Reversible hemianopsia in postpartum due to posterior reversible encephalopathy syndrome in pregnant with late eclampsia

Caso de hemianopsia reversível no pós-parto decorrente de síndrome da encefalopatia reversível posterior (PRES) em gestante com eclâmpsia tardia

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ABSTRACT

Objectives: To describe a case of Posterior Reversible Encephalopathy Syndrome diagnosed in pregnant women with late-eclampsia, as well as its clinical management. Case description: A 34 years old patient in her third pregnancy had started with high blood pressure levels during labor; after eleven days postpartum, she presented a decreased right visual acuity; subsequently one episode of seizure followed by partial loss of vision in the right eye. After conducting tests and ruled out stroke, the patient was diagnosed as Posterior Reversible Encephalopathy Syndrome (PRES). Established the clinical management of seizures and hypertensive crisis, there was complete remission of symptoms and reversal of the initial clinical picture. Conclusion: Once properly diagnosed and treated, the Posterior Reversible Encephalopathy Syndrome can present satisfactory progress, especially when associated with an acutely triggered factor, as eclampsia.

Keywords: brain edema; disease models, animal; eclampsia; hemianopsia; pre-eclampsia; hypertension; pregnancy complications.

RESUMO

Objetivos: Descrever um caso de Síndrome da Encefalopatia Reversível Posterior em gestante diagnosticada com eclâmpsia tardia, bem como seu manejo clínico. Descrição do caso: Paciente feminina, 34 anos, em sua terceira gestação, iniciou com aumento dos níveis tensionais durante o trabalho de parto e, após onze dias de puerpério, apresentou quadro de diminuição da acuidade visual à direita, seguido de crise convulsiva e subsequente perda parcial da visão do olho direito. Após a realização de exames de imagem e descartada a possibilidade de acidente vascular encefálico, a paciente foi diagnosticada com Síndrome da Encefalopatia Reversível Posterior (PRES). Instituído o manejo clínico das crises convulsivas e hipertensivas, houve remissão completa dos sintomas e reversão do quadro clínico inicial. Conclusão: Uma vez adequadamente diagnosticada e tratada, a Síndrome da Encefalopatia Reversível Posterior pode apresentar evolução satisfatória, especialmente quando associada a um fator desencadeado agutamente, como a eclâmpsia.

Palavras-chave: complicações na gravidez; eclâmpsia; edema encefálico; hemianopsia; hipertensão; modelos animais de doenças; pré-eclâmpsia.

INTRODUCTION

Posterior reversible encephalopathy syndrome (PRES), a condition of unknown pathophysiology, is diagnosed with the aid of clinical examination and imaging tests. Two theories have been postulated: the vasogenic theory and the cytotoxic theory.1 The first relates to vasogenic edema secondary to increases in blood pressure that compromise cerebral autoregulation and cause vasodilation and increased patency of the blood-brain barrier.2 The second explains the onset of cytotoxic edema by a sudden sharp increase in blood pressure leading to cerebral vasoconstriction followed by endothelial damage, hypoxia, and vasospasm.3

The following have been listed among the clinical findings of PRES: headache, altered consciousness, seizures, and visual disturbances. White matter vasogenic edema affecting mainly the occipital and parietal lobes has been observed in the computed tomography (CT) and nuclear magnetic resonance (NMR) scans of patients with PRES and used to diagnose the syndrome. Factors such as acute increases in blood pressure, autoimmune disease, loss of renal function, immunosuppressant therapy, and chemotherapy have been considered as potential inducers of PRES. Examples of
such factors include gestational hypertension, systemic lupus erythematosus, and immunosuppressant therapy used in organ transplantation.1,4

Pre-eclampsia (PE) significantly increases maternal and fetal morbidity and mortality. As a general rule, patients with PE present with hypertension at gestational ages ≥20 weeks accompanied by proteinuria.6 PE is believed to start with the inadequate implantation of the conceptus onto the uterine wall and the incomplete invasion of the trophoblast due to impaired spiral artery remodeling. These events lead to reduced uteroplacental blood flow followed by ischemia and endothelial dysfunction; the ensuing systemic involvement produces increased blood pressure levels.1

Eclampsia sets in when a patient with PE has seizures unrelated to neurologic impairment or other specific causes. Bouts usually occur within up to 48 hours of delivery, but there have been reports of patients suffering from the disease before delivery or during labor.1,7 In the past, seizures were described as local or generalized cerebral vasospasm. Today, increased perfusion pressure - also referred to as hypertensive encephalopathy - is deemed to be a primary cause of the disease for its effects on cerebral autoregulation and subsequent vasogenic edema.1,2

A possible association between PRES and pre-eclampsia/eclampsia has been suggested when severe hypertension causes cerebral vasospasm, resulting in edema and cytotoxic edema. This association has been pointed out when acute hypertension in patients with eclampsia impairs autoregulation and, consequently, causes the passive dilation of cerebral arterioles and the onset of cerebral vasogenic edema, also a landmark of PRES.

This report aimed to present a case of fully reversed PRES in a patient diagnosed with eclampsia 11 days after delivery.

CASE REPORT

A healthy 34-year-old housewife, pregnant for the third time, born in Alegrete, RS, with residence in Porto Alegre, RS, was seen at the emergency unit of the Hospital São Lucas at PUCRS (HSL/PUCRS). She was pregnant for 36 weeks and reported lower abdominal pain at admission. Her blood pressure was 130/86 mmHg and her baby had a heart rate of 136 beats per minute. She had an uneventful C-section on the day she was discharged from the emergency unit; the patient had previously undergone two C-sections and was diagnosed with oligohydramnios.

