Posterior Reversible Encephalopathy Syndrome (PRES): Series of cases

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Abstract

The hypertensive pregnancy disorders, including preeclampsia, complicate more than 10% of all pregnancies and are the main cause of maternal and perinatal morbidity and mortality around the world. A rare syndrome associated with hypertension, preeclampsia and eclampsia is the syndrome of reversible posterior encephalopathy (PRES) also known as reversible posterior leukoencephalopathy syndrome. In these series of cases we reported patients with preeclampsia or eclampsia complicated with PRES at Porto Alegre Clinical Hospital during three years (2012 to 2015). Our purpose is to discuss frequent signs and symptoms and encourage early diagnosis, since its potential reversibility once the right treatment began at time.

Keywords: Leukoencephalopathy; Eclampsia; Hospital; Syndrome

Introduction

Posterior reversible encephalopathy syndrome (PRES) is a clinic-radiological entity, described by Hinchey in 1996 based on fifteen cases, clinically characterized by signs and symptoms of a neurotoxic syndrome such as headache, vomiting, nausea, cognitive impairment (from confusion, somnolence, and lethargy to encephalopathy or coma), seizures, visual abnormalities, including cortical blindness and focal neurological sign, associated or not with blood pressure elevation.

PRES can develop in association with many clinical conditions, as hypertension, pre eclampsia, eclampsia, kidney disease, sepsis, solid organ and bone marrow transplantation, autoimmune disorders, thrombotic thrombocytopenic purpura and exposition to immunosuppressive and cytotoxic drugs. Preeclampsia is one of the most common situation associated with PRES. However, regardless the underlying cause, the main abnormality is cerebral vasogenic edema [1].

The physiopathology of PRES is still unclear, controversial and unproved. Increased systemic blood pressure exceeding cerebral vasculature autoregulatory device, resulting in fluid extravasation and edema is one theory often cited as the underlying mechanism. Parietal and occipital lobes predominate as the most common affected areas of the brain. However, researches demonstrate that it isn’t limited to the posterior occipital white matter and can occur simultaneously in the cortex, frontal lobes, basal ganglia, and brainstem. The differential diagnosis includes venous sinus thrombosis and progressive multifocal leukodystrophy [2].

The selective involvement of the posterior cerebral areas could reflect a watershed zone due to a less degree of adrenergic innervations and autoregulation mechanisms in this area. However, more recent neuroradiology research demonstrates that PRES is rarely limited to the posterior occipital white matter and can occur in the cortex, frontal lobes, basal ganglia, and brainstem, with a trend suggesting basal ganglia involvement in eclamptic patients. Other theories as to the pathophysiology of PRES have been proposed, suggesting endothelial dysfunction and cerebral vasospasms [3-4].

The diagnosis is made on clinical grounds and neuroimaging. Recognition of PRES has evolved with increasing availability of magnetic resonance imaging (MRI), which is the best exam to evaluate it nowadays. Clinical findings of PRES include blood pressure (BP) elevation, headache, decreased alertness, altered mental status, seizures, and visual loss. Once the cause is removed, cerebral vasogenic edema is commonly reversible equally among all patterns of affected areas. Focal zones of restricted diffusion or hemorrhage may remained as residual areas of encephalomalacia. Nevertheless, patients with severe manifestations of PRES, such as coma and/or "status epilepticus", may require admission to the intensive care unit. Moreover, permanent neurological impairment or death occurs in a minority of patients.

