

## Case Series:

# Posterior Reversible Encephalopathy Syndrome (PRES): A case series in postpartum patients

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### Abstract :

*Posterior reversible encephalopathy syndrome (PRES) refers to a clinicoradiologic entity with characteristic features on neuroimaging and nonspecific symptoms comprising headache, confusion, visual disturbances and seizures. The radiological findings in PRES are thought to be due to vasogenic edema, predominantly in the posterior cerebral hemispheres and are reversible with appropriate management. We report 4 cases of PRES diagnosed by MRI scan following uneventful caesarean section in previously normotensive patients who were successfully treated with antihypertensives, anticonvulsants and supportive treatment.*

**Key Words:** PRES, Posterior Reversible Encephalopathy Syndrome, Postpartum Hypertension.

### Introduction

Posterior reversible encephalopathy syndrome is a well recognized, clinical and neuro-radiological entity first described in 1996 by Hinchey et al.<sup>1</sup> This is characterized by headache, vomiting, confusion, visual abnormalities and motor signs. These transitory neurological disturbances are thought to be due to cerebral vasospasm causing ischemia of the involved territory. It describes a potentially reversible imaging appearance and may occur in diverse situations, including hypertension, eclampsia, pre-eclampsia, immunosuppressive medications such as cyclosporine, various antineoplastic agents, severe hypercalcaemia, thrombocytopenic syndromes, Henoch Schölein purpura, Systemic Lupus Erythematosus (SLE), hemolytic uramic syndrome, amyloid angiopathy, renal failure, post-transplantation, infection, sepsis (gram negative organisms predominate) and shock.<sup>2,3</sup> Importantly these changes appear to be completely reversible if the underlying cause is treated or the precipitating drug withdrawn early in the clinical course.<sup>1</sup> We report 4 cases of reversible encephalopathy syndrome, occurring in the postpartum period managed successfully.

### Case Report

#### Case -1:

A 21 year old female, 4 days post caesarean section presented with headache and sudden onset of focal seizure with secondary generalization. Examination revealed an afebrile patient, drowsy but arousable, blood pressure 150/100 mmHg, there was no focal neurological deficit and meningeal signs were absent. Fundoscopy did not reveal any papilloedema. CT brain and MR venogram were normal. MRI Brain showed hyperintensities in frontal, parietal & occipital lobes of both cerebral hemispheres predominantly involving the cortices on T2W and FLAIR images. She was treated with antihypertensive and anticonvulsant and recovered completely with no residual neurological deficit.

#### Case 2:

A 30 year-old-female, 8 days post caesarean section presented with severe headache, generalized tonic clonic seizure and disorientation for 1 day. Examination revealed that she was disoriented and afebrile with blood pressure of 180/110 mm of Hg. Her metabolic profile was normal and urine showed trace proteins. CSF study, was done to rule out infective causes, was normal. MRI brain showed hyperintensities mostly involving the cortices of frontal, parietal, temporal & occipital lobes of both cerebral hemisphere along with involvement of adjacent subcortical white matter (more remarkable in right frontal lobe) on T2W and FLAIR images. She was treated with antihypertensives and anticonvulsants. She got stabilized by the fourth day and her neurological symptoms resolved by 7days. She was discharged on antihypertensive and anticonvulsant medications.

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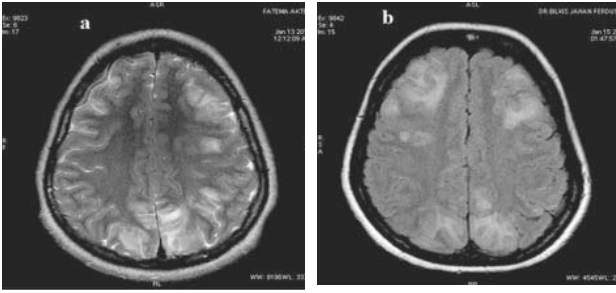
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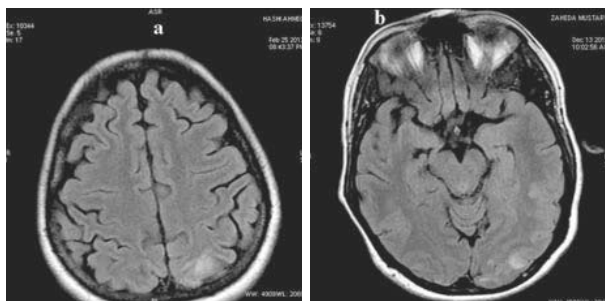
**Figure 1-**(a) T2weighted axial image shows hyperintensities in frontal, parietal & occipital lobes of both cerebral hemispheres predominantly involving the cortices. (b) T2 weighted axial image shows hyperintensities are mostly involving the cortices of frontal, parietal, temporal & occipital lobes of both cerebral hemisphere along with involvement of adjacent subcortical white matter (more remarkable in right frontal lobe)

### Case 3:

A 24 year-old-female, 4 days post caesarean section presented with headache, vomiting and blurring of vision for 1 day. Examination revealed a conscious, oriented and afebrile patient with moderate edema and blood pressure 170/110 mm of Hg. A central nervous system examination revealed diminution of vision with regard to perception of hand movement. Pupillary reactions and fundoscopic examination were normal. Urine examination revealed 2+ proteinuria. MRI brain showed hyperintensities along cortices of both parieto-occipital regions in T2-weighted and FLAIR sequences. She was started on antihypertensives and her vision improved to 6/6 after 48hours following commencement of treatment.

