“Double” Trouble in Postpartum State

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ABSTRACT

Background: Guillain–Barre syndrome (GBS) is a perplexing neurological disease of autoimmune etiology with an incidence of 1.2 to 1.9 cases/100,000 population. The overall incidence remains almost the same in pregnancy. Posterior reversible encephalopathy syndrome (PRES) is a clinoradiological entity most commonly encountered in peripartum preeclampsia; however, it can also be seen in other clinical scenarios such as immunosuppressant treatment, infection/sepsis, autoimmune diseases, and during cancer chemotherapy. GBS can be considered as an independent risk factor of PRES, due to acute dysautonomia and pain. Here we report the unique case of a patient who presented to the neurology department with flaccid quadriplegia in the immediate postpartum period, was diagnosed as having GBS and initiated on treatment for the same, and subsequently developed seizures secondary to PRES.

Case description: A 22-year-old patient, with no previously known comorbidities, was admitted to the obstetrics department in her 37th week of gestation in labor. The patient underwent an emergency lower segment cesarean section (LSCS) (previous LSCS in labor) with no periprocedural complications. On the next day, she developed progressive ascending quadriplegia sparing sensory and sphincters. Nerve conduction studies done showed features of GBS, and cerebrospinal fluid studies done showed albuminocytological dissociation. The patient was then initiated on plasmapheresis in the intensive care unit setting and closely monitored for any deterioration in overall neurological picture and bulbar symptoms. The patient remained stable and showed positive response to the plasmapheresis. On the morning of postpartum day 6, the patient complained of sudden painless bilateral complete loss of vision followed by an episode of generalized tonic–clonic seizures. She had labile blood pressure with reactive pupils, normal fundus, and no other long-tract signs. Magnetic resonance imaging (MRI) brain and magnetic resonance venogram were done. MRI brain done showed features suggestive of PRES. She was managed with antiepileptic drugs, blood pressure optimization, and supportive care with rapid recovery in vision in 2 days. She showed steady improvement in motor power following plasmapheresis and subsequently was managed with physiotherapy to full functional recovery.

Conclusion and clinical significance: GBS in pregnancy, PRES in the postpartum female, and PRES in GBS are all unique entities with no defined incidence and only documented case reports. GBS rarely complicates pregnancies; there are few case reports of the same, the exact incidence of which is not defined. Proposed mechanisms leading to PRES in patients with GBS are dysautonomia leading to acute rise in the blood pressure, activation of sympathetic nervous system secondary to pain, and intravenous immunoglobulin usage. Review of the literature shows that previous cases of PRES in combination with GBS is a very unique finding with only a few case reports making this case distinctive. In our patient who developed GBS in the peripartum period, the probable mechanism of PRES was the labile blood pressures secondary to dysautonomia. With management of GBS, a close watch on pressures and close neurological observation of the patient made a near-complete recovery.

Keywords: Case report, Guillain–Barre syndrome, Postpartum complications, Posterior reversible encephalopathy syndrome, Seizures, Sudden vision loss.

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INTRODUCTION

Guillain–Barre syndrome (GBS) is an acute flaccid paralysis syndrome with an incidence of 1.2 to 1.9 cases/100,000 population, and the incidence remains almost the same in pregnancy. However, in pregnancy it may be associated with increased maternal morbidity and mortality.¹

Posterior reversible encephalopathy syndrome (PRES) is a clinoradiological entity described by Hinchey et al. in 1996. PRES can have varied clinical presentations ranging from headache, nausea/vomiting, seizures, altered sensorium, visual abnormalities, and focal neurological deficits.² The most common predisposing situations are peripartum preeclampsia, posttransplant conditions, during immunosuppressant treatment, infection/sepsis, autoimmune diseases, and during cancer chemotherapy.³

Here we report the unique case of a patient who presented to the neurology department with flaccid quadriplegia in the immediate postpartum period, was diagnosed as having GBS and initiated on treatment for the same, and subsequently developed seizures secondary to posterior reversible leukoencephalopathy.

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CASE DESCRIPTION

A 22-year-old patient, with no previously known comorbidities, was admitted to the obstetrics department in her 37th week of gestation in labor. The patient underwent an emergency lower segment cesarean section (LSCS) (previous LSCS in labor) with no periprocedural complications. On the next day, she developed

progressive ascending quadriparesis sparing sensory and sphincters.

Nerve conduction studies done on the day of illness showed motor axonopathy of bilateral ulnar and common peroneal nerves (right > left) and conduction block in antecubital fossa of right median nerve (Figures 1 and 2).

Cerebrospinal fluid studies done showed albuminocytological dissociation—proteins of 88 mg/dL and cell count of 0.

