

CASE REPORT

Rare presentation of posterior reversible encephalopathy syndrome (PRES) and neurogenic stunned myocardium in a patient after emergency cesarean section

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ABSTRACT

Posterior Reversible Encephalopathy Syndrome (PRES) is clinically characterized by seizures, lethargy, nausea and visual impairment. These findings are thought to be due to vasogenic edema, predominantly in the posterior cerebral hemispheres and are reversible with appropriate management. Neurogenic-stunned myocardium is a syndrome of reversible left ventricular dysfunction, associated with excessive sympathetic discharge states like pheochromocytoma, high-grade subarachnoid hemorrhage (SAH), status epilepticus and significant emotional stress. Here, we report a case of PRES and Neurogenic-stunned myocardium occurring simultaneously in a 25 year old primigravida at 34 weeks of gestation with twin pregnancy who presented to the emergency department with eclampsia and fetal distress. A careful review of literature did not return any report where these two conditions co-existed in an obstetric patient.

Key words: Posterior Reversible Encephalopathy Syndrome; PRES; Neurogenic stunned myocardium; Eclampsia; Cesarean section

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INTRODUCTION

Posterior reversible encephalopathy syndrome (PRES) was first described in 1996 by Hinchey et al¹ as a clinical syndrome characterized by confusion or decreased level of consciousness, lethargy, nausea, headache, visual changes, and seizures. The pathogenesis of PRES is still unclear.⁵ The obstetric causes of PRES include preeclampsia, eclampsia and HELLP syndrome.⁴ Sepsis, use of immunosuppressive drugs, history of renal and autoimmune diseases, HIV syndrome, acute intermittent porphyria, organ transplantation and hypertensive encephalopathy are among the other causes of PRES.⁵ Early diagnosis and treatment of PRES helps to prevent secondary complications and may decrease mortality and morbidity.¹ The

myocardial injury and dysfunction occurring as a result of imbalance of autonomic nervous system is defined as neurogenic stunned myocardium.⁶ There is a reversible left ventricular dysfunction as a result of excessive sympathetic discharge states, like diverse types of acute brain injury, status epilepticus, high-grade subarachnoid hemorrhage (SAH), acute hydrocephalus, pheochromocytoma, and significant emotional stress.²

In this case report, we present a case of a 25 year old primigravida at 34 weeks of gestation with twin pregnancy, admitted through the emergency department with symptoms suggestive of eclampsia. She underwent an emergency cesarean section due to fetal distress. Postoperatively, she was shifted to the intensive care unit (ICU) due to deterioration in the level of consciousness; subsequent workup

revealed a severely reduced systolic function, deranged liver function and MRI findings suggestive of PRES.

CASE REPORT

Written consent was obtained from the patient for disclosure of her clinical information to the scientific journal. A 25 year old primigravida with no prior known co-morbid, presented to the emergency department with severe headache and an episode of convulsions at 34 weeks of gestation with twin pregnancy. On admission to the emergency room her heart rate was 140 beats per min, respiratory rate was 28 breaths/min, non-invasive blood pressure (NIBP) was 169/110 mmHg, oxygen saturation 98% on 5 lit of oxygen through face mask and core temperature was 37°C. On auscultation, her chest was clear. Neurological examination revealed a GCS of 15/15, but she was agitated and combative, pupils were bilaterally equal and reactive to light and deep tendon reflexes were brisk. Initial laboratory workup showed hemoglobin of 13.9 g/dl with hematocrit of 41%, platelets count of 19,400/mm³, total leucocyte count 11.6/mm³ and random blood sugar level was 71 mg/dl. Renal and liver function tests were within normal range. She was immediately taken to the operating room for an emergency cesarean section due to fetal distress. In view of the previous history of an eclamptic seizure, surgery was conducted under general anesthesia. A baby girl (weight 1700 g) and a baby boy (weight 2100 g) were born with an Apgar score of 3 and 4 respectively at first min and 8 and 7 at ten min. The surgery remained uneventful with an estimated blood loss of 700 ml. Patient was successfully extubated upon meeting extubation criterion. In the postanesthesia care unit (PACU) she became disoriented and agitated. On examination her GCS was 12/15, SpO₂ was 99% on 5 lit/min oxygen by face mask, blood pressure 130/90 mmHg and heart rate 130 beats/min. Within 30 min of shifting to PACU her GCS dropped to 6/15. Arterial blood gas analysis showed a pH 7.35, PCO₂ 25.1, PO₂ 65.7, HCO₃ 13.8, B.E -9.7 SaO₂ 91.6% (at 5 lit/min oxygen supplementation by Hudson mask). In view of deterioration in her level of consciousness and hypoxia, she was intubated and shifted to ICU for hemodynamic and neurologic monitoring and supportive care. In the ICU she remained stable and tolerated gradual weaning from the ventilatory support, but her neurological status did not improve and a neurology consult was requested. MRI findings were suggestive of PRES. Further laboratory workup revealed deranged liver