### Case 4:

A 37 year old female, 6 days post caesarean section presented with 2 episodes of convulsion for 1 day. Examination revealed a drowsy but arousable patient with blood pressure 160/120 mm of Hg and plantars were bilateral extensor. Her metabolic parameters, CSF study were normal and urine examination revealed trace proteins. MRI brain showed bilateral corticosubcortical hyperintensities involving parieto-occipital lobes, more remarkable on the left in T2 weighted and FLAIR sequences. She was treated with anticonvulsant and antihypertensive medications. She became stable by the 3rd day with no residual neurological deficit.



**Figure 2-**(a) T2-weighted axial image shows hyperintense signal in parieto-occipital region.(b) T2-weighted axial image shows hyperintense signal in left parieto-occipital region.

## Discussion

PRES has become better recognized with the progress made in imaging modalities. Most cases are associated with hypertensive emergencies, allogenic bone marrow transplantation, solid organ transplantation, autoimmune disease, and high dose chemotherapy, toxemia of pregnancy, cryoglobulinemia and thrombotic thrombocytopenic purpura.<sup>2</sup>

Rapid diagnosis of PRES is essential to prevent complications such as infarction and hemorrhage. Proper diagnosis requires careful attention to the clinical and radiographic presentation. Pathogenesis of PRES has been suggested that there is a temporary failure of auto regulatory capabilities of the cerebral vessels leading to hyper perfusion, a breakdown of blood brain barrier and consequent vasogenic edema.<sup>4</sup>

Neuroimaging is essential to the diagnosis of PRES. Typical findings are symmetrical white matter edema in the posterior cerebral hemispheres, particularly the parieto-occipital regions with sparing of the calcarine and paramedian occipital lobe structures, differentiating it from bilateral posterior cerebral artery infarcts. Neuroradiographic abnormalities of PRES are often apparent on Computed tomography (CT) scans but are best depicted by magnetic resonance imaging (MRI). The most common abnormalities on MRI are punctate or confluent areas of increased signal on proton density and T2-weighted images.<sup>2,5</sup> When regions of the brain other than parieto-occipital lobes are predominantly involved, the syndrome can be called atypical. In such cases, a diffusion weighted MRI with ADC mapping shows increased ADC values representing vasogenic edema in these areas, thus differentiating atypical PRES from other brain disorders.<sup>6</sup> Most case reports suggest that posterior reversible encephalopathy syndrome (PRES) is usually benign. In many cases, it seems to be fully reversible within a period of days to weeks after removal of the inciting factor and control of blood pressure. Significant reversal of neuroradiological abnormalities with complete clinical recovery forms the diagnosis.

The differential diagnosis of postpartum seizure includes eclampsia, subarachnoid hemorrhage, intracerebral hemorrhage, thrombotic phenomena, intracranial neoplasm, head trauma, idiopathic epilepsy, infection (meningo-encephalitis), amniotic fluid embolism, postpartum angiopathy.<sup>1,7</sup> Brain MRI with venogram rule out intracranial bleed, ischemia secondary to thromboembolism, vasospasm or space occupying lesion. Amniotic fluid embolism presents with cardiopulmonary collapse and coagulopathy which are absent in our patients. Postpartum angiopathy is characterized by severe thunderclap headache, seizure, focal neurological deficits and segmental narrowing and dilatation of large and

medium sized arteries. Typically scanning reveals ischemic lesions but MRI findings are suggestive of PRES.<sup>1</sup>

A study of 76 patients by Alexander M. McKinney et al, showed that the incidence of regions involvement was parieto-occipital 98.7%, temporal 68.4%, thalamus 30.3%, cerebellum 34.2%, brainstem 18.4%, and basal ganglia 11%.<sup>8</sup> The incidence of less common manifestations was enhancement 37.7%, restricted diffusion 17.3%, hemorrhage 17.1% and a newly described unilateral variant 2.6%.<sup>8</sup> In our study, the most commonly involved location was the parieto-occipital region, which was seen in 4 cases (100%). This was followed by frontal lobe in 2 cases (50%) and temporal lobe in 1 case (25%).

### Conclusion

PRES is a rare but easily treatable condition and should be considered in differential diagnosis of postpartum seizure. Awareness of diverse clinical and radiological presentation of PRES is essential to avoid misdiagnosis and treatment delay. The syndrome of PRES is correctly recognized on neuro-imaging and potential complications can be avoided by appropriate therapy.

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