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PRES in Lupus Nephritis and Sickle Cell Trait: A Diagnostic and Therapeutic Challenge

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Abstract: Posterior reversible encephalopathy syndrome (PRES) is a rare, potentially reversible clinico-radiological condition affecting a small percentage of patients with systemic lupus erythematosus (SLE). This case report describes a 26-year-old female with SLE and type IV lupus nephritis who presented with seizures, altered sensorium, and hypertension. MRI revealed bilateral symmetrical T2/FLAIR hyperintensities in multiple brain regions, confirming PRES. Despite intensive management, including antihypertensive and anticonvulsant therapy and extraventricular drain placement for hydrocephalus, the patient developed multiorgan dysfunction and succumbed following cardiac arrest. This case underscores the importance of considering PRES in young females with SLE presenting with neurological symptoms and highlights the need for prompt radiological diagnosis and management to prevent irreversible damage or death.

Keywords: *Hypertension; Lupus Nephritis; Posterior Reversible Encephalopathy Syndrome.*

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I. INTRODUCTION

Posterior reversible encephalopathy syndrome (PRES) is a rare clinico-radiological entity characterized by vasogenic edema, predominantly affecting the posterior cerebral white matter. It occurs in approximately 1.4% of patients with systemic lupus erythematosus (SLE) [1], often associated with hypertension, antiphospholipid antibody positivity, or renal failure [1]**.** PRES is more common in females and presents with non-specific neurological symptoms, including seizures, headache, altered mental status, and visual disturbances [2]**.** Risk factors include severe hypertension, autoimmune disorders like SLE, eclampsia, and cytotoxic drugs [3]. Magnetic resonance imaging (MRI) is the gold standard for diagnosis, revealing characteristic T2/FLAIR hyperintensities [4]. This case report highlights the importance of early recognition and management of PRES in a young female with SLE to prevent adverse outcomes.

II. CASE REPORT

A 26-year-old female with a known history of SLE and histopathologically- proven type IV lupus nephritis presented to the ER of NRI Institute of Medical Sciences (NRIIMS), Visakhapatnam, India, with altered sensorium and generalized tonic-clonic seizures, as reported by her

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caregiver. The caregiver noted approximately 20 seizure episodes, each lasting 2–3 minutes, characterized by involuntary limb movements, eye-rolling, teeth clenching, and drooling. She had a one-day history of fever and vomiting prior to admission. No signs of neck rigidity were noted. The patient had been on oral Prednisolone 25 mg daily for four months following her SLE diagnosis and was known to have sickle cell trait.

On examination, she exhibited hypertension (BP: 210/110 mmHg), tachycardia (pulse: 110 bpm), fever (100°F), and bilateral forearm rashes. Due to unresponsiveness and absence of localizing signs, she was intubated, transferred to the intensive care unit (ICU), and started on antihypertensives and anticonvulsants. Laboratory investigations revealed leukocytosis, thrombocytopenia, elevated inflammatory markers (CRP, procalcitonin, ESR), proteinuria, hematuria, and deranged liver and renal function tests.

➢ *Radiological Findings*

- *Initial MRI Brain:*

MRI revealed bilateral symmetrical T2-weighted and FLAIR hyperintensities with high signal intensity on apparent diffusion coefficient (ADC) sequences, involving the cortical and subcortical regions of the frontal, parietal, temporal, occipital lobes, and cerebellar cortex, suggestive of vasogenic edema (Figure 1 & 2) . After excluding differentials such as lupus cerebritis and ischemic stroke, a diagnosis of PRES secondary to SLE was confirmed.

