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# Posterior reversible encephalopathy syndrome due to unilateral renal artery stenosis: A case report

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## Abstract:

A tricenarian female with a past medical history of status epilepticus secondary to posterior reversible encephalopathy syndrome (PRES) of unknown etiology presented with a 2-week history of double vision, dizziness, elevated blood pressure, and altered mental status. On hospital day 2, she experienced status epilepticus, during which her blood pressure rose to 240/160 from her baseline of around 140/90. The patient was subsequently intubated for airway protection and transferred to the intensive care unit, where she was started on a nicardipine drip. Due to her history of thrombotic microangiopathy, empiric treatment with plasma exchange and prednisone was started but discontinued when ADAMTS13 came back negative. Urine metanephrines also were found to be negative. Computed tomography angiography of the abdomen showed left renal artery stenosis and stent was placed. Remarkably, over the coming days, her blood pressure normalized, and her neurologic symptoms significantly improved. As a result, antihypertensive medications were titrated down, and the patient was finally provided with a cause of her repetitive, life-threatening episodes of PRES.

## Keywords:

Malignant hypertension, magnetic resonance imaging brain, posterior reversible encephalopathy syndrome, renal artery stenosis

## Introduction

Posterior reversible encephalopathy syndrome (PRES) is a complex disorder with many clinical associations, causative factors, and imaging manifestations. PRES is usually seen in patients with comorbidities such as labile hypertension, renal failure, immunosuppression, autoimmune disorders, and eclampsia,<sup>[1]</sup> but its pathophysiology remains a topic of debate.<sup>[2]</sup> PRES is a clinical syndrome characterized by reversible subcortical vasogenic brain edema secondary to endothelial dysfunction, predominantly involving the bilateral parieto-occipital regions.<sup>[1,3]</sup> PRES may present with a wide spectrum of

neurological symptoms including headache, altered mental status, visual changes, seizures, and focal neurological deficits, in addition to nonspecific symptoms such as nausea and vomiting.<sup>[3]</sup> Magnetic resonance imaging (MRI) of the brain must be performed as early as possible in patients with high suspicion of PRES. Based on the few studies from the literature, all of the patients were accurately diagnosed as having PRES, demonstrating the sensitivity of MRI to PRES was 100%.<sup>[4-6]</sup> Most cases of PRES are reversible, but delays in diagnosis and treatment may lead to irreversible lesions. Early diagnosis by MRI scan, and differentiation from other causes such as seizures, meningitis, and psychosis, is extremely important to initiate treatment and prevent further complications. We present the case of a young female with

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repeated seizures and hypertensive crises who was subsequently diagnosed with PRES. Workup revealed the cause of her hypertensive crises and ensuing PRES to be unilateral renal artery stenosis.

### Case Report

A 32-year-old female with a past medical history of hypothyroidism, hypertension, and status epilepticus secondary to PRES of unknown etiology presented with a 2-week history of double vision, dizziness, elevated blood pressure, and altered mental status. Three years ago, the patient was admitted to the hospital with similar complaints before going into life-threatening status epilepticus at that time. MRI of the brain on that visit was done and was positive for diffuse, posterior circulation predominant T2 hyperintense lesions consistent with PRES [Figure 1a-d]. At that time, the etiology of PRES was unclear despite an extensive workup. ADAMTS13 was negative, excluding thrombotic thrombocytopenic purpura (TTP). Likewise, normal urine metanephrines excluded pheochromocytoma. Magnetic resonance angiography of the abdomen was negative for any vascular abnormality but did incidentally note a right adrenal mass. However, biopsy of the mass was negative for pheochromocytoma. As such, on that prior visit, she was discharged with PRES of unknown etiology on antihypertensive medications.

