Cortical visual loss in posterior reversible encephalopathy syndrome in late postpartum eclampsia: Case series

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The purpose of this study was to determine the prevalence of visual disturbances in patients with posterior reversible encephalopathy syndrome (PRES) associated with late postpartum eclampsia. We retrospectively reviewed the clinical records of late postpartum eclampsia patients with features of PRES for the presence of visual disturbances and location of radiological abnormalities. We found a higher prevalence of cortical visual loss in patients with PRES associated with late postpartum eclampsia. Bilateral symmetrical vasogenic edema of the parieto-occipital lobe was the most common magnetic resonance imaging (MRI) abnormality noted. No significant differences were observed in the extent of edema in patients with and without visual loss.

Key words: Eclampsia, postpartum, reversible, visual loss

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Reversible cortical visual loss has been well documented with posterior reversible encephalopathy syndrome (PRES).[1] PRES is associated with diverse clinical etiologies and hypertensive encephalopathy is critical in the pathogenesis.[2] Pre-eclampsia and eclampsia are well-known risk factors of PRES. Late postpartum eclampsia accounts for 16% of cases of eclampsia[3] and has been known to be associated with PRES.[4]

Although the pathogenesis and imaging appearances are similar the clinical syndrome can be variable with symptoms ranging from mild blurring of vision to severe neurological deficit.

The objectives of this study were to determine (1) the prevalence of visual disturbances in late postpartum eclampsia patients with PRES (2) the location of MRI lesions (3) correlation between visual symptoms and imaging abnormalities.

To the best of our knowledge no such study has been reported from our population.

Materials and Methods

A retrospective review of clinical records of late postpartum eclampsia patients who had presented with acute onset of visual and/or neurological symptoms to the emergency department of a medical college hospital in South India between April 2009 and May 2013 was done. All patients had underwent MRI with diffusion weighted imaging (DWI) and apparent diffusion coefficient (ADC) mapping to differentiate vasogenic from cytotoxic cerebral edema. Clinical or radiological recovery had been noted in all patients.

Patient records were analyzed for presence of visual symptoms, peak systolic and diastolic blood pressures,

Table 1: Clinical profile of patients with PRES

Age	Blood pressure	Time of presentation postpartum (days)	Clinical features	Location of MRI lesions	Time to recovery (days)
25	170/90	9	Headache, visual loss	Bilateral parieto-occipital lobes	3
25	150/100	10	Headache, seizures	Bilateral posterior parietal lobe, occipital lobe, frontal lobe and right cerebellum	4
25	120/80	9	Seizures, altered sensorium	Bilateral parieto-occipital lobe	1
30	160/100	15	Headache, altered sensorium, seizures	Bilateral posterior parietal and occipital	5
24	150/90	10	Headache, seizures	Bilateral parieto-occipital, frontal and cerebellar hemispheres	2
22	180/100	4	Headache, altered sensorium, seizures, visual loss	Bilateral parieto-occipital, left cerebellum	1
21	120/100	5	Headache, seizures	Bilateral occipital, frontoparietal and cerebellar hemispheres	2
29	120/80	5	Headache, seizures, visual loss	Bilateral parieto-occipital	2
27	130/90	12	Headache, visual loss	Bilateral occipital, right cerebellum	2
32	140/100	6	Headache, seizures, visual loss	Bilateral parieto-occipital	4

PRES: Posterior reversible encephalopathy syndrome, MRI: Magnetic resonance imaging

associated neurological symptoms, location of MRI lesions and time taken for recovery. The MR images were graded for extent and severity of cortex and white matter vasogenic edema, degree of confluence, mass effect and ventricular distortion on a scale from 1 to 5.[5] This was done independently by two radiologists blinded to patients' clinical profile. Differences in edema grade was agreed upon by consensus. Edema grade results for patients with and without visual disturbances were compared.

Statistical analysis was performed using Chi-square test and a *P* value of less than 0.05 was considered to be statistically significant.

Results

Ten patients were included in the study. Clinical and imaging findings are summarized in Table 1.

