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POSTPARTUM POSTERIOR REVERSIBLE ENCEPHALOPATHY SYNDROME (PRES): A CASE REPORT AND A BRIEF REVIEW OF LITERATURE

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ABSTRACT

Posterior reversible encephalopathy syndrome (PRES) refers to a clinico-radiologic entity with characteristic features on neuroimaging and nonspecific symptoms comprising headache, confusion, visual disturbances and seizures. The radiological findings in PRES are thought to be due to vasogenic edema, predominantly in the posterior cerebral hemispheres and are reversible with appropriate management.We report a case of PRES diagnosed by MRI scan following uneventful caesarean section in previously normotensive patients who were succesfully treated with anticonvulsants and supportive treatment. This condition is important to recognize and needs to be treated promptly to prevent morbidity and mortality in pregnancy and postpartum.

KEYWORDS: PRES, Posterior Reversible Encephalopathy Syndrome, Postpartum.

INTRODUCTION

Posterior Reversible Encephalopathy Syndrome is an obstetric emergency frequently occurring in a postpartum period. Reversible posterior eukoencephalopathy syndrome (RPLS) or posterior reversible encephalopathy syndrome (PRES) was first described by Hinchey in 1996.[1] She described it as a reversible syndrome manifested as headache, altered mental functioning, seizures, and loss of vision associated with white matter changes,

suggestive of edema mainly in the posterior regions of the cerebral hemispheres, but also involving the brainstem, cerebellum, and other cerebral areas.[2] This is characterized by headache, vomiting, confusion, visual abnormalities and motor signs. These transitory neurological disturbances are thought to be due to cerebral vasospasm causing ischemia of the involved territory.[3] It describes a potentially reversible imaging appearance and may occur in diverse situations, including hypertension, eclampsia, pre-eclampsia, immunosuppressive medications such as cyclosporine, various antineoplastic agents, severe hypercalcaemia, thrombocytopenic syndromes, Henoch Scholein purpura, Systemic Lupus Erythematosus (SLE), hemolytic uramic syndrome, amyloid angiopathy, renal failure, post-transplantation, infection, sepsis (gram negative organisms predominate) and shock.[4] Importantly these changes appear to be completely reversible if the underlying cause is treated or the precipitating drug withdrawn early in the clinical course.

PRES is a very rare condition and so is usually not suspected. This causes delay in diagnosis and treatment, which can lead to permanent neurological damage.

We report a case of reversible encephalopathy syndrome, occurring in the postpartum period managed successfully.

CASE REPORT

A 23-year-old female, 10 days post caesarean section presented with 3-4 episodes of generalized tonic clonic seizures and disoriented for one day. Examination revealed that she was drowsy but arousable, disoriented and afebrile with pulse of 140/min, blood pressure of 120/80 mm of Hg and respiratory rate of 20/min. Patient was in post-ictal phase. Other medical history were unremarkable. No signs of preeclampsia, such as edema, proteinuria, or neurological complaints had been observed during pregnancy. Pupillary reactions were normal and fundoscopy revealed severe arterial attenuation. Results of abdominal, cardiovascular, and respiratory system examinations were unremarkable. Her complete blood picture, kidney function test, liver function test, clotting parameters, and electrocardiogram were within normal limits. Her metabolic profile and urine was normal. Electroencephalogram (EEG) record showing diffuse slowing s/o diffuse encephalopathy. MRI Brain (figure 1) showed multiple small patchy hyperintensities are seen involving cortex and subcortical white matter of bilateral frontal, parietal and parieto-occipital regions, corona radiate and the left centrum semiovale suggestive of Posterior Reversible Encephalopathy Syndrome (PRES). She was treated with antibiotics, anticonvulsants and

supportive therapy. She got stabilized by the fourth day and her neurological symptoms resolved by seven days. She was discharged on anticonvulsants and supportive medications. She completed her treatment and under regular follow up and was free of neurological symptoms.

