Postpartum posterior reversible encephalopathy syndrome (PRES) in a twin pregnancy complicated by preeclampsia-eclampsia: case report

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Summary

This is the second case in literature of posterior reversible encephalopathy syndrome (PRES) in a twin pregnancy complicated by preeclampsia-eclampsia. A 27-year-old primigravida with dichorionic diamniotic twin pregnancy was admitted at 36 weeks of gestation for induction of labour due to preeclampsia. On the second day postpartum, the patient developed severe hypertension, visual symptoms, confusion, headache, and eclamptic fits. Head computed tomography (CT) showed hypodense basal ganglia lesions. The patient was treated in the intensive treatment unit with hydralazine and labetalol infusions and anticonvulsants. Five days later, there was complete clinical improvement and follow-up magnetic resonance imaging (MRI) was normal. The patient was discharged 11 days postdelivery. Diagnosis of PRES is based on the presence of clinical features of acute neurologic compromise, abnormal neuroimaging findings, and complete reversibility of findings after prompt treatment. Early recognition and proper treatment result in complete reversibility of this condition.

Key words: PRES; Preeclampsia; Eclampsia; Twin pregnancy.

Introduction

Posterior reversible encephalopathy syndrome (PRES) is a clinical entity that presents in a wide variety of conditions. It was first described by Hinchey *et al.* in 1996 [1] but the currently used term "PRES" was proposed by Casey *et al.* in 2000 [2]. Diagnosis of PRES is based on the simultaneous presence of clinical features of acute neurologic compromise, brain neuroimaging findings, and clinical/radiologic proof of reversibility [3]. Despite the increasing number of reports indicating awareness of PRES among clinicians [4], its prevalence is still unknown [5,6].

The authors present the case of a primigravid woman with a twin pregnancy who was induced at 36 weeks of gestation due to preeclampsia and was diagnosed postpartum with PRES. To the best of the authors' knowledge, this is the second case report on the association between twin pregnancy and PRES.

Case Report

A 27-year-old primigravida, who conceived via assisted conception treatment was diagnosed with a dichorionic-diamniotic twin pregnancy at her dating scan. Her antenatal course was unremarkable with normal growth of both fetuses until she was admitted at 36 weeks of gestation with a diagnosis of preeclampsia. On admission, the patient complained of visual disturbances and headache and had increased blood pressure of 150/90 mmHg with 2+ proteinuria. Induction of labour was planned with dinoprostone. While in labour the suspicion of chorioamnionitis was raised due to a maternal temperature of 38.1°C and fetal tachycardia and the patient received intrapartum antibiotics. Pain relief was provided with epidural analgesia. Several hours later, two healthy babies were born both by ventouse delivery because of delayed second stage. The patient was subsequently taken to the postnatal ward.

On second day postpartum, the patient developed a high blood pressure (210/120 mmHg), with severe photophobia, confusion, and headache. Hypertension was treated with hydralazine and magnesium sulphate was given for seizure prevention. Once stabilised, a head computed tomography (CT) was performed that showed areas of low attenuation in the basal ganglia with no evidence of intracranial haemorrhage (Figure 1). Differential diagnosis included embolic events and dural venous sinus thrombosis. Immediately after the CT examination, the patient suffered two generalised eclamptic fits. Blood pressure was 214/140 mmHg (mean Arterial Pressure = 171) and oxygen saturation was 87%. The patient was intubated and transferred to the intensive treatment unit (ITU) for further management with the diagnosis of PRES.

The patient remained in the ITU for five days and was aggressively treated with hydralazine and labetalol infusions and magnesium sulphate. The patient was extubated the second day and was treated with clonazepam. There was clinical improvement and a magnetic resonance imaging (MRI) done five days following the CT scan showed no abnormalities at all (Figure 2).

The patient was transferred to the postnatal ward and was discharged eleven days post-delivery.

Discussion

PRES syndrome has been associated with a variety of conditions. The main obstetric precipitating factor is preeclampsia-eclampsia [7] with cases of PRES having been observed both postpartum and antepartum [4, 5, 8, 9]. Other associa-

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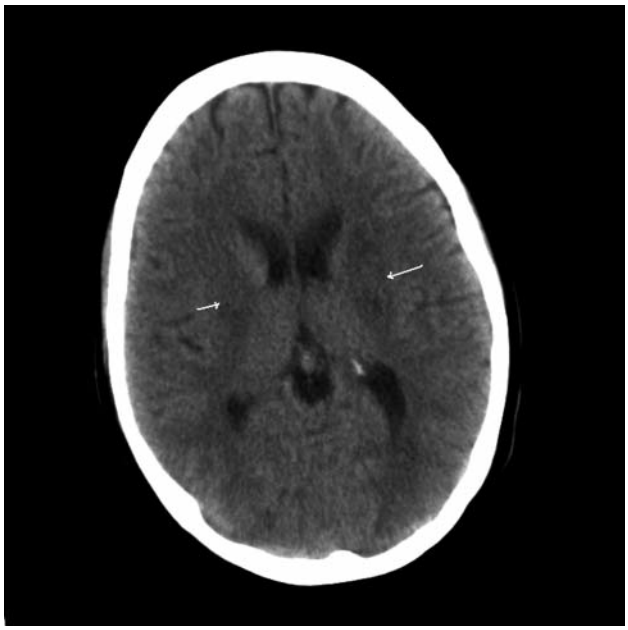

Figure 1. — Transverse head CT scan obtained without intravenous contrast medium on day two postpartum. There are areas of low attenuation (hypodense) in the left ganglionic region and to a lesser extent to the right (white arrows). There is no intracranial hemorrhage. Differential diagnosis includes an embolic event and dural venous sinus thrombosis.

