REVERSIBLE POSTERIOR LEUKOENCEPHALOPATHY SYNDROME IN PRE-ECLAMPSIA - A CASE REPORT

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ABSTRACT

BACKGROUND
A 26-year-old primigravida with 34-week pregnancy admitted in Medicine ICU with complaint of recurrent seizure, progressive bilateral visual loss with rapid deterioration of sensorium. Examination revealed accelerated hypertension, proteinuria and cortical blindness. Patient was managed with anti-decongestive, antihypertensive, antiepileptic and induction of labour. She delivered a still born baby. She regained consciousness 48 hours after seizure control with complete visual restoration at 3 weeks.

KEYWORDS
PRES - Posterior Reversible Leukoencephalopathy Syndrome, CT Computed Tomography, MRI - Magnetic Resonance Imaging.


BACKGROUND
Although visual disturbances are quite common, complete blindness is rare with an incidence of 1 - 3% in patients of preeclampsia and eclampsia.1 Blindness in preeclampsia/eclampsia syndrome can be due to the involvement of the occipital cortex, retina or optic nerve. Most cases of blindness in preeclampsia and eclampsia were commonly attributed to retinal pathology including vascular abnormalities, oedema or detachment and acute ischaemic optic neuropathy as a result of decreased blood supply to the pre-laminar portion of the optic nerve. Cortical blindness is another leading cause of blindness in toxemia of pregnancy.2

Reversible posterior leukoencephalopathy syndrome (PRES) is a clinical and radiological syndrome, first described by Hinchey et al in 1996.[1] The main causes include hypertensive crisis, toxemia of pregnancy, renal failure, fluid retention and some immunosuppressive drugs toxicity, post-transplantation stage of liver diseases, haemolytic uraemic syndrome, acute intermittent porphyria, malignancies, vasculitis, transfusion and erythropoietin, oxybutynin or intravenous immunoglobulin (IVIG) treatment.3[3][4][5]

The most common clinical manifestations of PRES are seizures, headache, nausea and vomiting, altered mental status, decreased alertness, cortical blindness and transient motor deficits. The main finding in neuroimaging is posterior white matter oedema, which is predominating in the occipital and parietal lobes and posterior fossa structures.[4]

CASE HISTORY
A 26-year-old primigravida, at 34 week of pregnancy presented with persistent headache, nausea for last 3 weeks and progressive visual loss for last 7 days and she had 2 episodes of generalised tonic-clonic seizure with progressive cognitive decline for last 3 days.

Physical examination revealed an unconscious patient with GCS-M5E2V3, pulse rate of 90/minute, BP of 200/120 mm Hg and bilateral pedal oedema. CNS examination - grade 4 hypertensive retinopathy with lack of meningeal signs or focal neurological deficit. Per abdominal examination - fundal height - 34 weeks with longitudinal lie in cephalic presentation with non-audible foetal heart rate with stethoscope.

Haematological Investigation
Serum bilirubin - 2.2 mg/dL, SGOT - 92 IU/L, SGPT - 102 IU/L, Creatinine - 1.2 mg/dL, Urea - 36 mg/dL, Urine Protein - ++

Imaging
USG - Single dead intrauterine pregnancy of 32 weeks gestational age. Immediate CT scan (head) revealed bilateral white matter hypodensities in parieto-occipital regions. MRI revealed hyperintensity in bilateral parieto-occipital region in T2 weighted and FLAIR sequences. Repeat MRI brain after 3 weeks showed significant resolution of white matter lesions.

Treatment
She was managed with injection phenytoin, mannitol and lasix with antihypertensive measures. The labour was induced and she delivered a stillborn male baby of 1.4 kg. She regained consciousness after 48 hours of last ictus. Her BP became normal 2 weeks after delivery.

Followup
Patient’s vision also improved completely (6/6 both eye) at 3 weeks with normalisation of blood pressure.
Hinchey et al while drawing attention to a reversible posterior leukoencephalopathy syndrome associated with hypertensive encephalopathy, eclampsia and immunosuppressive drugs such as cyclosporine as major aetiologies. In these case reported here, patient developed RPLE following eclampsia, which is a well-known and important aetiological factor for this syndrome. In this patient, clinical findings was generalised seizures and visual abnormalities in the form of cortical blindness. Abrupt hypertension as a cause of RPLE syndrome, is a result of capillary leak syndrome. Neuroimaging in RPLE syndrome include non-enhancing white matter abnormalities that appear as areas of low attenuation on CT scan and appear hypointense on T1WI, MRI and hyperintense on T2WI. The lesions are seen mainly in the posterior regions of the cerebral hemispheres. These abnormalities partially or completely resolve on followup scanning, thereby suggesting subcortical oedema without infarction.

It is important to note that neuroimaging usually reveals sparing of the calcarine and paramedian occipital lobe structures, a fact that distinguishes RPLE from bilateral infarction of the posterior cerebral artery territory, significant reversal of neuroradiological abnormalities coupled with complete clinical recovery suggests the diagnosis. Sudden elevation of blood pressure disrupts the autoregulatory mechanisms of CNS vasculature, leading to development of areas of vasoconstriction and vasodilatation, breakdown of blood brain barrier, focal transudation of fluid and petechial haemorrhages. Hypertensive encephalopathy and eclampsia share similar pathophysiology. Intensive investigations and long-term antiepileptic therapy is not warranted as RPLE is essentially reversible.

CONCLUSION
Reversible posterior leukoencephalopathy syndrome (PRES) is a clinical and radiological syndrome characterised by seizures, headache, nausea and vomiting, altered mental status, cortical blindness and transient motor deficits with unique neuroimaging feature consisting of posterior white matter oedema involving parieto-occipital lobes and posterior fossa structures.

REFERENCES