



## Posterior Reversible Encephalopathy Syndrome in Patient of Postpartum Preeclampsia: A Case Report

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### Abstract

Posterior Reversible Encephalopathy Syndrome (PRES) is a rare clinical-radiologic condition, not commonly reported in the literature. PRES is an uncommon complication of preeclampsia/eclampsia. The authors present a case of posterior reversible encephalopathy syndrome in patient of postpartum preeclampsia with unilateral papillary edema and pulsatile tinnitus, without seizures. The neuroradiological investigation observed the concomitance and/or development of a cavernous carotid aneurysm. Knowledge about PRES of postpartum patients is indispensable in everyday neurological practice. This condition should always be considered in pregnant with altered consciousness, visual abnormalities, increased blood pressure and headache, once seizures, as in our reported case, may be absent.

**Keywords:** Pregnancy; Brain diseases; Blood pressure; Ischemia; Infarction

### Introduction

Posterior Reversible Encephalopathy Syndrome (PRES) is a well-recognized, clinical and neuroradiological entity first described in 1996 by Hinchey et al. [1]. It is a rare disorder that is caused by endothelial dysfunction resulting in a failure of autoregulation of the posterior circulation in the brain with associated edema and encephalopathy. The immune system is also considered to play a role in the development of this disorder [2]. The PRES is a clinical and radiological entity associating varying degrees, headaches, impaired consciousness, seizures and visual disturbances to neurological and radiological abnormalities of the parietal-occipital white matter [3].

### Case Presentation

A 40-year-old right-handed woman, G2P1, without any pathological past history, with good prenatal care, with a normal prenatal analysis, blood pressure during the follow-up was normal, admitted for elective caesarean section at 38 weeks of amenorrhea. In post-partum, after 24 hrs of the caesarean section. The lady presented a severe abdominal pain, were to take abdominal ultrasound that found liquid in the cavity. She was referred to the surgical center, but in the abdominal inventory, nothing was found abnormal. Returned to the room without pain, only a little confused, being discharged 24 hrs later. After four days at home, she began to feel a headache, in which she noticed that the arterial pressure was elevated (180/110 mm of Hg) and noticed also that she had begun to swell the legs and the face. Simultaneously, she began to get confused, with tinnitus and seeing little with her left eye and lowering of the level of wakefulness. Her obstetrician started methyldopa 250 mg t.i.d. and hydrochlorothiazide 25 mg b.i.d., she had improvement of blood pressure levels and edema of the lower limbs, but not of clinical symptoms, such as tinnitus, left eye vision and mild mental confusion was requested which revealed increased lactic dehydrogenase, liver enzymes, but blood count was normal. The 24-hr urine test revealed proteinuria. After 40 days of the second surgery, as there was no improvement in pulsatile tinnitus on the left and left visual loss, the obstetrician referred to the neurologist. The examination revealed left papilledema, left eyelid ptosis and mild ocular hyperemia. We asked for Magnetic Resonance Imaging (MRI), and an emergency cerebral arteriography, which revealed the presence of cavernous carotid fistula direct to the left, high flow that promoted circulatory "robble" in the cerebral hemisphere (Figure 1,2). She had been referred to the neurosurgery service that realized coaxial system using microcatheter and microguy, catheterizing selectively the fistula and with the release of decorative balloons; it was possible to occlude it without intercorrences. After two days, the symptoms improved.

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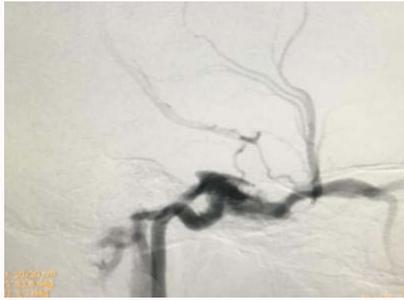


Figure 1: Lateral view of cavernous internal carotid aneurism.



Figure 2: Internal carotid aneurism.