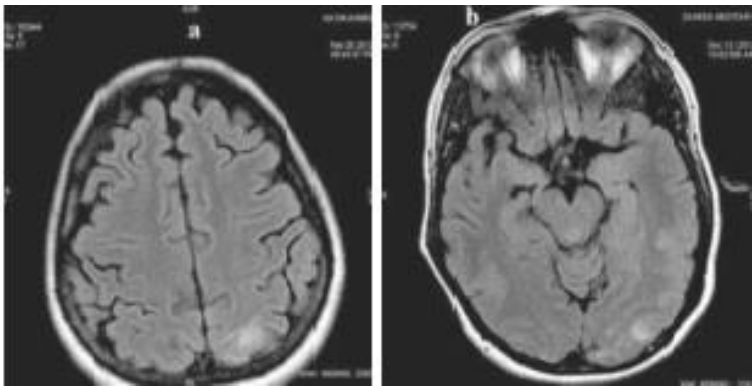


Figure 1

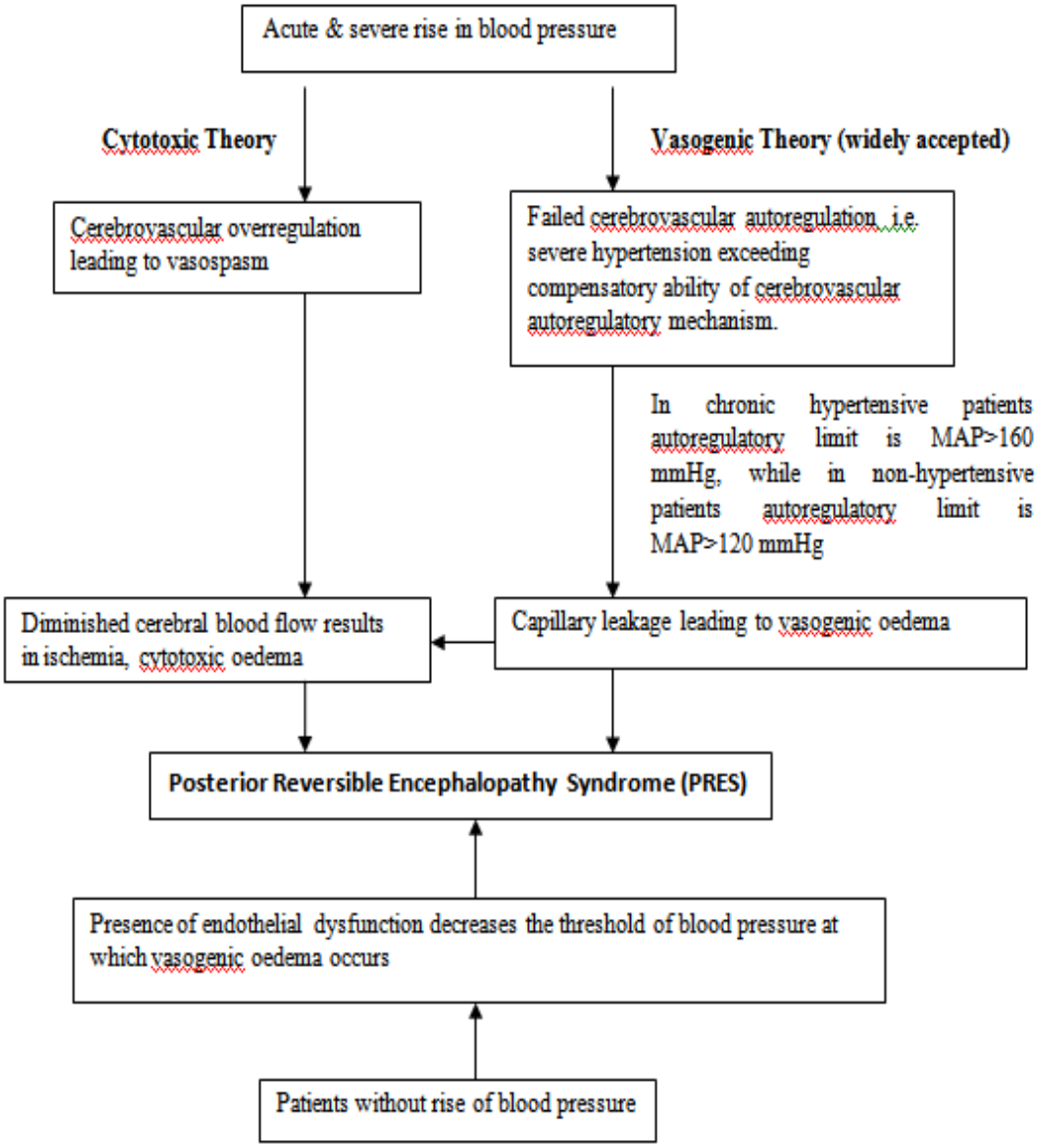
DISCUSSION

PRES was first described by Hinchey & colleagues in 1996 as Reversible Posterior Leukoencephalopathy Syndrome. PRES has been described as various non-specific symptoms of intense headache, visual problems (e.g. blurring of vision, hemianopia, cortical blindness, hemineglect), altered consciousness and generalised seizures. Rarely the patients may develop focal neurological deficits like paresis. PRES may develop suddenly or over several days.[5]

The term PRES is a misnomer as it occurs not always in posterior cerebral circulation but may occur primarily in anterior cerebral circulation. Also, PRES is not always reversible; it can be irreversible if the aetiology is not treated. In addition, patient in PRES may not always present with an encephalopathy, instead patient may have various nonspecific clinical features other than encephalopathy. In PRES white matter is mostly involved but it may involve deep white and gray matters.[6]

