Case Report

A case of the first presentation of pre-eclampsia and eclampsia as posterior reversible leukoencephalopathy syndrome: A case report

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Abstract

Posterior reversible leukoencephalopathy syndrome [PRES] is a rare disorder which is usually associated with a wide variety of diseases including renal diseases, autoimmune diseases, hypertension, pre-eclampsia and immunosuppressive therapy. The exact aetiology remains unknown. Even though there are cases of PRES occurring in patients with pre-eclampsia, cases of the first presentation of pre-eclampsia and eclampsia as PRES are rare.

Here we report a case of a previously healthy primigravida presenting with generalized tonic-clonic convulsions. Her blood pressure had been normal throughout pregnancy as well as on admission. Following admission there were two high blood pressure recordings. Pre-eclamptic toxaemia [PET] screening reported as positive, indicating pre-eclampsia. Magnetic resonance imaging [MRI] brain was suggestive of PRES. She was treated with magnesium sulphate, anticonvulsants and antihypertensives after which seizures were controlled and blood pressure was normalized. Emergency lower segment caesarean section was performed and the baby was delivered without any complications.

Therefore, it is important to consider PRES as a differential diagnosis in a pregnant patient presenting with convulsions even in the absence of a background history of pre-eclampsia. Early recognition and treatment of PRES will enhance prognosis.

Keywords: Posterior reversible leukoencephalopathy syndrome, Pre-eclampsia, Eclampsia, Seizures, Anticonvulsants

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Introduction

Posterior reversible leukoencephalopathy syndrome (PRES) is a clinical disorder that presents with a combination of symptoms, including visual disturbance, seizures, headache, and altered level of consciousness [1]. The cause of PRES is not clearly understood, even though there are several suggested theories. A wide variety of conditions, including hypertension, pre-eclampsia, renal diseases, autoimmune diseases, and immunosuppressive therapy, are known to be associated with PRES [2].

Here we report a case of a primigravida presenting at 32 weeks of gestation with generalized tonic-clonic convulsions who was diagnosed as having pre-eclampsia
and eclampsia complicated with PRES. Even though there are several case reports of PRES developing in patients with pre-eclampsia and during the post-partum period, cases of the first presentation of pre-eclampsia and eclampsia as PRES are rare.

**Case presentation**

An 18-year-old primigravida with an unremarkable medical history presented to National Hospital, Kandy, Sri Lanka, at 32 weeks of gestation with two episodes of generalized tonic-clonic convulsions, each lasting for 3 minutes duration. She had associated headaches, vomiting, back pain, and blurred vision. She complained of facial and bilateral lower limb swelling for three days duration prior to the onset of seizures. There was no history of fever or altered behaviour. Blood pressure and blood sugar levels have been normal throughout the pregnancy. An ultrasound scan done one week prior to the admission had revealed a normal placenta. There was no past history or family history of neurological diseases, autoimmune diseases, and eclampsia/pre-eclampsia.

Physical examination on admission did not reveal neck stiffness, and the Kernig sign was negative. She had periorbital oedema. She did not show alopecia, oral ulcers, or skin rashes. Central nervous system examination was normal, and reflexes were not exaggerated. The fundoscopic examination did not reveal papilledema or sub-retinal haemorrhages. Blood pressure was 120/80 mmHg. Other system examinations were normal. Within 24 hours following admission, high blood pressure recordings of 180/110 mmHg and 170/100 mmHg had been recorded.

Full blood count revealed a white cell count of 7.9 x 10^9 /L (neutrophils 62.7%, lymphocytes 25.7% and eosinophils 1.9%), haemoglobin of 10.1 g/dL (normal range: 13.6-17.2 g/dL), mean corpuscular volume of 90.1 fL (normal range: 80-96 fL) and a platelet count of 108 x 10^9/L (normal range: 150-400 x 10^9/L). The blood picture was reported as normal for pregnancy. Capillary blood glucose was 100 mg/dL (normal range: 70-130 mg/dL). Erythrocyte sedimentation rate (ESR) was 78 mm/hr. Blood and urine cultures were normal. Urine full report revealed 1+ proteinuria. Liver function tests and enzymes were normal except for albumin of 3 g/dL (normal range: 3.5-5.5 g/dL) and alkaline phosphatase of 184.2 u/L (normal range: 44-147 u/L). Renal function tests, serum electrolytes, serum ionized calcium, and magnesium were normal. Clotting profile and autoimmune screening were normal. Lactate dehydrogenase level, reticulocyte count, and direct antoglobulin test were normal. Electroencephalography revealed evidence of mild generalized cerebral dysfunction suggestive of encephalopathy or post-ictal phase. Non-contrast Computed Tomography (NCCT) of the brain was reported as normal. An ultrasound scan of the abdomen revealed moderate ascites with right-side pleural effusion and a single live foetus. Magnetic resonance imaging (MRI) brain revealed areas of white matter oedema in posterior cerebral hemispheres involving parieto-occipital regions sparing the calcareous and paramedian parts of the occipital lobe, suggestive of PRES.

