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Posterior Reversible Encephalopathy Syndrome (PRES) Associated with Eclampsia: A Case Study

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ABSTRACT

The occurrence of posterior reversible encephalopathy syndrome (PRES) in patients with eclampsia is a rare condition. PRES is a reversible syndrome characterized by headache, seizure, altered mentation and loss of vision associated with white matter changes on imaging. The lesions in PRES are thought to be due to vasogenic oedema, predominantly in the posterior cerebral hemispheres. This study reports a 16-year-old pregnant woman who presented with blindness and seizure. The MRI of her brain showed abnormal signal intensity in the white matter of the occipital and frontal lobes. She was treated successfully with pregnancy termination, anti-hypertensives, anticonvulsants, and supportive care. It is concluded that early diagnosis is important to prevent permanent neurologic damage and mortality.

Keywords: Posterior reversible encephalopathy syndrome, eclampsia, visual loss, brain MRI

INTRODUCTION

Posterior reversible encephalopathy syndrome (PRES) is a clinic-radiological entity first described by Hinchey, et al. [1,2]. PRES which is also known as reversible posterior leukoencephalopathy syndrome presents with rapid onset of symptoms such as: nausea, headache, altered consciousness, cephalalgia, visual disturbance, cortical blindness, blurred vision, photophobia, hemianopia, and other focal neurologic deficits such as paresis, dysesthesia, or dysphasia as well as seizure [3]. PRES is also a misnomer because the image changes and clinical features may not be limited to the posterior cerebral hemispheres [4]. Also, the reversibility of PRES may be clinically or radiologically incomplete; the condition may be complicated by ischemic or haemorrhagic stroke, and may lead to a chronic seizure disorder or death [5].

The global incidence of PRES is unknown [6]. It has been reported in patients aged 4 to 90 years, most cases occur in young to middle aged adults and death has been reported in up to 15% [7].

PRES occurs in a large array of clinical conditions, predisposing disease, and factors such as toxaemia of pregnancy, arterial hypertension, organ transplantation, autoimmune disease, conditions with renal failure as well as cytotoxic and immunosuppressive medication [3].

Although the underlying pathophysiological mechanisms are still debated, the main hypotheses imply both endothelial dysfunction and failure of cerebral auto-vasoregulation [3].

Image diagnostics such as CT and MRI of CNS performed on patients with pre-eclampsia and eclampsia, have revealed PRES in several cases. MRI, is the golden standard and CT scan only revealed 50% of the lesions. White matter lesions in the occipital lobes, posterior parietal lobes and posterior temporal lobes are classic findings. Lesions may be seen in the frontal lobes, cerebellum, and pons, but seem to be minor and only visible in addition to injuries in the other brain structures mentioned above [8].

This syndrome is oedema without infusion, so early diagnosis resolves the cause and prevents permanent damage and death. This study describes a case of PRES in association with eclampsia in a 37-week pregnant woman.

CASE PRESENTATION

A 16-year-old Iranian woman, primigravida, with 37 weeks of pregnancy, presented with complaints of visual loss, nausea and vomiting to the emergency department. Her pregnancy was wanted and in spontaneous conception but her medical and drug history were unremarkable. Her antenatal blood pressure was reported to be normal, and no neurologic complaints were observed during pregnancy. The patient experienced blurred vision, nausea and vomiting for 12 h. Her blurred vision gradually progressed to blindness with no complaints of headache and seizure.

On examination, her reading was 15/15, as measured on the Glasgow Coma Scale. She had several projectile vomiting in the emergency room. On arrival, her vital signs were: Blood pressure: 160/110 mmHg, Heart Rate: 100 beats per minute, Respiratory Rate: 20 breaths per minute, Temperature: 37°C. On general examination, the following were recorded: Foetal heart rate: 146, Uterine height: 38 cm, Cervix: closed, uterine contractions: negative, Chest: clear on auscultation, abdominal examination: Normal, Her legs: Bilateral 2+ pitting oedema without size differences. On examination of the central nervous system, on fundoscopic examination, both optic discs were normal and there was no focal neurological deficit.

She was admitted and a magnesium bolus of 4 g was given; thereafter, continuous infusion was initiated at 2 g per hour (to prevent seizure). Her blood pressure was controlled by administering labetalol intravenously; systolic blood pressure: 140 mmHg and diastolic blood pressure: 90 mmHg. On investigation, Hgb: 13.1%, Platelet count: 222000 per microliter, Urine Protein: 4+. Liver function test, kidney function test, electrolytes and clotting parameters were normal. The electrocardiogram was normal. In addition, a neurological consult and neuroimaging is ordered. She was managed conservatively in our centre by administering magnesium intravenously and controlling blood pressure, thereafter she was referred to another centre due to the absence of neuroimaging devices and neurologist in our centre on the same day. Differential diagnosis was: PRES, haemorrhagic or ischemic CVA of occipital lobe, and retinal detachment. The initial impression was PRES and the brain MRI revealed abnormal signal intensity in the superior area of both occipital lobes and similar changes in the right frontal lobe (Figures 1 and 2).