PRES a recently described clinical neuroradiological term that is associated with several medical condition besides preeclampsia/eclampsia and hypertension e.g. Renal failure, Post-transplantation (Allogeneic bone marrow transplantation; Solid organ transplantation), Immunosuppressive therapy (Cyclosporine; Tacrolimus) Autoimmune diseases (SLE; Systemic Sclerosis; Wegener's Granulomatosis; Poly Arteritis Nodosa), Post-cancer chemotherapy and has recently shown to be associated with infection, sepsis, and shock.[7]

The pathophysiology of PRES is not well understood. Vasogenic theory i.e. hypertension with loss of autoregulation remains a widely accepted consideration for the development of brain oedema.[8] Sometimes patients with normal blood pressure may develop PRES if they have substantial rise in blood pressure which is considered to be within the range of normal blood pressure. This is believed to be due to some neurotoxic substances.[9] PRES is seen in the absence of hypertension in 20%–40% of patients.[5] Alternatively, endothelial dysfunction/injury, hypoperfusion, and vasoconstriction may lead to altered integrity of the blood-brain barrier.[10] In the latter, though some degree of hypertension is present, reported blood pressure usually do not reach the limit of autoregulation (mean arterial pressure >160 mm Hg).



Pathophysiology of PRES[11]

Neuroimaging is essential to the diagnosis of PRES. Typical findings are symmetrical white matter edema in the posterior cerebral hemispheres, particularly the parieto-occipital regions with sparing of the calcarine and paramedian occipital lobe structures, differentiating it from bilateral posterior cerebral artery infracts.[12]

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Neuroradiographic abnormalities of PRES are often apparent on Computed tomography (CT) scans but are best depicted by magnetic resonance imaging (MRI). The most common abnormalities on MRI are punctate or confluent areas of increased signal on proton density and T2 -weighted images.[13] When regions of the brain other than parieto-occipital lobes are predominantly involved, the syndrome can be called atypical. In such cases, a diffusion weighted MRI with ADC mapping shows increased ADC values representing vasogenic edema in these areas, thus differentiating atypical PRES from other brain disorders. Most case reports suggest that posterior reversible encephalopathy syndrome (PRES) is usually benign. In many cases, it seems to be fully reversible within a period of days to weeks after removal of the inciting factor and control of blood pressure. Significant reversal of neuroradiologocal abnormalities with complete clinical recovery forms the diagnosis.[14]

The differential diagnosis of postpartum seizure includes eclampsia, subarachnoid hemorrhage, intracerebral hemorrhage, thrombotic phenomena, intracranial neoplasm, head trauma, idiopathic epilepsy, infection (meningo-encephalitis), amniotic fluid embolism, postpartum angiopathy.[3] Brain MRI with venogram rule out intracranial bleed, ischemia secondary to thromboembolism, vasospasm or space occupying lesion. Amniotic fluid embolism presents with cardiopulmonary collapse and coagulopathy which are absent in our patients.[15] Postpartum angiopathy is characterized by severe thunderclap headache, seizure, focal neurological deficits and segmental narrowing and dilatation of large and medium sized arteries. Typically scanning reveals ischemic lesions but MRI findings are suggestive of PRES.[16]

A study of 76 patients by Alexander M. McKinney et al, showed that the incidence of regions involvement was parieto-occipital 98.7%, temporal 68.4%, thalamus 30.3%,

cerebellum34.2%, brainstem 18.4%, and basal ganglia 11%[17] The incidence of less common manifestations was enhancement 37.7%, restricted diffusion 17.3%, hemorrhage 17.1% and a newly described unilateral variant 2.6%. In our study, the most commonly involed location was the parieto-occipital region, which was seen in 4 cases (100%). This was followed by frontal lobe in 2cases (50%) and temporal lobe in 1 case (25%).

The objective of the treatment of PRES is directed at the underlying aetiology e.g., control of blood pressure, reducing the dose or withholding the offending drug in patients undergoing chemotherapy or immunosuppression. Some physicians use antiepileptic drugs or Magnesium sulphate to avoid the progress of seizures, but their role is controversial or yet to be established.[18]

The clinical outcome is variable but is mostly favourable with prompt treatment of the underlying cause and repeat neuroimaging may not be necessary. Immediate action to identify potential triggering drugs, controlling hypertension, and treating aetiology of PRES can lead to complete reversal of radiological and neurological findings.[19] However, in few patients, PRES progresses to ischemia, infarction, or death.

Postpartum Posterior Reversible Encephalopathy Syndrome (PRES) is a rare condition usually detected after uneventful pregnancy. This condition is important to recognize and needs to be treated promptly to prevent morbidity and mortality in pregnancy and postpartum.

CONCLUSION

PRES is a rare but easily treatable condition and should be considered in differential diagnosis of postpartum seizure. Awareness of diverse clinical and radiological presentation of PRES is essential to avoid misdiagnosis and treatment delay. The syndrome of PRES is correctly recognized on neuro-imaging and potential complications can be avoided by appropriate therapy.

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