

Posterior Reversible Encephalopathy Syndrome in a Parturient Posted For Cesarean Section

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Abstract: We report a 28-year-old primigravida who presented in 34 weeks of pregnancy with seizures, unconsciousness, hemiparesis and blindness, scheduled for emergency caesarean section. She was managed under general anaesthesia and shifted to intensive care unit. Postoperative computed tomography brain revealed an intra-axially podensity involving predominantly white matter regions of bilateral parieto-occipital lobes, caudate nucleus and cerebellum, suggestive of PRES. MRI confirmed the diagnosis. Clinical improvement with complete resolution of visual disturbances was observed with supportive treatment. The importance of prompt suspicion and management in preventing short- and long-term neurological deficits in reversible condition like PRES is highlighted.

Keywords: Leukoencephalopathy, posterior leukoencephalopathy syndrome, posterior reversible encephalopathy syndrome.

I. INTRODUCTION

Posterior reversible encephalopathy syndrome (PRES) is characterized by headache, altered mental status, visual disturbances, and seizures. Radiological features typically include edema of the posterior cerebral regions, especially of the parietooccipital lobes. Atypical imaging features, such as involvement of anterior cerebral regions, deep white matter, and the brain stem are also frequently seen. Vasoconstriction is common in vascular imaging. Different conditions have been associated with PRES, but toxemia of pregnancy, solid organ or bone marrow transplantation, immunosuppressive treatment, cancer chemotherapy, autoimmune diseases, and hypertension are most commonly described. The pathophysiology of PRES is unclear and different hypotheses are being discussed. Posterior reversible encephalopathy syndrome is best managed by monitoring and treatment in the setting of a neurointensive care unit. The prognosis is usually benign with complete reversal of clinical symptoms within several days, when adequate treatment is immediately initiated. Treatment of severe hypertension, seizures, and withdrawal of causative agents represent the hallmarks of specific therapy in PRES. Delay in diagnosis and treatment may lead to permanent neurological sequelae. Therefore, awareness of PRES is of crucial importance for the intensivist.

II. CASE STUDY

A 28-year-old primigravida at 34 weeks gestation, weighing 50 kg, presented in unconscious condition. Her relatives give history of acute onset headache, loss of vision, altered sensorium and one episode of seizure following which she was taken to primary health care centre from where she was referred to our hospital labeling as eclampsia. On her way to hospital she experienced 3 more episodes of generalized tonic clonic seizures in one hour. There was no past history of hypertension, cardiac diseases, vision abnormalities or seizures. She was treated with with IV magnesium sulphate (MgSO₄) and posted for emergency cesarean section in view of variable fetal heart sounds. Patient had 2 more episodes of GTC in ward. On arrival, in preop room the patient was unconscious with Glasgow Coma score of 7/15. She was moving right upper and lower limbs to painful stimuli but not left indicating hemiparesis of that side. There was bilateral pedal oedema and Blood pressure was 170/110mmHg with a heart rate of 100 beats per minute. Her respiratory rate was

16 breaths per minute and chest was clear on auscultation. Room air saturation was 98%. Pupils were equal and reactive to light. Plantar reflex was upgoing on left. Other system examinations were normal. Complete blood picture (Hb of 10.8 g %), renal and liver function tests, clotting parameters and electrocardiogram were normal. Urine analysis revealed proteinuria 2+. In the preop room and on operating table she developed 2 more episodes of generalised tonic-clonic seizure, following which her trachea was intubated with rapid sequence induction using thiopentone and succinylcholine. Anaesthesia was maintained with oxygen, nitrous oxide and isoflurane. Intraoperative hypertension was managed with IV labetalol 10 mg with non invasive blood pressure monitoring. A 2.2 kg baby with APGAR score 9 was extracted and transferred to a neonatal intensive care unit. Analgesia was achieved with 100 µg of IV fentanyl, sedation with 1 mg midazolam and muscle relaxation with inj atracurium 20 mg. A total of 1000 ml of Ringer's lactate was infused during the entire procedure which lasted for 45 minutes. Urine output was 200 ml introp and blood loss was approximately 500ml-600ml. In immediate postop period she started responding to verbal stimuli with spontaneous eye opening. But response was sluggish and was not consistent. Postoperatively, she was shifted to an intensive care unit (ICU) for planned mechanical ventilation. Medical management included magnesium sulphate, labetalol, mannitol. Neurology consultation was obtained in the immediate postoperative period and computed tomography (CT) brain revealed an intra-axial hypodensity involving predominantly white matter regions of bilateral parieto-occipital lobes. MRI with T2 weighted image confirmed the diagnosis.

Her trachea was extubated as her sensorium, vision, cardiorespiratory parameters improved on the second postoperative day. On the third postoperative day, there was complete recovery of vision (6/6) as well as motor and sensory system. Then she was shifted to ward. Rest of her hospital stay was uneventful till she was discharged on 7th postop day.

