CASE REPORT

Unusual presentation of PRES in the postnatal period

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SUMMARY

Posterior reversible encephalopathy syndrome (PRES) is a clinicoradiological entity, associated with a vast array of medical conditions and a variety of presenting symptoms. There is a characteristic pattern of radiographic features alongside suggestive clinical manifestations, which lead to a diagnosis of PRES. This report describes the case of a 39 years old, previously normotensive woman, who presented on day 7 postpartum with generalised tonic clonic seizures, reduced conscious level and a history of blurred vision and headache. She was treated immediately as eclamptic and transferred to the intensive care unit for stabilisation. Following an inconclusive CT result, an MRI was performed 2 days after presentation, which demonstrated white matter changes consistent with those found in PRES. She made a full recovery and a repeat MRI scan 7 weeks later showed no progression of the lesions noted on the original scan.

BACKGROUND

Posterior reversible encephalopathy syndrome (PRES) is an 'encephalopathic' entity which was first described by Hinchley *et al* in 1996 based on 15 cases.¹ Its global incidence is unknown. It has been assigned a variety of names: reversible posterior leukoencephalopathy syndrome, reversible posterior cerebral oedema syndrome and reversible occipital parietal encephalopathy. This reflects the lack of consensus on the diagnosis of PRES. Indeed, this term itself has recently been questioned based on the risk of permanent neurological impairment and a mortality rate of up to 15%.^{2,3}

The intensity and severity of clinical manifestations and cerebral imaging abnormalities vary widely, making the diagnosis very difficult. A high index of suspicion, in the presence of suggestive clinical manifestations and radiological criteria, will ultimately establish the diagnosis of PRES and allow appropriate treatment to be initiated.⁴

CASE PRESENTATION

This case refers to a 39-year-old primigravida who had an uneventful antenatal period (including a booking blood pressure of 100/70 mm Hg at 12 weeks pregnancy). She was delivered by emergency caesarean section at 40 weeks and 5 days gestation due to a pathological cardiotocography and failure to progress in the first stage of labour. The caesarean section was complicated by uterine angle extension and subsequent haemorrhage. The patient, therefore, needed a postoperative blood transfusion of three units red cell concentrate but

made an otherwise uneventful postoperative recovery. She was discharged home from hospital on the 4th postoperative day without fever, and with a blood pressure 127/73, haemoglobin 99 g/L, haematocrit 0.303 and a normal surgical wound.

The patient was admitted via the A&E department 1 week postpartum with witnessed tonic clonic seizures. She had reported headache and blurred vision earlier that morning. On examination she had brisk reflexes, but no other obvious focal neurology, a raised blood pressure of 148/94 and 3+ protein on urine dipstick. A preliminary diagnosis of postpartum eclampsia was made. Treatment as per eclampsia protocol was started which included magnesium sulphate 4 g given intravenously and then 1 g/h over 24 h. Intravenous labetalol infusion was also started with a strict fluid balance monitoring.

INVESTIGATIONS

Initial investigations revealed a full blood count with evidence of haemoconcentration (platelet count 446×10^9 , haemoglobin 128 g/L, haematocrit 0.403, mean cell haemoglobin concentration 25.8 pg), normal clotting (international normalised ratio 0.9 and activated partial thromboplastin time ratio 0.98), normal liver function tests and urea and electrolytes. The urine protein/creatinine ratio was raised at 367 mg/mmol. She was transferred to intensive therapy unit (ITU) for stabilisation where an urgent CT scan was requested to exclude subarachnoid haemorrhage.

The CT head demonstrated a small right-sided acute occipital infarct and recommended CT venography. This was duly performed and showed no evidence of cerebral venous thrombosis, but confirmed a small area of low attenuation within the right occipital cortex and subcortical white matter, thought to most likely reflect an acute infarct. An MRI head scan was done 1 day later. This showed small areas of increased signal within the periventricular deep white matter, but no convincing acute infarct was seen within the right occipital lobe.

DIFFERENTIAL DIAGNOSIS

Late postpartum eclampsia (ie, eclamptic seizures developing greater than 48 h, but less than 4 weeks postpartum) accounts for approximately 13% to 16% of all cases of eclampsia, and often lacks a diagnosis of pre-eclampsia in the antenatal or intrapartum period.⁵ At the time of presentation it would have been important to consider alternative diagnoses such as cerebrovascular accident, sagittal sinus thrombosis and nephrotic syndrome, all of

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which could present similarly, but would have additional biochemical and/or neurological findings. However, given her recent pregnancy, the presence of classical prodromal symptoms and significant proteinuria, the decision was made to treat for eclampsia until further results were available. The subsequent response to treatment, alongside the radiological findings, led to the diagnosis of PRES.

