



REVERSIBLE POSTERIOR LEUKOENCEPHALOPATHY SYNDROME OR PRES IN A PRIMIGRAVIDA WITH TWIN PREGNANCY WITH POSTPARTUM ECCLAMPSIA (PPE): A CASE REPORT

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ABSTRACT

Reversible posterior leukoencephalopathy syndrome (RPLS), also known as posterior reversible encephalopathy syndrome (PRES), is a rare neurological disorder and patients with eclampsia, suffering from it has a high probability. PRES is a clinico-radiological syndrome characterised by headache, seizures, altered mental status, and disturbed vision. It involves white matter vasogenic edema that affects the posterior occipital and parietal lobes of the brain. In the present study, we report a case of a 24-year-old primigravida with twin pregnancy (dichorionic diamniotic) of 36 weeks and four days associated with mild pregnancy induced hypertension leading to the development of PRES after PPE postpartum eclampsia.

KEYWORDS

Reversible posterior leukoencephalopathy syndrome, posterior reversible encephalopathy syndrome, primigravida, dichorionic diamniotic

INTRODUCTION

Reversible posterior leukoencephalopathy syndrome (RPLS), also known as posterior reversible encephalopathy syndrome (PRES), is an uncommon neurological condition (Allen et al., 2006). Although projected as a rare condition, eclampsia patients have a very high chance (greater than 90%) of developing PRES (Hinduja, 2020). PRES is a clinico-radiological syndrome characterised by headache, seizures, altered mental status, and disturbed vision. It is primarily caused by white matter vasogenic edema, seen on both computed tomography (CT) and magnetic resonance imaging (MRI), that affects the posterior occipital and parietal lobes of the brain (Liman et al., 2012). The mechanism involves loss of autoregulation, breakdown of the blood brain barrier (BBB), and consequent vasogenic edema surrounding cerebral arteries and arterioles.

The prognosis of this syndrome is good and majority of patients recover within a few days to weeks (Pilato et al., 2020). 20% of PRES patients had associated eclampsia (Mahendra et al., 2021). Early identification and treatment of pre-eclampsia and eclampsia is the key to prevent there consequences such as cerebral ischaemia, intracerebral haemorrhage etc (Lam et al., 2017). The recovery happens in a week if it is identified and treated expeditely (Fugate et al., 2015). In the present study, we report a case of a 24-year-old primigravida with twin pregnancy (dichorionic diamniotic) of 36 weeks and four days associated with mild pregnancy induced hypertension (PIH) leading to the development of PPE followed by PRES.

Case Vignette

A 24-year-old primigravida with twin pregnancy (dichorionic diamniotic) of 36 weeks and four days, associated with mild pregnancy induced hypertension reported to the OPD with complaints of malaise and reduced foetal movement. Her blood pressure was 170/100 mmHg at the time of admission (missed medication) and her urine contained no protein. Prior to this her blood and urine tests conducted four days back were also normal and her family history was unremarkable. She was admitted for PIH management and expedited delivery to improve fetal outcome. The patient was conscious and oriented with a blood pressure 170/100 mm Hg, pulse rate 90/min and respiratory rate 18/min at the time of admission. Her respiratory and cardiovascular examination were within normal limits and abdominal examination revealed twin pregnancy with normal heart beats.

Investigation

The patient was admitted in the hospital and antihypertensive drugs were started to manage high blood pressure. She delivered two babies through caesarean section after stabilisation of blood pressure. Her blood pressure in the post-operative period returned to normal i.e 130/70 (mmHg) and the urine strip test for protein was found 3+. Immediately after delivery there were no signs of headache or eclampsia. The blood pressure after 8 hours post-delivery was 150/100 (mmHg) and later the patient complaint of severe headache, blurred

vision and threw a convulsion in the postnatal ward and later in the ICU.

Differential Diagnosis

Due to the sudden development of headache and blurred vision with elevated blood pressure and Eclampsia the patient was suspected to have either intracranial haemorrhage or PRES and later MRI scan confirmed that the patient had PRES. MRI reported hyperintensities in bilateral posterior parietal subcortical white matter suggesting edema as seen in **Fig. 1**. Further investigations were done and the patient was found to have high urine protein creatinine ratio of 0.95, with low platelets of 131, low hemoglobin of 8.5, elevated uric acid of 6.3 and LDH of 308 which were in line with the diagnosis of PRES and eclampsia.