III. DISCUSSION

A primigravida presenting at term with seizures and blindness puts every clinician in dilemma of varied possibilities. The possibilities could be cerebrovascular accidents complicating pregnancy, eclampsia and clinical syndromes like PRES.

PRES constitutes a clinical syndrome characterized by headache, altered mental status, seizures. Altered mental status could range from lethargy, somnolence, restlessness, agitation, confusion to stupor and coma. Multiple seizures over shorter span are more common as compared to eclampsia.

Multiple theories have been proposed on the pathophysiology of PRES, the most accepted being the vasogenic oedema. Cerebral autoregulation maintains a constant blood flow to the brain despite alterations in the systemic pressures. Once this mechanism gets disrupted, increased perfusion pressure is sufficient to overcome the blood-brain barrier, allowing extravasation of fluid, macromolecules and even red blood cells. So, PRES represents vasogenic rather than cytotoxic oedema in the majority of cases. But cytotoxic edema is also causative factor as PRES has been described with cytotoxic drugs.

Hinchey et al. first reported a 'reversible posterior leukoencephalopathy syndrome' in 1996 [2]. This name was superseded by 'posterior reversible encephalopathy syndrome' in 2000, which is now the most widely accepted terminology [1]. Most cases of PRES are associated with hypertensive disorders, particularly those of pregnancy. In most obstetric cases, there is a history of pre-eclampsia and, unlike in the case reported here, PRES usually develops only after delivery. It has also been described in normotensive patients in association with drug-induced or HIV-associated immunosuppression, with thrombotic thrombocytopenic purpura / haemolytic uraemic syndrome and with acute intermittent porphyria [3-6].

Onset of symptoms is sudden and acute and include blindness, generalised or focal seizures, impairment of consciousness. The presentation described here of complete loss of consciousness without a contributory history is unusual. Altered mental status could range from lethargy, somnolence, restlessness, agitation, confusion to stupor and coma. Multiple seizures over shorter span are more common as compared to eclampsia as seen in our case. Blindness in severe preeclampsia is mostly associated with cortical aetiology. Cortical blindness is a clinical syndrome characterized by intact papillary reflexes and normal fundoscopic findings. The lost vision is usually regained over a period of a week. [15]

Clinical improvement always follows the treatment of elevated blood pressure and withdrawal of offending agents. Magnesium therapy should be initiated as soon as eclampsia or PRES in pregnancy is suspected, as it treats both seizures and hypertension. Finsterer and associates found that treating pre-eclamptic patients with nitroglycerin infusion needs caution, as it may worsen PRES. [7] Distinguishing PRES from acute stroke is especially important to guide treatment.

Mild-to-moderate hypertension should not be treated in ischaemic stroke, but must be managed aggressively in PRES [8] Early treatment usually results in complete reversal of the deficits over few days to several weeks.

CT findings are of bilateral low attenuation changes predominantly in the white matter of the parieto-occipital region, and may lead to the diagnosis. [9]MRI, however, is the imaging modality of choice as it differentiates subcortical vasogenic oedema of PRES from the cytotoxic oedema of acute cerebral infarction. This is seen as increased T2 or fluid-attenuated inversion recovery (FLAIR) sequence signal without reduction in apparent diffusion coefficient (ADC) signal on diffusion-weighted imaging. [10] Some recommend MRI not only for symptomatic patients with suspected PRES, but also for asymptomatic patients with severe pregnancy-induced hypertension so that urgent delivery can be carried out [14]. But financial restraints may prevent this in many developing countries.

Earlier term 'leukoencephalopathy' suggesting that only white matter is involved is quite misleading since grey matter lesions are present in as many as 94% of cases. Nonetheless, 'posterior reversible encephalopathy syndrome' is also a misnomer, in several respects. Firstly, the imaging and clinical features may not be limited to the posterior cerebral hemispheres. The brain stem is involved in more than half of cases and the anterior and middle cerebral artery territories are involved in over 60%. Posterior circulation changes do, however, predominate, most likely due to the relative lack of efferent sympathetic innervation in this region [11]. Also, reversibility of PRES may be clinically or radiologically incomplete [12], and the condition may be complicated by ischaemic or haemorrhagic stroke, and may lead to a chronic seizure disorder or death. Narbone et al. [13] have argued therefore that 'potentially' should replace 'posterior' in the term.

IV. SUMMARY

Though the association of PRES a rare neurooradiological syndrome and pregnancy induced hypertension is well documented, the cause and effect relationship is not well established. A pregnant patient presenting with seizures and blindness may not necessarily eclamptic; possible presence of PRES must need a consideration. This case report emphasizes the need for early diagnosis and prompt treatment of PRES to prevent short- and long-term neurological sequelae such as brain damage, haemorrhage and infarction.

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