

Case Report

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# Posterior reversible encephalopathy syndrome in a adult female

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## Abstract

Posterior reversible encephalopathy syndrome is a clinico-neuroradiologic diagnosis, with rapidly evolving neurologic condition, characterized by headache, confusion, altered mental status, seizures, cortical blindness, lethargy, stupor, and occasionally, focal neurological signs accompanied by a typical computed tomography or magnetic resonance imaging pattern. With early recognition and treatment, complete resolution of symptoms occurs. Typical imaging findings characteristically involve the white matter bilaterally in the parieto-occipital regions. Atypical imaging finding of contrast enhancement of lesion can occur, but is less common. A 20-year-old primiparous lady presented with posterior reversible encephalopathy syndrome. To the best of our knowledge, this is the first documented case in Nigeria. This case-report highlights the importance of recognizing the salient imaging features in this lethal but reversible entity with prompt management.

**Keywords:** Female, leukoencephalopathy, Nigerian, posterior, reversible

## Résumé

Syndrome d'encéphalopathie réversible postérieure est un diagnostic clinico-neuroradiologiques, avec l'évolution rapide des conditions neurologiques, caractérisée par des maux de tête, confusion, état mental altéré, convulsions, une cécité corticale, léthargie, stupeur, et, parfois, des signes neurologiques focaux accompagnée d'une structure calculée typique de tomодensitométrie ou imagerie par résonance magnétique. La reconnaissance précoce et le traitement, une résolution complète des symptômes se produit. Les résultats d'imagerie typiques impliquent typiquement la substance blanche sur le plan bilatéral dans les régions pariéto-occipitale. Atypique conclusion d'imagerie de l'amélioration du contraste de la lésion peut survenir, mais est moins fréquente. Une dame primipare de 20 ans, présentée avec syndrome d'encéphalopathie postérieure réversible. Au meilleur de notre connaissance, c'est le premier cas documenté au Nigeria. Cet affaire-rapport met en évidence l'importance de reconnaître le saillant d'imagerie caractéristiques dans cette entité létale mais réversible avec gestion rapide.

**Mots-clés:** Femelle, leucoencéphalopathie, nigérian, postérieure, réversible

## Introduction

Posterior reversible encephalopathy syndrome (PRES) is a clinico-neuroradiologic diagnosis, which was first described by Hinchey in 1996.<sup>[1]</sup> This rapidly

evolving neurologic condition is characterized by headache, confusion, altered mental status, seizures, cortical blindness, lethargy, stupor, and occasionally, focal neurological signs accompanied by a typical CT or MR imaging pattern.<sup>[2,3]</sup>

Classic neuroimaging applied to patients shows edema involving the white matter in the posterior portions of the cerebral hemispheres, especially bilaterally in the parieto-occipital regions.<sup>[4]</sup> Atypical imaging findings include contrast enhancement, hemorrhage, and restricted diffusion on MRI.

Significant reversal of neuroradiologic abnormalities, along with complete clinical recovery, is suggestive of the diagnosis<sup>[5]</sup> as in this case report.

We report a case of PRES in a woman with postpartum eclampsia, highlighting the imaging findings, and emphasizing the need for prompt diagnosis and treatment.

## Case Report

A 20-year-old primipara was referred to University of Ilorin Teaching Hospital, (UITH), on account of post-partum eclampsia from a private hospital.

Patient was booked and received antenatal care at the private hospital, which was uneventful. However, she had an episode of intrapartum generalized tonic-clonic seizure, which was aborted with 10 mg of diazepam, and she delivered a 2.7 kg live male baby via spontaneous vaginal delivery (SVD).

She remained stable and was discharged 5-days after delivery. Two days after discharge, she developed generalized headache, dizziness, and blurring of vision. She was, therefore, referred to UITH.

On physical examination, she was found to be drowsy but arousable, pale, anicteric, afebrile. BP = 160/100 mmHg, urinalysis showed protein++ (300 mg/dl).

