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Case Report

Posterior Reversible Encephalopathy Syndrome in a 20-Year-Old Female Admitted into the Intensive Care Unit

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Abstract

Posterior Reversible Encephalopathy Syndrome (PRES) is a rare clinico-neuroradiological syndrome. Here we present a case of a 20-year-old lady who was admitted into our Intensive Care Unit (ICU) with respiratory distress, limb weakness, blurring of vision, four episodes of generalized tonic-clonic seizures, altered sensorium and headache on the14th day following Lower Segment Caesarian Section (LSCS) due to 38 weeks of pregnancy with severe pre-eclamptic toxaemia (PET). She was diagnosed as a case of PRES based on the clinical features along with findings on Magnetic Resonance Imaging (MRI). She was treated conservatively. Clinical condition of the patient was improved within 3 days. So, we conclude early recognition and proper management helps to reduce permanent neurological damage and thereby reduce morbidity and mortality.

Key words: *Posterior reversible encephalopathy syndrome; Reversible posterior leukoencephalopathy syndrome; Postpartum seizure; Pre-eclamptic toxaemia (PET); Vasogenic oedema*

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Introduction

Posterior Reversible Encephalopathy Syndrome (PRES) is a transient clinico-neuroradiological[1] condition characterized by cerebral endo-theliopathy with successive disruption of the blood–brain barrier and reversible vasogenic oedema.[2,3] Patients frequently present with headache, seizure, impaired consciousness, visual disturbances, paresis, nausea, vomiting and focal neurological signs.[4-6]

The risk factors for developing PRES usually associated with fluctuations of blood pressure, pre eclamptic toxaemia (PET), eclampsia, sepsis, renal failure, cytotoxic agents, autoimmune disease, bone marrow or stem cell transplantation, thrombotic thrombocytopenic purpura, Henoch-Schonlein purpura, leukaemia, lymphomas, sickle cell anaemia, hemolytic uremic syndrome, pheochromocytoma, primary aldosteronism, electrolyte disturbances, blood transfusion etc.[7,8]

Though pathogenesis of PRES still remains unclear two theories are popular, one is the hyper perfusion

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theory or vasogenic theory and another one is the hypo perfusion or ischaemic theory which is also known as cytotoxic theory. The vasogenic theory suggests that due to the error of CNS auto regulation and sympathetic innervations of vessels emanating from basilar and vertebral arteries, resulting in increased cerebral blood flow. This leads to raise capillary filtration pressure and damage to capillary wall which causes increased blood-brain barrier permeability finally leads to cerebral vasogenic oedema.[9] The second theory suggests that T-cell activation and release of cytokine leads to endothelial dysfunction, deranged auto-regulatory response. Further, activation of argentine vasopressin (AVP) leads to vasoconstriction and cerebral ischaemia which potentiates development of PRES.[10,11]

The disease is usually diagnosed by high clinical suspicion and neuro-imaging especially MRI of the brain.[12] The classic imaging patterns usually shows reversible vasogenic oedema in the posterior circulation territories but sometimes anterior circulation territories may also be involved.[13] T2-weighted and Fluid Attenuated Inversion Recovery (FLAIR) sequences helps in detecting cortical and subcortical lesion related to PRES. In addition, Diffusion Weighted Imaging (DWI) and Apparent Diffusion Coefficient (ADC) mapping sequences are helpful in differentiating cytotoxic from vasogenic oedema and ischaemic lesions.[10]

The management of PRES usually starts from early diagnosis, symptomatic treatment, correction of specific causes and removal or reduction of the triggering factors such as stopping of chemotherapy or immunosuppressive agents, adequate control of blood pressure in hypertensive patients, use of antiepileptic drugs to attenuate seizure, correction of electrolyte imbalances, maintenance of proper hydration etc.[10,14]

Case report

A 20-year-old lady was admitted into the ICU of Enam Medical College and Hospital with the history of respiratory distress, limb weakness, blurring of vision, four episodes of generalized tonic-clonic seizures, altered sensorium for 5 hours and headache for 8 days on the 14th day following LSCS due to 38 weeks' pregnancy with severe PET. Initially, she was taken to a local hospital where she was diagnosed as a case of seizure disorder with postpartum psychosis with urinary tract infection. After initial management she was referred to our hospital. She was normotensive, non-diabetic, however she had a history of having bronchial asthma since childhood.

On examination, we found her Glasgow Coma Scale (GCS) was 9 out of 15, oxygen saturation was 95% on room air, vitals were normal, rhonchi were present on auscultation of Lungs. Planter reflexes were withdrawal on left side and 'no response' on right side. Deep tendon reflexes were sluggish on both limbs on right side however were normal on left side. In addition, she did not have any 'muscle power' on both limbs on right side, yet were normal on left side. Both pupils were normal in size, reacting to light and bilaterally symmetrical. Kernig's and Brudzinski's sign were negative.

Initial laboratory investigations revealed mild anaemia (Hb 9.6 gm/dl), raised White cell count (15,120/ccm), raised D-dimer (338 ng/mL), and significant amount of pus cell on urine R/M/E. However, bed side heat coagulation test was not remarkable. Serum creatinine, electrolytes, calcium, magnesium, albumin, SGPT, lactate and arterial blood gas was normal. Further, Real Time Polymerase Chain Reaction (RT-PCR) for SARS-COVID-19 was also negative. Chest radiograph showed pulmonary hyperinflation with congestion. Echocardiogram and computed tomography (CT) scan of the brain appeared normal.