A baby girl was born weighing 2,720 grams, with 1st and 5th minute Apgar scores of 8 and 9, respectively. The patient was well when she was discharged 72 hours after delivery. A week after the birth of her baby, she sought care at the emergency obstetrics unit complaining of lower limb edema and a holocranial headache against which oral painkillers were ineffective. The patient was suspected for postpartum pre-eclampsia and was promptly hospitalized. Her urine protein to creatinine ratio was 1.76 g/g on admission; serum creatinine: 0.8 mg/dL; prothrombin time: 104%; INR: 0.97; activated partial thromboplastin time: 23.25 seconds; fibrinogen: 257 g/L; platelet count: 318,000 platelets/mm³; uric acid: 5.5 mg/dL; serum glutamic oxaloacetic transaminase: 32 UK; glutamic-pyruvic transaminase: 42 UK.

Eight days after admission the patient’s blood pressure peaked at 180/100 mmHg, and improved with the prescription of a single dose of an oral ACE inhibitor (captopril, 25 mg). Three days later, her blood pressure shot up to 220/110 mmHg (measured in both arms). The patient was given oral captopril again. One hour later her blood pressure was at 200/100 mmHg, and she was prescribed oral hydralazine 25 mg. Her blood pressure dropped to 160/100 mmHg. After another hour, the patient complained of a visual disturbance in her right eye.

The attending nephrology team then decided for the following therapy: enalapril 10 mg, two pills administered orally twice a day; amlodipine 5 mg, two pills administered orally twice a day; and clonidine, when needed. Minutes after the onset of the new symptom, the patient had a seizure and was promptly seen by the medical team. She was given 5 ml of IV hydralazine and started on an infusion of magnesium sulfate, which led to the cessation of symptoms within 15 minutes. Three hours later the patient complained of blurry vision on her right eye. A head NMR scan was ordered as she was suspected for stroke. The symptoms improved the next day. Her systemic blood pressure was gradually corrected, but she was kept on close monitoring to avoid sudden decreases in her blood pressure levels.

The patient still had hemianopia and headaches for another day, although her blood pressure was stabilized (126/76 mmHg). The NMR scans revealed signs of involvement in the right occipital lobe secondary to vasogenic edema (Figure 1), a finding consistent with PRES. On that day, the patient had no complaints, was not affected by hemianopia, and her blood pressure was under control at 120/70 mmHg. A second series of
NMR scans showed considerable improvements in her radiologic findings (Figure 2).

**Figure 1.** Magnetic nuclear resonance scan of a patient suspected for PRES: signs of involvement in the right occipital lobe secondary to vasogenic edema.

**Figure 2.** Magnetic nuclear resonance scan taken five days after the scan shown in Figure 1 showing considerable improvement in radiologic findings.

The patient was discharged with a stable blood pressure level. She was advised to have her blood pressure checked daily, and was prescribed enalapril 10 mg 1 pill twice a day, amlodipine 5 mg 1 pill twice a day. The patient was scheduled to return for a visit with the nephrology and obstetrics teams. On the following month her enalapril prescription was wound down to 5 mg 1 pill twice a day, and on the third month she was taken off her antihypertensive medication.

**Discussion**

PRES affects individuals of all ages. Uremic encephalopathy, nephrotoxicity caused by calcineurin inhibitors, pre-eclampsia, and eclampsia rank among the predisposing factors for the syndrome.8-10

PRES symptoms include headache, visual disturbances, altered consciousness, and seizures. The name of the syndrome prompts one to consider involvement of the posterior lobe, as injuries are present in this region, located in the vertebrobasilar territory, due to the absence of baroreceptors. However, in some cases the anterior lobe is also involved. These are usually more severe cases with concurrent involvement of the posterior circulation.11 NMR and CT scans from published case reports have shown subcortical edema involving white and grey matter in the parietal and occipital lobes.12

Patients may be suspected for PRES based on their clinical history. However, before diagnostic NMR scans are ordered, many other conditions are considered due to the nonspecific nature of clinical findings. CT scans may show hypodense lesions in the posterior brain, but NMR is the gold standard imaging technique. The advantage in using NMR imaging lies in the possibility of differentiating between vasogenic edema - a reversible condition - and cytotoxic edema - an irreversible condition.13

Eclampsia is known to occur in up to 48 hours from delivery in 30%-35% of the cases. Fifteen to 25% of the cases may occur even after 48 hours - late onset postpartum eclampsia - with or without the characteristic signs of eclampsia. In patients with eclampsia, PRES is more likely to manifest after delivery, as patients accumulate more fluid and are more prone to having cerebral edema.13,14 In this case report, the patient had eclampsia 11 days after delivery.

In the case described herein, PRES was associated with postpartum eclampsia. The patient improved satisfactorily as her seizures and hypertension were properly managed. The prognosis for individuals with PRES is good. Once hypertension is under control and cerebral edema subsides, most patients tend to improve significantly.15

The case described in this report is an example of a transient increase in blood pressure leading to edema
and PRES, a condition still without a satisfactory animal model. Our group has worked to develop an animal model for PRES, with promising results.9 Our studies have used a reduced uterine perfusion pressure (RUPP) rat model. Abitbol16 first described the rat model in 1982; female rats are currently considered the best animal model for this type of experiment. The procedure consists of clipping the aorta (below the renal arteries) and the uterine arteries of the rats to reduce uteroplacental perfusion.

The decrease in uteroplacental perfusion causes systemic blood pressure to increase, for reasons yet unknown. This model was used by Granger et al.17 in a study on gestational hypertension. The acute blood pressure increases produced by this procedure made it ideal to investigate the repercussions on the vascular bed of the central nervous system and changes in the blood-brain barrier.9

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