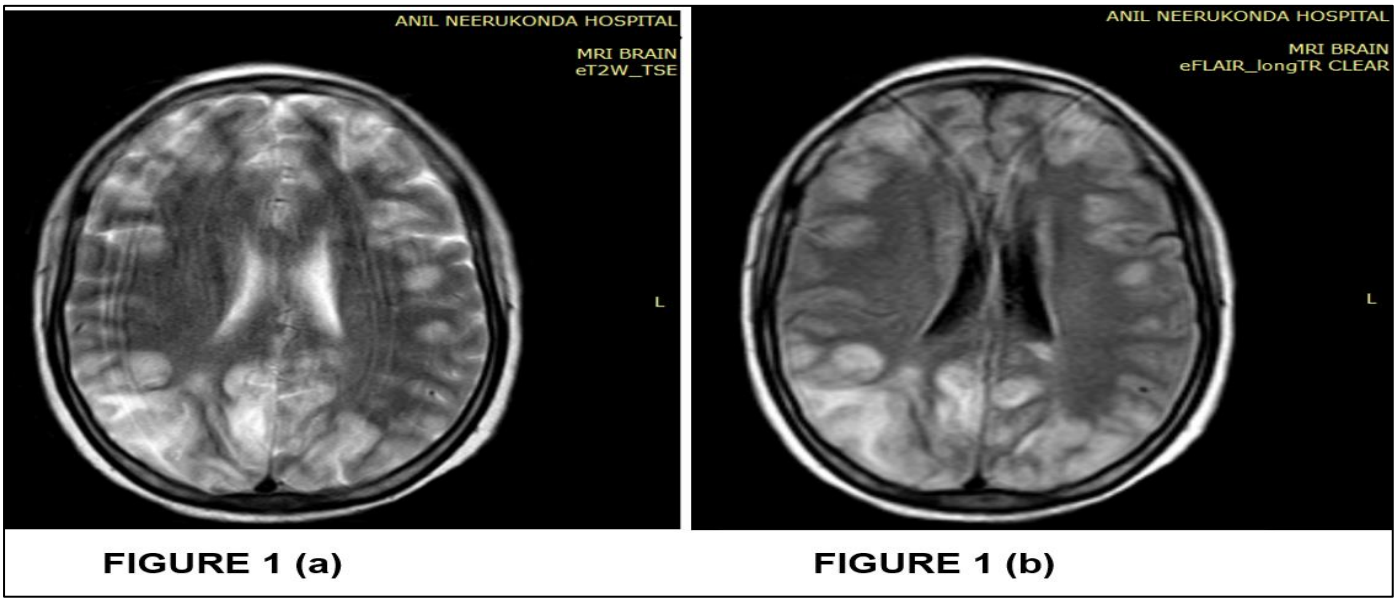

Fig 1 Axial T2-weighted (a) and FLAIR (b) MRI sequences showing bilateral symmetrical hyperintensities in the cortical and subcortical regions of the frontal and parietal lobes, indicative of vasogenic edema consistent with PRES.

