Case Report

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Cortical Blindness in a Postpartum Woman due to Posterior Reversible Encephalopathy Syndrome without Hypertension and with Uncommon Features

K. Gunasekaran[1], S. Eswari[2], S. Sivakumar[3], K. Thiruvarutchelvan[4]

Abstract

Cortical blindness is loss of vision with intact pupillary reflexes and normal optic fundus, due to damage to primary visual cortex. Cortical blindness in postpartum period is an uncommon event. Possible etiologies are cerebrovascular disease complicating pregnancy, eclampsia, clinical syndrome like posterior reversible encephalopathy syndrome (PRES), etc. Common among these is PRES. The common visual disturbances seen in patients with posterior reversible encephalopathy syndrome (PRES) are blurred vision, visual field deficits like quadrantanopia and hemianopia, visual hallucinations and rarely PRES produces complete loss of vision. Here we report a postpartum woman who developed complete loss of vision due to PRES. Interestingly her blood pressure was normal, though PRES is commonly associated with elevated BP. In addition to that she demonstrated uncommon site of edema in neuroimaging. To add on, her MRI brain showed high signal in DWI image which usually occurs with ischemic infarct. She was managed conservatively with excellent recovery of her vision.

Keywords: Posterior Reversible Encephalopathy Syndrome; Cortical Blindness; Post-Partum.

[1]Assistant Professor
[3]Professor & Head
[4]Associate Professor,
Department of Neurology,
[2]Post Graduate Student,
Department of Obstetrics and Gynecology,
Government Mohan Kumaramangalam Medical College Hospital, Salem, Tamil Nadu, 636001, India.

Corresponding Author:
K. Gunasekaran,
Assistant Professor,
Department of Neurology,
GovernmentMohan Kumaramangalam Medical College Hospital, Salem, Tamil Nadu, 636001, India.
E-mail
drgunasekaranneuro@gmail.com

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Introduction

Cortical blindness is a clinical syndrome characterised by loss of vision with intact pupillary reflexes and normal optic fundoscopy, due to damage to primary visual cortex (area V1). This may be permanent or transient depending on the cause. Causes for permanent cortical blindness are anoxia, stroke involving the posterior or middle cerebral arteries, tumour involving occipital lobes, traumatic brain injury, etc. Transient loss of vision occurs in conditions like migraine, posterior reversible encephalopathy syndrome (PRES), cerebral hypoperfusion (reduced cardiac output, systemic hypotension), occipital epilepsy, etc. Among these, posterior reversible encephalopathy syndrome (PRES) constitutes the important cause of cortical blindness, especially in situations likeperipartum period. Here we describe a post partum woman who developed cortical blindness due to PRES.

The posterior reversible encephalopathy syndrome is a neuroradiological syndrome of subacute onset. Various causes of PRES are hypertension, preeclampsia/eclampsia, bone marrow transplantation, organ transplantation, sepsis, renal failure, autoimmune diseases and chemotherapy. PRES is characterised by seizure, headache, vomiting, visual disturbances, altered mentation, confusion andrarely focal neurological deficits. Common visual symptoms seen in PRES are blurring of vision, visual field deficits like quadrantanopia and hemianopia, visual

hallucination, visual neglect and rarely cortical blindness [1]. Incidence of visual symptoms in PRES is 20% to 50%, depends on the etiology of PRES. That is, incidence is more in 'eclampsia related PRES' when compared to 'PRES due to various etiologies'. For example, a study which analysed' eclampsia related PRES' shows visual symptoms in 50% of their patients.[1] Whereas a study by Fugate et al shows visual disturbances in 20% of their patients [2] and a study by Liman et al shows visual symptoms in 27.8% of their patients [3] (these two studies analysed 'PRES due to various etiologies').

In this case report, we describe a postpartum woman who developed cortical blindness due to PRES without any other features of PRES and recovered vision completely within 24 hours with conservative management. And interestingly there were some uncommon features noticed in this case. Firstly, there was no significant elevation of blood pressure though PRES is commonly associated with increased BP. Secondly, neuroimaging demonstrated vasogenic edema in uncommon site. Finally, there was no residual sequelae in spite of MRI brain demonstrated diffusion restriction in diffusion-weighted (DWI) image which usually indicates ischemic infarct.

