Case Report:
A Rare Case of Reversible Encephalopathy Syndrome Accompanying Late Postpartum Eclampsia or Hypertensive Encephalopathy-A Clinical Dilemma

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Abstract: Posterior Reversible Encephalopathy Syndrome (PRES) refers to a clinic-radiologic diagnosis. Clinically it is characterized by non specific symptoms such as headache, confusion, visual disturbances and seizures. The radiological findings in PRES are thought to be due to vasogenic oedema, predominantly in the posterior cerebral hemispheres, and are reversible with appropriate management. We report a case of reversible encephalopathy diagnosed by MRI scan occurring in atypical areas like the caudate and lentiform nuclei of the brain following an uneventful lower segment caesarean section in a normotensive patient, who was successfully treated with anti-hypertensives, anticonvulsants and supportive treatment. The differential diagnosis of convulsions in the post-partum period is discussed.

Key Words: PRES; Vasogenic edema; Postpartum; Normotensive.

Introduction:
Posterior reversible encephalopathy syndrome (PRES) is a well-recognized, clinical and neuro-radiological entity first described in 1996 by Hinchey et al.(1) This is characterized by headache, vomiting, confusion, seizures, visual abnormalities and motor signs. These transitory neurological disturbances are thought to be due to cerebral vasospasm causing ischemia of the involved territory. The ensuing cerebral ischemia has a characteristic imaging pattern on MRI scan. It is associated with a variety of underlying conditions including hypertensive encephalopathy, preeclampsia, systemic lupus erythematosus, thrombotic thrombocytopenia purpura, treatment with immunosuppressants, renal failure and central nervous system infections. Importantly, these changes appear to be completely reversible if the underlying cause is treated or the precipitating drug withdrawn early in the clinical course.(1) We report a case of reversible encephalopathy syndrome, occurring in the post partum period managed successfully.

Case Report:
A 24 years old multi-parous lady underwent an elective lower segment caesarean section under spinal anaesthesia. She was normotensive at presentation and had no other contributing risk factors. Her blood pressure was within normal limits throughout the surgery and in the post-operative recovery period.

On the fourth postoperative day she complained of bifrontal headache which was constricting in type especially on standing, suggestive of post spinal headache. Her blood pressure was in the normotensive range and she responded to paracetamol with domperidone combination and good hydration.

On the fifth postoperative day she developed severe headache along with visual disturbances followed by generalised tonic and clonic convulsions and postictal confusion. Her blood pressure was 220/120 mm Hg. A provisional diagnosis of postpartum eclampsia was thought off. Blood pressure could not be controlled even after starting her on magnesium sulphate and calcium channel blockers. She was shifted to medical intensive care unit by the physicians who suspected post partum cerebral venous thrombo-embolism. Her investigations including serum electrolytes, serum calcium, serum magnesium, liver function tests and coagulation profile were within normal limits. There was no evidence of urinary proteinuria. Her Serum uric acid was raised, which measured 9.8 mg/dl with changes in serum creatinine-3.4 mg/dl and blood urea- 77mg/dl. Antinuclear antibody evaluation was negative. Fundoscopy did not reveal any papilloedema. CT brain and MRI venogram (Figure-1) were normal.

On brain MRI scanning the T2 weighted axial image showed hyperintense areas within the cerebellar hemispheres, gray and subcortical white matter of bilateral frontal, parietal, occipital and temporal regions, left lentiform nucleus and right caudate nucleus suggesting the possibility of posterior reversible encephalopathy syndrome(Figure 2 and 3).

She was started on anti-hypertensives, anticonvulsants, anti-edema measures, proton pump inhibitors and antibiotics. The patient stabilized by the fourth day. Her neurological symptoms completely resolved by 7 days. She was discharged on anti-hypertensive and anti-convulsive medications. She completed her treatment and followed up for almost 15 months and was free of neurological symptoms.
Discussion:
The differential diagnosis for seizures in the late post-partum period includes eclampsia, subarachnoid haemorrhage, intracerebral haemorrhage, thrombotic phenomena, intracranial neoplasm, head trauma, idiopathic epilepsy, infection (meningoencephalitis), amniotic fluid embolism, postpartum angiopathy.(1,2)

There was no past history of epilepsy or head injury. Infection due to dural puncture was a possibility, but the total count was 11,500 cells per cu mm which was within normal limits. Post dural puncture spinal headache usually presents within 24 to 48 hours. Brain MRI with venogram ruled out intracranial bleed, ischaemia secondary to thromboembolism, vasospasm or space occupying lesion. Amniotic fluid embolism rarely occurs after 48 hours post partum and generally presents with cardiopulmonary collapse and coagulopathy which was not seen in our patient. Possibility of systemic lupus erythematosis with renal involvement was considered, but the Anti-nuclear antibody evaluation was negative.

An alternative explanation is the possibility of post-partum angiopathy. This diagnosis should be considered in a post-partum patient with hypertension and headache but no proteinuria as it was in this case. Post-partum angiopathy is a form of reversible cerebral segmental vasoconstriction characterised by severe “thunderclap” headaches, seizures, focal neurological deficits and segmental narrowing and dilatation of large and medium sized arteries. Typically, scanning reveals ischaemic lesions but MRI findings consistent with reversible posterior leucoencephalopathy syndrome have been reported.(1)

Posterior reversible encephalopathy syndrome mostly secondary to late post partum eclampsia was suggested, as she presented with headache, followed by seizures , with raised serum uric acid levels 9.8 mg/dl (normal 3.8-7.0 mg/dl), hypertension without proteinuria, in uremia mostly secondary to hypertensive encephalopathy itself. The clinical dilemma we faced was: is it eclampsia or hypertensive encephalopathy due to other reasons? What is the ideal treatment? With multi-disciplinary approach we were able to scale down the differential diagnosis. MRI was typically consistent with vasogenic oedema, but which was involving the atypical areas like cerebellum, frontal, left lentiform nucleus and the right caudate nucleus along with typical areas, bilateral occipital, parietal and temporal. Usually it involves the posterior occipito-parietal lobes and hence the nomenclature posterior reversible encephalopathy syndrome.(3)

PRES is still an under recognised and untreated condition. The incidence in the peripartum setting is not known. After reviewing the literature we could get access to 18 cases reported out of which 3 cases manifested as late post partum eclampsia and only one among them was normotensive with no abnormality in haemogram or biochemical findings before the onset of convulsions.(4,5) So this is the second case reported in the English literature, in a normotensive multiparous lady following uneventful caesarean section where post dural puncture headache was not contributory, presenting as reversible encephalopathy syndrome involving atypical areas like caudate nucleus and lentiform nucleus accompanied with late postpartum eclampsia or hypertensive encephalopathy. Due to prompt intervention and supportive therapy, this woman recovered within a week and did not have any neurologic deficits at the time of discharge. She continued with antihypertensives and anticonvulsants and stopped these medications after consultation and counselling by neurologist and was clinically stable for the last 15 months.

Postpartum atypical reversible encephalopathy syndrome (PRES) following an uneventful pregnancy is a very rare clinical entity. Recognition at the earliest and prompt initiation of the supportive measures can prevent permanent neurologic damage and thereby the associated morbidity. Multidisciplinary care forms the corner stone to achieve a safe motherhood in these women.
References: