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Case Report

PRES, a diagnostic dilemma in pregnancy: three case series with unusual presentation

Sudhir Mansingh, Gunjan Rai*, Bikram Bhardwaj

Department of Obstetrics and Gynecology, Command Hospital Chandimandir, Haryana, India

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***Correspondence:**

Dr. Gunjan Rai,

E-mail: gunjanraims24@gmail.com

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ABSTRACT

Authors report a series of three cases of unusual presentation of posterior reversible encephalopathy syndrome in pregnancy. First patient, 29 years old G2P1L1, who was a booked case at our hospital, presented with complain of no fetal movement perception for 3 days at 27 weeks of period of gestation. No history of hypertension and even no record of hypertension after admission. On USG detected to have severe early onset IUGR and AEDF in Umbilical artery doppler. Went in to spontaneous labor and delivered vaginally a preterm neonate of birth weight of 740 gms at 27 weeks 06 days of period of gestation. Postpartum period was uneventful till day four and on day five of postpartum she developed severe headache and seizure. MRI done which was suggestive of PRES. Second patient 27 years old primi gravida with 37 weeks 01 day, booked at our hospital with regular ANC visit brought with history of headache, vomiting with semi-conscious state with diminution of vision till finger count only. She developed seizure thrice while examination. Antenatal period was uneventful with no history of hypertension. Underwent emergency LSCS on same day and delivered a 2.8 kg healthy female neonate. Patient treated as a case of eclampsia and later MRI findings were suggestive of PRES. Third patient 19 years old primigravida booked ANC case at another hospital. She underwent emergency LSCS at 39 weeks POG for fetal distress at same hospital. Antenatally no history of hypertension or any other co-morbidity. On fourth post-op day, she developed headache and vomiting followed by one episode of seizure and after initial management she transferred to our hospital for further management. When we received patient, she was on Magsulph infusion considering postnatal eclampsia. We managed with Inj Lorazepam 2 mg intravenous and later with Inj Levetiracetam. Final diagnosis has been made as PRES after MRI and MRV brain. We found very atypical presentation of all three cases with difficulty in diagnosis and challenging management, so we are reporting these cases.

Keywords: Posterior Reversible Encephalopathy Syndrome, Reversible posterior leukoencephalopathy syndrome, Systemic hypertension

INTRODUCTION

When pregnant woman presents with hypertension, seizure and blindness at term or near term is always a diagnostic dilemma. The differential diagnosis to be kept in mind includes eclampsia, cerebrovascular accidents, and PRES. Posterior reversible encephalopathy syndrome

(PRES) is a clinical-neuroradiological entity. This syndrome initially described in 1996 by Hinchey and co-workers, as reversible posterior leukoencephalopathy syndrome. This syndrome is characterized by headache, visual disturbances, seizures, altered mental status and radiological findings of edema in the white matter of the brain areas perfused by the posterior brain circulation. While most cases are due to systemic hypertension

(HTN), other conditions also contribute as etiology or risk factors in the absence of HTN, are immunosuppressant drugs use, nephrotic state, sepsis, and systemic lupus erythematosus (SLE).¹

CASE REPORTS

Case presentation 1

A 29-year-old women G2P1, a booked case with unremarkable previous medical history reported in OPD with history of no perception of fetal movements for last three days at 27

weeks of period of gestation. USG level II was normal done at 18 weeks. Urgently USG obstetrics with doppler of umbilical artery and biophysical profile was done. Biophysical profile was 3 and umbilical artery was showing AEDF. Inj Dexamethasone started and was planned to follow with ductus venosus doppler, but she underwent preterm labour next day and was advised for LSCS as fetus was premature and there was uteroplacental insufficiency but both parents were unwilling for LSCS. She was monitored continuously on CTG during whole labour course, she delivered a live female baby with birth weight of 740 gms, cried well and had a good apgar score, shifted to NICU.

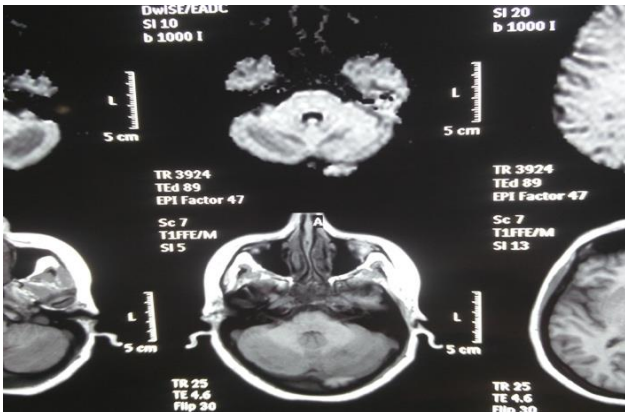


Figure 1: Diffusion weighted and FLAIR axial images at the level of inferior cerebellar peduncle show white matter FLAIR hyperintensities without any restricted diffusion, in bilateral cerebellar lobes (left>right).

Post-delivery period was uneventful of mother when she developed hypertension and started on tab amlodipine 10 mg OD and became normotensive after 3 to 4 days. 7th post-partum day she had severe headache followed by seizure. Physical examination revealed that she was not oriented to time place and person, temp 98.60F, blood pressure 150/100mm Hg, other examination of CVS, respiratory system and abdominal system was within normal limits. Inj lorazepam was given and planned for NCCT Brain. Ill-defined hypo densities (Avg HU-20) are seen in bilateral high parietal region, bilateral cerebellar lobes with sparing of the vermis with minimal mass effect seen in front of cerebellar foliae. Findings were

suggestive of Posterior reversible encephalopathy syndrome.

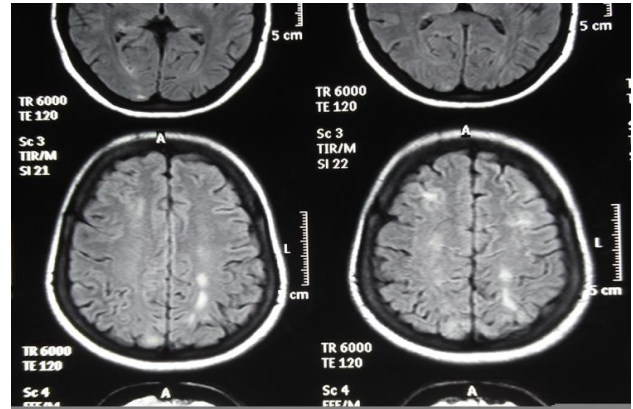


Figure 2: Patchy hyperintensities involving subcortical and deep white matters of bilateral frontal and parietal lobes with cortical sparing.

MRI Brain and MR Venography done after 3 days. Bilateral patchy focal areas of altered signal intensity areas are seen in the sub cortical as well as deep white matter of bilateral parieto-occipital, frontal and bilateral cerebellar hemisphere with cortical sparing. These areas appear hyperintense on FLAIR images. No restriction of diffusion is seen on DWI. Increased signal is seen in affected areas on ADC map suggestive of vasogenic edema. There is no significant mass effect. MRV reveals no abnormality. Others were within normal limits. In the given clinical setting findings were suggestive of Posterior reversible encephalopathy syndrome.

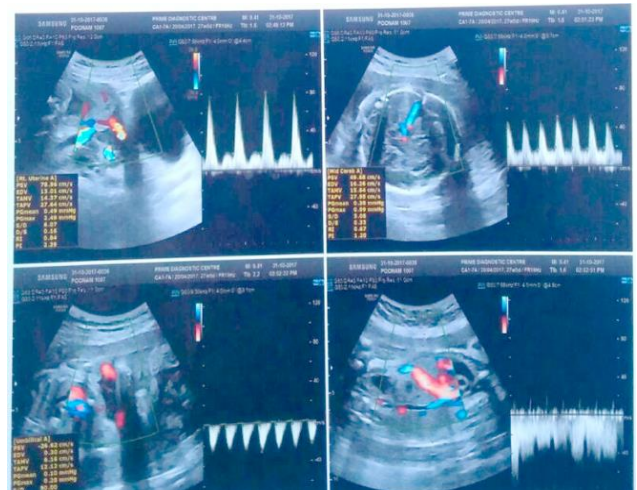


Figure 3: Spectral traces through uterine artery shows resistance to diastolic flow in the form of a diastolic notch, through the umbilical artery shows absent end diastolic flow.

