

# International Journal of Clinical Obstetrics and Gynaecology

ISSN (P): 2522-6614  
ISSN (E): 2522-6622  
© Gynaecology Journal  
[www.gynaecologyjournal.com](http://www.gynaecologyjournal.com)  
2023; 7(5): 41-43  
Received: 22-06-2023  
Accepted: 26-07-2023

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## Expect the unexpected: A rare presentation of posterior reversible encephalopathy syndrome in late post-partum period

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**DOI:** <https://doi.org/10.33545/gynae.2023.v7.i5a.1378>

### Abstract

Posterior reversible encephalopathy syndrome (PRES) is an acute neurotoxic Syndrome characterized by a headache, seizures, altered mental status and visual loss and characterized by white matter vasogenic edema affecting the posterior occipital and parietal lobes of the brain predominantly. PRES is reversible when recognized early and treated promptly by treating the underlying causative factor. Common triggering factors of PRES include blood pressure fluctuations, preeclampsia/eclampsia, renal failure, cytotoxic agents, and autoimmune conditions. Here we present a case of young woman who presented with headache, generalized tonic-clonic seizures and cortical blindness in the late postpartum stage without any significant prior history in antenatal and postoperative period. Timely intervention, prompt management and diagnosis resulted in complete subsidence of symptoms pointing towards the reversible nature of the disease.

**Keywords:** Encephalopathy, post-partum period, timely intervention, prompt management

### Introduction

No one is so brave that he is not disturbed by something unexpected and to expect the unexpected is thoroughly a modern intellect. This stands true for late postpartum hypertensive convulsive disorder Eclampsia, which accounts for less than 16% of postpartum eclampsia. Late postpartum eclampsia occurring simultaneously with PRES, though rare but a proven entity that entails re-hospitalization, as suggested by recent evidences.

Posterior reversible encephalopathy syndrome (PRES) is a rare but serious clinical-neuroradiological entity, with manifestation of a series of nonspecific clinical signs and symptoms, including headache, vomiting, visual disturbances, altered mental status, seizures, and unconsciousness.

The etiology of PRES is not clear. However there are certain predisposing factors that are thought to be associated with PRES are hypertension, pre-eclampsia/eclampsia, immunosuppressive agents, cancer chemotherapy, autoimmune diseases, and sepsis. Among these factors, pre-eclampsia and eclampsia (3%-8.5%) are most commonly reported in the literature. There are a few atypical cases without the appearance of hypertension or proteinuria. Here we report a rare case of PRES with late postpartum eclampsia presenting on 7<sup>th</sup> postoperative day of LSCS, with no history of hypertension, proteinuria or any neurological ailment in the antenatal or immediate postoperative and convalescent period and was discharged from the hospital on 4<sup>th</sup> post-operative day.

### Case report

A 32 yrs woman, gravida 2 para 2 with history of one previous LSCS for CPD 6 yrs back, booked patient with regular antenatal check-up, underwent LSCS at 38 weeks of gestation, indication being malposition with oligohydramnios. She had an otherwise uneventful pregnancy except for hypothyroidism for which she was being treated with 125 mcg of levo-thyroxin. All her antenatal blood pressure recordings were within normal limits. Her blood sugar was normal. Family history was unremarkable. She never smoked and consumed alcohol. Her post-operative period was uneventful and she was discharged on 4<sup>th</sup> postop day. She presented on 7<sup>th</sup> postop day with complaint of severe headache and diminution of vision in both eyes.

On examination she was fully alert and oriented. Her temperature was 98.4 F, pulse rate 90 min and respiratory rate 16/min. blood pressure was 135/85 mm Hg at the time of presentation to the hospital but there was a history of two recordings of severely raised BP equivalent to 175/95 recorded at an interval of 30 min apart at home, Respiratory and cardiovascular examination were within normal limits. Abdominal examination revealed supra pubic tenderness. Renal angles were non-tender.

