CASE REPORT

Posterior reversible encephalopathy syndrome (PRES) in a patient with late postpartum eclampsia

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SUMMARY

Posterior reversible encephalopathy syndrome (PRES) is a reversible neurological entity characterised 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 vasogenic cerebral oedema of occipital and parietal lobes of the brain. We present here a young woman with headache, generalised 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.

BACKGROUND

Posterior reversible encephalopathy syndrome (PRES) was first described by Hinchey *et al*¹ in 1996. It is characterised 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 oedema 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

We report a case of a 28-year-old woman who developed late postpartum eclampsia complicated by the development of PRES.

CASE PRESENTATION

A 28-year-old woman, gravida 2 para 2, with an unremarkable medical history presented to the emergency department with symptoms of abdominal pain and distension, 2 weeks after caesarean section with total abdominal hysterectomy performed for placenta accreta. She had an otherwise uneventful pregnancy and she was breast-feeding her newborn male normally. All her antenatal blood pressure recordings were within normal limits. She also gave a history of a few episodes of nausea and vomiting. Family history was unremarkable. She never smoked and consumed alcohol only on rare occasions.

Physical examination revealed that she was fully alert and oriented. She had a temperature of 38.1°C,

blood pressure 135/85 mm Hg, pulse rate 90/min and respiratory rate 16/min. Respiratory and cardiovascular examination was within normal limits. Abdominal examination revealed suprapubic tenderness. Renal angles were non-tender.

Further evaluation with a CT of abdomen revealed a diagnosis of retrovesicular abscess which was drained percutaneously under fluoroscopic guidance. Intravenous antibiotics were given and the patient showed marked clinical improvement.

On the fourth day of hospitalisation, the patient developed one episode of witnessed generalised tonic-clonic seizure. Shortly after resolution of seizures, while the patient was regaining consciousness, she started to report of headache and a bilateral loss of vision. She was found to have an elevated BP of 180/100 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 hand movement. Pupils were normally reactive to light and fundus examination was unremarkable. 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 downgoing bilaterally.

INVESTIGATIONS

Laboratory findings were significant for an elevated white cell count of 17 000, haemoglobin 13, platelets 230 000. Urinalysis was remarkable for 3+protein. PT, PTT, INR and liver profiles were within normal limits. T2-weighted and fluid-attenuated inversion recovery images of brain MRI showed bilateral posterior parietooccipital hyperdensities in the cortex and subcortical white matter consistent with posterior reversible leukoencephalopathy syndrome (figure 1).

DIFFERENTIAL DIAGNOSIS

With the sudden development of headache and bilateral vision loss with elevated blood pressure, we considered an initial diagnosis of ischaemic stroke, cerebral haemorrhage and cerebral venous thrombosis, PRES and hypertensive emergency with retinal haemorrhage. A normal fundus examination essentially ruled out retinal haemorrhage, while the reversibility of symptoms with characteristic MRI findings led us to a diagnosis of PRES.

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Figure 1 Fluid-attenuated inversion recovery image of brain MRI showing hyperdensities in the cortex and subcortical white matter of right parieto-occipital lobe consistent with posterior reversible leucoencephalopathy syndrome.

TREATMENT

With the impression of late postpartum eclampsia, the patient was transferred to the intensive care unit. Labetalol infusion was started at a rate of 1 mg/min with close monitoring of blood pressure. Meanwhile phenytoin infusion was started at a rate of 50 mg/min. Magnesium sulfate was given in a loading dose of 6 g intravenously over 15 min followed by a maintenance dose of 2 g/h with monitoring of respiratory rate and patellar reflex. The patient was continuously monitored for haemodynamic stability. After MRI result consistent with PRES, magnesium and phenytoin were discontinued.

OUTCOME AND FOLLOW-UP

The patient's headache rapidly resolved and her vision improved to 20/20 after about 3 h of the onset of symptoms. She was observed in the intensive care unit for 24 h. She continued to be symptom free and was later transferred to a regular floor. The patient continued to improve clinically and was discharged home symptom-free on the sixth day of hospitalisation.

DISCUSSION

PRES is a reversible neurological entity characterised by the presence of white matter oedema affecting the occipital and parietal lobes. The exact incidence of PRES is unknown.⁶ Patients with renal transplantation undergoing calcineurin inhibitor therapy develop PRES syndrome in about 4–8% of the cases.⁷ 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

Pre-eclampsia and eclampsia are common medical disorders affecting pregnancy with significant maternal and fetal morbidity and mortality.⁸ Hypertension and proteinuria are hallmarks for the diagnosis of pre-eclampsia, whereas seizures are typical of eclampsia.⁸ Pre-eclampsia/eclampsia usually occurs between 20 weeks of pregnancy to 48 h postpartum.⁹ The term late postpartum eclampsia (PPE) is used when eclamptic events occur between 48 h and 4 weeks after pregnancy.¹⁰ A large observational study suggested that late PPE involves about 14% of cases of eclampsia.¹⁰

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.¹¹ Other reported causes include sepsis, autoimmune diseases such as systemic lupus erythematosus, systemic sclerosis, tumour lysis syndrome, Guillain-Barres syndrome, AIDS, thrombotic thrombocytopenic purpura and acute intermittent porphyria.^{2 12 13} PRES in association with late postpartum eclampsia has been reported before.^{12 14–17} Although the exact prevalence of PRES in LPE is unknown, a recent study suggested it could be more common than expected.¹⁸

Clinically, PRES presents with headache, seizures, encephalopathy, visual disturbances and focal neurological symptoms.¹⁹ 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).^{20 21} Moreover, permanent neurological impairment or death occurs in a minority of patients.^{22–24}

Differential diagnosis of PRES includes stroke, meningo-encephalitis, demyelinating lesions of the brain and cerebral venous thrombosis. Early imaging is crucial to make this distinction. MRI is the imaging modality of choice.²⁵ 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.¹¹ 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.

The management of PRES involves early diagnosis, treatment of symptomatology and correction of the causative factor.^{20 21} As indicated by its name, appropriate treatment is expected to ensure a full recovery. However, permanent complications and fatalities have been reported.²⁶ Recurrence of symptoms has been observed in 8% of the cases.⁶

Learning points

- ▶ When a postpartum patient presents with focal neurological deficits, the differential diagnoses to be considered includes haemorrhage, infarction, venous thrombosis, vasculitis, pontine or extrapontine myelinolysis and posterior reversible encephalopathy syndrome (PRES).
- ▶ MRI of the brain is crucial to make this distinction.
- ▶ PRES syndrome is a reversible entity when the aetiology is properly recognised and quickly treated; and if not, it can lead to irreversible neurological sequelae and even death.

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Competing interests None.

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