Case presentation 2

23 years old primigravida with period of gestation of 37 weeks 01 day, brought in casualty with history of sudden

onset of occipital headache for 6 hours followed by vomiting three times, non-projectile, non-bilious with semi-conscious state with diminution of vision till finger count only.

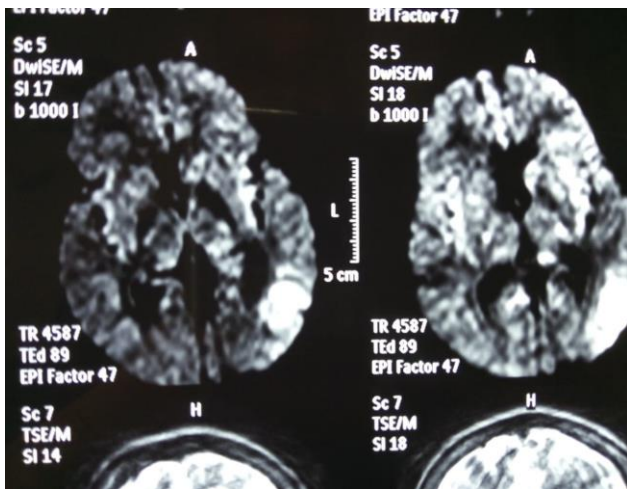


Figure 4: Diffusion weighted axial images show restricted diffusion in Left temporal lobe.

Antenatal period was uneventful with no history of hypertension, diabetes mellitus, or IUGR. On examination pulse 85 per minute regular, BP 152/90 mm Hg, no pallor, no icterus, pupil normal size, normal reactive. While examination she developed seizure three times with an interval of 20 to 30 minutes and her blood pressure remained between 140/90 to 150/90 mm Hg. Inj Lorazepam 2 mg IV given after first episode of seizure and repeated subsequently during other two episodes of seizure. Urine dipstick test was negative for protein, even then anticipating eclampsia she was started with Magsulph 4 gm slow IV followed by 1 gm per hour by controlled infusion.

There was no history of hypertension and urine protein was negative so keeping intracerebral bleed/thrombosis as a differential diagnosis a MRI brain was planned. During this management BPP was 10/10 and obstetric USG was performed with no IUGR and umbilical S/D ratio was WNL. MRI brain was suggestive of multifocal infarct in parieto-occipital and rt temporal region. She was started with inj Levetiracetam 1 gm iv bd and was planned for LSCS as she had poor Bishop score and unstable lie. On same day emergency LSCS was done and she delivered a healthy female neonate with birth weight of 2.8 kg and patient was shifted in ICU.

Postop period was uneventful and was continued with levetiracetam, her vision improved to normal by second post op day. A repeat MRI was performed on 6th post op day which revealed ill-defined areas of altered signal intensity in Lt parieto-occipital region, rt parietal region and post aspect of right external capsule, appearing hyperintense on FLAIR and T2W1 and hypotense on T1W1 causing mass effect in the form of effacement of

adjacent sulcal spaces. Normal MR Venography. Findings were suggestive of PRES. Pt was discharged on 7th post op day on oral levetiracetam and was advised to taper dose on OPD basis.

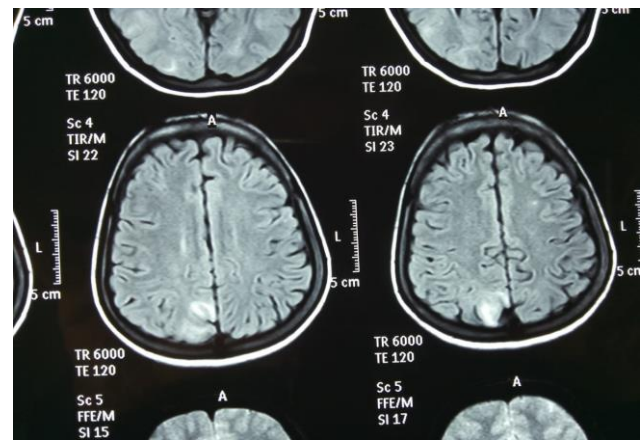


Figure 5: FLAIR axial image shows subcortical white matter hyperintensities in Right occipital lobe.