## Discussion

Posterior Reversible Encephalopathy Syndrome (PRES) is defined as a clinical and radiological syndrome characterized by elevation of blood pressure with a headache, nausea and vomiting, confusion, seizures, visual disturbances and radiological abnormalities of the parietal-occipital white matter. Although moderately common, PRES associated with postpartum is widely unknown and is poorly described in the literature [4]. Its early diagnosis is important due to reversibility after prompt reduction of blood pressure. Delayed recognition and treatment can cause a risk of a permanent brain injury [5]. Its pathophysiology remains controversial. Differential diagnosis includes cerebrovascular disorders and severe neurological conditions [3]. In this case report, we report a case of posterior reversible encephalopathy syndrome in a patient of postpartum preeclampsia. We also highlight the importance of rapid blood pressure control and radiological investigations, discuss the pathophysiology and clinical manifestations and identify the differential diagnosis of this rare condition.

## Physiopathology

Posterior Reversible Encephalopathy Syndrome (PRES) is a rare entity often associated with other diseases, including preeclampsia, eclampsia, hypertensive encephalopathy, and renal failure. Treatment with cytotoxic and immunosuppressive drugs, such as cyclosporine, tacrolimus, and interferon alfa [2,3]. Other reported causes are shown in Table 1. PRES during postpartum and pregnancy it is commonly allied to predisposing factors such as preeclampsia and eclampsia. This clinical state is responsible for high indices of perinatal and maternal death, and preeclampsia can be aggravated by hemolysis, elevated liver enzyme levels, low platelet count syndrome (HELLP syndrome) during postpartum period in about one-third of cases [3].

Table 1: Other reported cases of posterior reversible encephalopathy syndrome [1,3].

S. No	Following organ transplantation
1	Thrombotic thrombocytopenic purpura
2	HIV syndrome
3	Shock, sepsis and infection
4	Blood transfusion

This syndrome is clinically featured by seizures, headaches, confusional states, vomiting, visual abnormalities, nausea, consciousness disturbance and rarely focal sensory and motor neurological deficits [4,6]. Usually, seizures are generalized, tonic-clonic and precede all other manifestations, starting with occipital lobe, preceded by an aura [5]. Seizures are the dominant symptom [2], other manifestations include alterations in alertness activity, temporary agitation alternating with lethargy inconstancy, as coma and stupor. There also may be lower motor responses, and disturbances of vision such as visual neglect, hemianopia and cortical blindness (which can evolve to Anton's syndrome, defined as the denial of blindness). Fundus examination and pupillary responses' abnormalities are uncommon, and only a low number of patients relate lower limbs in coordination and weakness [5]. Laboratory results vary according to the different associated conditions [3]. Despite the reversibility of these symptoms, they can persist if hemorrhage and ischemia are present [6].

The pathophysiology of PRES remains uncertain and involves multifactorial etiologies. The most accepted etiology suggests that a rapid elevation in blood pressure causes cerebral arterioles dilatation, ensuring a breakdown in cerebral auto-regulation [3]. This dysfunction of the blood-brain barrier allows the displacement of fluid from the intravascular to interstitial compartments, resulting in vasogenic edema predominantly in the white matter [1,3]. In cases of peripartum PRES syndrome, frequently there are other mechanisms associated, as hyperpermeability of capillaries during pregnancy, modulation of vascular tone with endogenous vasopressor agents, and low levels of placental prostaglandins and cytotoxic factors, causing hypersensitiveness and endothelial dysfunction [1]. PRES occurs during puerperium rather than during pregnancy because fluid accumulation in this period increases the tendency for development of brain edema [5]. Parietal and occipital lobes are the most affected due to the amount of sympathetic innervation [7]. Vertebrobasilar system is less supplied by adrenergic innervation, so the capacity of vasoconstriction is lower in posterior lobe in comparison to anterior cerebral areas [5]. According to these hypotheses, the occipital topographic plan is the most susceptible to infarction and ischemia [4].