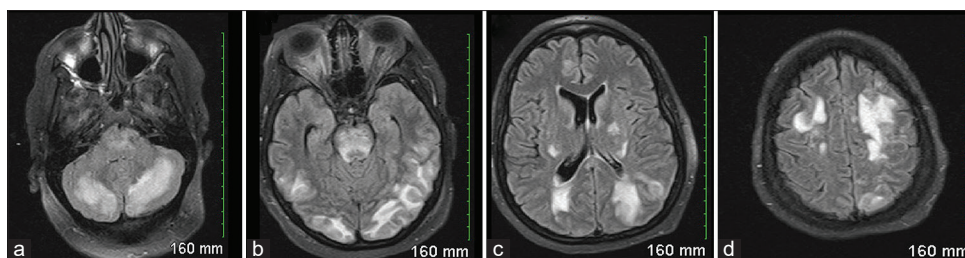
On this admission, the patient was initially hypertensive to 172/90 mmHg. Other vital signs were unremarkable. Physical examination revealed dysmetria on finger-to-nose testing and vertical nystagmus bilaterally. On the following day, her blood pressure rose to 240/160 mmHg, and she sustained a generalized, tonic-clonic seizure. The patient was given lorazepam, levetiracetam, and lacosamide without improvement. Oxygen saturation was found to be in the lower 80s, and the patient was intubated and sedated. She was subsequently transferred to the neuro-intensive care unit (ICU) for further management. Computed tomography (CT) scan of the head showed developing hypointensities in the left temporal and occipital lobes [Figure 2]. Unfortunately, the patient was too unstable for MRI.

Chest X-ray and CT scan of the chest showed extensive consolidations at the bases of the lungs bilaterally, consistent with a severe aspiration-induced respiratory failure [Figure 3a and b]. Transthoracic echocardiogram showed mild left ventricular hypertrophy without any other structural abnormalities and with normal ejection fraction.

Due to schistocytes on peripheral blood smear and a history of multiple episodes of hypertensive emergencies, and exclusion of other causes, both TTP and malignant hypertension were considered in the main differential diagnosis as causing PRES in this patient. Empiric treatment with plasma exchange and intravenous steroids was initiated while awaiting further workup. ADAMTS13 and urine metanephrines eventually came back normal. However, because of persistently elevated blood pressure and elevated creatinine, CT angiography of the abdomen and pelvis was ordered, which showed 80% stenosis of the proximal left renal artery [Figure 4a]. Subsequent catheter angiography confirmed this finding [Figure 4b]. Vascular surgery was consulted, and a stent was placed in the left renal artery [Figure 4c]. Remarkably, over the coming days, her creatinine levels and blood pressure were normalized, and her neurologic symptoms were significantly improved. As a result, antihypertensive medications were titrated down, and the patient was finally provided with a cause of her repetitive, life-threatening episodes of PRES.

### Discussion and Conclusion

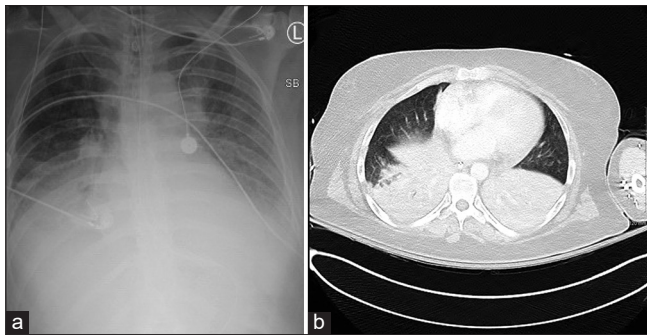
We report a case of PRES in the setting of a severe renovascular hypertension from unilateral renal artery stenosis. Clinically, PRES includes several types of clinical signs and symptoms with many different inciting factors. Renovascular hypertension is more commonly caused by atherosclerotic disease rather than fibromuscular dysplasia.<sup>[7]</sup> Atherosclerosis primarily affects male patients over the age of 45 years. This disorder is particularly common in patients who have atherosclerosis; however, it can also occur as a relatively isolated renal lesion.<sup>[8]</sup> In contrast to atherosclerosis, fibromuscular dysplasia most often affects women younger than the