Case presentation 3

19 years old primigravida a booked ANC in another hospital. Antenatal period was uneventful with no history of Hypertension, Diabetes Mellitus. She underwent emergency LSCS at 39 weeks of period of gestation with an indication of decreased fetal movement perception with non-reassuring fetal heart rate with poor biophysical profile.

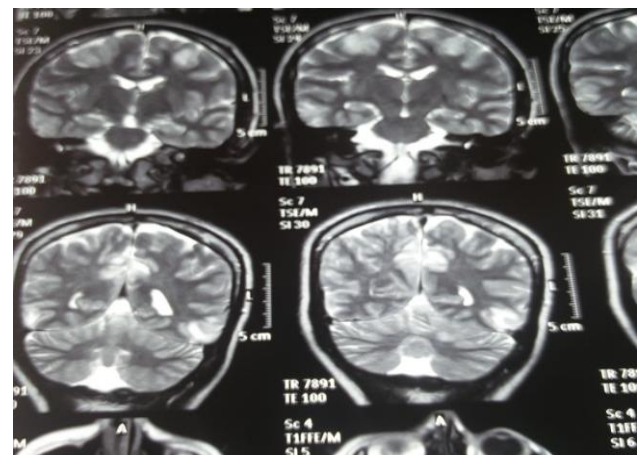


Figure 6: T2 weighted coronal images show T2 hyperintensities along subcortical and deep white matter of bilateral parietal and temporal lobes.

Immediate postpartum period was uneventful. On fourth post-operative day patient developed headache which was in occipito-frontal area and had vomiting four times which was nonprojectile in nature. Patient vitals were within normal limits. After four hours of headache she developed one episode of seizure and it was generalized tonic clonic type. Inj Lorazepam 2 mg intravenous given

and started with Magsulph infusion keeping postnatal eclampsia in mind, and a MRI brain has been recommended which was suggestive of Cerebral venous thrombosis and there was recommendation of MRV brain on MRI report. Patient has been transferred to us with same diagnosis. She has been started with Inj low molecular weight heparin 40 mg subcutaneously twice a day, Inj Levetiracetam 2mg intravenous bolus followed by 500 mg intravenous twice a day.

MRI brain and MR angiography, venography done which shows hyperintensities involving the grey and deep cortical white matter in bilateral frontal lobe, bilateral precentral gyri, bilateral high parietal bilateral occipital lobe and bilateral temporal lobe and final impression was given as posterior reversible encephalopathy syndrome. Later she was switched over to tab levetiracetam 500 mg oral once a day. Patient has been managed in obstetric ward with no further seizure episode. Patient discharged after 10 days and was called after six weeks in OPD for review.

Differential diagnosis

During pregnancy or postpartum, sudden onset of headache with seizure possibility of eclampsia comes first in mind but both patients had no history of hypertension and proteinuria during pregnancy and during postpartum period there was no proteinuria associated with hypertension, so chances of eclampsia is least likely.

We assumed a diagnosis of ischemic stroke, cerebral hemorrhage or cerebral venous thrombosis in all cases, but finally with the help of MRI, led us to a diagnosis of PRES.

Treatment

Patients has been shifted in ICU and treatment started in strict observation and with seizure precaution. Fundoscopy was normal in all patients.

Inj Levetiracetam 1 gm loading intravenous and followed by 500 mg bd iv and furthure, oral 500 mg Bd for six weeks. Patients has been discharged after, without any neurological sequelae.

Outcome and follow-up

Patients became symptom free after Levetiracetam. There was no recurrence of seizure after starting of this medicine. Patients were discharged without any neurological sequelae. Patient became asymptomatic when started with levetiracetam, but first patient discharged after five weeks as baby was in NICU being IUGR with preterm, second patient was discharged on 7th post op day and third patient discharged after 10 days. Repeat MRI done for all after 12 weeks and these were within normal limits with no focal abnormality.