After doing brain imaging (2 h after admission), the patient had one episode of generalized tonic-clonic seizure but it was for about one minute. After stabilization, an immediate caesarean section was performed. Surgery was done under general anaesthesia and a male infant weighing 3300 g, with APGAR score of 9 and 10 respectively in the first and fifth minutes was delivered by vertex. The patient was stable during operation and intraoperative hypertension was managed with intravenous labetalol. Thereafter, she was transferred to the ICU for supportive management and close monitoring. She was treated with anticonvulsant (magnesium infusion 2 g per hour) for 48 h and her blood pressure was controlled tightly with intravenous labetalol. Serial laboratory data were normal in the postpartum period. Her retinal examination was normal and did not have retinal detachment. Visual loss improved within 6 days and there was complete recovery of vision (6/6). She was shifted out of ICU and discharged 7 days after operation, symptom free. When discharged home, she did not receive antihypertensive medications. As a result of complete clinical improvement, follow up imaging was not done.

DISCUSSION

Preeclampsia is a systemic syndrome in pregnancy characterized by hypertension and proteinuria. The diagnosis of eclampsia is established if seizure occurs together with preeclampsia which cannot be explained by other causes. The incidence of eclampsia in developed countries average 1 in 2000 to 3000 deliveries [9,10]. PRES with eclampsia is a rare condition and is associated with headache, seizure, altered mentation and visual disturbances. It can present with focal neurologic deficits, mimicking a stroke and can also present atypically. The major causes of this syndrome are severe hypertension, preeclampsia, eclampsia, renal failure, vasculitis, TTP, immunosuppressive treatment, sepsis and hyperammonaemia. The pathogenesis of this syndrome is the inability of the brain circulation to auto regulate, in response to acute changes in blood pressure. Vasogenic oedema occur due to disruption of the blood brain barrier. When unrecognized, irreversible cytotoxic oedema may occur [11].

PRES can present during pregnancy and late in the postpartum period after an uneventful pregnancy. It is usually associated with late eclampsia and a high rate of cortical visual loss in these patients [12]. A high degree of suspicion is required in patients of late postpartum eclampsia because it occurs between 48 h postpartum and 1 month after delivery, frequently in women who have had a normal pregnancy, delivery, and no signs of preeclamptic syndrome [1]. Visual complications in preeclampsia are: hypertensive encephalopathy, retinal detachment, and cortical blindness while visual abnormalities in PRES are: blurred vision, visual neglect, homonymous hemianopia, visual

hallucinations, and cortical blindness [6]. Roth et al. reported a higher percentage of disturbed vision in preeclamptic-eclamptic–related PRES [13]. Patients with PRES due to other aetiologies, more often present with severe symptoms like altered mental status or neurological deficit and lesser visual disturbances [14]. Cortical blindness is a clinical syndrome characterized by intact papillary reflexes and normal fundoscopic finding [4]. The lost vision is usually regained within 4 h to 8 days [15]. In this case, PRES presented with acute visual loss (cortical blindness) and seizure, before delivery at term gestational age and lasted for 6 days. Seizure is recorded in about 92% of cases [15]. Multiple seizures are more common than a single event [4]. In this case, a single seizure occurred.

Since it can be easily misdiagnosed, proper diagnosis requires careful attention to clinical and radiographic presentation [16]. The diagnostic criteria for posterior reversible encephalopathy syndrome are [17]:

1) The presence of neurologic symptoms or findings.

2) Presence of risk factors for PRES.

3) Absence of other possible causes of encephalopathy.

4) Reversible course on follow up.

The radiologic finding is vasogenic oedema, and it is most common in the occipital and parietal regions. The preferential involvement of the parietal and occipital lobes is thought to be related to the relatively poor sympathetic innervation of the posterior circulation. The feature along with predominant involvement of the white matter distinguishes this syndrome from bilateral posterior cerebral artery territory infarction [1].

Brain MRI shows signs of subcortical and cortical oedema with hypersignals in T2/fluid attenuated inversion recovery (FLAIR) or diffusion sequences. In the study by Alexander, et al. [18] in 67 patients with PRES, the incidence of regions of involvement was parieto-occipital, 98.7%; posterior frontal, 78.9%; temporal, 68.4%; thalamus, 30.3%; cerebellum, 34.2%; brain stem, 18.4% and basal ganglia, 11.8%. Divya Karuppannasamy, et al. [12] evaluated the correlation between visual symptoms and imaging abnormalities. They found that bilateral symmetrical vasogenic oedema of parieto-occipital lobe was the most MRI abnormality and no significant differences were seen in the severity of oedema between patients with and without visual loss [12]. In this case MRI showed oedema on occipital and frontal lobes (Figures 1 and 2).