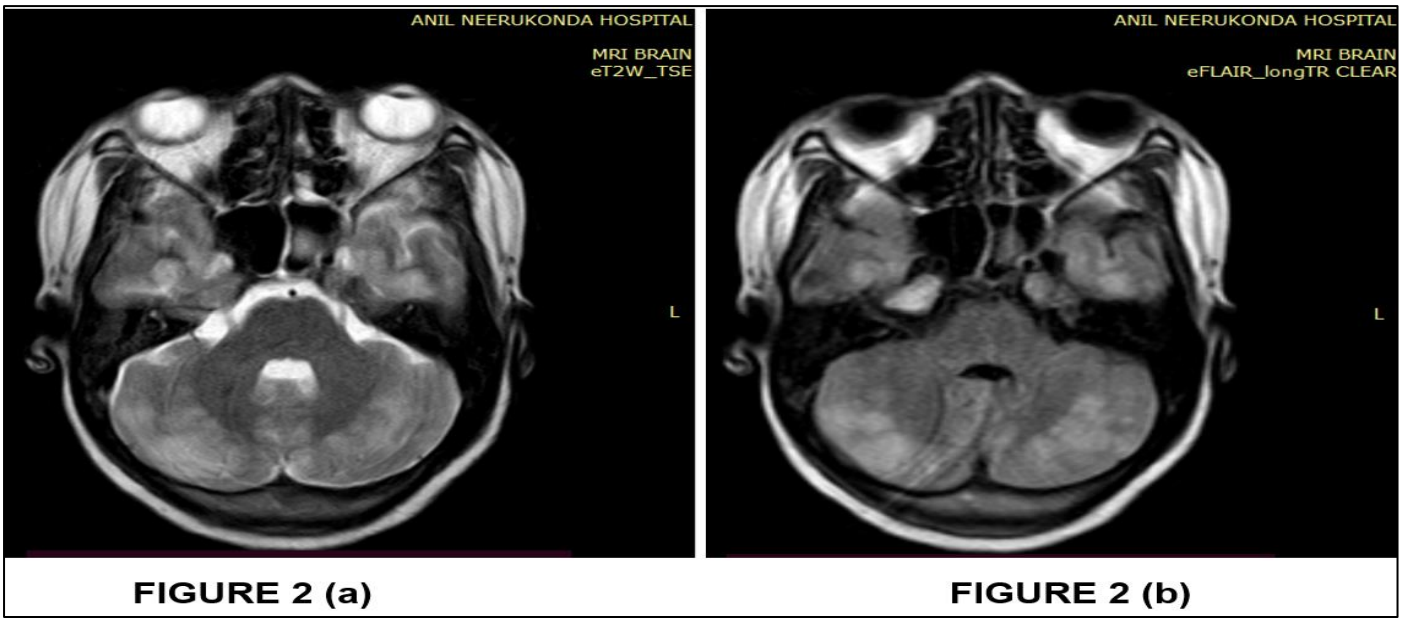

Fig 2 Axial T2-weighted (a) and FLAIR (b) MRI sequences showing bilateral symmetrical hyperintensities showing bilateral symmetrical hyperintensities in cerebellar hemispheres respectively.

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- *Subsequent CT Brain:*

Due to clinical deterioration despite medical management, a non-contrast CT (NCCT) brain was performed. It revealed ill-defined hypodensities in the bilateral frontal, high parietal, posterior temporal, occipital lobes, and cerebellar hemispheres, causing mass effect on the fourth ventricle and secondary hydrocephalus (Figure 3). These findings indicated advanced PRES with raised intracranial pressure. An emergency extraventricular drain (EVD) was placed.

