Posterior Reversible Encephalopathy Syndrome: A case report with 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 non-specific symptoms comprising headache, confusion, visual disturbances and seizures. The lesions in PRES are thought to be due to vasogenic oedema. Here we are reporting a postpartum case of 18 year old primi with PIH who presented to our emergency department with detorioration of consciousness, headache and respiratory distress. Patient also had an episode of GTCS (Generalised Tonic Clonic Seizure) on admission. However, on the basis of MRI finding PRES was diagnosed and treated successfully in critical Care. Due to timely intervention patient had recovered without any neurological deficit.

Keywords: Posterior reversible encephalopathy syndrome, Preeclampsia, Postpartum seizure.

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Introduction

Posterior reversible encephalopathy syndrome (PRES) is described as a clinicoradiological entity that was well described by Hinchey et al.[1] in 1996, shortly after two other small case-series were published.[2,3] This condition has been designated by a variety of names (reversible posterior leukoencephalopathy syndrome, reversible posterior cerebral edema syndrome, and reversible occipital parietal encephalopathy).PRES is now the accepted term but has been challenged recently based on the risk of neurological impairment and up to 15% mortality rate. PRES is characterized by variable associations of seizure activity, consciousness impairment, headaches, visual abnormalities, nausea/ vomiting, and focal neurological signs.[4]

PRES can develop in association with various conditions. However, regardless of the underlying cause or associated condition, the main abnormality is cerebral vasogenic edema, the pathogenesis of which is still not clear.[4] PRES is supposed to be typically reversible once the cause is removed. However, patients with severe manifestations of PRES, such as coma and/or status epilepticus, may require admission to the intensive care unit (ICU).[5,6] In our case also patient needed intensive care admission and care. Moreover, permanent neurological impairment or death occurs in a minority of patients.[4]

Most cases occur in young to middle-aged adults. However, there is a marked female predominance.[1,4]

Our Case

An 18 year old primipara (P1L1) with history of preeclampsia was referred from outside health centre post normal vaginal delivery with complaints of headache, deterioration of consciousness and respiratory distress. She had delivered a still born baby girl at her 9 months of gestation. There was no history of bleeding per vaginum or foul smelling discharge. There was no history of any fever or cough. On examination she was pale, mild icterus was present. On admission she had one episode of GTCS (Generalised Tonic Clonic Seizure) and BP was 150/100 mm Hg.Injection labetolol was given to control the BP. Seizure was controlled with Injection Midazolam 2 mg and Phenytoin Loading dose was given and maintenance dose was advised. Urine output was within normal. Neurological examination could not reveal any abnormality. Patient was intubated in view of respiratory distress and GTCS and put on ventilator. Diffuse slowing without epileptiform discharges was detected in Electroencephalography. There was no papilledema Chest X-ray showed basal infiltrate and PaO_2/FiO_2 was 200. Blood investigations were within normal limits. BP was controlled. MRI (T2 FLAIR) was done and revealed thickening of cortex and hyperintensity in the bilateral superior and middle frontal gyri. Hyperintensity is also noted in the subcortical white matter. (Fig. 1), which was suggestive of PRES. This patient was gradually weaned off from ventilator and extubated after 5 days. Injectables were converted to tablet forms. Patient was shifted toward after 6 days. There was no residual neurological deficit.

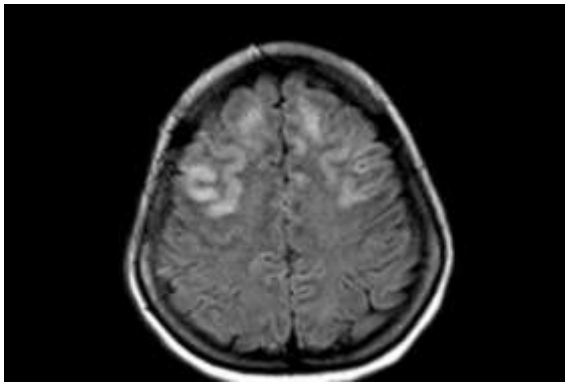

Fig. 1: MRI (T2 FLAIR) was done & revealed thickening of cortex and hyperintensity in the bilateral superior & middle frontal gyri. Hyperintensity is also noted in the subcortical white matter

Discussion

PRES can develop in association with a vast array of conditions. However, regardless of the underlying cause, the main abnormality is cerebral vasogenic edema, the pathogenesis of which is still under debate.[4] PRES is typically reversible once the cause is removed. However, patients with severe manifestations of PRES, such as coma and/or status epilepticus, may require admission to the intensive care unit (ICU).[5,6] Moreover, permanent neurological impairment or death occurs in a minority of patients.5 In our case patient had recovered without any neurological deficit.

Even though high BP is not usually described among the main signs of PRES. However, it is reported in most studies in 67% to 80% of patients.[1-3,7,8] In our case also the presentation was with deterioration of consciousness with high BP, which was controlled in the right time and thus helped the patient to recover.

There are two main hypotheses for other PRES which contradict each other. One involves impaired cerebral and endothelial dysfunction with cerebral hypo perfusion is another one. Both hypotheses result in blood-brain barrier dysfunction with cerebral edema.[4]

In our case we had supported the ventilation and controlled the cerebral edema with injection mannitol and mild hypocapnia, as well as control of hypertension collectively increased the chance of better outcome of the patient. Most of the cases of PRES is diagnosis of exclusion, however we should always keep this condition in mind while treating patients with such symptoms.

Thus, in patients with PRES a timely intervention of anti-hypertensives, anti-cerebral edema measures as well as management of other associated symptoms like seizures can make this condition truly reversible.

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