function tests (SGPT= 2437 IU/lit, ALP= 232 IU/lit) and low platelets (53000/mm³). Echocardiogram showed severely reduced left ventricular systolic function and visually estimated ejection fraction of approximately 20% along with wall motion abnormality. She was given supportive treatment and gradually regained consciousness in the next 48 hours and was extubated and shifted to step down special care unit after 72 hours of ICU stay. She was discharged on 5th post-operative day in stable condition with normal cardiac function and without any neurological sequel.

DISCUSSION

The co-existence of posterior reversible encephalopathy syndrome with neurogenic stunned myocardium has been reported only once in the literature in a female patient with chronic backache and to date remains an extremely rare combination of medically significant systemic involvement.² To the best of our knowledge the coexistence of these two diverse entities in an obstetric patient is still a first to be reported. Both posterior reversible encephalopathy syndrome and neurogenic stunned myocardium have a diverse spectrum of symptoms which need to be accounted for the proper management of this potentially lethal combination. PRES is associated with characteristic neuroimaging findings of posterior cerebral white matter edema.⁷ However, it is often not confined to either the white matter or the posterior regions of the brain which is consistent with the findings in this case where the MRI showed not only the involvement of the occipital lobes but also the frontal and the parietal lobes. The preponderance of this condition in females is a known fact and should be considered in patients on immunosuppressive therapies, renal disease and hypertensive disorders.⁵ PRES is associated with disordered cerebral autoregulation and endothelial dysfunction, however, the precise pathogenesis still remains unclear.³ PRES is a clinico-radiographic diagnosis and there are no specific diagnostic criteria. Headache is not localized, is constant, moderate to severe, and unresponsive to analgesia.⁸ Conscious level may range from mild somnolence to confusion and agitation, progressing to stupor or coma. The neurologic syndrome may be preceded by hypertensive crisis. Although the patient had stable hemodynamics preoperatively, the alteration in the GCS could be a delayed response to the hypertensive, eclamptic episodes. The exact duration from hypertensive crisis to progression to a full blown neurological compromise is unknown. In this patient the evidence of myocardial

dysfunction was confirmed by an echocardiogram which showed severely reduced systolic function along with akinesia of the anterior and posterior segments with only the basal segments contracting with preserved function. The concurrent presence of mildly elevated cardiac enzymes suggested the diagnosis of neurogenic stunned myocardium which can further be extrapolated to be linked with the hypertensive disorder of the pregnancy that could have potentially lead to the co-existence of these diverse entities.

Treatment recommendations are somewhat limited because the available evidence is limited to case reports and case series. Management is based on control of seizures, hemodynamic support and supportive treatment. The therapeutic effect of MgSO₄ in eclampsia is multifactorial.⁹ It has central anticonvulsant activity and acts by decreasing the peripheral vascular resistance and prevents vasoconstriction, preserves blood-brain barrier and prevents the development of cerebral edema. Use of antihypertensive drugs, like labetalol, nicardipine and hydralazine, is recommended.¹⁰ Intravenous dexamethasone is also being used especially in patients with intracranial masses as it decreases vasogenic peri-tumor oedema.¹¹

Differential diagnosis for PRES syndrome

includes acute cerebrovascular accident, meningoencephalitis, demyelinating lesions of the brain, cerebral venous thrombosis, electrolyte imbalance (hyponatremia) and hypoglycemia.¹² Distinguishing PRES from acute stroke is very important especially in uncommon cases such as described here in which patient's GCS deteriorated in the immediate postoperative period.

The management of PRES involves early diagnosis, symptomatic treatment and correction of the causative factors.

PRES, as well as neurogenic stunned myocardium itself, carry a relatively good prognosis with partial to complete resolution of neuroimaging abnormalities and partial to complete resolution of visual manifestations and reversal of cardiac dysfunction. The overall prognosis of the patient is individualized and based upon the inciting factors.

CONCLUSION

PRES syndrome is an uncommonly encountered condition in parturients and carries a significant morbidity and mortality if it goes unrecognized. It's a potentially reversible condition and requires supportive care till it resolves. It is necessary to have a high index of suspicion in susceptible patients in order to ensure a positive outcome.

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