

Posterior Reversible Encephalopathy Syndrome in Pregnancy - Relevance in Diagnosis and Management

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ABSTRACT

Posterior reversible encephalopathy syndrome (PRES) is a cliniconeuroradiological syndrome, which may present with headache, encephalopathy, seizures, visual disturbances, and blindness. The lesions in PRES are due to vasogenic oedema in posterior cerebral hemispheres and can be reversible if managed accurately. MRI shows diffuse abnormal signal intensities involving deep white matter of occipital lobes. Reporting 4 cases of PRES diagnosed in our Lady Goschen Hospital, attached to KMC Mangalore in a duration of two years. In all four cases, PRES was suspected when patients presented with eclampsia persistent in spite of administration of Pritchard's regimen. MRI confirmed diagnosis. Three of them recovered while one of the women succumbed due to status eclampticus, respiratory failure and shock. Clinical improvement with complete resolution of visual disturbances was observed with supportive treatment in the other 3 patients. In order to prevent irreversible brain damage, correct clinical suspicion and timely diagnosis with apt management is essential. If adequate blood pressure-titrated treatment is followed, there may be speedy recovery from eclampsia and hence improvement in maternal condition.

Keywords

Posterior reversible encephalopathy syndrome, Fetus, Antihypertensives.

Introduction

Posterior reversible encephalopathy syndrome (PRES) is a cliniconeuroradiological syndrome which generally presents as headache, encephalopathy, convulsions, vision disturbances and even blindness [1]. The lesions in PRES are because of vasogenic oedema in posterior cerebral hemispheres and if managed correctly, may be reversible. MRI shows diffuse abnormal signal intensities involving deep white matter of occipital lobes. Majority of abnormalities are recognised in posterior cerebral hemispheres. Almost always, symptoms resolve partially or completely during check-ups which helps in noting the predominance of subcortical oedema without infarction. Conclusions derived show that changes in the posterior cerebrum is generally reversible and can be correlated with improvement clinically. We hereby are reporting

4 cases of PRES diagnosed in Lady Goschen Hospital, attached to KMC Mangalore in a duration of two years.

Case Series

Between 2015-2017, 4 patients who presented with eclampsia with MRI reports suggestive of PRES were studied. Relevant clinical history, physical examination was done. Antihypertensives were administered. Delivery was expedited in antenatal patients. MRI was done. Patients were followed up for symptoms and BP recordings.

Case Report 1

32-year-old Primigravida was admitted at 35 weeks 6 days with fetal growth restriction for evaluation. She had 4 episodes of seizures and pregnancy was terminated in view of status eclampticus and non-reassuring heart status of the fetus. Pritchard's regimen was started. She underwent emergency LSCS and delivered a live term baby. The lady was shifted to ICU and ventilated. Her MRI

showed abnormal signal intensities involving frontoparietal region and occipital lobes suggestive of PRES. The patient was ventilated and deteriorated in spite of ventilator and circulatory assistance. The patient succumbed in view of multiple seizures, respiratory failure and shock. The baby recovered with 5 days of NICU stay and was discharged.

Case Report 2

28-year Primigravida at 34 weeks 5 days was referred to our hospital in view of VDRL positive status, mild anaemia and high BP recordings. VDRL subsequently done in our hospital was negative. After a battery of supplementary tests, A PLA was suspected. Antihypertensives were started for her antenatally. She underwent an emergency LSCS in view of oligohydramnios. On POD 3, she had an episode of convulsions. She was started on Pritchards Regimen and recovered symptomatically. On POD 7 she had another 2 episodes of convulsions and was shifted to ICU care, where an MRI revealed abnormalities in temporal, subcortical areas and in occipital lobes, suggestive of PRES and hence patient was managed with antihypertensives and anti-oedema measures. On a follow up MRI with MR Venogram after 40 days, complete resolution of abnormal signals and normalcy of dural venous sinuses noted. The resolution of abnormalities in the MRI correlated with BP control using antihypertensives. Baby recovered after 3 days of NICU care. Both the baby and the mother were discharged after 42 days. 3 months later, her follow up showed normal BPs and no residual symptoms.

Case Report 3

26-year Primipara on POD2 following emergency LSCS was referred to our hospital with 2 episodes of convulsions and on Pritchard's regimen. LSCS was done in view of DCDA twins in labour at 35 weeks 6 days with first twin in breech presentation. In view of persistent disorientation, MRI done showed abnormal lesions in thalamus and parieto-occipital lesions suggestive of PRES. With investigations she was diagnosed to have HELLP syndrome. She was managed with blood and platelet transfusions, antibiotics and antihypertensives. By POD-18, she recovered and she and the babies were discharged. 1 month later follow up showed normal BPs and no residual symptoms.

