

Case Reports**Nalini Sharma, MS**

Department of Obstetrics & Gynaecology
North Eastern Indira Gandhi Regional Institute of Health
& Medical Sciences
Shillong, India

Shri R Sharma, MD, DM

Department of Medicine
North Eastern Indira Gandhi Regional Institute of Health
& Medical Sciences
Shillong, India

Kyrshanlang G Lynrah, MD

Department of Medicine
North Eastern Indira Gandhi Regional Institute of Health
& Medical Sciences
Shillong, India

Monaliza Lyngdoh, MD

Department of Medicine
North Eastern Indira Gandhi Regional Institute of Health
& Medical Sciences
Shillong, India

Mesashbha Rupsi, MD

Department of Medicine
North Eastern Indira Gandhi Regional Institute of Health
& Medical Sciences
Shillong, India

Address for correspondence:

Nalini Sharma, MS
Department of Medicine
North Eastern Indira Gandhi Regional Institute of Health
& Medical Sciences
Shillong, India

Email: srmsims_sharma@rediffmail.com

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Reversible Posterior leukoencephalopathy syndrome (RPLS) is a proposed cliniconeuroradiological entity with a vast spectrum of different aetiologies.^{2,4} The clinical hallmark of this syndrome are headache, confusion, seizures, cortical visual disturbances or blindness and, less common, other focal neurological signs. A variety of causes are known, like hypertension, pre-eclampsia/eclampsia, cyclosporin A or tacrolimus neurotoxicity, uraemia and porphyria.^{6,7,8,9} Vision loss in pregnancy is a rare occurrence. Reversible Posterior leukoencephalopathy syndrome (RPLS) is a very rare cause of sudden vision loss in pregnancy.⁵ We report a case of 36 years old female who presented to us with sudden onset vision loss with Reversible Posterior leukoencephalopathy syndrome (RPLS).

Reversible Posterior Leukoencephalopathy Syndrome: A Rare Cause of Sudden Onset Vision Loss in Pregnancy

Reversible Posterior leukoencephalopathy syndrome is a proposed cliniconeuroradiological entity characterized by headache, altered mental status, cortical blindness, seizures, and other focal neurological signs, and a diagnostic magnetic resonance imaging picture. A variety of different etiologies have been reported like hypertension, pre-eclampsia/eclampsia, cyclosporin A or tacrolimus neurotoxicity, uraemia and porphyria. With early diagnosis and prompt treatment, the syndrome is usually fully reversible. Reversible Posterior leukoencephalopathy Syndrome (RPLS)) is a very rare cause of sudden onset vision loss in pregnancy. There are only few case reports in literature. We report a case of RPLS with sudden onset vision loss in pregnancy. Clinicians as well as radiologists must be familiar with this clinically frightening, under diagnosed condition to assure timely diagnosis and treatment to prevent persistent deficits

Key words: pregnancy, reversible leukoencephalopathy, vision loss

Case Report

A thirty six years old eleventh gravid at 36 weeks of gestation presented to emergency department of our hospital. She had chief complaints of sudden loss of vision, holocranial headache, vomiting and altered sensorium and right-sided hemiparesis for 5 hours. Her past, personal and drug history were not significant. Her blood pressure was 180/120 mm of Hg. On neurological examination, the patient was found to have a confusional state, right-sided hemiparesis of grade 3/5. Her visual acuity examination revealed no perception of light bilaterally with bilateral reactive equal pupils. Funduscopy was normal and did not reveal any signs of papilledema or hypertensive retinopathy. On investigations there was 1+ proteinuria.

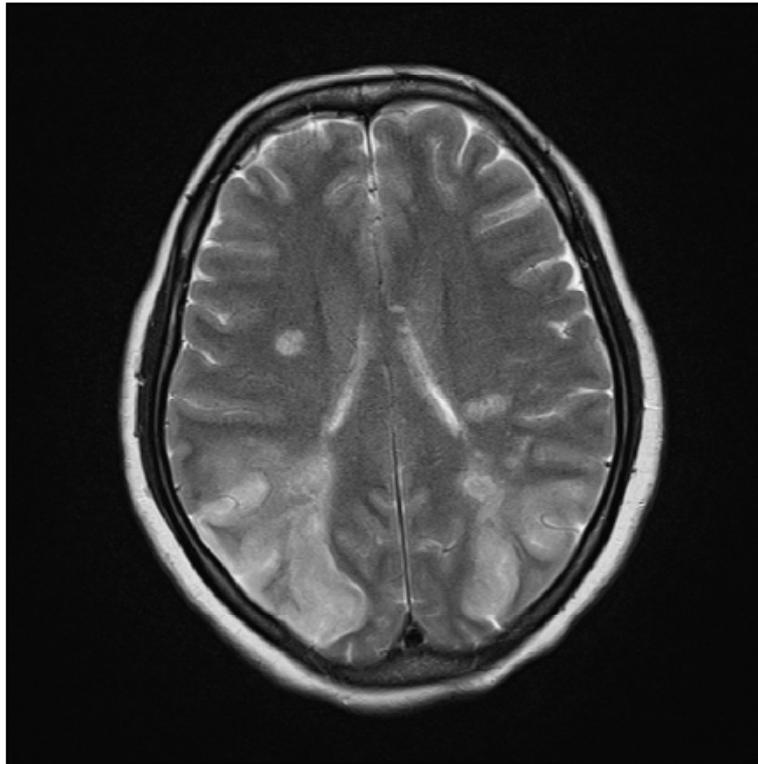


Figure 1: T2 WI MRI images demonstrating symmetric hyperintensities in bilateral parietal and occipital lobes; characteristic of Reversible Posterior leukoencephalopathy syndrome.

