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Case Report
Published: 20 Jul, 2020

Journal of Neurology Forecast

Posterior Reversible Encephalopathy Syndrome (PRES) in a Patient with Late Postpartum with Probable SLE

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Abstract

Posterior Reversible Encephalopathy Syndrome (PRES) is a reversible clinic-radiological entity characterized by seizure, headaches, visual symptoms, impaired consciousness and other focal neurological findings. It is caused by a wide variety of causes ultimately leading to a white matter vasogenic cerebral edema of occipital and parietal lobes of the brain predominantly. We present here a young woman with headache, generalized tonic-clonic seizures and cortical blindness in a late postpartum stage. Reversibility of the symptoms and characteristic imaging findings led us to a diagnosis of PRES in our patient. While searching for etiology her ANA and lupus anti coagulant came positive with presence of dysmorphic RBC in urine. The patient improved after management with intravenous fluids, antibiotics, antiepileptics and monitoring of blood pressure. If recognized and treated early, the clinical syndrome commonly resolves within a week. A high index of suspicion and prompt treatment can reduce morbidity, mortality and pave the path for early recovery.

Keywords: PRES white matter; Vasogenic edema; Seizure; Cortical blindness

Introduction

Posterior Reversible Encephalopathy Syndrome (PRES) was first described by Hinchey et al., [1] in 1996. It is characterized by a milieu of seizure activity, impaired consciousness, headaches, visual symptoms, nausea/vomiting and focal neurological signs [2]. PRES can be associated with a number of conditions, all of which result in cerebral vasogenic edema which seems to be the crucial pathogenic mechanism [3,4]. As the name suggests, it is typically reversible once the underlying cause is removed. The global incidence of PRES is unknown. It has been reported in patients ranging from 4 to 90 years of age, with most cases occurring in young-aged to middle-aged adults [2]. A marked female preponderance is observed which may reflect some of the underlying causes [2]. PRES occurs in association with a number of causes, most commonly hypertension, pre-eclampsia/eclampsia and immunosuppressive agents [5]. This condition has been known by various names previously (reversible posterior leukoencephalopathy syndrome, reversible posterior cerebral edema syndrome and reversible occipital parietal encephalopathy). PRES is now the widely accepted term [3]. It is commonly but not always associated with acute hypertension [6]. This clinical syndrome is increasingly recognized, commonly because of improvement and availability of brain imaging. The major clinical conditions associated with PRES are represented in table 1. There is wide variation in the severity of clinical symptoms, i.e., the visual disturbance can vary from blurred vision, homonymous hemianopsia to cortical blindness. Altered consciousness may vary from mild confusion or agitation to coma. Other symptoms include nausea, vomiting and brainstem deficits. Seizures and status epilepticus are common, while non-convulsive status epilepticus may be more common than generalized status epilepticus. Non convulsive status should be cautiously observed in patients with prolonged altered consciousness, which may be mistaken commonly for postictal confusion. Signs include stereotypic movements like staring, head turning, eye blinking. Post ictal confusion usually lasts for hours, but PRES and convulsive status can last for many days and can be mistaken for drug intoxication psychosis or psychogenic states [7]. If recognized and treated early, the clinical syndrome commonly resolves within a week as in our patient.

Case Report

Mrs. X, a 19 year old primi, hailing from Dohar, Dhaka not known to have diabetes mellitus, hypertension and bronchial asthma presented at Ad din Women's Medical College Hospital, Dhaka 7 days after LUCS due to prolonged labour with disorientation with restlessness and bilateral loss of vision for 1 day. She had H/O witnessed generalized tonic clonic seizure for four episodes

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Received Date: 12 Jun 2020
Accepted Date: 15 Jul 2020
Published Date: 20 Jul 2020

Citation: Majumder A, Akter KT, Ronald Gomes R. Posterior Reversible Encephalopathy Syndrome (PRES) in a Patient with Late Postpartum with Probable SLE. J Neurol Forecast. 2020; 3(1): 1006.

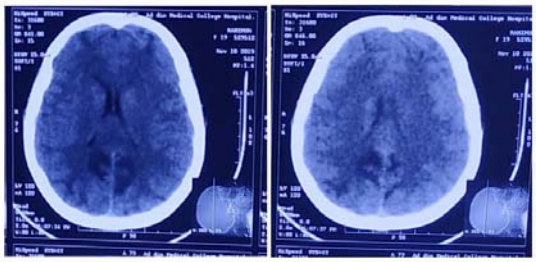

Figure 1 and 2: CT Brain showing bilateral diffuse cerebral edema involving white matter and infarct in both occipital lobes consistent with posterior reversible leukoencephalopathy syndrome.