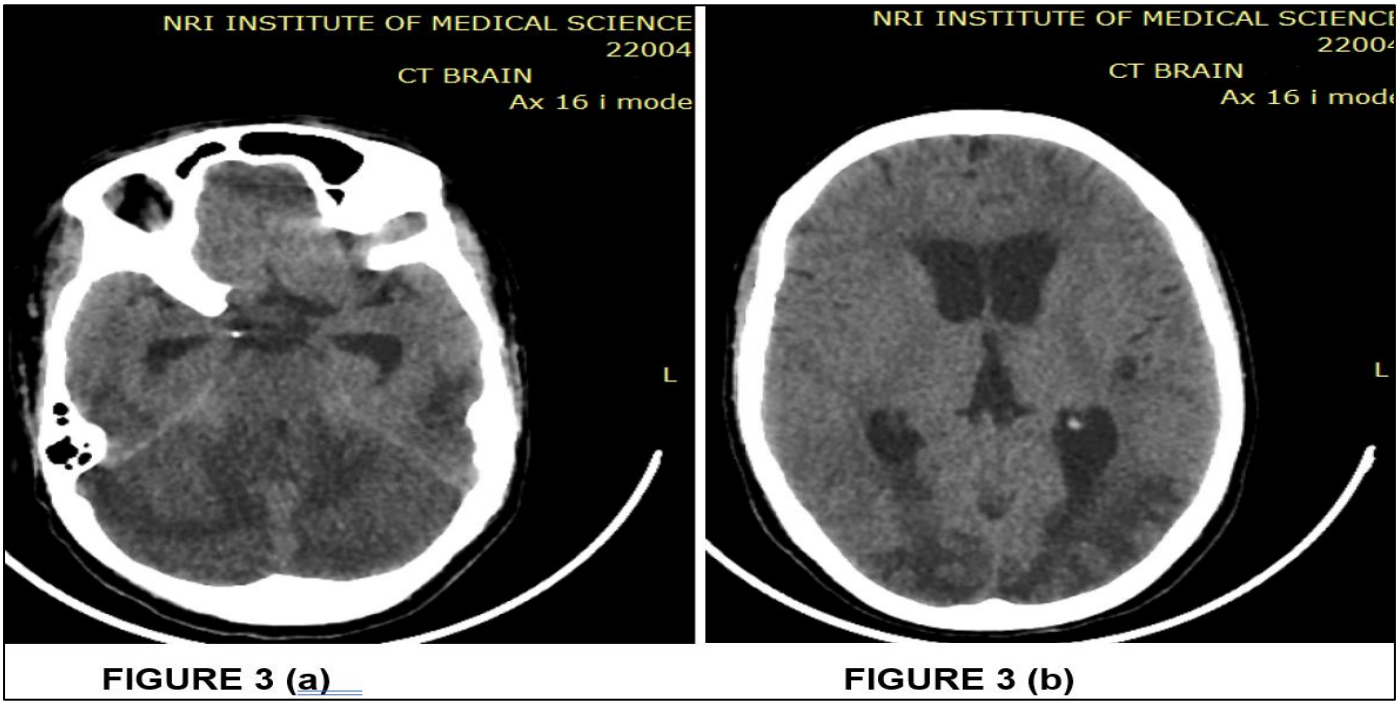

Fig 3 Axial CT brain showing hypodensities in bilateral cerebellar hemispheres with mass effect on the fourth ventricle (a) and early ventriculomegaly with an Evans index of ~0.3, with hypodensities in the white matter of bilateral posterior occipital lobes (b).

- *Follow-Up CT Brain (Day 5 Post-EVD):*

The follow-up CT showed complete resolution of hypodensities and normalization of ventricular size, indicating radiological reversibility of PRES (Figure 4).

