

# A case of recurrent posterior reversible encephalopathy syndrome

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## Summary

**Purpose:** To report a case of recurrent posterior reversible encephalopathy syndrome (PRES) after post-partum eclampsia. **Case Report:** A 22-year-old woman, para 3+0, presented with headaches, decrease level of consciousness, and convulsions after home delivery. Her second delivery was an emergency cesarean section due to eclampsia with PRES at 30 weeks of gestation. On examination she was unconscious with blood pressure of 220/120. She was intubated and transferred to the intensive care unit. Computer tomography scans confirmed the diagnosis of recurrent PRES. She recovered completely with supportive treatment and was discharged home one week after admission. **Conclusion:** Posterior reversible encephalopathy syndrome may recur in a subsequent pregnancy.

**Key words:** Posterior reversible encephalopathy syndrome; Recurrence.

## Introduction

Posterior reversible encephalopathy syndrome (PRES) is a rapid onset, clinicoradiological syndrome characterized by a range of neurological symptoms. This acute disorder normally manifests over a few hours and includes symptoms such as headache, confusion, seizures, and visual disturbances [1]. Typically, PRES is associated with acute hypertension resulting in subcortical vasogenic edema, of which the posterior regions of the cerebral hemispheres appear to be particularly susceptible. In 20-30% of cases, however, PRES can develop in normotensive individuals and is believed to be triggered by endothelial dysfunction caused by endogenous/exogenous toxins [2]. In very rare cases, PRES can also involve the medulla and/or cervical cord [3]. PRES, fortunately, is a reversible condition, and if promptly identified and treated, the disorder can usually be

resolved within a week [4]. The authors report a case of PRES after post-partum eclampsia in a 22-year-old woman.

## Case Report

A 22-year-old woman from Burkina Faso with an obstetric history of three term births, 0 pre-term births, 0 abortions, and three living children (T3, P0, A0, L3), presented to the emergency room with headaches, a decreased level of consciousness, and convulsions after delivering at home. She was unregistered in the present hospital and had not received proper antenatal care. Her previous first birth was a spontaneous vaginal delivery at full term and the second delivery was an emergency cesarean section due to eclampsia with PRES at 30 weeks of gestation. Axial and sagittal CT scans taken at this initial presentation of PRES displayed left occipital cortical and sub-cortical ill-defined low density (Figures 1 and 2). A follow-up axial T2 FLAIR sequence MRI scan performed two weeks later displayed no abnormal signal, hence indicating the resolution of PRES. Upon the home de-

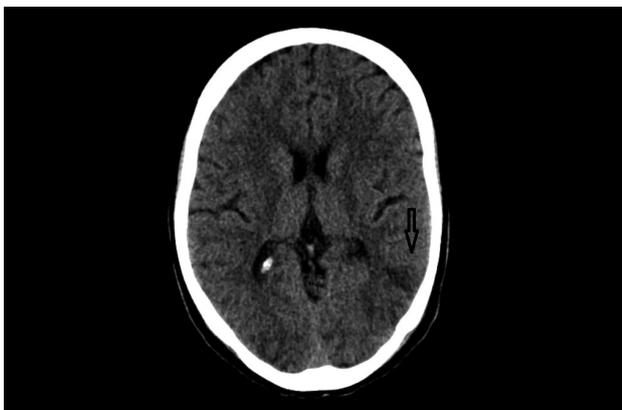


Figure 1. — Axial CT scan showing left occipital cortical and sub-cortical ill-defined low density (arrow).

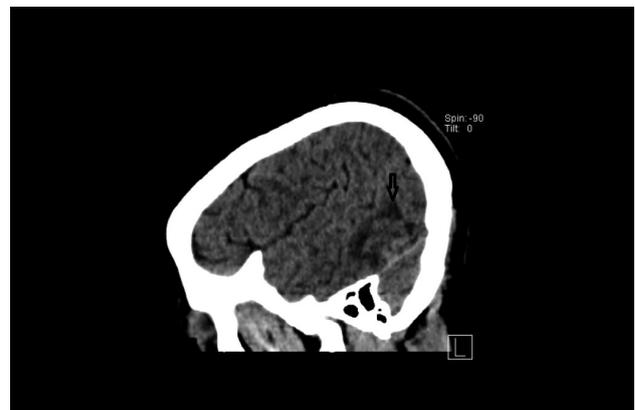


Figure 2. — Sagittal CT scan of the brain displaying left occipital cortical and sub-cortical ill-defined low density (arrow).

Revised manuscript accepted for publication May 25, 2017



Figure 3. — Axial CT brain scan depicting bilateral occipital vasogenic edema (arrow).