Table 1: PRES associated clinical conditions.

1.	Preeclampsia
2.	Eclampsia
3.	Infection/ Sepsis/Shock
4.	Autoimmune disease- SLE
5.	Cancer chemotherapy
6.	Transplantation including stem cell transplantation
7.	Hypertension

prior to the day of presentation. Seizure was associated with urinary incontinence and lip injury. She also had headache prior to onset of seizure which was initially mild but was progressively increasing not subsided even after taking NSAIDs. Headache was associated with nausea and vomiting but there was no fever, photo or phonophobia. She also denied any joint pain, rash, oral ulcer. She had uneventful pregnancy with all her ante natal blood pressure recordings were in normal limits. Family history was unremarkable and she was breast feeding her male newborn child normally. On examination she was drowsy (GCS 9/15), not obeying commands but withdrew limbs to painful stimuli, a febrile with an elevated BP of 130/80 mm Hg; rest of the vital signs were within normal limits. An ocular examination revealed a diminution of vision of bilateral eyes to perception of light. Pupils were normally reactive to light and fundus examination was unremarkable with no evidence of papillodema. Rest of the cranial nerve examination was unremarkable. Power was 5/5 across all major joints and sensory function was intact all over the body. Cerebellar signs were intact and there was no evidence of meningeal signs such as nuchal rigidity or Kernig's/Brudzinski's sign. Plantars were down going bilaterally. Laboratory findings were significant for an elevated white cell count of 19000, N-85.4%, haemoglobin 10.7, platelets 334000. CRP 55.26.Urinalysis was remarkable for 3+protein and plenty of pus cells and RBC 12-15/HPF. Phase contrast microscopy revealed 2% of dysmorphic RBC in urine. No Casts were present. PT, PTT, INR and liver functions, renal functions including serum electrolytes were within normal limits. ANA was found positive with 25 u/ml while anti dS DNA was negative. Lupusanti coagulant was positive. As there was financial constraints, MRI of brain could not be done rather CT brain was done which revealed showed bilateral diffuse cerebral edema involving white matter and infarct in both occipital lobes consistent with posterior reversible leukoencephalopathy syndrome (Figure 1 and 2). The patient was started treatment with intravenous fluid, broad spectrum antibiotics, anti epileptic phenytoin and Inj. Dexamethasone. The patient was continuously monitored for haemodynamic stability. The patient's headache rapidly resolved and her vision improved. The patient continued to improve clinically and her vision improved gradually from perception to light to finger counting to normal visual acuity of 6/6 and was discharged home symptom-free on the sixth day of hospitalisation. There is a plan to follow her up at regular interval for the development of definite/classical SLE with repetition of lupus anticoagulant 12 weeks later.

Table 2: Imaging pattern in PRES.

1.	Holohemispheric watershed
2.	Superior frontal sulcus
3.	Dominant parietal/occipital
4.	Partial and/or asymmetric PRES

Discussion

PRES is a reversible neurological entity characterised by the presence of white matter oedema affecting the occipital and parietal lobes [8,9]. The exact incidence of PRES is unknown [10]. Patients with renal transplantation undergoing calcineuron inhibitor therapy develop PRES syndrome in about 4-8% of the cases [11]. It can occur at any age and most commonly affects females. This probably reflects the fact that one of the common causes of PRES is pre-eclampsia/eclampsia developing during pregnancy [2]. The exact pathophysiological mechanism of PRES is still unclear [12-14]. Three hypotheses have been proposed till now, which include 1) cerebral vasoconstriction causing subsequent infarcts in the brain, 2) failure of cerebral auto-regulation with vasogenic edema, and 3) endothelial damage with blood-brain barrier disruption further leading to fluid and protein transudation in the brain [12-14]. The distinct imaging patterns in PRES are represented in table 2. The reversible nature of PRES has been challenged recently based on new reports of permanent neurological impairment and mortality reaching 15% [15,16]. Pre-eclampsia and eclampsia are common medical disorders affecting pregnancy with significant maternal and fetal morbidity and mortality [17]. Hypertension and proteinuria are hallmarks for the diagnosis of pre-eclampsia, whereas seizures are typical of eclampsia [17]. Pre-eclampsia/eclampsia usually occurs between 20 weeks of pregnancy to 48 h postpartum [11]. The term late Postpartum Eclampsia (PPE) is used when eclamptic events occur between 48 h and 4 weeks after pregnancy [18]. A large observational study suggested that late PPE involves about 14% of cases of eclampsia [18]. A variety of clinical conditions are associated with the development of PRES. Among the reported causes, common ones include hypertensive emergency, renal disease, pre-eclampsia/eclampsia and immunosuppressive agents [19]. Other reported causes include sepsis, autoimmune diseases such as systemic lupus erythematosus, systemic sclerosis, tumour lysis syndrome, Gullain-Barre syndrome, AIDS, thrombotic thrombocytopenic purpura and acute intermittent porphyria [2,20,21]. PRES in association with late postpartum eclampsia has been reported before [20,22-25]. Although the exact prevalence of PRES in LPE is unknown, a recent study suggested it could be more common than expected [26]. Clinically, PRES presents with headache, seizures, encephalopathy, visual disturbances and focal neurological symptoms [9]. As the name suggests, reversibility of these symptoms is one of the hallmarks of the disease. However, some patients with severe manifestations of PRES, such as coma and/or status epilepticus, may require admission to the Intensive Care Unit (ICU) [8,27]. Moreover, permanent neurological impairment or death occurs in a minority of patients [8,28,29]. Subjective cognitive problems, development of chronic epilepsy, and progress to irreversible (partial) blindness can be long-time consequences

