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Case Report

Posterior Reversible Encephalopathy Syndrome (PRES): Case Report in a Woman with Pregnancy-Induced Hypertension

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ABSTRACT

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Posterior reversible encephalopathy syndrome is a clinical-radiological syndrome characterized by headache, visual disturbances, seizures, altered mental status and white matter vasogenic edema affecting the posterior occipital and parietal lobes of the brain predominantly. Its pathogenesis is not fully understood and two opposite theories have been proposed, both based on the central role of hypertension. Here we present a case of a 28-year-old primigravida, at 37 weeks, with an unremarkable history presented to the Emergency Department showing convulsions and cognitive impairment. Magnetic Resonance imaging showed cortical-subcortical hyperintense areas on T2-weighted and fluid attenuated inversion recovery sequences bilaterally involving parietal and occipital lobes.

INTRODUCTION

The Posterior Reversible Encephalopathy Syndrome (PRES) is a clinical-radiological syndrome characterized by headache, altered consciousness, seizures and visual disturbances [1,2]. It was first described in 1996 by Hinchey [3]. This syndrome has been known by various names previously (reversible occipital-parietal encephalopathy, reversible posterior leukoencephalopathy syndrome and reversible posterior cerebral edema syndrom). PRES is nowadays the widely accepted term. It is often associated with acute hypertension. Currently this syndrome is increasingly recognized because of improvement and availability of brain imaging. The major clinical conditions associated with PRES are:

- Hypertensive encephalopathy
- Preclampsia/eclampsia/HELLP syndrome
- Immunosuppressive/cytotoxic drugs (e.g., cyclosporin, antineoplastic drugs, interferon-α, antiretroviral therapy)
- Acute or chronic renal diseases
- Thrombotic thrombocytopenic purpura/haemolyticuraemic syndrome
- High-dose steroid therapy
- Liver failure
- Endocrine dysfunctions (e.g., primary aldosteronism, pheochromocitoma)
- Hypercalcemia/hyperparathyroidism
- Bone marrow transplantation
- Massive blood transfusion/erythropoietin therapy
- Porphyria



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There is wide variation in the severity of clinical symptoms. Visual disturbances can vary from blurred vision, homonymous hemianopsia to cortical blindness. Altered consciousness may vary from mild confusion or agitation to coma. Other symptoms include nausea, vomiting and brainstem deficits. Seizures and status epilepticus are common, while non-convulsive status should be cautiously observed in patients with prolonged altered consciousness, which may be mistaken commonly for postictal confusion. Signs include stereotypic movements like staring, head turning or eye blinking. Postictal confusion usually lasts for hours, but PRES and non-convulsive status can last for many days and can be mistaken for drug intoxication, psychosis or psychogenic states. It is caused by a wide variety of causes ultimately leading to a vasogenic cerebral oedema of occipital and parietal lobes of the brain. However, as the name suggests, it is typically reversible within one or two weeks once the underlying cause is identified and early treated [4-6].

CASE REPORT

Here we present the case of a 28-years-old woman, primigravida, at 37 weeks, with an unremarkable medical history presented to our Emergency Department (ED) showing tonic-clonic convulsions and cognitive impairment. Her relatives referred symptoms of visual disturbances (blurring) and headache started one day before. She did not have a past history of hypertension, vision abnormalities or seizures, nor other neurological diseases.

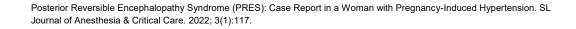
Physical examination in ED revealed that she was unconscious, with left gaze deviation, Glasgow Come Scale (GCS) 6 (Eye 1, Verbal 1, Movement 4). Pupils were bilaterally isochoric, isocyclic and normally reactive to the light stimulation. Blood pressure was 150/80 mmHg with a heart rate of 80 bpm. Her respiratory rate was 20 breaths per minute with a room air saturation of 97%. Body temperature was 37°C. Respiratory and cardiovascular physical examination was within normal limits. Blood count, renal and liver function tests, clotting parameters, glycemia and electrocardiogram were normal. Her arterial blood gas analysis showed: pH 7.07, PO₂ 89,5 mmHg, pCO₂ 45 mmHg, HCO₃ 13,3 mmol/l.

A provisional diagnosis of eclampsia was made with the gynecologist and an intravenous dose of magnesium sulphate 4g was administered in 10 minutes. The patient was intubated with a rapid sequence induction and immediately shifted for an emergency caesarean section. A normal-weight baby was extracted (APGAR 8 after 1 minute) and transferred in neonatal intensive care unit. Postoperatively, after a head Computerized Tomography (CT) scan, the woman was shifted to the intensive care unit to continue analgosedation, mechanical ventilation and invasive monitoring. Her pharmacological treatment included magnesium sulphate infusion (1g/h), clonidine, α -methyldopa and mannitol (1 g/kg/die).

