

Posterior reversible encephalopathy syndrome presenting as bilateral blindness in a post partum female

Abstract

Posterior reversible encephalopathy syndrome (PRES) previously is an europhthalmic disorder characterized by painless loss of vision, seizures, headache, and altered mental status and is associated with white matter vasogenic edema predominantly affecting the occipital and parietal lobes of the brain. It has several etiologies but one of the most common includes eclampsia. This case report describes a rare scenario where a patient presented with complete bilateral loss of vision due to PRES syndrome. She was 3 days postpartum and had undergone caesarian section for pre-eclampsia. Her condition recovered completely with treatment within a week of presentation.

Keywords: posterior reversible encephalopathy syndrome, eclampsia, cortical blindness

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Introduction

Posterior reversible encephalopathy syndrome (PRES) is a clinic radiological entity that was first reported by Hinchey et al.,¹ in 1996 based on 15 cases. He described it as white matter edema presenting with headache, altered mental functioning, seizures, and abnormalities of visual perception. This condition has been called previously by several terms such as reversible posterior cerebral edema syndrome, reversible posterior leukoencephalopathy syndrome, and reversible occipital parietal encephalopathy. It was Stott et al.,² who proposed the term Posterior reversible encephalopathy syndrome (PRES) for the condition. PRES is characterized by visual abnormalities, loss of consciousness, seizures, consciousness, headaches and focal neurological signs. Causes of PRES are diverse and include pre-eclampsia, eclampsia, renal insufficiency, solid organ transplantation and immunosuppressive therapy.³ Some relatively rarer causes include use of bevacizumab, head injury, seizure and migraine.⁴ Though several cases of PRES have been reported in neurology and neuroradiology, ophthalmic literature has had limited exposure and this case report tries to describe a common presentation of PRES along with its management.

Case report

A 20year old female patient presented to the emergency department with history of bilateral loss of vision since three days. She complained of painless non progressive complete loss of vision, which was sudden in onset in both the eyes immediately after undergoing lower segment caesarian section under spinal anesthesia for pre-eclampsia in another center. There was no history of nausea, vomiting or convulsions. On examination, her visual acuity was perception of light with inaccurate projection of rays bilaterally. Pupils were 3mm in size, normally reactive and afferent papillary defect was absent. Anterior segment examination was unremarkable. No abnormality was detected on fundus examination. Her neurological examination was also within normal limits. Magnetic Resonance Imaging (MRI) T2 and Fluid-Attenuated Inversion Recovery (FLAIR) sequences showed bilateral hyper-intensities primarily located in the cortex and adjacent sub-cortex of occipital and temporal lobes (Figure 1) (Figure 2). Blood pressure was 142/96mmHg on presentation. Other systemic findings including central nervous system examination was within normal limits.

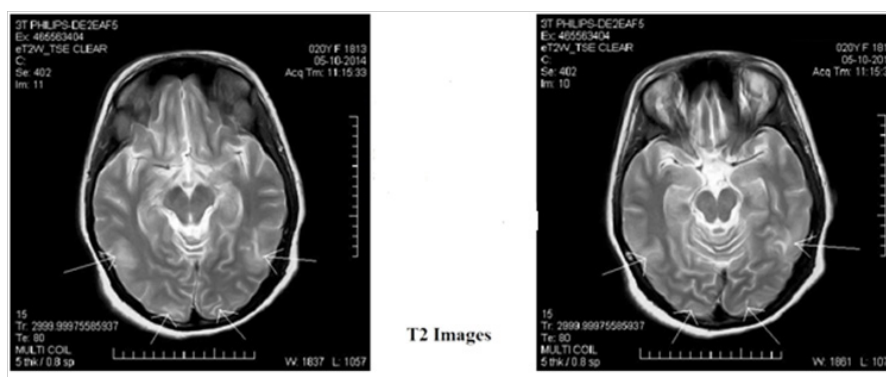


Figure 1 Bilateral hyperintensities primarily located in the cortex and adjacent sub-context of occipital and temporal lobes.

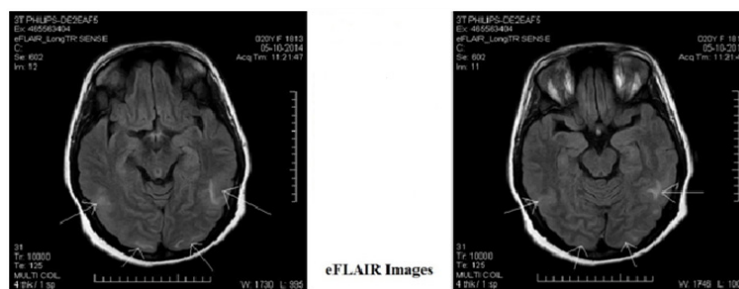


Figure 2 Bilateral hyperintensities primarily located in the cortex and adjacent sub cortex of occipital and temporal lobes.

With all these clinical and MRI findings, the patient was diagnosed as a case of Posterior reversible encephalopathy syndrome (PRES). She was admitted and treated with Intravenous Mannitol 1g/kg body wt., Intravenous Dexamethasone 10mg and Oral Nifedipine 60mg (Procardia XL, Pfizer Inc.). After 2 days of treatment, her best corrected visual acuity improved to counting fingers at 5 meters distance in right eye and 6/60 in left eye, colour vision could be recorded at this stage and was found to be normal. One week later, the patient was discharged from the hospital with complete visual recovery (best corrected visual acuity of 6/6 in both the eyes).