Series of Cases

Case 1

Female, 17 years, first pregnancy, 33 weeks. Patient was hospitalized on January 1, 2015, with hypertension and history of seizure at home. At admission, the protein/creatinine (P/C) ratio was 0,46 and therapy with magnesium sulfate was initiated. Cesarean delivery was indicated after patient stabilization, on the same day. At postpartum, the patient remained with hypertension, headache and a new seizure episode was reported four days after delivery. A new PC ratio was 9,9 and a magnetic resonance image (MRI) was requested, due to suspected of posterior reversible encephalopathy syndrome. The MRI demonstrated extensive vasogenic edema in the frontal, parietal and occipital lobes, bilaterally, consistent with PRES. Adjustment in the antihypertensive regimen was performed and better blood pressure levels was obtained. Patient was discharged on January 15, 2015.
Case 2

Female, 15 years, 4 days postpartum started with headache accompanied by vomiting. Prenatal and childbirth without complications. Managed as post-raquianalgesia headache. Six days after delivery, started with bilateral amaurosis and tonic-clonic seizure. P/C ratio was 0.4. CT showed hypodense areas partially defined in tempo-occipital regions bilaterally with focal loss of differentiation between gray and white matter. MRI suggestive of PRES. Initiated magnesium sulfate infusion and blood pressure control was obtained. Patient has complete visual recovery and partial lacunar amnesia, went home using antihypertensive [5].

Case 3

Female, 25 years, 34 weeks of pregnancy, started with reduction of fetal movements and visual blurring for 3 days. Presented generalized edema and blood pressure of 190 / 120mmHg. P/C ratio was 6.43 and uric acid 8.2mg/dL. Performed emergency cesarean for fetal condition non-reassuring. Received magnesium sulfate for 24 hours. On the 3rd postoperative day, presented hypertensive crises refractory to medication, behavioral change and visual hallucinations. Handled with sodium nitroprusside. She remained in intensive care due to hypertension difficult to control. MRI showed a hypertensive lesion in the left parietal lobe, with restrict diffusion of water molecules and discreet breaking blood-brain barrier, suggesting PRES. Good progress with gradual withdrawal of antihypertensives.

Case 4

Female,19 years, first pregnancy, 35 weeks, reported occipital headache started this afternoon. Wake up with bilateral amaurosis. She denied other premonitory symptoms. No obstetric complaints. P/C ratio was 3.34. Two hours after entering in the Hospital the patient remained with amaurosis, no fetal movement and showed an increase in blood pressure levels (150x90 mmHg). Performed CT scan with no abnormalities. Performed IMR that showed increased pituitary volume, compatible with pregnancy. Grooves brain injury, ventricles and cisterns of the base within the normal. Void of the presence of normal flow dural venous sinuses and caliber arteries of the brain. Mucosal thickening lobed in the right maxillary sinus. Orbital contents and rocks within the usual. notes up in and flair hyperintense t2 no right cerebellar hemisphere, hyperintense and hypointense adc on the map in broadcast, featuring facilitation of infusion, find that can suggest a suspected clinic back reversible encephalopathy. Initiated infusion of magnesium sulfate. Performed cesarean delivery. Patient recovered the vision two hours post cesarean operation [6].

Discussion

Since 1996, when Hinchey described the syndrome, many papers were published; however the precise physiopathology mechanism remains unclear. Few papers analyzed if the syndrome presented in pregnant patients was the same described in the other patients with PRES.

Roth studied 21 patients and described few differences between pregnant patients and non pregnant patients in a paper published on 2009. Headache was more reported by pregnant patients (87.5%) than non-pregnant patients (30.8%) and visual disturbance was also more reported in pregnant patients than in non-pregnant patients (75% vs 46.2%). The blood pressures of both groups were very similar².

Marrone et al., find a difference in creatinine and demonstrated that pregnant patients present breakdown of cerebral autoregulation with lower mean arterial blood pressure³.

Liman et al. described that preeclampsia-eclampsia patients had significantly less severe edema, less cytotoxic edema, hemorrhage and contrast enhancement, while more frequent complete resolution of edema and less frequent residual structural lesions were seen on follow-up imaging.

Conclusion

The syndrome of reversible posterior encephalopathy (PRES) remains a rare, but important complication of prevalent obstetrics pathologies as hypertension, eclampsia and pre-eclampsia. The terminology once created by Hinchey in 1996 no longer embrace all the different clinical-radiological manifestations associated with the syndrome. Through this series of cases and articles review, we point the diverse patterns of this disease in order to demonstrate its relevance and favorable outcome when premature diagnose.

References