Head CT scan revealed two slightly hypodense areas in the parietal lobes. A brain Magnetic Resonance (MR) imaging was performed after few hours. It showed cortico-subcortical hyperintense areas on T2-weighted and Fluid Attenuated Inversion Recovery (FLAIR) sequences bilaterally involving parietal and occipital lobes (Figures 1,2). It was also associated to an important diffusion restriction in Diffusion-Weighted Imaging (DWI). MR angiogram and MR venogram were normal. These findings were in line with the diagnosis of PRES. Since the patient had regained a normal sensorium and good gas exchange, she was extubated on second postoperative day. She had no visual disturbances, no headache, no seizures. After 24hours of spontaneous breathing, considering the clinical stability, the patient was transferred to the obstetrician department. The patient continued to get better and was then discharged home symptom-free on the seventh day of hospitalization. In a follow-up MR imaging performed ten days later, the areas of hyperintensity in parietal and occipital lobes bilaterally disappeared.



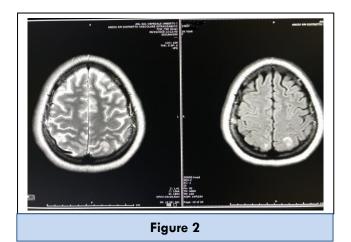
Figure 1





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DISCUSSION

The term PRES has been used based on the similarity in the appearance on imaging, the common location of the parietaloccipital lobe or 'posterior' location of the lesions. The exact pathophysiological mechanism of PRES is still unclear. Three hypotheses have been proposed till now, which include (i) cerebral vasoconstriction causing subsequent infarcts in the brain, (ii) failure of cerebral autoregulation with vasogenic edema, and (iii) endothelial damage with blood-brain barrier disruption further leading to fluid and protein transudation in the brain. The reversible nature of PRES has been challenged recently based on new reports of permanent neurological impairment and mortality reaching 15% [7]. No clinical studies are available till now regarding patients with PRES needing life-sustaining treatments. The improved knowledge and research about factors influencing the outcome of PRES will result in better early management, less morbidity and mortality. According to studies, delayed diagnosis and treatment may lead to mortality or irreversible neurological deficit. In hypertension associated or drug-induced PRES, the effective therapy includes withdrawal of offending agent, immediate control of blood pressure, anti-convulsive therapy and temporary renal replacement therapy (hemodialysis/peritoneal dialysis), if required. Corticosteroids may improve vasogenic edema, but there is no solid evidence for usage in PRES.

A high index of suspicion and prompt treatment can reduce morbidity, mortality and pave the path for early recovery. Clinical improvement always follows the treatment of elevated blood pressure. The therapy with magnesium should be started as soon as eclampsia or PRES in pregnancy is suspected, as it treats both seizures and hypertension. Differential diagnosis of PRES includes stroke, meningoencephalitis, demyelinating lesions of the brain and cerebral venous thrombosis. Early imaging is crucial to make this distinction. MR imaging is the imaging tool of first choice. PRES appears as high signal intensity predominantly in the posterior regions of the brain. DWI helps to distinguish the vasogenico edema from cytotoxic oedema, which is characteristic of this disease.

CONCLUSION

Even if MR imaging is helpful to detect this syndrome, only the reversibility of this condition and the improvement of the clinical picture allow a definitive diagnosis of PRES. As the name suggests, reversibility of these symptoms is one of the hallmarks of the disease. Early treatment usually results in complete reversal of the deficits over few days to several weeks. However, some patients with severe manifestations of PRES, such as coma, can develop a permanent neurological impairment. In conclusion, the case described here a pregnant woman presented to the ED with high blood pressure, seizures and visual disturbances. Initial diagnosis of eclampsia was made and managed promptly with magnesium sulphate and caesarean section. However, postoperative CT and overall brain MR imaging helped us to diagnose. This case report emphasizes the need for early diagnosis and prompt treatment of PRES to avert short- and long-term neurological sequelae and ensure a full recovery. It is very important that intensivists and all physicians be well aware of this syndrome since prompt recognition and precocious treatment have prognostic implications. There are no large studies in literature about PRES and its etiopathogenesis is still uncertain despite several hypotheses. This is just a case report with all its intrinsic limitations, but it could be a further stimulus for more and larger studies about PRES to better understand this insidious morbid condition.

DISCLOSURES

Authors have no conflict of interest to declare, nor funding. An informed written consent was obtained before publication. The authors declare that this submission is in accordance with the principles laid down by the Responsible Research Publication Position Statements as developed at the 2nd World Conference on Research Integrity in Singapore, 2010.

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