after years from acute episode [30]. Early and late complication such as pulmonary edema, dissection of extracranial internal left carotid artery, cerebral herniation, short term memory loss, subarachnoid hemorrhage, permanent mild dysmetria, visual impairment, and death have been described [24,31-33]. Differential diagnosis of PRES includes stroke, meningoencephalitis, demyelinating lesions of the brain and cerebral venous thrombosis. Early imaging is crucial to make this distinction. MRI is the imaging modality of choice [34]. PRES appears as high signal intensity predominantly in the posterior regions of the brain. Diffusion-weighted MRI helps to distinguish the vasogenic oedema from cytotoxic oedema, which is characteristic of this disease [18]. Our patient presented with headache, generalised tonic-clonic seizure and cortical blindness in a late postpartum stage posing a diagnostic dilemma. But the reversibility of the condition and the imaging finding guided us to a diagnosis of PRES. Nowadays the hypothesis of endothelial dysfunction in the pathophysiology of PRES is also proposed. For this reason monitoring LDH serum level as marker of endothelial dysfunction could be useful [35]. It is mandatory to remember that there are many severe obstetric complications that could be caused by endothelial dysfunction as preeclampsia, and so in these patients an isolated monitoring of LDH is not recommended, but a full screening for serum marker of preeclampsia. We revealed an increasing in two of three cases of PRES: in one patient the elevated LDH level is associated with thrombocytopenia, elevated liver enzymes, and increasing in markers of hemolysis and first depended on the developing of HELLP syndrome in a preeclamptic woman; meanwhile the other patients showed an isolated increasing in LDH level that could be linked to the developing of PRES, as reported in other cases in literature [35]. Approaching a woman suffering from headache after CS or a VD with intrapartum epidural a close monitoring is necessary in order to have a quick intervention in case of development of PRES.

The management of PRES involves early diagnosis, treatment of symptomatology and correction of the causative factor [36,37]. As indicated by its name, appropriate treatment is expected to ensure a full recovery. However, permanent complications and fatalities have been reported [27]. Recurrence of symptoms has been observed in 8% of the cases [6]. In hypertension-associated or drug-induced PRES, the effective therapy includes withdrawal of offending agent, immediate control of blood pressure, anti-convulsive therapy and temporary renal replacement therapy (hemodialysis/peritoneal dialysis) if required. In Systemic Lupus Erythematosus (SLE)-related PRES, aggressive treatment with corticosteroids and cyclophosphamide is effective. Corticosteroids may improve vasogenic edema, but there is no solid evidence for usage in PRES. Blood transfusion may cause a rapid increase in total blood volume, which further leads to cerebral blood flow overload. Abrupt or acute cerebral hyperperfusion exceeding the capacity of auto-regulation of cerebral capillary perfusion pressure might result in vasogenic edema found in PRES. The possibility of severe anemia as the predisposing factor, due to inadequate supply of oxygen to the brain may result in dysfunction of endothelial cells, further causing a functional loss or damage to the integrity of the blood-brain barrier in capillary circulation which cannot be ruled out [27]. Our patient had no hypertension even transiently during the whole episode.

Conclusion

PRES syndrome should always be considered in women associated with epileptic seizures or other neurological symptoms during pregnancy and in the postpartum. In our cases the patient obtained a complete remission of symptoms due to the early diagnosis and the sudden therapy. Our review stated the necessity to perform an instrumental diagnosis, using MRI as diagnostic gold standard tool and an adequate pharmacological and life support therapy in order to avoid any delay in diagnosis and treatment that may results in death due to transtentorial herniation or in irreversible neurological sequelae due to cerebral infarction or hemorrhage.

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