Her hemoglobin was 9.6 gm% with normal platelet count and normal liver and renal function tests. She was treated on the line of preeclampsia with Inj. Labetalol 20 mg IV and prophylactic Magnesium sulfate to prevent eclampsia. She had a normal vaginal delivery within four hours of admission which was uneventful. Postnatally A magnetic resonance imaging (MRI) scan revealed high signal intensities on T2-weighted images, which were most pronounced in the parieto-occipital cortices in both hemispheres (**Figure 1**). A Reversible Posterior leukoencephalopathy syndrome (RPLS) was suspected and managed conservatively. The patient started improving within the following days and she recovered completely by the seventh day.

Discussion

Common causes of visual loss in pregnancy are varied and includes thrombotic disorders of eyes and central nervous system, coagulation disorders, hyperemesis gravidarum, eclampsia and preeclampsia, central serous retinopathy and optic neuritis syndrome Reversible Posterior leukoencephalopathy syndrome (RPLS) is a very rare cause of sudden vision loss in pregnancy.⁵ RPLS can be diagnosed with (massive) reversible hyper intensities

on T2-weighted cranial MR-images. Predominantly, it affects the territory of the posterior circulation and the clinical hallmarks are headache, confusion, seizures, cortical visual disturbances or blindness and, less common, other focal neurological signs Modern imaging modalities (diffusion weighted imaging (DWI) with apparent diffusion coefficients (ADC-maps)) could prove that most cases of RPLS are caused by vasogenic rather than cytotoxic oedema.^{6,1} The pathogenesis is attributed to a failure of cerebral autoregulation that probably is facilitated in posterior brain regions due to a sparse sympathetic innervation of the vertebrobasilar vascular system.⁶ In any case, the absence of abnormalities on DWI (with increased ADC) can be interpreted as a favorable prognostic finding whereas increased signal on DWI or reduced ADC point towards frank ischaemia with cytotoxicity and cell loss.^{1,3} In our case DWI did not show any abnormality, ADC values were increased in the lesions and neurological symptoms as well as T2-hyperintensities were fully reversible. A variety of causes are known, like hypertension, pre-eclampsia/eclampsia, cyclosporin A or tacrolimus neurotoxicity, uraemia and porphyria.^{6,7,8,9} Visual changes range from loss of acuity, visual neglect, or homonymous hemianopia to complete cortical blindness. Reversible Posterior leukoencephalopathy

syndrome (RPLS) manifests on CT as hypodensities of the posterior white and gray matter. Lesions are generally bilateral and parieto-occipital, but may involve temporal or frontal lobes, brainstem or cerebellum.¹⁰ Treatment of arterial blood pressure or withdrawal of the offending drug(s) is the treatment of choice and usually results in incomplete resolution of the deficits over several days to several weeks, although partial resolution has been reported and the disease can be fatal. The key to diagnosis in RPLS naturally is the image, but suspicion must be raised by the clinician. Both should be familiar with this underdiagnosed, clinically frightening syndrome to avoid persistent deficits. It mostly is a benign, reversible condition, especially once the causative factor (e.g. hypertension) can be eliminated.

References

1. Ay H *et al.* Posterior leukoencephalopathy without severe hypertension: utility of diffusion-weighted MRI. **Neurology** **51**: 1369–1376, 1998
2. Casey SO. Posterior reversible encephalopathy syndrome: utility of fluid-attenuated inversion recovery MR imaging in the detection of cortical and subcortical lesions. **Am J Neuroradiol** **21**: 1199–1206, 2000
3. Hagemann G, Ugur T, Witte O W , Fitzek C. Recurrent posterior reversible encephalopathy syndrome (PRES). **Journal of Human Hypertension** **18**: 287–289, 2004
4. Hinchey J, Chaves C, Appignani B. A reversible posterior leukoencephalopathy syndrome. **N Engl J Med** **334**: 494–500, 1996
5. Chaudhary R, Dadhich S, Vyas J. Posterior reversible encephalopathy syndrome:—A rare cause of sudden onset vision loss in pregnancy. **NJOG** **6**: 51-52, 2011
6. Schwartz RB *et al.* Diffusion-weighted MR imaging in hypertensive encephalopathy: clues to pathogenesis. **Am J Neuroradiol** **19**: 859–862, 1998
7. Schwartz RB. Preeclampsia–eclampsia: clinical and neuroradiographic correlates and insights into the pathogenesis of hypertensive encephalopathy. **Radiology** **217**: 371–376, 2000
8. Truwit CL. MR imaging of reversible cyclosporin A-induced neurotoxicity. **Am J Neuroradiol** **12**: 651–659, 1991
9. Utz N. MR imaging of acute intermittent porphyria mimicking reversible posterior leukoencephalopathy syndrome. **Neuroradiology** **43**: 1059–1062, 2001
10. Witlin AG, Friedman SA, Egerman RS, Frangieh AY, Sibai BM. Cerebrovascular disorders complicating pregnancy: beyond eclampsia. **Am J Obstet Gynecol** **176**: 1139–1148, 1997