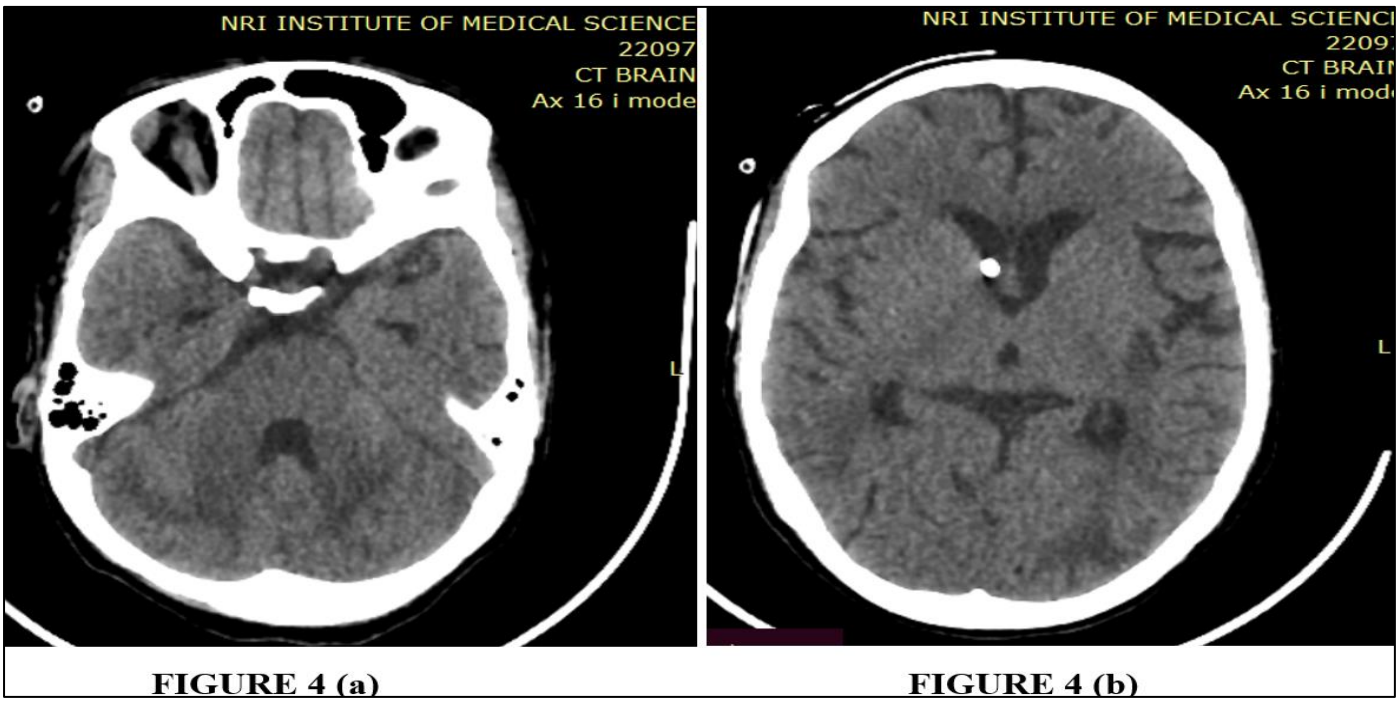

Fig 4 Axial CT brain showing resolution of hypodensities in bilateral cerebellar hemispheres with a normal fourth ventricle (a) and extraventricular drain in the right lateral ventricle with normal-sized ventricles (b).

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➢ *Hospital Course*

The patient remained in the ICU, with complications including renal failure, persistent hypertension, severe anemia, and thrombocytopenia. She subsequently developed hypotension and bradycardia, leading to cardiac arrest. Resuscitation was unsuccessful, and the patient succumbed.

III. DISCUSSION

Posterior reversible encephalopathy syndrome (PRES), first described by Hinchey et al. in 1996, is a rare neurological disorder characterized by vasogenic edema due to impaired cerebral autoregulation or endothelial dysfunction, often triggered by acute hypertension or autoimmune conditions like SLE [5].

The term PRES may be a misnomer, as lesions are not always located posteriorly and reversibility is not always present. Some cases of fatal outcome or irreversible neurological damage following PRES have been reported. This neurological deficit is also widely misdiagnosed and presents with nonspecific symptoms, including headaches, seizures, altered mental state, visual disturbance, ataxia, vertigo, or even a coma [6].

PRES is a clinical and radiological entity, associating a pathognomonic triad, characterized with neurological symptoms, typical imaging findings and a rapidly reversible evolution as soon as the causing factor is managed [7].

This case illustrates a severe presentation of PRES in a young female with SLE, complicated by lupus nephritis and sickle cell trait, culminating in a fatal outcome.

PRES in SLE is associated with risk factors such as hypertension, renal dysfunction, and immunosuppressive therapy [1]. Our patient's uncontrolled hypertension (BP: 210/110 mmHg) and active lupus nephritis likely contributed to the development of PRES. The sickle cell trait may have exacerbated microvascular dysfunction, though its role in PRES remains underexplored [8]. Differential diagnoses in SLE patients with neurological symptoms include lupus cerebritis, neuropsychiatric SLE, ischemic stroke, and meningitis [7]. Table 1 outlines key distinguishing features [7].

Radiologically, MRI is the gold standard, showing bilateral symmetrical T2/FLAIR hyperintensities with high ADC signal, distinguishing vasogenic edema in PRES from cytotoxic edema in ischemia [7]. CT is useful in emergencies to detect complications like hydrocephalus, as seen in our case, where CT-guided EVD placement was critical. The resolution of radiological findings post-EVD underscores PRES's potential reversibility, though clinical outcomes depend on timely intervention. Delayed diagnosis or management, as occurred in this case due to rapid multiorgan dysfunction, can lead to irreversible damage or death [9,10].

This case highlights the need for heightened suspicion of PRES in SLE patients with neurological symptoms, particularly young females with hypertension or renal involvement. The fatal outcome emphasizes the importance of aggressive management of underlying triggers and complications [9,10].