OUTCOME AND FOLLOW-UP

The patient continued to be managed as a case of postpartum eclampsia under the joint care of the obstetric and intensive care teams. She developed sepsis secondary to a lower respiratory tract infection with subsequent pleural effusions. This responded well to piperacillin/tazobactam and careful fluid balance. She was discharged from ITU 5 days later on nimodipine and lamotrigine. Her blood pressure was well controlled on oral antihypertensives and she continued to make a full recovery with ongoing physiotherapy input. She was followed up by the neurology team and a repeat MRI head 7 weeks later demonstrated multiple foci of signal abnormality in the periventricular deep white matter, most likely to be ischaemic in nature. This was unchanged compared with the previous scan. No new abnormality was noted and no focal abnormality was seen in the right occipital region.

DISCUSSION

PRES is a clinoradiological syndrome, the global incidence of which is unknown. Most cases of PRES occur in young to middle aged adults, with a marked female preponderance that may reflect some of the causes.⁴

The pathophysiology of PRES is controversial and focuses around two main hypotheses. The first and most widely accepted theory states that rapidly developing hypertension leads to impaired cerebral autoregulation, followed by cerebral hyperperfusion. This increased cerebral blood flow causes the extravasation of protein and fluid which produces focal vasogenic oedema.⁶ The second hypothesis implicates endothelial dysfunction with cerebral hypoperfusion, but this may be more relevant where PRES is associated with cytotoxic therapy, and not in the presence of raised blood pressure.⁴ Regardless of the mechanism, the end result is that of altered cerebral blood perfusion, resulting in blood–brain barrier dysfunction and cerebral vasogenic oedema.

PRES can develop in association with a vast array of complex medical conditions including sepsis, autoimmune diseases, high-dose chemotherapy, eclampsia, pre-eclampsia and delayed eclampsia (PRES within several weeks after delivery). The length of time taken for symptoms to develop varies from acute onset to several days.

Clinical manifestations of PRES vary widely, but typical symptoms include headache, visual disturbances, paresis, nausea and vomiting, impaired consciousness and altered mentation.⁷ Generalised seizures are common and may lead to coma. Hypertension is not essential for a diagnosis of PRES to be made. In fact, moderate-to-severe hypertension is only present in 70–80% of patients⁸ and it is interesting to note that this has not been shown to be significantly associated with the severity of PRES.⁴

The cerebral imaging findings also vary, but until recently were thought to mainly constitute symmetrical vasogenic oedema localised to the posterior white matter. However, the advancement of neuroimaging techniques has led to four radiological patterns of PRES being described in the literature, according to the distribution of oedema: holohemispheric

watershed pattern; superior frontal sulcus pattern; dominant parieto-occipital pattern; and partial or asymmetric expression of the primary patterns.⁴ The involvement of parieto-occipital regions is more common, as in this case.

The development of MRI with diffusion-weighted sequences has improved the diagnosis of PRES and indeed MRI is superior to CT for this purpose. Diffusion-weighted images or fluid-attenuated inversion recovery sequences may show foci of high signal intensity in cortex that is either undergoing infarction or at high risk of infarction.⁸ CT findings are often normal or non-specific; however, there is some evidence to suggest that repeated CT scanning may be helpful when MRI is unavailable.⁹ PRES is typically reversible once the underlying cause is removed. Therefore repeated imaging several weeks later is recommended.

Appropriate therapy usually results in complete resolution of the deficits over several days to weeks,⁶ although permanent neurological impairment has been reported and the disease can be fatal.^{3 10} Where hypertension is present, immediate treatment with antihypertensives, and in cases of eclampsia, control and prevention of seizures with magnesium sulphate or phenytoin or withdrawal of the trigger factor, is the treatment of choice.¹¹

Learning points

- ▶ Posterior reversible encephalopathy syndrome (PRES) is a recognised complication of eclamptic and non-eclamptic fits.
- ▶ PRES can also be a consequence of cytotoxic therapy.
- ▶ Awareness of the vague early clinical signs together with MRI findings is likely to elicit an early diagnosis and management and may prevent permanent damage in this otherwise typically reversible condition.
- ▶ Multidisciplinary input in such cases is likely to lead to improvements in case morbidity.

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Competing interests None.

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