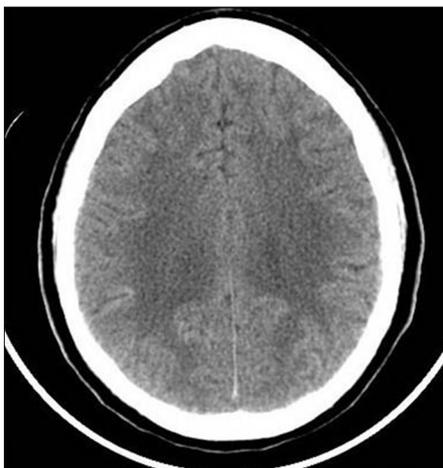
A diagnosis of post-partum eclampsia was made, and she was commenced on magnesium sulfate infusion, intermittent hydralazine infusion, and oral moduretic. On the second day of admission, she started talking irrationally and incoherently. The neurologist was invited to review the patient. On neurological examination, left lower limb was laterally rotated. There was global hypotonia and brisk tendon reflexes. Power was zero in the left upper limb (LUL) and left lower limb (LLL) and 3/5 in other limbs. There were no signs of meningitis. The pupils were equal in size (4 mm) bilaterally and reactive to light. The rest of systemic examinations were normal.

She later complained of severe headache and visual disturbances that had been increasing in severity over the preceding two days. She was agitated, disorientated, and confused. An impression of right hemispheric stroke to exclude venous sinus thrombosis was made by the clinician at this stage.

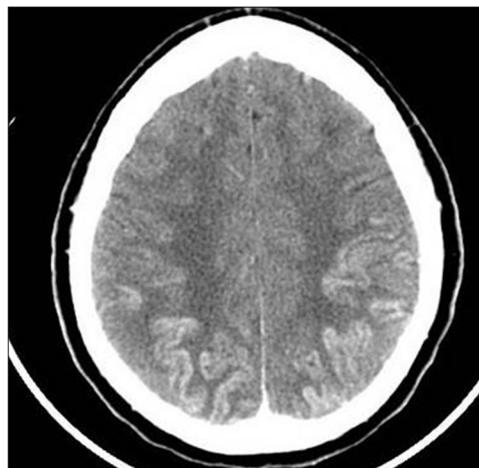
During ophthalmologic review, fundoscopic examination was unremarkable. Diagnosis of cortical blindness was, however, made 5<sup>th</sup> day post-admission. She had since been co-managed by neurologist, ophthalmologist, and physiotherapist. After three days on admission, the BP was maintained between 120/90 and 110/80 mmHg. Full blood count, urea, and creatinine were within normal limits.

Cranial CT scan done appeared normal on pre-contrast scan [Figure 1], but showed relatively bilaterally symmetrical posterior parieto-occipital gyriform enhancement on post-contrast scan [Figure 2]. There was no evidence of intracranial hemorrhage or cerebral infarction.

Based on the findings, a diagnosis of posterior reversible encephalopathy syndrome (PRES) was entertained.



**Figure 1:** Non-contrast CT (axial view) showing no obvious abnormality



**Figure 2:** Contrast CT (axial view) showing bilateral gyriform enhancement in the posterior cerebral hemispheres

Protein became negative 10<sup>th</sup> day post-admission undetectable by dipstick. Thereafter, the vision and the power in all the limbs improved progressively. She became ambulant and could count fingers and identify colors at 23 day post-admission. She made full recovery with no neurological sequelae, thus confirming the radiologic diagnosis of PRES. She was discharged after 25 days on admission to be followed up in the clinic. Repeat cranial CT after recovery could not be done due to financial constraint. She had been seen at follow-up clinic twice, with full recovery of vision including ability to read, and also full functions of all the limbs.

## Discussion

The terms posterior reversible leukoencephalopathy, reversible posterior cerebral edema syndrome, or posterior reversible encephalopathy syndrome (PRES) refer to a clinico-radiologic entity characterized by headaches, confusion, visual disturbances, seizures, and posterior transient changes on neuroimaging.<sup>[6]</sup> PRES is usually associated with toxemia of pregnancy, acute hypertension, renal insufficiency, or immunosuppressive therapy.

