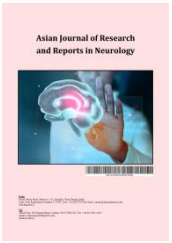


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# Severe form of Reversible Posterior Encephalopathy Syndrome (PRES) in a Patient with Lupus

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***Authors' contributions***

*This work was carried out in collaboration among all authors. All authors read and approved the final manuscript.*

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***Case Study***

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## ABSTRACT

PRES represents a clinico-radiological syndrome indicating spontaneously reversible cerebral distress. It associates predominantly neurological manifestations with white matter signal abnormalities on brain magnetic resonance imaging.
The purpose of our work is to highlight the clinical and evolutionary characteristics of severe forms of PRES, aiming to derive therapeutic and prognostic considerations.
We present a case of a patient with systemic lupus erythematosus complicated by nephropathy, in whom the diagnosis of PRES was established, with a favourable outcome under treatment.

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*Keywords: Pres; lupus; reversible cerebral distress.*

## 1. INTRODUCTION

Reversible Posterior Encephalopathy Syndrome (PRES), a clinico-radiological entity, was recently characterized in 1996. In the context of neurological studies, brain imaging, particularly magnetic resonance imaging (MRI), reveals multifocal lesions of the white matter, mainly localized in the parieto-occipital region of the cerebral hemispheres [1,2]. The pathophysiology of this syndrome, however, remains poorly understood. The aim of this work is to emphasize the clinical and evolutionary features of severe forms of PRES, providing therapeutic and prognostic insights [3-5]. Labelling a neurological condition requiring pathophysiological treatment as PRES could potentially deprive many patients of recovery chances or even survival (e.g., thrombolysis for CVAs) [6-8]. On the other hand, ignoring PRES could be detrimental to the patient's condition and recurrence risks. We report a case of severe PRES in a patient who underwent surgical intensive care.

## 2. CASE PRESENTATION

A 30-year-old female patient with systemic lupus erythematosus complicated by nephropathy was urgently hospitalized due to sudden-onset generalized seizures preceded by intense headaches two hours earlier. Clinical examination revealed a confusional state, right hemiparesis, GCS of 13, body temperature of 37.8°C, blood pressure of 200/130 mmHg, and heart rate of 100 bpm. After treatment with calcium channel blockers for hypertensive peak control, a brain CT scan (Fig. 1) showed multiple ill-defined hypodense lesions in bilateral parieto-occipital regions, resembling venous ischemic strokes.

A treatment regimen involving antihypertensives, anticoagulants, and antiepileptics was initiated. The patient's condition improved under treatment, becoming conscious without seizures and stabilized blood pressure. Follow-up brain magnetic resonance imaging (MRI) performed 11 days later revealed no parenchymal lesions, with complete regression of cerebral ischemic lesions (Fig. 2).

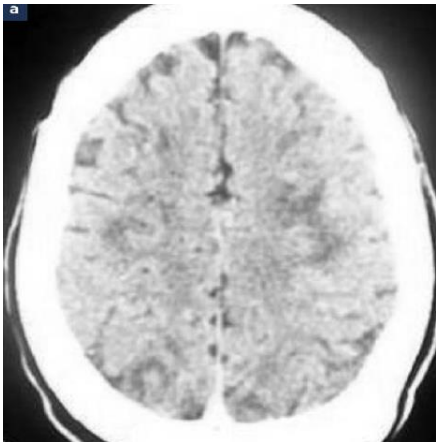

**Fig. 1. A brain CT scan**

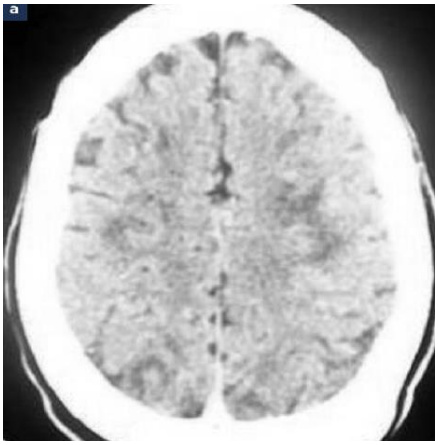

**Fig. 2. Brain magnetic resonance imaging (MRI) of cerebral ischemic lesions**

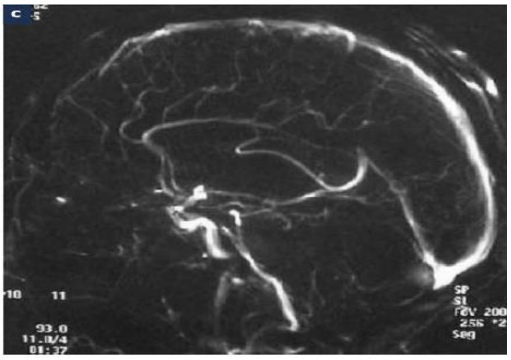

**Fig. 3. Angio-MRI demonstrated normal patency of venous sinuses**

## 3. DISCUSSION

PRES represents a clinico-radiological syndrome indicating spontaneously reversible cerebral distress. The true incidence of PRES remains uncertain due to limited published data, diverse etiologies, various nomenclatures, and the lack of awareness among clinicians [1-2]. The typical clinical presentation includes thunderclap headaches recurring over one to three weeks,