On next day MRI of the brain was done which revealed bilateral symmetrical cortical and sub-cortical T1W1 hypo, T2W1 and FLAIR hyper intense signal changes were seen in parieto-occipital regions. Few areas showed diffusion restriction in DWI images (Fig 1). So, her diagnosis was confirmed as PRES.

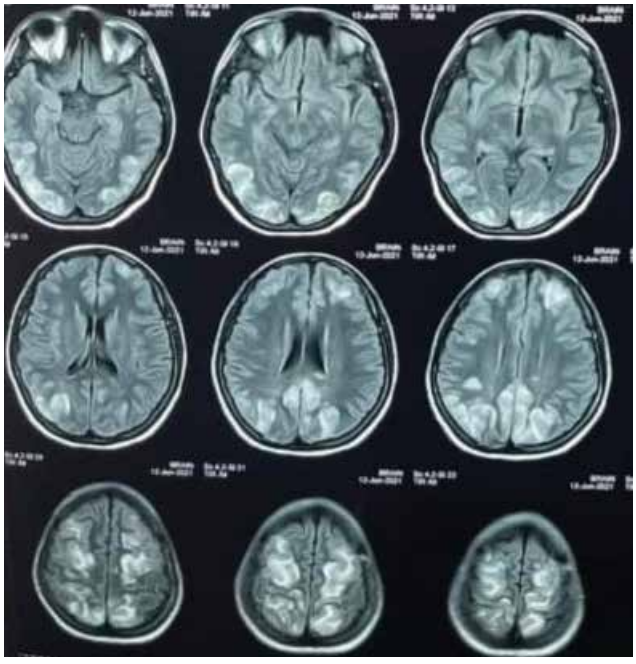

Fig 1. MRI of brain showing bilateral symmetrical cortical and sub-cortical hyper intense signal changes in parieto- occipital regions.

Rapid symptomatic management was started with intravenous fluids, antibiotics, anti-epileptics, steroid therapy, proton pump inhibitors, nebulization, and physiotherapy along with strict monitoring of patient's vitals as per ICU protocol. Patient was able to maintain adequate oxygen saturation by bag mask ventilation. The condition of the patient improved rapidly within three days so she was shifted to neuromedicine ward and later she was discharged on family's request on day 6. On follow up visit after one week she was found completely normal.

Discussion

Previously PRES was known as reversible posterior leukoencephalopathy syndrome, reversible posterior cerebral oedema syndrome or reversible occipital parietal encephalopathy.[15] It was first described as PRES by Hinchey et al[16] in 1996 after observing on several cases with headaches, altered mental status, seizures, visual loss, and radiologic findings of reversible symmetric posterior cerebral white-matter abnormalities on MRI[16]. In addition to this focal neurological deficit[17] may be observed, Legriel et al[18] found 19% of the patients in their study with focal neurological deficit similar to the finding of our patient.

Besides these usual symptoms sometimes patient may present without seizure with prolonged altered consciousness, drug intoxication and psychosis.[15] Our patient was also in non convulsive state for the initial 8 days for which she was diagnosed as a case of postpartum psychosis. PRES can be found several weeks after parturition and the clinical presentation is often confused with other diseases. Survillo et al[19] found two cases of PRES who were admitted into the ICU in their late postpartum period and Rijal et al[20] reported a case who was admitted into the ICU two weeks following LSCS which is also similar to our case.

The global incidence of PRES is unknown. It can occur in patients ranging from children to elderly. Mostly, cases occur in young to middle aged adults with significant female dominance.[20] In a study by Hinchey et al[16] found 13 females and only 2 males among 15 cases and age ranged from 15 to 62 years. Similarly, our case is also a young lady. Besides this, several authors such as Wen et al[21], Herrera et al [22], Fahmida et al[8], Nielsen et al[9] reported PRES on young female. On the other hand, Fonseca et al[11] reported a case about PRES on a 6 years old girl and Cherniwasky et al[4] reported a 58 years old female with PRES.

Several conditions are associated with the development of PRES such as hypertensive emergency, renal disease, sepsis, PET, eclampsia and immunosuppressive agents etc.[20] Among these, preeclampsia/eclampsia is the most common.[23] In our case there is also a history of severe PET as a triggering agent for developing PRES associated with urinary tract infection.

Patients with severe symptoms generally require ICU admission. Common indications for ICU admission include encephalopathy, seizures, status epilepticus, and respiratory failure.[10] Proper managements can ensure a complete recovery within a short period of time, usually within one week. Continued progression to several weeks is uncommon. Troung et al[6], Fahmida et al[8], Nielsen et al[9], Rijal et al[20] showed early recovery. Similarly, our case also recovered early. However,

Islam et al[24] reported delayed recovery for their case.

Recurrence of PRES can occur after resolution of the symptoms even after several years. Roth & Ferbert[25] found 8% recurrence cases in a study over 6 years of time. Delayed or misdiagnosis and inadequate management may lead to irreversible neurological deficit[26] or death upto19%[27]. Early detection and intensive management reduces the rate of chronic neurological sequel and fatal outcome.[28,29]

Conclusion

Since, PRES is a newly recognized disease and the symptoms are similar to many other neurological disorders, so the diagnosis is challenging. Further clinical and radiological evaluation and longtime follow up is needed for better understanding the pathogenesis of the disease and to reduce morbidity and mortality.

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