Case Report

Mrs. S., a 19 years old, house wife, admitted to our institute's labour ward with labour pain. She was referred from a nearby PHC with h/o labour pain with meconium stained liquor. She was a primigravida, in term pregnancy. On examination, patient was in normal sensorium, BPwas 110/70 mmhg, body weight was 58 kg, minimal pedal edema was present. PV examination showed well effaced, fully dilated cervix. 45 min after admission, patient delivered an alive boy baby with birth weight of 2.81 kg with Apgar8/10, by normal vaginal delivery with episiotomy. 15 min before delivery patient noticed blurring of vision. BP at that time was 110/70 mmhg. After delivery, she lost her vision completely, could not see her baby, not able to perceive even light. There was no relevant associated symptoms like headache, vomiting, altered sensorium or seizure. There were no significant past medical history like hypertension, heart disease, diabetes, visual impairment or seizure. BP at the time of admission, that is 45 min before delivery was 110/70 mmhg. After delivery, when she was complaining about loss of vision, BP was 110/80 mmhg. Pulse rate, respiratory rate and temperature were normal. Pedal edema +. As there was no other alarming signs patient was started on intravenous fluids,antibiotics and supportive care with reassurance. Ophthalmologist examined her and found normal fundus. On neurological examination, patient was conscious, oriented, obeying commands. Comprehensionwas good. Word output was normal. No hallucinations or delusions were present. No meningeal signs were present. Pupils on both sides were 3mm and light reflex was normal. Acuity: complete loss of vision +, not even light perception was present. Optic fundus normal. Extra ocular movements were full. Otherwise neurological examination was unremarkable. Other systems examination were normal. With these findings and clinical picture diagnosis was made as cortical blindness. Few hours later, that is 12 hrs after the onset of symptoms patient underwent CT brain which revealed hypodensity in bilateral parieto occipital region and right basal ganglia, suggestive of PRES (Figure 1 and 2). Her investigation reports were as follows: Blood: Hb: 8.9 gm %, PCV: 26%, basic blood and urine investigations were normal. Blood sugar: 99 mg %, urea: 16 mg %, creatinine: 0.6 mg%. HIV – Nonreactive.

Twenty four hours after the onset of visual loss patient completely regained vision (6/6) without any specific treatment. At that time pupils were normal, 2.5 mm in size, light reflex present. Upto 48 hrs after delivery, BP was within 110/80 mmhg. On third day there was mildly elevated BP to 130/90 mmhg and

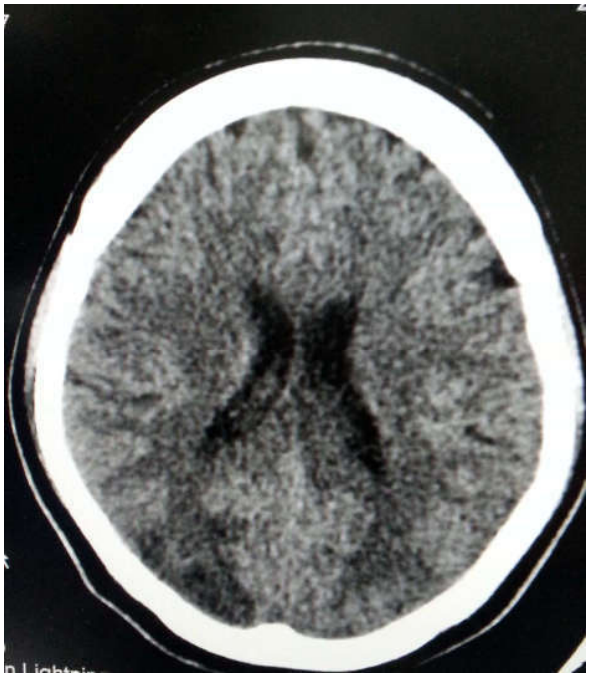

Fig. 1: CT brain axial view showing hypodensity in bilateral parietooccipital region