DISCUSSION

Posterior reversible encephalopathy syndrome [PRES (also known as reversible posterior leukoencephalopathy syndrome)] presents with rapid onset of symptoms including headache, seizures, altered consciousness, and visual disturbance.² It is often, but by no means always, associated with acute hypertension.² If promptly recognized and treated, the clinical syndrome usually resolves within a week, and the changes seen in magnetic resonance imaging (MRI) resolve over days to weeks.³

Chronic kidney disease and acute kidney injury are both commonly present in patients with PRES, and PRES is strongly associated with conditions that co-exist in patients with renal disease, such as hypertension, vascular and autoimmune diseases, exposure to immunosuppressive drugs, and organ transplantation. It is therefore important to consider PRES in the differential diagnosis of patients with renal disease and rapidly progressive neurologic symptoms. Posterior reversible encephalopathy syndrome is an increasingly recognized disorder, with a wide clinical spectrum of both symptoms and triggers, and yet it remains poorly understood.

Normally there is a compensatory mechanism in CNS which avoid fluid leakage from intravascular space to interstitium when there is sudden increase in blood pressure. When there is sudden HTN, sympathetic tone increases resulting in vasoconstriction and facilitate this autoregulation mechanism.⁴ This autoregulation becomes abnormal in patients of PRES resulting in breakdown of normal blood brain barrier and finally leading to vasogenic brain edema.⁵ There is an apparent predisposition for edema to occur in the posterior CNS areas, particularly in the occipitoparietal areas.⁶

Vasogenic edema results from the combination of HTN and endothelial injury. Since cases of PRES without HTN have been reported, endothelial dysfunction may represent a common pathway in the pathogenesis of PRES, regardless of etiology. Therefore, endothelial damage due to other risk factors such as diabetes mellitus, dyslipidemia, and smoking may indirectly play a role in the pathogenesis of PRES.

Headache is the most common presenting symptom, but it may not be a presenting symptom in all.⁶ Other presenting features are altered sensorium, lethargy, stupor somnolence and can be brought up in coma. Visual disturbances range from blurred vision to cortical blindness, and permanent visual field defects have been reported.⁷ These patients can present with seizure or repeated episodes of seizure as first clinical history. Very rarely can present with motor function disorder such as hemiparesis, dystonia, dysarthria and dysmetria.⁸ Sluggish pupillary reflexes or frank myosis can be part of the clinical picture.⁹ Brain stem involvement manifestations comprise dyspnea, anarthria, and dysphagia.¹⁰ There is association of intracranial HTN and

PRES so on funduscopy examination papilledema and hemorrhages can be noted.¹¹ Recurrence is another clinical situation that could be present in 3.8% of the cases.¹² Histopathological changes seen in PRES include hydropic axonal swelling and myelin edema which is shown as myelin pallor without tissue destruction.¹³

The neuroimaging differential diagnosis of PRES include neoplasms, encephalitis, inflammatory and infectious processes, demyelinating pathology and cerebrovascular accidents.¹⁴ Diffusion-weighted magnetic resonance imaging (DWI) is the study of choice in PRES to discriminate between vasogenic and cytotoxic edema.

The treatment of PRES, as a secondary pathology, depends upon the determination of the underlying contributing condition; however, palliative therapy for symptoms that might worsen the outcome (e.g. seizures) must be provided, as well as strictly monitored BP control. If treatment started within time patients improves without any neurological sequel.

Hemispheric PRES patterns have been reported as well as isolated reversible brain stem and cerebellar edema, occasionally resulting in obstructive hydrocephalus.

CONCLUSION

To conclude PRES as an entity should be entertained whenever a pregnant lady presents with headache of sudden onset, altered sensorium and visual disturbances. If not diagnosed in time it may prove detrimental to both mother and fetus. Diffusion weighted MRI is the gold standard investigation for diagnosing PRES. Possibility of recurrence is to be entertained in subsequent pregnancy. Treatment of PRES depends on underlying cause. However symptomatic treatment and strict blood pressure control to be started immediately to avoid long term neurological sequelae.

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Ethical approval: The study was approved by the Institutional Ethics Committee

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