## Diagnosis

Diagnosis of PRES in postpartum can be both clinical and radiological [6]. Lesions are best visualized with Magnetic Resonance Imaging (MRI), which is the golden standard for diagnosis. T2 weighted MR images, and especially on Fluid-Attenuated Inversion Recovery (FLAIR) acquisition, show diffuse hyperintensity involving the white matter of parieto-occipital areas, evidencing edema. T1 weighted images show isointensity to hypointensity. These abnormalities are often symmetrical and may involve cerebellum, basal ganglia, frontal lobe, and brainstem in patients with serious clinical conditions. In unusual cases, grey matter can also be affected [3,5]. Computer Tomography (CT) is useful to detect hypodense lesions

of posterior leukoencephalopathy [5]. However, CT abnormalities appear in 50% of the cases of PRES. MRI provides information about cerebral abnormalities earlier than CT and is an indispensable tool in differential diagnosis [3].

The differential diagnosis of PRES during pregnancy and puerperium period is challenging and difficult to distinguish [3]. It includes eclampsia, ischemic and hemorrhagic stroke, idiopathic epilepsy, infections, vasculitis or other metabolic disorders [3-5]. The distinction between cases of ischemic stroke and PRES is of paramount importance, due to the treatment of PRES recommends an instantaneous reduction of blood pressure levels [5]. Another fundamental differential diagnosis of PRES is progressive multifocal (PML) syndrome. Their imaging picture is similar, but seizures are infrequent. PML symptoms include gait difficulty, altered mental status, speech, and visual disturbances, which tend to deteriorate progressively and lead the patient to death within six months. These two pathological conditions may be distinguished on clinical grounds and neuroimaging [2].

## Treatment

If promptly diagnosed and properly treated, PRES in postpartum is considered a reversible pathology [4]. The primary treatment consists in control of elevated blood pressure, ischemia, infarct, and vasospasm [3]. After resolution, patients who presented seizures do not develop chronic epilepsy [5]. Magnesium sulfate (MgSO<sub>4</sub>) improves both seizures and hypertension, being the first-line therapy. Nonetheless, patients must be treated concomitantly with antihypertensive calcium antagonists [3].

Clinical outcomes related to this postpartum syndrome are generally favorable after immediate blood pressure control [4]. Symptoms usually disappear in 3 days to 8 days and imaging abnormalities return to normal in a few weeks. However, PRES can culminate in a wide variety of clinical disorders when not appropriately treated, progressing to ischemia and infarction [3,5]. About 5% to 12% of cases may evolve to irreversible brain injury with severe neurologic deficit and death [3].

Our patient presented confusion, headaches, decreased left vision, increased levels of liver enzymes, pulsatile tinnitus and absence of seizures. Tinnitus can be a consequence of an abnormal vascular structure and generated by the increase of turbulent blood flow, being present in high jugular bulb, intracranial hypertension, arteriovenous malformations and atherosclerosis of the carotid artery. When characterized by pulsatile tinnitus, may indicate a neurological pathology [6]. Because of the pulsatile nature of tinnitus,

it seems more likely that in the case report this symptom was caused by the increase of intracranial pressure levels frequently present in PRES syndrome conditions.

In the case report, our patient was administrated immediately methyldopa 250mg t.i.d. and hydrochlorothiazide 25 mg b.i.d., which improved blood pressure, levels, but not other clinical symptoms. Her MRI findings showed a cavernous carotid aneurysm [8].

## Conclusion

In conclusion, knowledge about posterior reversible encephalopathy syndrome of postpartum patients is indispensable in everyday neurological practice. This clinical condition should be always considered in pregnant with symptoms such as altered consciousness, visual abnormalities, increased blood pressure and headaches, once seizures, as in our reported case, may be absent. Diagnosis involves magnetic resonance imaging. PRES can be reversible if blood pressure levels are immediately treated. Delayed treatment usually may lead to ischemia and infarction.

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