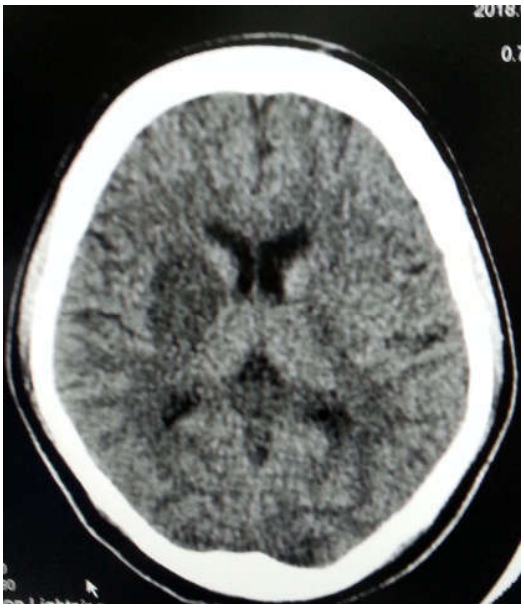

Fig. 2: CT brain axial view showing hypodensity in right basal ganglia

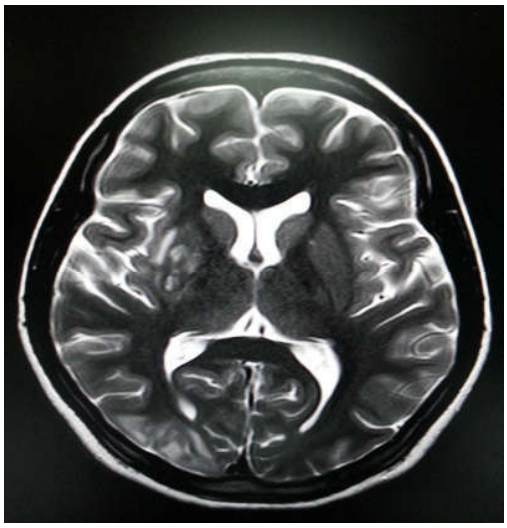

Fig. 3: MRI brain axial view T2 image showing hyperintensity in right basal ganglia

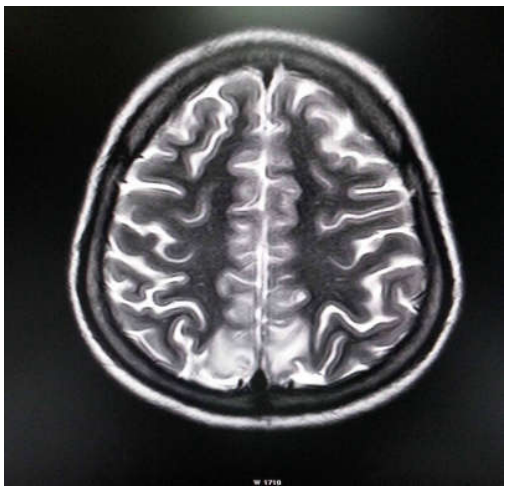

Fig. 4: MRI brain axial view T2 image showing hyperintensity in bilateral parietooccipital region

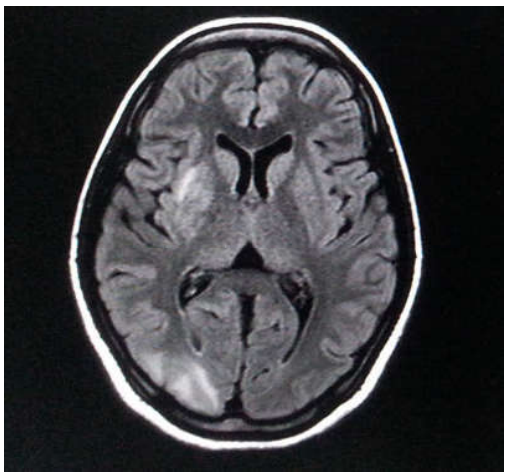

Fig. 5: MRI brain axial view FLAIR image showing hyperintensity in right parietooccipital region and right basal ganglia

140/80 mmhg. Hence patient was started on tab labetolol 100 mg twice a day. At that time her blood and urine investigations were repeated, which were normal except Urine albumin 1+. ECG - normal. No LVH or ischemic changes, rate - 80/min.