accompanied by nausea and vomiting. A diastolic blood pressure exceeding 120 mmHg is usually observed [3]. Status epilepticus is possible. The most frequent complications are subarachnoid hemorrhages (20-25%) and parenchymal vascular events (5-10%), such as hematomas or infarctions. Clinical and radiological events manifest at different times during the disease course. Hemorrhages (subarachnoid hemorrhages and hematomas) as well as reversible posterior encephalopathy syndromes (PRES) occur early, usually within the first week. In contrast, ischemic complications like transient ischemic attacks and infarctions manifest significantly later, around the second week. Diagnosis requires demonstrating multifocal and segmental cerebral vasoconstriction through angiography (magnetic resonance angiography, CT angiography, or conventional angiography) and proving the reversibility of these anomalies through follow-up angiography within 12 weeks of onset. The pathogenesis of this syndrome remains unknown [4,5]. Three theories attempt to better understand the pathophysiological mechanisms:

The first theory, and the most common [6,7], is based on elevated blood pressure on one hand and rapid symptom resolution upon stabilization of blood pressure.

The second theory focuses on cerebral vasoconstriction resulting from high blood pressure or a certain systemic process. The consequence of this autoregulation phenomenon is decreased cerebral perfusion, leading to vasogenic edema [4,8,9].

The third theory implicates endothelial dysfunction, emphasizing the role of systemic inflammation and endothelial activation, potentially related to medications, infections, or underlying diseases [5].

Therapeutic strategies depend on the etiology and clinical presentation of PRES. The primary treatment goals include:

- Blood pressure control: Controlling hypertension is of paramount importance and involves regular antihypertensive agents. Calcium channel blockers (nicardipine or diltiazem), beta-blockers (e.g., labetalol), and diuretics are commonly used. Arterial vasodilators like sodium nitroprusside and diazoxide are second-line options. Selective neuroprotective calcium channel blockers like nimodipine are useful in preventing cerebral vasospasms, particularly in hypertensive encephalopathy during pregnancy [10].
- Seizure cessation: Benzodiazepines (clonazepam or diazepam) should be administered intravenously as first-line treatment. Phenytoin or phenobarbital can be used as second-line treatments or for status epilepticus. Valproic acid is a therapeutic option, especially in heart failure, elderly patients, and neurological intensive care units. Continuous magnesium sulfate infusion is recommended for pregnant women.
- Anti-edema agents: Anti-edema treatment (mannitol) and corticosteroid administration should be considered on a case-by-case basis and may be beneficial in certain situations [10].

Although treatment led to a positive outcome, there remains a risk of recurrence; however, their frequency and timing remain unknown. Hence, the use of sympathomimetics and serotonergic agents should be definitively discouraged.

## 4. CONCLUSION

Reversible Posterior Encephalopathy Syndrome (PRES) is a clinico-radiological entity denoting spontaneously reversible cerebral distress, not widely recognized by many clinicians. The diagnosis of severity is necessary for timely management of associated complications, ensuring the reversibility of the syndrome and preventing permanent neurological sequelae. Therefore, considering the diagnosis of PRES in any acute encephalopathy in the presence of hypertension or predisposing factors, such as eclampsia, systemic diseases, or renal insufficiency, is crucial for prompt diagnosis and administration of appropriate therapies. Brain MRI is the best diagnostic tool, especially for ruling out ischemic stroke. PRES lesions appear as diffuse posterior hyperintensities in white matter on T2 and FLAIR sequences, with diffusion hypointensity and a high ADC coefficient. The primary therapeutic action for PRES aims to eliminate potential triggering or exacerbating factors and, based on observed symptoms, implement treatment ranging from antihypertensives and anticonvulsants to mechanical ventilation and neurorehabilitation in severe cases.

## CONSENT

As per international standard or university standard, patients' written consent has been collected and preserved by the author(s).

## ETHICAL APPROVAL

As per international standard or university standard guideline, ethical approval has been collected and preserved by the authors.

## ACKNOWLEDGEMENTS

Chouikh Chakib, Benani Mohamed, and Hanae El Ghiati examined the patient and drafted the manuscript. Touab Rida, Bouayida Ayoub, and Benani Mohamed evaluated the results and provided important clinical insights. Mohamed Rabii Andaloussi, Chakib Chouikh, Khalil Mounir, and Hicham Balkhi contributed to the case report's conception and assisted in manuscript writing. All authors have read and approved the final manuscript.

## COMPETING INTERESTS

Authors have declared that no competing interests exist.

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