Table 1 Differential Diagnosis for Neurological Symptoms in SLE

Condition	Key Features	Imaging Findings
PRES	Seizures, hypertension, reversible edema	T2/FLAIR hyperintensities, vasogenic edema
Lupus Cerebritis	Focal deficits, cognitive impairment	Patchy enhancement, infarcts
Neuropsychiatric SLE	Psychiatric symptoms, seizures	Variable, often normal or non-specific
Ischemic Stroke	Focal neurological deficits	DWI restriction, cytotoxic edema
Meningitis	Fever, neck rigidity, headache	Meningeal enhancement, CSF findings

IV. CONCLUSION

Posterior Reversible Encephalopathy Syndrome (PRES) is a critical yet underrecognized complication in systemic lupus erythematosus (SLE). This case highlights the following key points:

- **Diagnostic Approach**: PRES should be suspected in SLE patients presenting with seizures, altered sensorium, and hypertension.
- **Imaging Interpretation**: MRI with T2/FLAIR sequences is the gold standard, showing symmetrical vasogenic edema; CT is valuable for detecting complications like hydrocephalus.
- **Management Urgency**: Prompt antihypertensive and anticonvulsant therapy, along with interventions like extraventricular drainage, are essential to mitigate complications.
- **Prognostic Markers**: Uncontrolled hypertension and multiorgan dysfunction are associated with poor outcomes, emphasizing the need for early intervention.

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DISCLOSURES

- **Conflict of Interest**: None declared.
- **Funding**: No external funding was received.
- **Institutional**: NRI Institute of Medical Sciences, Visakhapatnam, India.
- **Ethical Committee**: Not applicable. This anonymized case report, based on de-identified clinical findings from

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a deceased patient, does not require Institutional Ethics Committee approval.

- **Approval Number**: N/A.
- **Informed Consent**: Informed written consent for publication of clinical details and images was obtained from the patient's next of kin.
- **Data Availability**: All data related to this case report are included in the manuscript.

REFERENCES

[1]. Liu B, Zhang X, Zhang FC, et al. Posterior reversible encephalopathy syndrome could be an underestimated variant of "reversible neurological deficits" in systemic lupus erythematosus. *BMC Neurol*. 2012;12:152. doi:10.1186/1471-2377-12-152.

[2]. Mikdashi J, Nived O. Measuring disease activity in adults with systemic lupus erythematosus: the challenges of administrative burden and responsiveness to patient concerns in clinical research. *Arthritis Res Ther*. 2015;17:1-10. doi:10.1186/s13075-015-0702-6.

[3]. Merayo-Chalico J, Barragán-Martínez C, Rodríguez-Jiménez P, et al. Clinical outcomes and risk factors for posterior reversible encephalopathy syndrome in systemic lupus erythematosus: a multicentric case–control study. *J Neurol Neurosurg Psychiatry*. 2016;87(3):287-94. doi:10.1136/jnnp-2014-310404.

[4]. Vaysman T, Xu P, Vartanian T, et al. "Highlighting" red nuclei by atypical posterior reversible encephalopathy syndrome in a patient with systemic lupus erythematosus. *Clin Case Rep*. 2019;7(7):1404-8. doi:10.1002/ccr3.2247.

[5]. Hinchey J, Chaves C, Appignani B, et al. A reversible posterior leukoencephalopathy syndrome. *N Engl J Med*. 1996;334(8):494-500. doi:10.1056/NEJM199602223340803.

[6]. Ellis CA, McClelland AC, Mohan S, et al. Cerebrospinal fluid in posterior reversible encephalopathy syndrome: implications of elevated protein and pleocytosis. *Neurohospitalist*. 2019;9(2):58-64. doi:10.1177/1941874418802061.

[7]. Afilal I, Nasri S, Bendaoud M, et al. Fatal outcome of posterior reversible encephalopathy syndrome (PRES) in a lupus nephropathy patient: a case report. *Radiol Case Rep*. 2022;17(6):2215-9. doi:10.1016/j.radcr.2022.03.084.

[8]. Thust SC, Burke C, Siddiqui A. Neuroimaging findings in sickle cell disease. *Br J Radiol*. 2014;87(1040):20130699. doi:10.1259/bjr.20130699.

[9]. Kwon S, Koo J, Lee S. Clinical spectrum of reversible posterior leukoencephalopathy syndrome. *Pediatr Neurol*. 2001;24(5):361-364. doi:10.1016/S0887-8994(01)00265-X.

[10]. Antunes NL, Small TN, George D, et al. Posterior leukoencephalopathy syndrome may not be reversible. *Pediatr Neurol*. 1999;20(3):241-3. doi:10.1016/S0887-8994(98)00244-2.