The 10 patients ranged in age from 21 to 32 years (Average-26 ± 3.49 years). Mean peak systolic and diastolic blood pressures were 144 ± 21.71 and 93 ± 9.19 mm of Hg, respectively. Average duration between delivery and onset of clinical symptoms was 8.5 ± 3.5 days. Nine patients had headache while seizures was noted in eight patients. Bilateral visual loss was noted in five patients of whom three patients were able to perceive hand movements and two patients had only perception of light. Ophthalmic examination revealed normal pupils and fundi suggestive of cortical blindness. Altered sensorium was observed in three patients. None of the patients had focal neurological deficit.

The most common abnormality noted on MRI included bilateral symmetrical hyperintensities on T2-weighted images and fluid attenuated inversion recovery (FLAIR) sequences in the parieto-occipital regions [Figs. 1 and 2]. DWI showed high signal intensity with no areas of restricted diffusion while ADC mapping did not show corresponding low signal intensity thus suggesting vasogenic edema.

Table 2: Vasogenic edema grade in patients with and without visual loss

Late postpartum eclampsia patients with PRES	Vasogenic edema grade (number of patients)					Group average
	Grade 1	Grade 2	Grade 3	Grade 4	Grade 5	
With visual loss	1	1	1	1	1	3
Without visual loss	2	2	1			1.8

PRES: Posterior reversible encephalopathy syndrome

Five patients had additional abnormalities in the cerebellar hemispheres and three patients had involvement of frontal lobes. Average time taken for visual recovery was 2.6 ± 1.34 days. All subjects made a complete visual recovery and visual acuity was restored to 20/20. Visual fields performed after recovery did not reveal any field defect.

Follow-up MRI in four patients after 1 month revealed complete resolution of these abnormalities. In the remaining, clinical symptoms and signs resolved completely. Follow-up scans were not performed due to financial constraints.

The extent of cerebral edema in patients with and without visual loss was compared and results summarized in Table 2. No statistically significant differences were observed in the severity of edema in both groups.

Discussion

Posterior reversible encephalopathy syndrome (PRES) is a rare neurotoxic state that presents with altered mental status, headache, seizures, and visual disturbances along with neuroimaging features of vasogenic edema involving the posterior cerebral circulation. Although several cases of PRES have been reported in neurology and neuroradiology

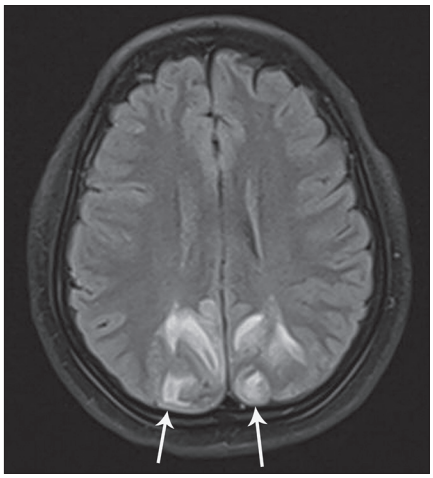

Figure 1: Axial MR image (fluid attenuated inversion recovery) showing confluent edema in subcortical and deep white matter (Grade 4) seen as hyperintensities in bilateral parieto-occipital lobes (patient 6)

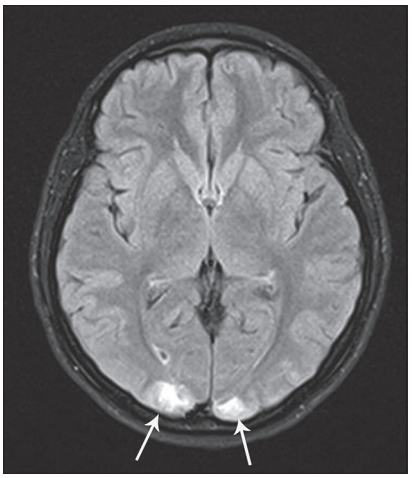

Figure 2: Axial MR image (fluid attenuation inversion recovery) showing edema in subcortical white matter (Grade 2) seen as hyperintensities in bilateral occipital lobes (patient 9)

literature, exposure in ophthalmic literature has been rather limited.

