Posterior Reversible Encephalopathy Syndrome in Pregnancy without Pre-Existing Preeclampsia: Cases of Two Primiparae

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Abstract

Reversible posterior encephalopathy syndrome (PRES) is a neurological condition that occurs in pregnant and immediate postpartum women, usually associated to eclampsia or preeclampsia. We report two cases of patients who developed PRES without any indicators for preeclampsia-eclampsia in her pregnancy, and describe clinical and instrumental features.

Keywords: Posterior reversible encephalopathy syndrome (PRES)

Introduction

Pregnancy is characterized by high energy requirements, and several changes take place in terms of hormones, hemodynamics and blood vessel structure in order to meet the new metabolic needs. However, the inadequate action of complex compensatory mechanisms may lead to a multisystem disorder, clinically characterized by hypertension, edema and proteinuria. This condition is known as preeclampsia. During the peripartum period, this condition leads to an increased susceptibility to neurovascular disorders. Several pathophysiological mechanisms are included: structural changes in the vessel walls, dysfunctions in the thrombotic system and altered endothelial permeability. Posterior reversible encephalopathy syndrome (PRES) is generally a reversible neurological condition, usually associated to preeclampsia. We report the cases of two primiparae with PRES-related clinical and instrumental features in the peripartum period, in the absence of further clinical criteria of eclampsia-preeclampsia. These cases show that PRES is related to the systemic pathophysiological mechanisms of pregnancy, which are not necessarily related to preeclampsia.

Case 1

A patient aged 16 was at the 39th week of physiological pregnancy, with no history of neurological disorders; she had a sudden headache associated with visual disorders (scotomas and dimming of sight) and vomit. Blood pressure was within normal limits (AP 130/70mmHg). A few minutes after the above symptoms, she had generalized tonic-clonic seizures. The patient was taken to the delivery room and an emergency C-section was performed with general anesthesia. The patient was then moved to the ICU and the following treatment was administered: propofol 0.4mg/kg/h, levetiracetam 500mg/TID. The physical examination did not show significant neurological impairment. Brain MR examination showed multiple bilateral cortical-subcortical signal changes in the parietal-occipital and frontal-polar areas, characterized by hyper-intensity in FLAIR T2 (Figure 1a & 1b). Post-contrast T1 images show leptomeningeal impregnation and cortical-subcortical impregnation areas on the above areas (Figure 2c & 2d). During hospitalization, CSF and hematocrit level examinations were performed to exclude brain infections. Urine examination was within normal limits.
limits and blood pressure levels were always stable. She was extubated after 1 day, with no neurological deficits. Follow up MR examination was performed with and without contrast agent after 12 days from the acute event; it did not display areas of altered signal, described in the previous MR examination (Figure 1e & 1f); after IV administration of paramagnetic contrast agent, no impregnation areas were observed.

Figure 1: Multiple areas of cortical-subcortical on FLAIR T2 weighted images in the occipital lobes (a), frontal and parietal lobes (b) which show contrast enhancement (c,d). Follow up at 12 days, FLAIR T2 weighted axial images show no areas of altered signal intensity (e, f).

Case 2

A patient aged 30 was at the 38th week of physiological twin pregnancy; she underwent C-section with spinal anesthesia. After 18 hours from surgery, patient experienced a sudden headache, followed by generalized tonic-clonic seizures and blood pressure increase (AP 150/80mmHg). She was immediately sedated with 5mg midazolam. Seizures ended, and she was moved to the ICU for vital parameters monitoring, where she was treated with valproic acid in continuous infusion at 4mg/kg/h. Orotracheal intubation was not required. Medical history, laboratory and hormonal examination did not detect pathological conditions that could explain the epileptic seizures. Brain MR examination showed two areas of altered signal intensity in the cortical-subcortical cerebellar hemispheres in FLAIR T2 images, and a further smaller area with the same features was observed in the subcortical white matter of the right frontal lobes (Figure 2a & 2b). These areas did not show impregnation after IV administration of contrast agent. Antiepileptic treatment was interrupted the day after because of good clinical conditions and normal blood pressure levels; there were no other seizures during hospitalization. The follow-up MR examination at 36 days showed no areas of altered signal intensity that were previously detected (Figure 2c & 2d).

Figure 2: FLAIR T2 weighted axial images showing areas of altered signal intensity at the cerebellar hemispheres (a) and the right frontal subcortical white matter (arrow, b). Follow-up MR examination at 36 day, FLAIR T2 weighted images show no areas of altered signal intensity (c,d).

Discussion

PRES is related to brain vasogenic edema within the framework of systemic toxicity. This syndrome is characterized by neuroradiological changes associated with several clinical features. The theory that PRES is related to changes in vessel self-regulation caused by a severe blood pressure increase in pregnancy shows that hypertensive preeclampsia is the pathophysiological mechanism underlying PRES. However, our cases of normotensive patients with preeclampsia show that PRES can be triggered by pathophysiological mechanisms that can overlap preeclampsia, but are not necessarily its consequence [1].

During pregnancy, the cardiovascular system adapts to the increased metabolic requirements through complex hormonal and hemodynamic changes. High estroprogestin levels lead to an increase of renin activity, which leads to a progressive increase of plasmatic volume. Despite the fact that this state is associated to an increase of the hematic corpuscular volume, it leads to hemodilution-related anemia. The heart activity compensates for fetal metabolic requirements and hypervolemia with an increase of output and frequency, ranging from 30% to 50%. Moreover, due to prostacyclin increase and circle redistribution within low resistance and high flow districts, a fall in peripheral vascular resistances is observed. This condition implies that the initial reduction of blood pressure levels observed from week 20 to 32 is followed by a recovery, or a slight increase of pre-
pregnancy pressure levels [2,3]. During pregnancy vessel walls are remodeled; a reduction of elastin and collagen is observed, followed by loss of distensibility.

These adaptations entail a demodulation of vessel self-regulation, and some areas may easily develop vasogenic edema, also in case of blood pressure levels within normal limits. In some cases, inappropriate endothelial activation follows; this triggers a cascade of events, including the release of inflammatory cytokines [4]. The latter are supposed to cause the production and release of strong vasopressor substances. Endothelial activation may cause systemic pressure instability, as well as an abnormal response to vasopressor stimulation, leading to vasogenic edema. This model that defines the induction process of preeclampsia may be extended to PRES.

Therefore, PRES in pregnancy may be the expression of systemic changes in brain circulation observed in preeclampsia. The fact that almost every patient with eclampsia has PRES signs at neuro imaging, shows that this syndrome is the first sign of suffering of the central nervous system during eclampsia [5,6], rather than its direct consequence [7]. Moreover, it was observed that PRES-related MR changes may occur before the development of eclampsia [8]. Therefore, the theory that makes PRES as the manifestation of hypertension with eclampsia is not exhaustive, due to the presence of blood pressure levels within normal limits. The observation of 2 consecutive cases of PRES in the absence of hypertensive stress should point out that endothelial changes might be significant in the development of vasogenic edema, by reducing the critical interval between the effect of pressure level increase and the effect of the blood-brain barrier. In line with this interpretation, the description of the above reported cases might be included within the framework of normotensive eclampsia [9,10]. Complete remission observed in these cases may also suggest that normal pressure levels may be prognostic indexes of positive outcome, combined with a minor incidence of neurological sequelae [11].

References