Two days after delivery ophthalmologist reviewed her and opined normal optic fundus with normal acuity (6/6). Two days after starting labetolol, BP reduced to 110/70 mmhg hence over next two days labetolol was tapered and stopped. Three days after delivery, MRI brain done which showed T2/FLAIR hyperintense signals in bilateral parietoccipital subcortical white matter and right basal ganglia (Figure 3,4,5,6 and 7). Tiny foci of diffusion restriction seen in right basal ganglia and bilateral parietal cortices, (Figure 8 and 9) features suggestive of PRES. Supportive care continued without antihypertensive drugs and one week later ophthalmologist reviewed her again and reported no papilledema, no retinopathy. Rest of her hospital stay was uneventful. She was discharged 15 days after admission with normal vision and normal neurological status. One day before discharge MRI Brain was repeated which revealed complete remission (Figure 10, 11, 12 and 13). No evidence of PRES or any other abnormalities.

Discussion

A primigravida presenting at term or immediate postpartum period with complaints of blindness

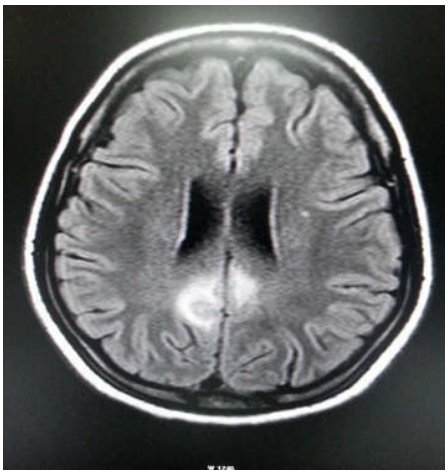

Fig. 6: MRI brain axial view FLAIR image showing hyperintensity in bilateral parietooccipital region

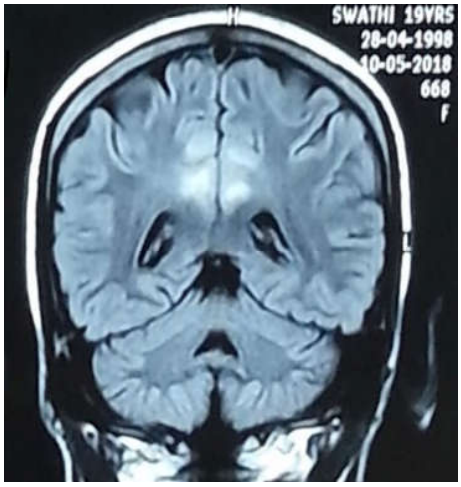

Fig. 7: MRI brain coronal view FLAIR image showing hyperintensity in bilateral parietooccipital region

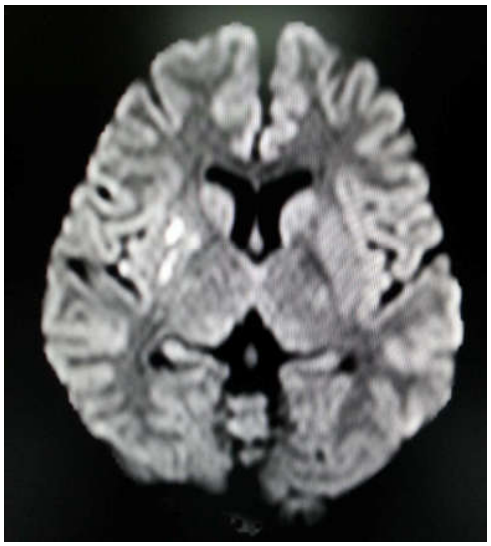

Fig. 8: MRI brain axial view DWI showing restriction in right basal ganglia

puts the treating obstetrician in diagnostic dilemma as there are various possibilities. The possible etiologies are cerebrovascular disease complicating pregnancy, eclampsia and clinical syndrome like posterior reversible encephalopathy syndrome (PRES). Common among these is PRES. This syndrome was first described in 1996 by Hinchey and colleagues [4].

