several weeks.<sup>3</sup>

Due to its reversibility and complete recovery, it is important to diagnose and treat PRES cases in the early stages. Previously very few cases were reported as post-partum PRES.<sup>3-5</sup> Again, to the best of our knowledge, only one case had been reported with an unusual delayed recovery from PRES due to co-morbid conditions (acute renal failure with electrolyte imbalance).<sup>4</sup> Therefore, we present this rare case of PRES with delayed reversal.

### Case report

A 29-year-old normotensive, non-diabetic lady in her post-partum period presented with seizure and impaired consciousness. Seizure was sudden onset, generalized tonic-clonic, single episode followed by loss of consciousness. On admission at emergency department, the Glasgow Coma Scale (GCS) was 6 (E1V2M3) (E= eye opening, V= verbal Response, M= motor response), pupil was normal-reactive and symmetrical, respiration was normal and blood pressure (BP) was 210/110 mm Hg. On neurological examination, there was equivocal bilateral plantar response. Bilateral papilloedema (Grade 2) was found on ophthalmoscopic examination. Other systemic examination revealed no abnormality.

Initial routine investigation revealed mild anemia (hemoglobin 10.1 gm/dl), slightly raised serum

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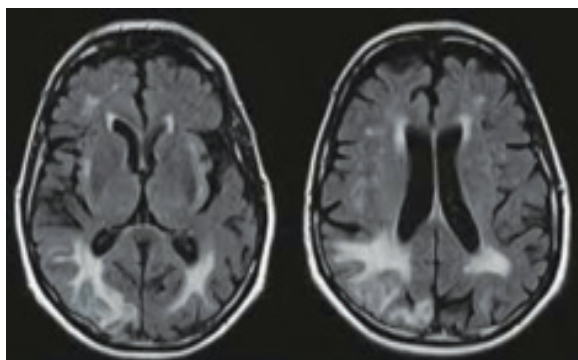
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creatinine (1.4 mg/dl), hyperglycemia (random blood glucose 11.4 mmol/l) and hyponatremia (serum sodium 130 meq/l). Immediate computed tomography (CT) scan of brain appeared normal. Magnetic resonance imaging (MRI) of brain obtained after 18 hours revealed hyper-intense lesion in the bilateral occipital lobes (Figure 1). Diffusion weighted imaging (DWI) showed restricted diffusion in occipital region.

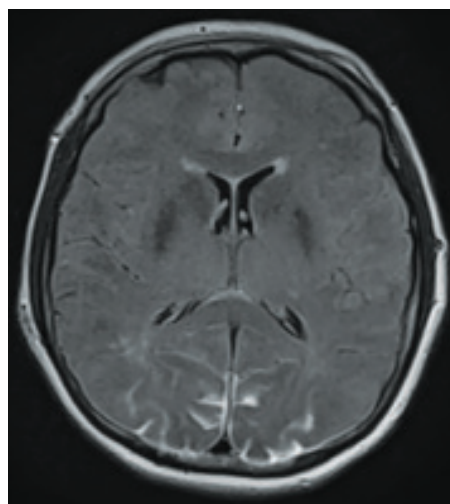


**Figure 1** MRI of brain (FLAIR) showing hyper-intense lesion in parieto-occipital region (after 18 hours of admission)

The patient was diagnosed and treated as hypertensive encephalopathy. The management goal was to treat the hypertension, hyperglycemia, electrolyte imbalance and prevention of complications. The management was continued by transferring the patient to a higher center where she was managed in high dependency unit (HDU) with oral combined anti-hypertensive drugs (amlodipine 5 mg and olmesartan 20 mg daily through nasogastric tube) and continuous monitoring of intake output, blood glucose and vital signs. Regular follow up was given with correction of hyperglycemia, electrolyte imbalance and pulmonary infection. The medical team (Neurologist, Internist and Physiotherapist) decided to continue the treatment in HDU setting. At the 4<sup>th</sup> week, the patient developed aspiration pneumonia and pressure sore (sacral) which was diagnosed by routine blood tests, chest x-ray and culture (blood and pus) and managed successfully (antibiotics and regular dressing). There was no improvement of GCS (less than 8) during the first 6 weeks.

The GCS of the patient started to improve gradually at 7<sup>th</sup> week of her admission. The GCS was found 8 (E1V3M4) at 7<sup>th</sup> week and 11 (E2V4M5) at the 9<sup>th</sup>

week. At the end of 9<sup>th</sup> week, the patient was able to communicate fully with GCS 12 (E3V4M5) and was transferred from HDU to Physical Medicine Department for rehabilitation. Clinically, the patient was functionally independent at 11<sup>th</sup> week. Follow up MRI of brain showed significant resolution of parieto-occipital lesion after week 9 (Figure 2).



**Figure 2** Follow up MRI of brain (FLAIR) showing marked decrease of hyper-intensity in parieto-occipital region (at 9<sup>th</sup> week)

### Discussion

PRES is a reversible condition which involves mainly the posterior part of the brain.<sup>1,2</sup> PRES was first reported and described in 1996 as a reversible encephalopathy.<sup>2,5</sup> This syndrome was first reported as reversible posterior leukoencephalopathy syndrome (RPLS) which was re-named as PRES in the year 2000.<sup>2,6</sup> Although the mechanism or pathogenesis of this reversible syndrome is still not clear, brain autoregulation mechanism and cerebral blood vessel endothelium dysfunction may have key role for this disease.<sup>1,6</sup> Sudden hypertensive episode can lead to failure of cerebral autoregulation mechanism. This causes acute disruption of blood brain barrier which causes reversible cerebral edema.<sup>6</sup>

MRI of brain can be the diagnostic imaging modality for PRES. T2-weighted images and fluid attenuated inversion recovery (FLAIR) images in MRI can detect the vasogenic cerebral oedema by hyper-intense signal change at posterior subcortical region.<sup>7</sup> Parietal and occipital lobes are commonly involved in PRES. But,

presented with altered consciousness for an unusual longer period of time (7 weeks) and eventually recovered both clinically and radiologically. Although the recovery in our case was incomplete due to multiple risk factors (prolonged stress hyperglycemia and pulmonary infection) and very long disease course, the reversibility was marked after a period of no response.

The overall outcome of PRES depends on abnormalities in imaging like infarction, hemorrhage and brain edema (mainly vasogenic edema).<sup>8</sup> Hyperglycemia is an important factor for poor outcome in PRES.<sup>2,8</sup> Septicemia and dementia can also play significant role in the overall outcome in PRES.<sup>9</sup> The lesion size in MRI corresponds with the overall outcome in PRES.<sup>9</sup> In our case, multiple factors like hyperglycemia, chest infection and large lesion size (parietal and occipital lobe) might be the reason for the delayed recovery.

Complete recovery is found in more than 50% cases in PRES. The mortality is high (15%) if the treatment is delayed or inadequate.<sup>8,9</sup> In severe PRES, the reversibility can be complete if diagnosed early, treated and rehabilitated correctly.<sup>9,10</sup> Underdiagnosed or misdiagnosed cases can lead to massive ischemic changes in the brain which is irreversible and carries poor prognosis.<sup>10</sup>

### **Conclusion**

It is absolutely crucial to diagnose and treat PRES early because of its complete reversible outcome. As reversibility can be delayed for more than 2 months, high index suspicion and long term follow up care with multidisciplinary specialty team is necessary. Preventing poor prognostic indicators can ensure good outcome. Further clinical and radiological evaluation is needed for better understanding the mechanism, treatment

modalities and prognosis of atypical, delayed reversal cases of PRES.

**Conflict of interest:** Nothing to declare.

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