Treatment

Eight hours post-delivery the patient developed single episode of severe frontal headache with convulsions and the patient was transferred to the intensive care unit (ICU). Magnesium sulphate infusion was started at a rate of 1 g/hour after a loading dose of 4gms with close monitoring of blood pressure along with oral Labetalol to control BP. Following the diagnosis of PRES, patient was given IV Dexamethasone 4mg twice daily. After 24 hours of eclampsia, the patient's blood pressure again started increasing and this time infusion of IV Labetalol was required to control the blood pressure. After 48 hours, Magnesium sulphate was stopped and patient was put on oral Amlodipine. The IV dexamethasone was also discontinued.

Outcome and follow-up

After four days of diagnosis of PRES, the patient's headache subsided and her vision returned back. She was transferred from ICU to the ward. On the sixth day of hospitalisation her BP normalized and her vision returned to normal as well. The electroencephalogram (EEG) and fundus examinations proved normal and no neurological deficit was detected. The patient was discharged after she became ambulatory and able to initiate breast feeding to her babies.

DISCUSSION/CONCLUSION

In PRES the occipital and parietal lobes of the brain exhibit white matter oedema and it is a reversible neurological condition. Cordelli et al. (2017) explained that PRES can occur in anyone at any age, but has a female preponderance. About 4-8% of patients of renal transplants on Calcineurin inhibitor therapy developed PRES syndrome (Cordelli et al., 2017) and in our study a pregnant female of 24 years developed PRES.

Phipps et al. (2019) reported that Pre-eclampsia and Eclampsia are widely prevalent medical conditions associated with pregnancy and causing significant maternal and foetal morbidity and mortality. Pre-eclampsia is diagnosed by the presence of hypertension with proteinuria, whereas Eclampsia is characterised by seizures. Pre-eclampsia and Eclampsia usually develop after 20 weeks of pregnancy and 48 hours after delivery respectively. When episodes of Eclampsia

take place between 48 hours and 4 weeks following delivery, the condition is referred to as late postpartum eclampsia (PPE). In our study, the patient developed pre-eclampsia after 36 weeks and 4 days of pregnancy and eclampsia developed at 8 hours after delivery.

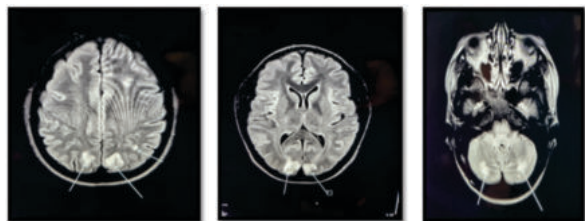
Several clinical disorders predispose patient to PRES, and common causes include pre-eclampsia/eclampsia, renal illness, hypertensive emergencies, and immunosuppressive medications. Sepsis, autoimmune conditions including systemic lupus erythematosus and systemic sclerosis, tumour lysis syndrome, Guillain-Barres syndrome, AIDS, thrombotic thrombocytopenic purpura, and acute intermittent porphyria are some of the other causes as reported by Khan et al.(2022), however in present case hypertension was the sole putative cause behind genesis of PRES.

Anderson et al. (2020) reported that PRES exhibits focal neurological signs and symptoms such as headache, seizures, encephalopathy, and visual abnormalities however in our study the patient reported sudden development of headache and blurred vision with elevated blood pressure.

Gao et al. (2018) reported reversibility of the symptoms as the chief characteristic of this syndrome. However, certain PRES patients need to be admitted to the intensive care unit if they are comatose or have status epilepticus (ICU). A small percentage of individuals experience permanent neurological deficit or death, however in our study the patient was admitted to the intensive care unit on complaints of severe headache, blurred vision and high blood pressure and Ecclamptic fit. The patient's vision gradually improved after four day of ICU stay and was discharged on sixth day.

Adam et al. (2013) and Fischer and Schmutzhard (2017) discussed that stroke, meningoencephalitis, demyelinating diseases and cerebral venous thrombosis are the differential diagnoses for PRES. To establish diagnosis, early imaging is prerequisite, MRI being the preferred modality and our study also used the same imaging to arrive at diagnosis.

Early diagnosis followed by symptomatic treatment, and treatment of causative factors are all integral parts in management of PRES (Kozak et al., 2007). Early and effective treatment aids full recovery, however, fatalities and long-term problems have been documented (Narbone et al., 2006) and in 8% of cases, symptoms have recurred (Roth & Ferbert, 2010).



(a) (b) (c)

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