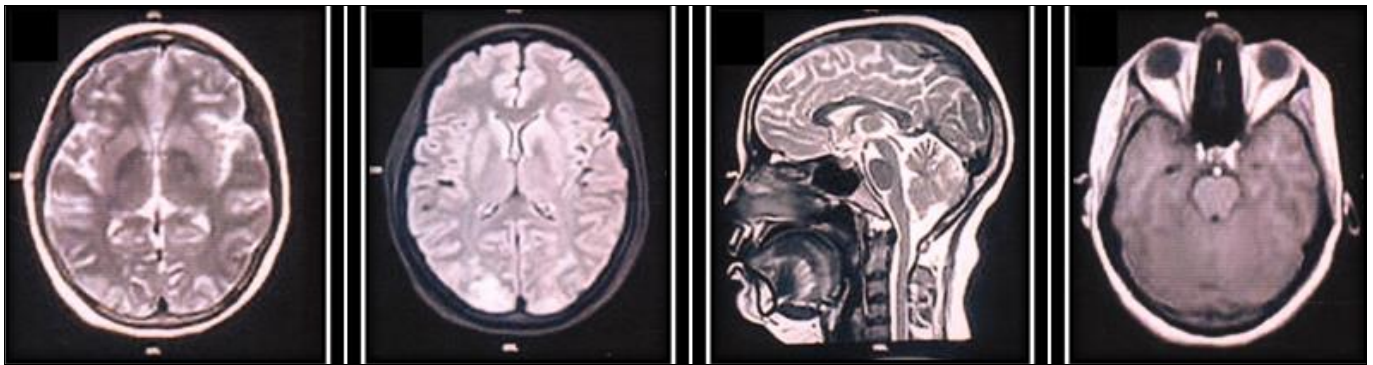
Two hours after hospitalization, she developed an episode of generalized tonic clonic convulsions following which she complained of loss of vision in both eyes. She was given supportive therapy and a neurophysician and ophthalmologist reference was made. Neurological examination revealed no signs of meningeal irritation, no sensory-motor deficit, no cerebellar signs and flexor planter reflex. Ophthalmological examination revealed normal tonometry and fundus examination showed normal optic disc, normal macula, foveal reflex present and vascular arcade was normal.

On investigation her haemogram showed Hb 12.9, WBC 11130 with neutrophils 84.6%, platelet count 310000, LFT and KFT are normal, and urinary protein was nil. High resolution MRI of brain revealed patchy areas of gyriform/subcortical symmetrical

areas of altered signal intensity in bilateral frontal, parieto-occipital lobes and bilateral cerebellar hemispheres. Signal changes are ISO to hypointense on T1Wt images and hyperintense on T2Wt images / FLAIR images. Basal ganglia, thalami, brain stem, and cerebellum show normal morphology and signal intensity. Visualized orbits are normal. The findings are in favour of Posterior reversible encephalopathy syndrome (PRES).

### Treatment

Since the patient presented to us late in the postpartum period with an acute episode of headache, convulsions and visual diminution, she was admitted in intensive care unit. Supportive therapy was given. Loading dose of phenytoin 1000 mg in 100 ml saline followed by 100 mg 8 hourly started along with levipil 500 mg 8 hourly given. Antibiotics were started on account of the raised WBC counts. The patient responded to the treatment and became capable to see in 36 hours. After the MRI Brain report consistent with PRES, the patient was assured of the reversible nature of the pathology and shifted to her room and discharged on 5<sup>th</sup> day with an advice to monitor her BP regularly. No antihypertensive was given as the BP remained normal throughout her hospital stay.



**Fig. 1:** Axial: T2Wt turbo-spin-echo (TSE), T2 and T1w FLAIR, DWI, SWI, Coronal: T2 TSE, Sagittal: T2 TSE. Findings as described are in favor of posterior reversible encephalopathy syndrome (PRES) with images showing Patchy areas of gyriform / subcortical nearly symmetrical areas of altered signal intensity seen in bilateral frontal, parieto-occipital lobes and bilateral cerebellar hemispheres. Signal changes are iso to hypo intense on T1Wt images and hyperintense on T2Wt/FLAIR images. No abnormal SWI blooming is seen. Rest of the bilateral cerebral hemispheres show normal differentiation of grey and white matter. Mild diffuse cerebral edema is seen with partial effacement of ventricular system and cortical sulci. Basal ganglia, thalami, brain stem and cerebellum show normal MR morphology and signal intensity. Sella, parasellar and suprasellar area are normal. Visualized orbit are normal.