The association of PRES with toxemia of pregnancy is well-established.<sup>[7]</sup> Before PRES was first explicitly characterized in 1996, a number of case reports described the MRI finding of posterior leukoencephalopathy as an interesting anomaly in the eclamptic patient. It has thereafter been reported severally in association with postpartum eclampsia, after an uncomplicated vaginal delivery,<sup>[8]</sup> as in this case report. Other well-recognized underlying causes associated with PRES include uremia, hemolytic-uremic syndrome, SLE, medications such as cyclosporine and tacrolimus, and chemotherapeutic agents such as cisplatin, interferon alpha, and intrathecal methotrexate.<sup>[9,10]</sup> Autoimmune connective tissue diseases, thrombotic thrombocytopenic purpura, HIV syndrome, acute intermittent porphyria, organ transplantation have also been reported.<sup>[5,11]</sup>

The pathophysiology of PRES has been a source of extensive debate among many investigators; however, it is thought to be related to hypertension-induced uncontrolled vasospasm, coupled with autoregulatory failure, and endothelial cell dysfunction/injury leading to blood-brain barrier leakage, with resultant cortical and sub-cortical vasogenic edema.<sup>[2,12,13]</sup>

The vulnerability of the posterior cerebral circulation compared with the anterior brain circulation may be related to differences in autonomic innervations. Sympathetically mediated vasoconstriction protects the anterior circulation from over-perfusion during acute hypertension and vasospasms secondary to sudden and severe increase in blood pressure or brain ischemia.<sup>[12]</sup>

There is a paucity of information on PRES in Africa possibly because of under-diagnosis in view of the need to have neuroimaging for confirmation. Araqi-Houssaini and colleagues reported a case series of 13 patients with eclampsia managed over a period of 5 years with total regression of symptoms in a significant number of them.<sup>[14]</sup>

The clinical findings are often non-specific; so, the diagnosis may be difficult to establish, particularly in patients who have other illness. Various neurologic conditions such as stroke, intracranial hemorrhage, venous thrombosis, and encephalitis could mimic PRES.<sup>[9]</sup>

The most characteristic imaging pattern in PRES is the presence of edema involving the white matter of the posterior portions of both cerebral hemispheres, especially the parieto-occipital regions, in a relatively symmetric pattern that spares the calcarine and para-median parts of the occipital lobes.<sup>[13]</sup> Asymmetric appearance had also been noted.<sup>[15]</sup> Other structures such as the brain stem, cerebellum, and frontal and temporal lobes) may also be involved, and although the abnormality primarily affects the sub-cortical white matter, the cortex and the basal ganglia may also be involved.<sup>[13,16,17]</sup>

Occasionally focal areas of edema may be beyond the resolution of CT scan, and this could account for the apparently normal CT in this patient.<sup>[15]</sup> Although they are rare, gyriform enhancement or parenchymal hemorrhage can occur in complicated cases.<sup>[6,13]</sup> Gyriform enhancement on contrast-enhanced CT as seen in this case, parenchymal hemorrhage and restricted diffusion on MRI, have all been described as atypical findings.<sup>[16]</sup> Although not widely described in the literature, incidence varying between 33% and 37% has been reported.<sup>[16,17]</sup>

Reasons proffered for these discrepancies in imaging findings in PRES include contrast bolus timing, amount, and type, and may also be caused by lack of contrast use in many studies, possibly because contrast-enhanced imaging may not be necessary to diagnose PRES.<sup>[11]</sup>

Furthermore, imaging findings in PRES have been characteristically described on MRI, which remains the investigation of choice. Conventional MRI shows high signal intensity on T2-weighted and FLAIR images. The lesions usually do not enhance on post-contrast T1-weighted MR images.<sup>[9]</sup>

Diffusion-Weighted Imaging (DWI) will show isointensity and increased signal intensity on (Attenuation Diffusion Coefficient) ADC values in all cases, indicating vasogenic edema.<sup>[11,16,17]</sup> DWI

is useful in PRES because it can distinguish between vasogenic and cytotoxic edema.<sup>[10,12,13,16]</sup> Although MRI was recommended for this patient, was not done due to lack of funds.

Symptoms usually resolve rapidly once appropriate treatment is started,<sup>[3]</sup> but delayed diagnosis and treatment could result in permanent neurological sequelae and death.<sup>[9]</sup>

## Conclusion

PRES is potentially a reversible condition despite its dramatic presentation. We will advocate routine use of contrast in suspected case of PRES on CT, especially in a developing country like Nigeria where MRI is expensive and relatively unavailable. CT may provide the only clue to diagnosis as presented in this case. Recognizing this syndrome will enable physicians to avoid a delay in diagnosis and institute treatment promptly to avoid permanent neurological sequelae.

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