Reversible posterior leukoencephalopathy syndrome in pregnancy: a case report

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Summary

Posterior reversible encephalopathy syndrome (PRES), is an acute, neurotoxic state. It is a very rare clinico-neuroradiological entity, and it is a complication of multiple clinical conditions. The association of PRES with toxemia in pregnancy is established. In this article, the authors discuss the case of a 22-year-old woman, gravida 1, 36-week pregnant, with extensive, bilateral white matter hypodensity, predominantly involving the parieto-occipital lobes region. These changes were highly suggestive of posterior reversible encephalopathy. This case report demonstrates that early treatment with control of blood pressure seizures can reverse this condition and also prevent progression to an irreversible damage, thus emphasizing the need for early diagnosis and treatment.

Key words: Reversible posterior leukoencephalopathy syndrome; Hypertension in pregnancy; Eclampsia; Brain edema.

Introduction

Reversible posterior leukoencephalopathy syndrome (RPLS) or posterior reversible encephalopathy syndrome (PRES) was first described by Hinchey in 1996 [1].

The syndrome is acute with diverse clinical presentations and characteristic computed tomography (CT) scan or magnetic resonance imaging (MRI) features.

This clinico-neuroradiological entity is a complication of multiple clinical conditions: hypertension, pre-eclampsia and eclampsia, renal failure, therapy with immunosuppressant or high dose of cytotoxic medications (cyclosporin A and tacrolimus) for autoimmune disease, and allogeneic bone marrow or organ transplantation. Other clinical conditions are characterized by uraemia and porphyria [2]. The association of PRES with toxemia of pregnancy is established [3].

The clinical hallmarks of this syndrome are: headache, altered mental functioning, seizures, and loss of vision associated with white matter changes. This changes are suggestive of edema mainly in the posterior regions of the cerebral hemispheres, but also involving the brainstem, cerebellum, and other cerebral areas [4]. The findings on neuroimaging in PRES include non-enhancing white matter abnormalities that appear as areas of low attenuation on CT scan and appear hypointense on T1-weighted imaging and hyperintense on T2-weighted MRI. The lesions are mainly seen in the posterior regions of the cerebral hemispheres. These abnormalities partially or completely resolve on follow-up scanning, thereby, suggesting subcortical edema without infarction [5].

The white matter is composed of myelinated-fiber tracts in a cellular matrix of glial cells, arterioles, and capillaries that makes it susceptible to the accumulation of fluid in the extracellular spaces [4]. It is suggested that vertebro-basilar territory, owing to its relatively sparse sympathetic innervation, may experience preferential disruption of autoregulatory mechanisms, leading to increased perfusion and edema [6].

Case Report

A 22-year-old woman, 36-week pregnant, weighing 63 kg, gravida 1, presented to the present department after she experienced headache, blurring of vision, and acute onset of generalized seizure.

The results of her general examination were unremarkable. Blood pressure was 140/90 mmHg with a heart rate of 95 beats per minute. Respiratory rate was 17 breaths per minute with an O_2 saturation of 99%. Body temperature was 36.3°C. Electrocardiogram was normal.

Her investigations included: haemochrome, serum electrolytes, serum calcium, serum magnesium, liver function tests, and coagulation profile were within normal limits. Biochemical values were: Hgb 11.7 g/dl, Htc 37.2%, WBC 13,130/mm^3; PLT 220,000/mm^3, AST 29 U/l, ALT 32 U/l, amylase 26 U/l, LDH 450 U/l; her coagulation parameters were: prothrombin time (PT): 113%, activated prothrombin time (APTT): 26 sec, INR 0,97. serum level of sodium was 138 mmol/l, potassium 3.6 mmol/l, and calcium 9.0 mmol/l. Renal function test and urine analysis were normal.

A diagnosis of eclampsia was made and the patient was transferred to the operating room where the patient underwent an emergency lower segment cesarian section under spinal anaesthesia. She gave birth to a healthy baby with a five- minute Apgar score of 9. Postoperatively she was transferred to the medical intensive care unit.

She underwent an invasive monitoring of vital parameters, assisted ventilation, neurological counselling, brain and thorax CT scans, and spinal tap. At the time, she was treated with nifepidine and fenobarbital.

Neurological examination, lumbar puncture, and thorax CT was normal. Brain CT showed extensive, bilateral white matter

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hypodensity, predominantly involving the parieto-occipital lobes region. These changes were highly suggestive of posterior reversible leukoencephalopathy. However atypical imaging findings can at times be misleading. On follow-up examination, patient showed marked clinical improvement with control of hypertension and was discharged in stable condition, as also confirmed by imaging . She was discharged from hospital on the nine post-operative day. At one month follow-up, the CT was completely normal.

Discussion

PRES is a very rare clinical entity. The differential diagnosis for seizures in pregnancy period includes: eclampsia, subarachnoid haemorrhage, intracerebral haemorrhage, thrombotic phenomena, intracranial neoplasm, head trauma, idiopathic epilepsy, infection (meningoencephalitis), and amniotic fluid embolism. PRES is still an under-recognised and untreated condition and the clinic-radiological hallmarks are to be established. There are no consensual guidelines to validate diagnosis of PRES [8].

Two theories have been proposed to explain the pathophysiology. The more popular theory suggests that hypertension leads to failure of autoregulation, subsequent hyper-perfusion, and vasogenic edema. The other theory suggests that vasoconstriction and hypoperfusion leads to brain ischemia and subsequent vasogenic edema [7].

PRES is a clinico-radiological entity. The combination of suggestive clinical manifestation and radiological criteria establishes the diagnosis of PRES.

PRES is reversible after appropriate treatment, which makes it important to recognize and treat the etiology to prevent its progression to irreversible damage.

This case report demonstrates that early treatment with control of blood pressure seizures can reverse this condition and also prevent progression to irreversible damage, thus emphasizing the need for early diagnosis and treatment [9, 10].

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