Discussion

PRES is associated with diverse clinical aetiologies and hypertensive encephalopathy is critical in the pathogenesis.¹ There are three proposed hypotheses of PRES pathophysiology: 1) cerebral vasoconstriction causing subsequent infarcts in the brain, 2) failure of cerebral autoregulation with vasogenic edema, and 3) endothelial damage with blood-brain barrier disruption further leading to fluid and protein transudation in the brain.⁵ Patients usually complain of headache and manifest fluctuating neurologic symptoms and signs, especially visual symptoms. Sometimes cerebral infarction ensues, but typically, the clinical and imaging findings suggest that ischemia reverses completely. MRI findings are characteristic with the edema present within the occipital lobes. Our patient also complained of headache, had loss of consciousness along with painless loss of vision. Two close differential diagnosis of this disorder are reversible cerebral vasoconstriction syndrome (RCVS) which typically presents with sudden, severe headache, sub arachnoid and intracerebral haemorrhage in patients with new-onset, severe hypertension⁴ and Bálint's syndrome characterized by inability to perceive the visual field simultaneously, difficulty in fixating the eyes and inability to move the hand to a specific object by using vision (optic ataxia) due to damage to the parieto-occipital lobes on both sides of the brain.⁶

Modern imaging techniques and experimental models suggest that vasogenic edema is typically the primary process leading to neurologic dysfunction; therefore, prompt recognition and management of this condition should allow for clinical recovery as long as superimposed hemorrhage or infarction has not occurred.⁴ In a review study by Seet et al.,⁷ 12 cases were identified that developed PRES following systemic bevacizumab treatment. All cases occurred secondary to rise in blood pressure due to bevacizumab treatment and responded well to blood pressure control and bevacizumab treatment. Bevacizumab is routinely used in high dose by oncologist for various cancer such as ovarian cancer, bowel cancer and the higher dose may be the factor responsible for PRES occurrence. It is known to cause rise in blood pressure in normotensive patients⁸ and maybe the precipitating

factor leading to vasogenic edema. As other anti-Vascular endothelial growth factor (Anti-VEGF) such as ranibizumab are not commonly used systematically, it is unknown if PRES develops only with bevacizumab or all anti-VEGF drugs.

Although elevated blood pressure is common in this condition, it can also occur secondary to calcineurin-inhibitor toxicity that is thought to occur secondary to breakdown of the blood brain barrier by dysfunction of the capillary endothelium itself.⁴ Pregnancy predisposes brain to vasogenic edema and this vasogenic edema of white matter may occur with normal or mildly elevated pressure due to endothelial dysfunction.⁹ PRES has been reported in patients aged 4 to 90 years, although most cases occur in young to middle-aged adults, the mean age ranging across case series from 39 to 47 years. There is a marked female predominance.³ In several studies, Pre-eclampsia/eclampsia has been observed as the etiological factor in 7% to 20%¹ of patients with PRES. The outcome in such cases has always been favorable. Our patient was a 20 year old female with pre-PRES due to pre-eclampsia.

Various studies have shown variable onset from 28 weeks gestational age to 13 days postpartum. High degree of suspicion is required in these patients.¹ The onset of PRES was immediate post-partum in our patient. Though it is a reversible phenomenon, as indicated by the name, recurrences have been reported in 6% of cases. In addition, if not recognized early and treated appropriately, permanent brain injury due to complications like intracerebral hemorrhage and infarction may ensue. Death has been reported in up to 15% of patients especially the cases developing PRES secondary to calcineurin inhibitors toxicity.¹⁰ The visual disturbances reported with PRES include cortical blindness, visual neglect, homonymous hemianopia and blurred vision and have been more common in patients developing PRES secondary to Pre-eclampsia/eclampsia.¹¹

MRI typically demonstrates bilateral symmetrical hyper intensities in the parieto-occipital regions on T2-weighted and FLAIR sequence images. Occipital lobe is commonly involved in PRES but spares the calcarine and paramedian occipital lobe. This along with major involvement of the white matter is a distinguishing feature from bilateral posterior cerebral artery territory infarction. Our MRI findings correlated with the findings of Karuppannasamy D et al.,¹² As systemic blood pressure rises, the brain's autoregulatory capability fails and it results in hyperperfusion via increased cerebral blood flow and resulting edema. Judicious control of hypertension has been found to be the primary treatment along with supportive treatment to lower the intracerebral pressure.⁴ Our patient recovered full vision of 6/6 within 1 week of treatment (Table 1) and was doing well on follow up at one month and three months. Her MRI did not show any lesions on follow up visits.

Table 1 Vision in both eyes from Day 1 to Day 7

Duration	Visual acuity in right eye	Visual acuity in left eye
Day 1	Perception of light present	Perception of light present
Day 2	Counting fingers at 5 meters	6/60
Day 3	6/60	6/60
Day 4	6/18	6/12
Day 5	6/6	6/6
Day 6	6/6	6/6
Day 7	6/6	6/6

We conclude that the knowledge about PRES is essential to ophthalmologists to manage this clinical entity that is reversible if treated promptly. MRI of the brain is an essential tool for diagnosis and management and requires the help of a neurologist. Emergency delivery and a good blood pressure control usually results in rapid and complete recovery.

Acknowledgements

None.

Conflict of interest

The author declares that there is no conflict of interest.

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