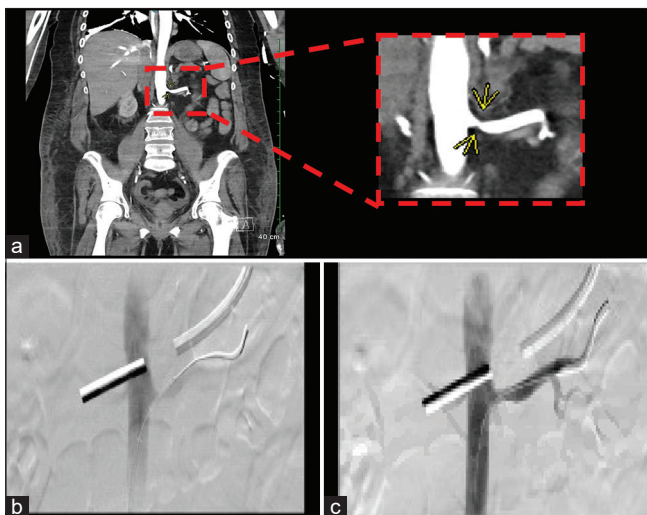
**Figure 1:** MRI of the brain. Multiple, confluent areas of white matter hyperintensity involving the cerebellar hemispheres bilaterally (a), pons (b), occipital lobes (c) and parietal lobes (d)



**Figure 2:** CT-scan of the brain. CT scan of the brain windowed at W: 40 L: 40 showing developing hypointensities in the left temporal and occipital lobes.



**Figure 3:** Chest X-ray. CT scan of the chest. (a) Chest X-ray and (b) CT scan of the chest showing extensive consolidations at the bases of the lungs bilaterally, consistent with a severe aspiration-induced respiratory failure.



**Figure 4:** CT and catheter angiography of the abdomen. (a) CT angiography showing a short segment severe stenosis just beyond the origin of the left renal artery. Estimated degree of stenosis is 80% diameter. Beyond that level, the vessel returns to normal caliber (b) Catheter angiography showing significantly decreased flow through the left renal artery. (c) Catheter angiography after stent placement showing normal flow through left renal artery.

age of 50 years.<sup>[8]</sup> Hemodynamically, significant renal artery stenosis usually results in severe, refractory hypertension and progressive renal insufficiency.<sup>[9]</sup> PRES is characterized by rapid-onset neurological changes in the setting of some acute stressor. The prompt diagnosis and treatment of the underlying cause will often result in the complete resolution of PRES, along with normalization of blood pressure, as observed in this patient. No clinical trials have evaluated the management of PRES, but rapid removal of the trigger appears to hasten recovery and to avoid complications: For example, aggressive blood pressure management, withdrawal of the offending drug, or delivery in eclampsia.<sup>[10]</sup> Antiepileptic drugs should be used to treat seizures, anesthesia, and ventilation should be used in obtunded patients. Corticosteroids should theoretically improve vasogenic edema, but unfortunately, there is no evidence for their use in PRES.<sup>[11]</sup> First-line agents for PRES-related hypertensive emergency include intravenous nicardipine (5–15 mg/h) and labetalol (2–3 mg/min). Nitroglycerine should be avoided in PRES patients, as it can worsen cerebral edema.<sup>[12]</sup> This case illustrates a unique, intense case of PRES secondary to severe, unilateral progressive renovascular hypertension. In the literature, it was described in just a few case reports of unilateral renal artery stenosis as a cause of PRES.<sup>[13–15]</sup> With the increased use of neuroimaging, especially MRI, more and more cases of PRES are being diagnosed. Patients in ICU may be ventilated and critical with a variety of etiologies, and intensivists should be aware about the possibility of PRES. A high index of suspicion in patients with hypertension along with neurological manifestations should prompt a radio imaging to confirm PRES and start immediate treatment.

### Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent form. In the form, the patient has given her consent for her images and other clinical information to be reported in the journal. The patient understand that name and initials will not be published and due efforts will be made to conceal identity, but anonymity cannot be guaranteed.

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### Conflicts of interest

There are no conflicts of interest.

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