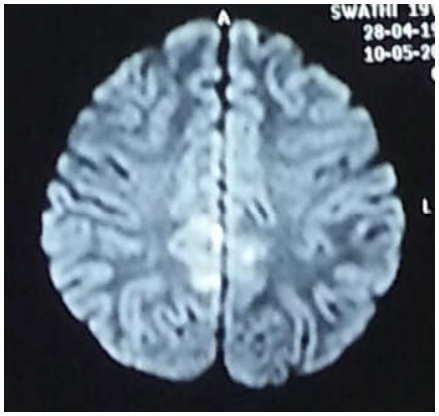

Fig. 9: MRI brain axial view DWI showing restriction in bilateral parietooccipital region

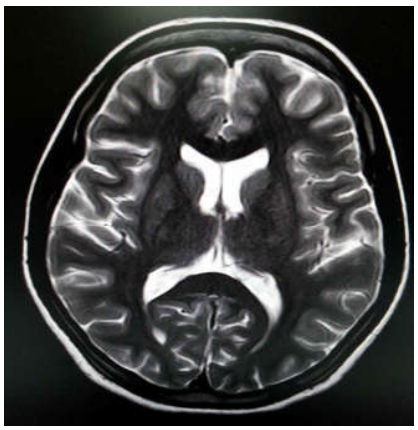

Fig. 10: MRI brain axial view T2 image showing normal right basal ganglia

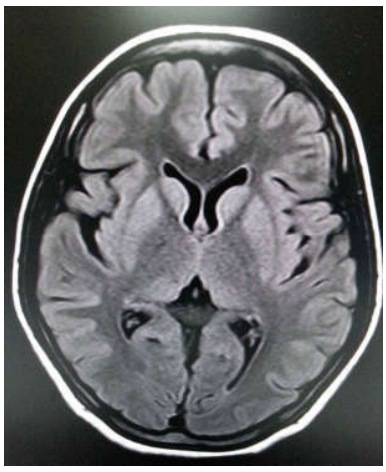

Fig. 11: MRI brain axial view FLAIR image showing no signal changes in right parietooccipital region and right basal ganglia

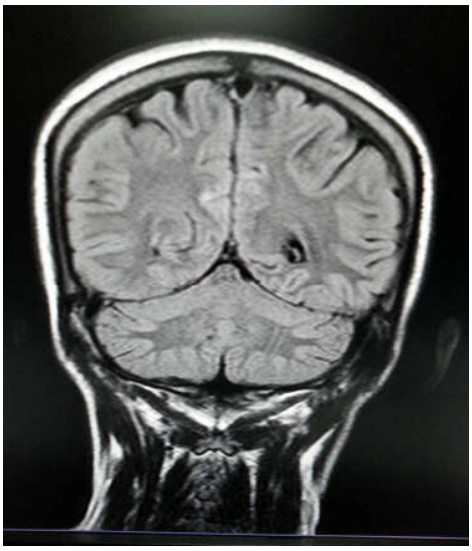

Fig. 12: MRI brain coronal view FLAIR image showing no signal changes in bilateral parietooccipital region

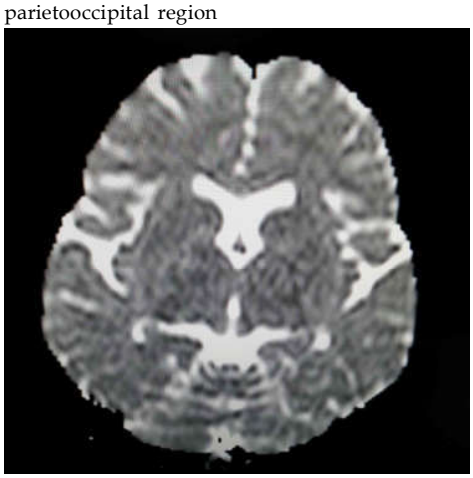

Fig. 13: MRI brain axial view DWI showing no restriction in right basal ganglia

There are two theories put forward for the pathophysiology of PRES [5]. According to the first hypothesis, elevation of blood pressure above the upper autoregulatory limit leads to cerebral hyperperfusion, which results in vascular leakage and vasogenicedema. Also the increased cerebral perfusion pressure contributes to blood brain barrier dysfunction producing extravasation of plasma and macromolecules through endothelial tight junction.