The causes of PRES are diverse and include pre-eclampsia, eclampsia, renal insufficiency, solid organ transplantation and immunosuppressive therapy.[6]

The pathogenesis of this syndrome is poorly understood. Hypertensive encephalopathy is said to be the cause of this syndrome which has been demonstrated by various clinical and experimental studies.[7] Patients with hypertensive encephalopathy have the same clinical signs as those with PRES and they also have rapid resolution of clinical and imaging abnormalities once the blood pressure is lowered. The most widely accepted theory states that sudden elevation of blood pressure causes failure of autoregulation in the cerebral blood vessels leading to hyperperfusion, breakdown of blood brain barrier, and vasogenic edema.

The posterior circulation is preferentially affected since it has less sympathetic innervation than the carotid circulation, thus rendering it less able to adjust to blood pressure fluctuations.[8]

However, this theory is not comprehensive because PRES can affect normotensives.[9] Three of our patients had peak systolic and diastolic blood pressures within normal limits.

High degree of suspicion is required in these patients of late postpartum eclampsia because it occurs between 48 hours postpartum and 1 month after delivery frequently in women who have had a normal pregnancy and delivery and no signs of a pre-eclamptic syndrome.[2] Vasogenic edema of white matter may occur with normal or mildly elevated pressure.

Alternative theory implicates endothelial dysfunction as the cause for occurrence of PRES in eclampsia and sepsis.[10,11] This is supported by the fact that elevations in markers of endothelial dysfunction such as lactate dehydrogenase and abnormal red blood cell fragmentation have been reported in PRES.[4] The presence of endothelial dysfunction decreases the threshold at which vasogenic edema occurs.[12]

Vasospasm with subsequent ischemia has also been hypothesized to cause loss of integrity of the blood brain barrier in eclampsia.[13]

Although reversible by definition early recognition and prompt treatment is essential to prevent secondary complications like intracerebral hemorrhage and infarction.

A large retrospective study of PRES in diverse clinical situations revealed the occurrence of visual symptoms in 20% and headache in 26% while seizures was observed in 74% of cases.[6]

The visual disturbances reported with PRES include cortical blindness, visual neglect, homonymous hemianopia and blurred vision.

Cortical visual loss was observed in five of our patients (50%) while the remaining denied any visual disturbances.

Our findings are in line with Liman *et al.,* who observed that visual disturbances like cortical blindness, blurred vision and hemianopia are more common in eclampsia related PRES (50%). Patients with PRES due to other etiologies more often present with severe symptoms like altered mental status or neurological deficit and lesser visual disturbances (27.8%).[14] Roth *et al.,* have also reported a higher percentage of disturbed vision in pre-eclampsia-eclampsia-related PRES.[15]

This could be because of the younger age and lesser associated comorbidities in these patients as compared to patients with PRES due to other etiologies.

All patients (with/without visual loss) demonstrated bilateral symmetrical hyperintensities in the parieto-occipital regions on T2-weighted images.

Lesion did not involve the calcarine cortex in all 10 patients. PRES lesions involving the occipital lobe spare the calcarine and paramedian occipital lobe structures. This feature along with predominant involvement of the white matter distinguishes this syndrome from bilateral posterior cerebral artery territory infarction.[2]

Although several studies have reported visual symptoms of varying degrees in PRES, to the best of our knowledge no

study has attempted to correlate the visual symptoms with imaging abnormalities.

The limitations of our study include its retrospective design, and smaller sample size of postpartum eclampsia patients. Further the timing of MR imaging differed in the acute phase. Because the edema is transient and reversible this might influence the results.

We conclude that there is a higher prevalence of cortical visual loss in patients with PRES associated with late postpartum eclampsia. Patients with higher degree of vasogenic edema of the posterior cerebral white matter might present with visual loss. Ophthalmologists should be aware of this clinical entity since it is reversible and readily treated by controlling the blood pressure.

Further prospective studies comprising larger sample size and different etiologies of PRES are warranted in this regard.

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