Figure 4. — Sagittal CT brain scan showing occipital vasogenic edema (arrow).

livery of her third child, 16 months later, the patient was taken to the emergency room. On initial examination, the patient was unconscious with a blood pressure reading of 220/120 mm Hg. She was intubated and transferred to the intensive care unit. Her coagulation profile and renal functions upon admission and in the subsequent days were all within the normal range. She had a hemoglobin level of 8.9 g/dL. The liver function tests were significantly deranged on admission, mainly with an elevated alkaline phosphatase at 309 U/L (normal range: 40–129), aspartate amino transferase at 526 U/L (10–40) alanine amino transferase at 181 U/L (0–41), and total bilirubin at 39  $\mu$ mol/L (3–17). Serum bicarbonates level was within the normal range. The patient underwent an urgent CT scan (Figures 3 and 4), which demonstrated the features characteristic of PRES.

The diagnosis of PRES was made based on the above findings, and her management was largely supportive. She was extubated on the third day after admission with rapid neurological recovery. Her liver function tests improved rapidly with third-day results showing alkaline phosphatase of 199 U/L, aspartate amino transferase at 96 U/L, alanine amino transferase at 100 U/L, and bilirubin at 21  $\mu$ mol/L. She recovered completely and was discharged home one week after admission.

## Discussion

From the current case study, it is clear that PRES may recur in a subsequent pregnancy. Fortunately, like the majority of PRES cases, the patient in question was correctly diagnosed and fully recovered within few days. Rapid withdrawal of the trigger usually accelerates the recovery; for example, delivery in eclampsia, blood pressure management or withdrawal from the offending drug [5]. In contrast, although prognosis is favorable in most cases, severe forms of PRES may result in acute hemorrhage or posterior fossa edema [6].

Currently, PRES is considered to be a rare disorder. However, epidemiological data must be interpreted with caution as this syndrome has likely been significantly underdiagnosed in the past [7].

Some case reports describing PRES in patients with eclampsia have been reported in the literature. In one report, three separate cases of women (19-, 24-, and 33-years of age) were diagnosed with eclampsia and also presented with acute PRES. All women were given antihypertensive drugs and were discharged within five to seven weeks after follow-up examination and neuro-imaging [8]. In a retrospective study of eclampsia with PRES cases managed between 2010 and 2014 at the Karnataka Institute of Health Sciences, Hubli, India, 11 out of 36,094 patients (0.03%) that delivered during this timeframe, were diagnosed with eclampsia complicated with PRES [9]. Furthermore, this complication was seen to be more common in post-partum eclampsia than antepartum (55% vs. 45%) and had a higher incidence in primigravida patients than multigravidas (81% vs. 19%) [9]. Also, the majority of cases (81%) were seen in young patients between 20–25 years. Interestingly, no cases were reported in teenage pregnancy nor patients over 30. In a similar study performed at the Mayo Clinic in Minnesota, USA between 2001 and 2008, 13 out of 17,317 women (0.075%) were diagnosed with eclampsia [10]. Upon review of their records, seven of these patients developed eclamptic seizures and underwent neuroimaging where radiologic evidence of PRES was found to be present in all seven women. In contrast to older reports, the lesions in these women were not predominately seen in the posterior cerebellum, but rather involved other regions of the brain. Patients in this study also developed PRES at a lower mean peak systolic blood pressure (SBP); 173 mmHg, than the mean peak SBP of 191 mmHg that was seen in the 113 PRES patients treated at the same clinic between 1999 and 2009 [10]. Of note, recurrent episodes of PRES are typically related to eclampsia and also, have an incidence that is proportional to recurrent eclampsia. In a retrospective review, three out of 78 patients (3.8%) developed recurrent PRES [11]. Infection was suspected in all

three patients during both episodes of PRES and evidence of endothelial injury was also reported in all patients. Furthermore, underlying conditions included antibody positive autoimmune disease, sickle cell disease, and allogenic bone marrow transplantation.

In all case studies of PRES and recurrent PRES reported, early diagnosis and treatment are highlighted as imperative measures for correct disease management. Furthermore, awareness of the diverse radiographic presentation of acute PRES is crucial to avoid misdiagnosis and treatment delay. Presently, MRI scans represent the most important diagnostic tools for this syndrome, and due to this neuroimaging technology, we may see an increase in the diagnosis of PRES in the future. In diffusion MRI images, evidence of PRES will be consistent with vasogenic edema and will show in a typical MRI as symmetric increase signals in the bilateral parietooccipital regions, subcortical white matter of the temporal and bilateral lobe, and in the posterior areas in T2A and FLAIR sequences [12].

To date, only two clinical trials have been attempted to evaluate the management of PRES. In 2013 a double-blind, placebo-controlled trial to determine if intravenous dexamethasone assists resolution of PRES encountered in eclamptic patients was attempted but had to be terminated due to a difficulty in recruiting participants (ClinicalTrials.gov identifier; NCT02027272). The second trial is currently recruiting and aims to describe the effects of different causes, imaging findings and laboratory parameters on the prognosis if this disorder (ClinicalTrials.gov identifier; NCT02665598). Hopefully, results from the clinical trial NCT02665598 and detailed case study reports will deepen our understanding of PRES and improve the management of this disorder.

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