### Discussion

Preeclampsia is a pregnancy specific hypertensive disorder with multi-systemic involvement and Eclampsia is one of the severe, convulsive manifestation of the hypertensive disorders of pregnancy. Eclampsia can occur antepartum (53%), intrapartum (19%) or postpartum (28%). Late postpartum eclampsia is the onset of convulsions more than 48 hrs. but less than 4 weeks after delivery. Rarely can it happen up to 6 weeks of delivery.

It has been seen that a significant proportion of women (20-38%) presenting with eclampsia may present without the classic signs of preeclampsia i.e., hypertension or proteinuria, similar to our patient who had no previous documentation of hypertension or proteinuria during any of her antenatal visits. The cerebral complications of preeclampsia are known but the underlying pathophysiological mechanism has not been fully elucidated and it is the endothelial dysfunction at the blood-brain barrier level resulting in cerebral blood flow alteration which could be the causative factor. Experts aren't sure what causes postpartum preeclampsia, but the American College of Obstetricians and Gynecologists (ACOG) points out that the fluid shifts that occur

after giving birth can possibly cause a rise in blood pressure levels during the three- to six-day period after labor and delivery.

Dynamic cerebral auto-regulation (DCA) is a physiological process that maintains cerebral blood flow relatively constant despite changes in blood pressure (BP). Altered DCA may cause over or under-perfusion injury and subsequent cerebral edema. The studies using transcranial Doppler ultrasound have consistently shown that there is an increased cerebral perfusion pressure (CPP) in preeclampsia.

Studies have demonstrated that the women with preeclampsia have depressed DCA compared with women with normal BP during pregnancy and this depressed DCA may contribute to cerebral complications such as cerebral edema, convulsions, and stroke.

Posterior reversible encephalopathy syndrome (PRES) is a clinico-radiological entity characterized by neurologic signs and symptoms such as vision loss or deficit (26%-67%), seizure (92%), headache (26%-56%), and consciousness impairment (13%-90%). Although suspicion for PRES is increased in the

setting of these clinical features, the diagnosis of PRES is made by the presence of vasogenic edema and FLAIR hyperintensities in the posterior aspects of the brain on magnetic resonance imaging involving the posterior circulation. An association between eclampsia and PRES was first described by Hinchey *et al.* in 1996. Women are particularly at risk of PRES in the settings of eclampsia and preeclampsia.

Other conditions that require to be distinguished in such presentations is - Reversible cerebral vasoconstriction syndrome and cerebral venous thrombosis. Reversible cerebral vasoconstriction syndrome, usually occurs postpartum, is characterized by the reversible multifocal narrowing of the arteries of the brain with signs and symptoms that typically include thunderclap headache, recurrent seizures and, focal neurologic deficits and the MRI shows cerebral vasoconstriction, string of bead appearance on angiography. However cerebral venous thrombosis can occur either in third trimester or postpartum with persistent and progressive headache, seizures, focal neurological deficits and MRI will show brain oedema or intracerebral haemorrhage.

Principles of management is mainly focused on symptomatic therapy along with medical control of hypertension, antiepileptic medication and long-term neurologic follow-up.

Prognosis of PRES is excellent and the symptoms usually resolves in 3-8 days (75%-90%), while MRI abnormalities may take longer to resolve - several days to week. Outcome may be guarded when there is associated co-morbidities like sepsis or intracranial haemorrhage. Recurrence (4%) can be there if hypertension is uncontrolled or the patient is on immunosuppressant therapy. Misdiagnosis and delayed treatment could lead to permanent neurological damage.

## Conclusion

Postpartum period, though joyous but an important period when the body undergoes physical and emotional changes. Monitoring of postpartum health is of utmost importance because of the added stress of breastfeeding the newborn and sleep deprivation. Mothers should be particularly advised to be vigilant of their physical changes and monitor temperature, blood pressure or any newly developed neuropsychiatric symptoms and refer to the hospital at the earliest. Post-partum eclampsia complicated with PRES is a known entity. MRI is the key to diagnosis and early recognition and treatment is the essence of complete recovery.

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### How to Cite This Article

Sinha R, Shukla L. Expect the unexpected: A rare presentation of posterior reversible encephalopathy syndrome in late post-partum period. *International Journal of Clinical Obstetrics and Gynaecology.* 2023;7(5):41-43.

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