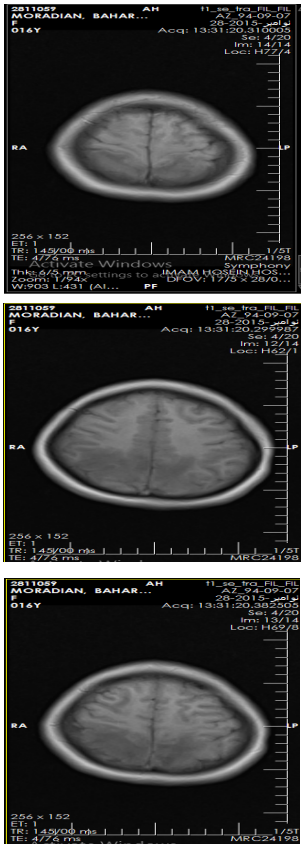

Figure 1 Cerebral magnetic resonance imaging in a patient with PRES. It shows bilateral abnormal signal foci in the occipital and frontal lobes

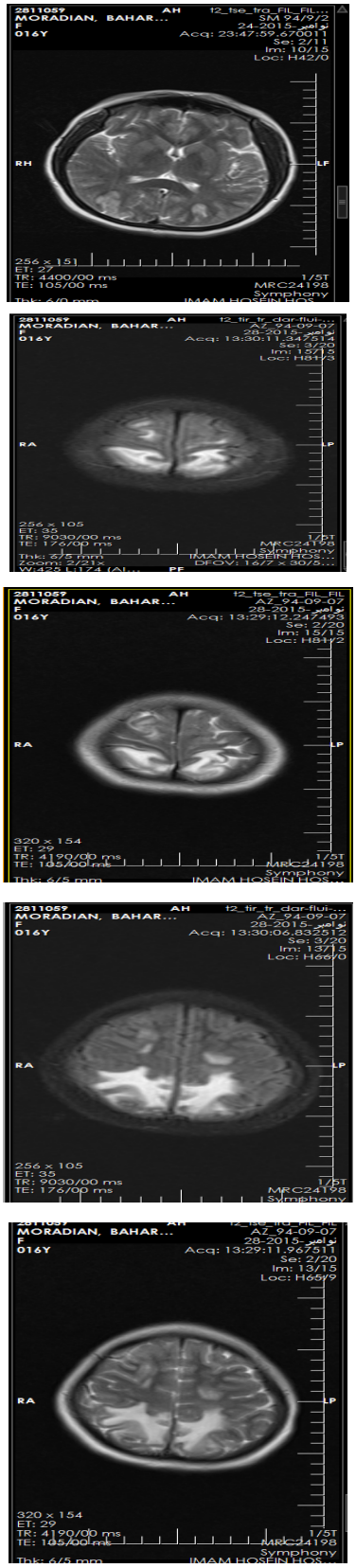

Figure 2 Cerebral magnetic resonance (FLAIR) imaging in a patient with PRES. It shows bilateral hyper-intense signal predominantly in occipital and frontal lobes

To prevent irreversible ischemic injury, early diagnosis and treatment is essential. A high degree of suspicion is needed for diagnosis. The diagnosis of PRES should be considered in all preeclamptic patients, especially in HELLP and eclampsia with neurological symptoms. Clinical symptoms can recover well within several days with early diagnosis and adequate treatment.

The standard treatment of neurologic findings is aggressive blood pressure control and management of underlying pathology [13]. The management of PRES is: ICU admission, continuous evaluation about need for upper airway protection in patients with consciousness impairment or seizure activity, correction of hypoglycaemia and electrolyte imbalance, thiamine in patients with evidence of vitamin B1 deficiency, anti-epileptic treatment, control of hypertension and correction of the underlying cause.

The essence of controlling hypertension is not to normalize the blood pressure but rather to decrease the mean arterial pressure (MAP) by 20% to 25% within the first 2 h [6,19,20]. More rapid blood pressure reduction is not recommended, since it can aggravate alterations in cerebral perfusion pressure and promote ischemia [21]. Intravenous antihypertensive drugs are necessary and appropriate choices include labetalol, nicardipine, or fenoldopam [6,21]. Finsterer, et al. found that controlling blood pressure by nitro-glycerine infusion may worsen PRES [22]. We used labetalol in this case. In most case reports, PRES had dramatic improvement in response to anti-hypertensive drugs, anticonvulsants, and supportive care [15,23-26].

CONCLUSION

This study presented the case of a pregnant woman with PRES. Its diagnosis is clinical and involved neuroimaging. The characteristic imaging findings helped in the diagnosis. Early diagnosis and treatment is essential to avoid permanent complications.

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