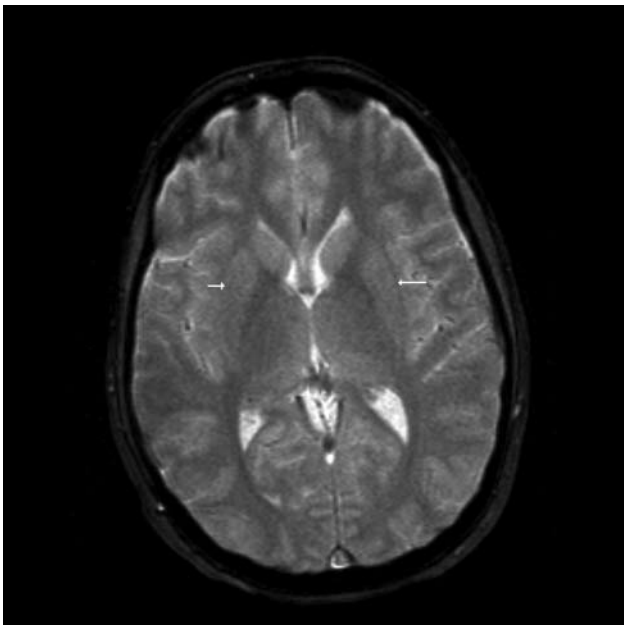

Figure 2. — T2-weighted MRI of the brain from the same patient five days after the head CT scan. There are no parenchymal abnormalities noted and no evidence of infarction or bleeding. The basal ganglia are normal (white arrows). The patient at this stage has clinically improved.

tions highlighted in literature include retained placental fragments leading to delayed eclampsia [10], dural puncture/regional anesthesia [11], infection/sepsis with gram-positive organisms [12], molar pregnancy [9,13] and only one case of twin pregnancy [14]. Even though features of PRES are almost universally present in eclamptic patients [15], it is yet unclear what proportion of eclamptic patients have PRES [6]. Non-obstetric conditions include abrupt hypertension, post-transplantation, autoimmune diseases, acute renal failure, blood transfusion, and immunodepressant drugs [7,16,17]. In a large case-series of n = 113 patients [3], etiologies included hypertension (61%), cytotoxic medication (19%), sepsis (7%) and preeclampsia-eclampsia (6%).

Clinical features of PRES include acute onset headache, visual symptoms, altered mental state, focal neurological deficits, and seizures [3,7,8]. In the largest case-series study of n = 27 women with preeclampsia-eclampsia and PRES [8], symptoms included seizures (62.5%), headache (58%), disturbed vision (50%), altered mental state (12.5%) and focal neurological deficits (4.1%). Laboratory findings indicative of endothelial injury are often seen such as thrombocytopenia and increased lactate dehydrogenase [7].

The "posterior" description in PRES has been suggested because of the involvement of the parietal-occipital lobes (83%-94%) [1-3,16]. Other brain regions include the frontal lobes (54.2%-77%) [3, 16], temporal lobes (64%) [3], cortex (8.3% - 90%) [7, 16], cerebellum (53%) [3], brainstem (27%) [3], and basal ganglia (4.2% - 34%) [3, 6-8, 16]. In preeclampsia-eclampsia, there is a trend for increased basal ganglia involvement [3,8], less severe oedematous lesions, and less occurrence of brain hemorrhage in comparison to other predisposing factors [8].

In the acute setting, diagnosis of PRES is more likely to be made by correlation of head CT findings and clinical features of the patient. Nevertheless, MRI imaging has the ability to detect subtle parenchymal changes [7] and distinguish vasogenic edema from cytotoxic edema that is associated with early onset infarcts [16]. To date, there is no clear correlation between clinical characteristics and severity/location of neuroimaging abnormalities in PRES [9].

The pathophysiology of PRES still remains unclear. An early theory suggests that there is vasoconstriction leading to brain hypoperfusion, subsequent ischemia, and cytotoxic oedema [3,18]. The current theory proposes that severe hypertension leads to a breakdown in cerebral autoregulation and leakage of serum into the cerebral interstitium producing focal vasogenic edema [3, 18, 19]. This vasogenic edema is seen as hypodense CT/MRI lesions [5] particularly in the posterior regions because of relatively less sympathetic innervation in the posterior circulation resulting in less protection against abrupt hypertension [3,9]. A third mechanism suggested is that of endothelial dysfunction in cases of preeclampsia-eclampsia and sepsis, leading to vascular instability, vasoconstriction and hypoperfusion [18].

Early recognition of PRES is vital since reversibility of PRES is not spontaneous but depends on prompt management of hypertension and withdrawal of precipitating factors [4,7,16]. If treated adequately, symptoms typically resolve within seven days [5]. In preeclampsia-eclampsia, there is less severe disease course with a shorter hospital stay and a better outcome in comparison to other etiologies [6].

Recurrent PRES has an incidence of 3.8% [20] and is associated with non-obstetric causes [6,7,20]. Even though recurrent eclampsia has an incidence of two percent [21], no recurrent PRES has yet to be reported in a subsequent pregnancy [6].

This is the second case report on the association between twin pregnancy and PRES. In the presented patient, precipitating factors such as preeclampsia, infection during labour and use of epidural anesthesia were present. Diagnosis was based on clinical and radiological findings and the complete recovery of the patient. Prompt recognition and aggressive treatment led to complete reversibility of this condition.

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