She developed another episode of seizures on the next day following admission, which was similar to previous episodes. A diagnosis of pre-eclampsia and eclampsia-associated PRES was made. She was started on a magnesium sulphate loading dose of 6 g followed by a continuous infusion of 2 g/hour. Meanwhile, respiratory rate and the patellar reflex were monitored. Since there were two high blood pressure recordings, she was started on labetalol 100 mg bd. The seizure episode was managed with an intravenous phenytoin infusion of 50 mg/min. Since this was considered a high-risk pregnancy, she underwent an emergency lower segment caesarian section (LSCS), and the baby was delivered without complications. Her blood pressure control was poor, and therefore nifedipine SR 20 mg bd was added. Following LSCS, intravenous phenytoin was withheld, and carbamazepine 200 mg bd was added. She developed one seizure episode 12 hours after LSCS, and carbamazepine was further increased to 300 mg bd.

Following that, blood pressure was well controlled, and she did not have further seizure episodes. Her symptoms resolved during the next three days. She was discharged with labetalol 100 mg bd, nifedipine SR 20 mg bd and carbamazepine 300 mg bd. Within the next three months, she did not develop seizures, and the blood pressure was well controlled. Repeat MRI brain was normal. Anticonvulsants and antihypertensive drugs were tailed off.

**Discussion**

Posterior reversible leukoencephalopathy syndrome is a rare disorder that usually occurs as a result of an underlying cause. As in our patient, seizures are often the presenting manifestation [3]. Associated visual conditions include visual hallucination, auras, hemianopia, visual neglect, and cortical blindness [3].
Even though hypertension is not essential for diagnosis, more than 70% of patients with PRES are hypertensive [3]. In a series of case recordings regarding associations of PRES, more than 50% of patients had chronic hypertension, and 38% had chronic kidney disease [4].

The exact cause for PRES is not known, but there are several suggested theories. There is suggested pathogenesis describing the temporary failure of autoregulatory mechanisms of cerebral vessels resulting in hyper-perfusion, breakdown of the blood-brain barrier, and vasogenic oedema. [5]. In cases of PRES associated with pre-eclampsia, it is believed that pregnancy itself predisposes to cerebral oedema. It is known that pregnancy has direct effects on perivascular innervations, causing nerve hypertrophy [6]. Even though there are reported cases of PRES in pregnancy and post-partum period in the available literature, cases of PRES as the first presentation of pre-eclampsia and eclampsia are rare.

Neuroimaging studies are essential to diagnose PRES. Even though radiographic abnormalities are often visible on computerized tomography (CT) scans, the investigation of choice is MRI [7]. Typically, white matter oedema is prominently noted in posterior cerebral hemispheres. Calcarine and paramedian parts of occipital lobes are generally spared, which makes it distinguishable from posterior cerebral artery infarction. Complications of PRES visible on MRI include ischaemia and intracranial haemorrhage [8]. Since the clinical and radiographic findings are not specific in PRES, other toxic and metabolic encephalopathies should be excluded. The differential diagnosis for PRES includes hypertensive encephalopathy, eclampsia, reversible cerebral vasoconstriction syndrome, auto-immune encephalitis, cerebral venous thrombosis, acute demyelinating encephalomyelitis, and malignancies such as central nervous system lymphoma. Usually, blood pressure in patients with PRES associated with eclampsia and pre-eclampsia is lower than in patients who develop PRES in other settings [9].

Early recognition of symptoms and prompt treatment is essential in order to prevent complications and improve morbidity and mortality. Management includes correcting the causative agent, managing hypertension and seizures. In malignant hypertension, the targeted diastolic blood pressure is 100-105 mmHg. Aggressive blood pressure lowering can lead to cerebral, coronary, and renal ischaemia. Recommended anti-hypertensives include clevidipine, nicardipine, labetalol, and nitroprusside [10]. Seizures are usually treated with anticonvulsants. The preferred drug varies depending on the patient’s comorbidities, renal function, and other side effects. In cases of pregnancy-associated with eclampsia and PRES, delivery of the placenta and baby is recommended. Magnesium is given to control seizures and is superior to anticonvulsants. Even though some cases report a continuation of treatment for 1-3 months, anti-epileptic drugs can be tailed off safely once the symptoms and neuroimaging findings resolve, generally after one to two weeks [11].