But our patient was having normal BP most of the days of hospitalisation, especially when she developed visual loss (CT brain demonstrated PRES at that time [Figure 1 and 2]) BP was normal. Two days after delivery, that is one day after completely regained her vision, showed minimally elevated BP, that too was within upper normal limit. Hence pathophysiology of PRES in this situation could be the second theory, that is endothelial dysfunction theory [5]. According to this, PRES is caused by endothelial dysfunction due to circulating endogenous (in conditions like preeclampsia/eclampsia, sepsis) or exogenous (chemotherapy, immunosuppressive agents) toxins. These toxins trigger vascular leakage and edema formation in addition to endothelial activation which results in release of immunogenic and vasoconstrictive substances. These vasoconstrictors produce cerebral vasospasm. Interestingly, blood pressure elevation occurs as a consequence of endothelial dysfunction instead of being the reason for occurrence of PRES. A variation on this "toxic/immunogenic" theory is that the trigger may be the excessive release of pro-inflammatory cytokines which result in endothelial activation, release of vasoactive agents, increased vascular permeability and edema formation. In our patient some endogenous toxins or pro-inflammatory cytokines which possibly has produced PRES and mild elevation of BP. Similar report is seen with Kunal Vakharia et al., study [6] in which BP was near normal and PRES resolved within 48 hours. According to W.S. Bartynski, in 20-30% of patients with PRES, BP is normal or only minimally elevated [7].

Secondly, as the name implies, posterior areas of the cerebral hemispheres are susceptible to PRES. Because there is reduced density of vasomotor sympathetic innervation in the posterior circulation compared to anterior which is densely innervated by the superior cervical ganglion. This prevents excessive vasodilatation, reduces the risk of cerebral hyperperfusion in anterior region.

But recently atypical distribution has been reported in this syndrome. That is, apart from common locations like occipital and parietal lobes, edema noticed in other areas like frontal, temporal lobes, cerebellum, basal ganglia, thalamus and brainstem. According to a report, frontal lobe is the commonest atypical site followed by basal ganglia [8]. This atypical distribution may give diagnostic confusion with other disorders like anoxic encephalopathy, central pontine myelinolysis, extrapontine myelinolysis, hypoglycemic encephalopathy and cortical vein thrombosis or deep venous thrombosis. Hence these atypical pattern represent a challenge for treating physician. But clinical correlation and follow up imaging will help in making appropriate diagnosis. Our patient demonstrated hypodensity in right basal ganglia in CT brain (Figure 1 and 2) as well as signal changes in same region in MRI brain, apart from bilateral parietooccipital region findings (Figure 3 and 5).

Thirdly, as PRES is due to reversible vasogenicedema, there should be high signal

intensity in T2/fluid attenuated inversion recovery (FLAIR) images of MRI brain and there should not be restriction in diffusion-weighted MR imaging (DWI). Because, high signal intensity in DWI indicates cerebral ischemia and conversion to infarction and irreversible tissue damage. But in our patient, in spite of MRI brain demonstrated high signal intensity in bilateral parietal cortices in DWI sequences (Figure 8 and 9), patient did not show clinical signs of cerebral infarction and repeat MRI brain was normal. Similar report is seen in Benziada-Boudour A et al. study [9]. Here the explanation is that this DWI signal abnormalities may be reversible until the effect of cytotoxic edema begins to dominate and ischemia ensues [8]. Also according to Ay et al, areas of massive vasogenic edema and increased tissue pressure eventually impairs the microcirculation and leads to ischemia and cytotoxic edema. once the vasogenicedema resolved, microcirculation improves and DWI changes may disappear.

Our patient's MRI brain showed restricted diffusion in bilateral parietal cortices in DWI probably indicates the severity of the condition. But fortunately patient recovered without sequelae and repeat MRI brain after two weeks demonstrated normal study (Figure 10,11,12 and 13).

Treatment of PRES is symptomatic, as there is no specific therapeutic strategy is currently available. Underlying disease or pathology leading to the development of PRES is to be treated. In general, prognosis of PRES is good since the neurological manifestations are reversible in majority of patients and the symptoms including the lost vision is usually regained within hours to a week. However, if accompanied by severe complications, neurological sequelae may persist. Our patient did not show any neurological deficits and she was managed conservatively with excellent recovery of her vision.

Conclusion

Though PRES is a condition commonly encountered in obstetrics ICU, cortical blindness is not seen as frequently as mild visual symptoms. Likewise PRES without elevated blood pressure and uncommon location of edema with DWI changes are rare. Hence this case is presented here to make the treating physician to be aware of the unusual features of PRES. PRES can be managed effectively by obstetrician along with neurophysician, radiologist and general physician.

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