Case Report 4

17-year unmarried Primigravida at 27 weeks 2 days, presented with 3 episodes of convulsions and disorientation. On examination, she was found to be hypertensive. After investigations, she was found to have mild anemia with thrombocytopenia. In view of disorientation, MRI taken showed abnormal lesions in occipital lobes. Medical management for termination of pregnancy failed and she underwent a hysterotomy under general anaesthesia and delivered a dead male fetus of 400g. She was shifted to ICU post operatively. Postnatally, she was discharged on antihypertensives and is yet to come for postnatal check-up.

Discussion

PRES is a neurological condition that is rare and mostly reversible that can occur at any age and affects females more commonly. This probably reflects the fact that one of the most common cause of PRES is pre-eclampsia or eclampsia and is characterised by presence of oedema at occipital and parietal lobes in the white matter. Early diagnosis of PRES is mandatory to avoid complications like haemorrhage and infarction.

Early diagnosis of posterior reversible encephalopathy syndrome is quintessential to prevent complications such as infarction and haemorrhage. Pathogenesis of PRES shows that there may be a temporary failure of capabilities of autoregulation of the cerebral vessels, leading to hyperperfusion, a breakdown of the blood brain barrier and consequent vasogenic oedema [2]. Most lesions are in parietal and occipital lobes, followed by the frontal lobes, the inferior temporal-occipital junction, and the cerebellum [3].

When regions of the brain excluding parieto-occipital lobes are majorly involved, the syndrome can be called atypical. Atypical imaging shows enhancement of contrast, haemorrhage and diffusion on MRI shows restriction. In such cases, a diffusion weighted MRI with ADC mapping shows increased ADC values. 5 Follow-up MRI after adequate therapy for the aetiology of PRES shows resolution of the lesions, unless the condition progresses to infarction or haemorrhage.

PRES lesions involving the occipital lobe spare the calcarine and paramedian occipital lobe and involves white matter mostly

Table 1. Comparison of factors associated in 4 cases of PRES

Factors	Case 1	Case 2	Case 3	Case 4
Age (years)	32	28	26	17
Parity	Primigravida	Primigravida	Primigravida	Primigravida
No. of fetus	1	1	2	1
Complaints	Headache	Blurring of vision	Seizures	Seizures
Associated risk factors	FGR	APLA	Thrombocytopenia	HELLP
Time of presentation	Antenatal	Antenatal	Postoperative	Antenatal
POG	35 weeks 6 days	34 weeks 5 days	POD2	27 weeks 2 days
Episodes of seizures	4	3	3	3
MRI	Frontoparietal region and occipital lobes	Temporal, subcortical and occipital lobes	Thalamus and parieto-occipital lobes	Occipital lobes
Maternal outcome	Death	Recovery	Recovery	Recovery
Fetal outcome	Recovery	Recovery	Recovery	Recovery

helps to differentiate the syndrome from bilateral infarctions of the posterior cerebral artery [4]. On CT SCAN, abnormalities are seen as symmetric, bilateral areas of low attenuation of white matter. Decreased perfusion has been noted on both brain single-photon emission CT technetium imaging and by MR perfusion [5]. Magnetic resonance angiography using 3D Time of flight (TOF) technique shows reversible vasoconstriction either focal or diffuse or pruning of vessels On Proton MR Spectroscopy reduced N-acetylaspartate:choline and N-acetylaspartate:creatinine ratios have been described in regions of PRES affliction [6].

In Liman TG *et al.*, mortality in 103 patients studied was 4.8%. Kalaiselvan et al showed a mortality of 7.4% due to infarction [7]. Out of the 29 patients studied by Virendra C Patil *et al.*, 10.34% of patients succumbed in which 1 of them was hypertensives [8]. In all of these studies, the rest of the patients recovered and had no residual disease.

Conclusion

In order to prevent irreversible brain damage correct clinical suspicion and timely diagnosis with apt management is essential. Suspicion should be thought of if convulsions persist in spite of initiation of treatment or multitude of seizures. If adequate blood pressure titrated treatment is followed, there may be speedier recovery from eclampsia and hence improve maternal condition. A multidisciplinary approach may be required for aggressive and adequate management of all cases and prevention of life threatening complications.

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