As suggested by most case series and case reports, PRES is usually benign and fully reversible within a period of days to weeks following removal of the causative factor and blood pressure control [12]. Cases of death have been reported secondary to cerebral oedema, intracerebral haemorrhages, and other complications [13]. Recurrence is infrequent in most cases [14].

Conclusions

When considering our case as well as available literature, PRES should be considered as a differential diagnosis in pregnant patients presenting with convulsions, even though there is no background history of pre-eclampsia. Early recognition and treatment avoid permanent brain damage with complete resolution of symptoms.

Informed Consent: Written informed consent was obtained from the patient.

References

1. McKinney AM, Short J, Truwit CL, McKinney ZJ, Kozak OS, SantaCruz KS, et al. Posterior reversible encephalopathy syndrome: incidence of atypical regions of involvement and imaging findings. AJR Am J Roentgenol. 2007;189(4):904–12. doi: 10.2214/AJR.07.2024.
2. Shah R, Kubisz-Pudelko A, Reid J. Posterior reversible encephalopathy syndrome following an inadvertent dural puncture during an emergency laparotomy for ischemic colitis – a case report. Local Reg Anesth. 2014;7:1-4. doi: 10.2147/LRA.S57660.
3. Hinchey J, Chaves C, Appignani B, Breen J, Pao L, Wang A, Pessin MS, Lamy C, Mas JL, Caplan LR. A reversible posterior leukoencephalopathy syndrome. N Engl J Med. 1996;334(8):494-500. doi: 10.1056/NEJM1996022233430803.

4. Fugate JE, Claassen DO, Cloft HJ, Kallmes DF, Kozak OS, Rabinstein AA. Posterior reversible encephalopathy syndrome: associated clinical and radiologic findings. Mayo Clin Proc 2010;85:427–32. doi:10.4065/mcp.2009.0590.

5. Lamy C, Oppenheim C, Méder JF, Mas JL. Neuroimaging in posterior reversible encephalopathy syndrome. J Neuroimaging. 2004;14(2):89-96.

6. Aukes AM, Vitullo L, Zeeman GG, Cipolla MJ. Pregnancy prevents hypertensive remodeling and decreases myogenic reactivity in posterior cerebral arteries from Dahl salt-sensitive rats: a role in eclampsia?. Am J Physiol Heart Circ Physiol. 2007;292(2):H1071-6. doi:10.1152/ajpheart.00980.2006.

7. Schwartz RB, Jones KM, Kalina P, Bajakian RL, Mantello MT, Grada B, et al. Hypertensive encephalopathy: findings on CT, MR imaging, and SPECT imaging in 14 cases. AJR Am J Roentgenol. 1992;159(2):379-83. doi:10.2214/ajr.159.2.1632361.

8. Covarrubias DJ, Luetmer PH, Campeau NG. Posterior reversible encephalopathy syndrome: prognostic utility of quantitative diffusion-weighted MR images. AJNR Am J Neuroradiol. 2002;23(6):1038-48.

9. Schwartz RB, Feske SK, Polak JF, DeGirolami U, Iaia A, Beckner KM, et al. Preeclampsia-eclampsia: clinical and neuroradiographic correlates and insights into the pathogenesis of hypertensive encephalopathy. Radiology. 2000;217(2):371-6. doi:10.1148/radiology.217.2.r00nv44371.

10. Hinchey J, Chaves C, Appignani B, Breen J, Pao L, Wang A, et al. A reversible posterior leukoencephalopathy syndrome. N Engl J Med.1996;334(8):494-500.doi: 10.1056/NEJM1996022233430803.

11. Bakshi R, Bates VE, Mechtler LL, Kinkel PR, Kinkel WR. Occipital lobe seizures as the major clinical manifestation of reversible posterior leukoencephalopathy syndrome: magnetic resonance imaging findings. Epilepsia. 1998;39(3):295-9. doi:10.1111/j.1528-1157.1998.tb01376.x.

12. Lee VH, Wijdicks EF, Manno EM, Rabinstein AA. Clinical spectrum of reversible posterior leukoencephalopathy syndrome. Arch Neurol. 2008;65(2):205-10. doi:10.1001/archneurol.2007.46.

13. Schweitzer AD, Parikh NS, Askin G, Nemade A, Lyo J, Karimi S, et al. Imaging characteristics associated with clinical outcomes in posterior reversible encephalopathy syndrome. Neuroradiology. 2017;59(4):379-86. doi:10.1007/s00234-017-1815-1.

14. Roth C, Ferbert A. Posterior reversible encephalopathy syndrome: long-term follow-up. J Neurol Neurosurg Psychiatry. 2010;81(7):773-7. doi:10.1136/jnnp.2009.189647.