The patient was then initiated on plasmapheresis in the intensive care unit setting and closely monitored for any deterioration in overall neurological picture and bulbar symptoms. The patient remained stable and showed positive response to the plasmapheresis. On the morning following the third cycle (postpartum day 6), the patient developed sudden painless loss of vision in both eyes soon followed by an episode of generalized tonic-clonic seizures. She had labile blood pressure at this time. Her pupils were equal and reactive; her optic fundi were normal.

MRI brain done showed altered signal intensities in bilateral fronto-parieto-occipital lobes and cerebellar hemispheres

predominantly in the cortical and subcortical white matter hyperintense on T2 and fluid-attenuated inversion recovery (FLAIR) sequences suggestive of PRES (Figures 3 and 4).

She was managed with antiepileptic drugs, blood pressure optimization, and supportive care with rapid recovery in vision in 2 days. She showed steady improvement in motor power following plasmapheresis and subsequently managed with physiotherapy to full functional recovery.

DISCUSSION

Neurological dysfunction in the postpartum period can be due to a new-onset disease manifesting secondary to the contributing factors encountered in a pregnancy. GBS in pregnancy, PRES in the postpartum female, and PRES in GBS are all unique entities, and extensive literature review did not reveal a definite incidence of the coexistence of all three.⁴

GBS rarely complicates pregnancies, and there are few case reports of the same, the exact incidence of which is not defined.⁵ Blood pressure fluctuations are a common feature in GBS: 61% of

Fig 1: Nerve conduction studies of bilateral upper limbs

Fig 2: Nerve conduction studies of bilateral lower limbs

the patients with GBS have transient hypertension lasting from 2 to 21 days and postural hypotension is found in 43% of patients.⁶

PRES is characterized by headache, seizures, encephalopathy, and a myriad of visual disturbances in the background of blood-brain barrier permeability changes in the posterior areas of the brain.⁷ About 90% of patients have seizures, commonly preceded by visual changes and/or headache,⁸ as was the clinical manifestation seen in our patient. It is often associated with hypertensive encephalopathy, preeclampsia, eclampsia, renal failure, immunosuppressive therapy, or chemotherapy. More rarely, it can be related to autoimmune disorders, thrombotic thrombocytopenic purpura, HIV syndrome, acute intermittent porphyria, blood transfusion, and electrolyte disturbance.^{2,7} Pathophysiology of PRES remains controversial and there are multiple theories. The preferred theory suggests that a rapid rise in blood pressure leads to a breakdown in cerebral autoregulation disrupting the blood-brain barrier, resulting in a potentially reversible vasogenic edema.⁹

Proposed mechanisms leading to PRES in patients with GBS are dysautonomia leading to acute rise in the blood pressure,

activation of sympathetic nervous system secondary to pain, and intravenous immunoglobulin usage.¹⁰ Review of the literature shows that previous cases of PRES in combination with GBS have been reported. The first case was reported by Del Giudice and Aicardi¹¹—a child who presented with hypertensive encephalopathy and GBS. The second case has been described by Okada et al.¹²—a 4-year-old child who had a severe GBS associated with continuous hypertension, flushing, excessive sweating, and tachycardia.

Management of our case fits both the criteria for GBS and PRES. This distinctively unique case reminds us of rare complications of GBS, which in itself is a rare disease. When managing a patient with GBS, we are well aware of the possible spectrum of complications that could occur. While dysautonomia is a commonly encountered problem, PRES occurring as a consequence of it is quite rare. However, in patients with labile blood pressures in GBS, this possibility must be thought of in the appropriate setting. Additionally, it is a known fact that the peripartum state itself is a predisposing factor for PRES, even in the absence of elevated blood

Fig 3: MRI brain (T2-weighted sequence)

pressure readings.⁷ Hence, in a postpartum patient with sudden loss of vision as seen in our patient, this possibility must always be kept in mind in order to initiate timely management of the same.

In our patient, timely diagnosis and management enabled complete functional recovery and ensured good outcomes.

The patient is on regular follow-up, has not had any further seizures, and has a normal visual acuity and motor power.

CONCLUSION

In conclusion, the association between GBS and PRES, no matter how rare, must always be kept in the clinician's mind. This will

prevent any diagnostic delay, thus improving outcomes. In patients with labile blood pressures, attempts to optimize the same should be made in a timely and aggressive fashion.

CLINICAL SIGNIFICANCE

Postpartum PRES is a commonly encountered scenario for both obstetricians and neurologists alike. However, this is a truly unique instance of two distinct pathological processes occurring in a single individual in a postpartum state that was successfully managed. This case highlights the importance of expecting the unexpected in a seemingly uncomplicated pregnancy. Aggressive and timely

Fig 4: MRI brain (FLAIR sequence)

intervention in this case not only saved the patient's life but also led to complete functional recovery.

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