Case report

A case of Guillain-Barre syndrome with pregnancy who delivered in ICU: a rare outcome of rare co-occurrence

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SUMMARY

Guillain-Barre syndrome (GBS) in pregnancy is a rare co-occurrence and it is generally accepted that it carries a high maternal risk. We report a 40-year-old housewife who developed GBS with respiratory failure during the third trimester (31 weeks of gestation). She had preterm vaginal delivery on ventilator support at 34 weeks of gestation in neurology intensive care unit. She managed successfully with ventilatory support, intravenous immunoglobulin and other supportive measures.

BACKGROUND

The reported incidence rates of Guillain-Barre syndrome (GBS) in the general population usually range from 1 to 2 per 100 000.¹ GBS in pregnancy is a rare co-occurrence and is associated with high maternal and perinatal morbidity.

Maternal mortality is usually due to respiratory complications, and perinatal mortality is due to preterm labour and delivery.² The maternal and perinatal mortality rate of >10% is associated with GBS.³ We report a case of GBS with respiratory failure in pregnant woman, in her third trimester (31 weeks of gestation), who delivered in intensive care unit (ICU), and successfully managed with intravenous immunoglobulin (IVIG) and ventilatory support.

CASE PRESENTATION

A 40-year-old, gravida two and para one at 31 weeks of gestation in the third trimester, presented with rapidly progressive symmetric, areflexic, ascending quadriplegia with respiratory failure within 48 hours of onset of symptoms. There was no history of preceding diarrhoea, respiratory tract infection, vaccination or travel to Zika virus endemic area preceding these symptoms. A clinical diagnosis of GBS with GBS disability score of 5 was made.

INVESTIGATIONS

Nerve conduction studies in laterality with clinical picture showed pure axonal motor affection of nerves with markedly reduced/absent compound muscle action potential (CMAP) amplitudes.

Haematological and biochemical investigations including complete blood counts, liver and renal function tests, and blood sugar level and serum electrolytes were normal altogether. Serological studies for HIV, Hepatitis B surface antigen (HBsAg),

anti-Hepatitis C virus (HCV) antibody, IgA level and Venereal Disease Research Laboratory were negative. A spot urine test for porphobilinogen was negative in ruling out porphyria. Ultrasound of the abdomen was done for assessment of fetal well-being showed a live fetus, 31 weeks appropriate for gestational age.

TREATMENT

In view of bulbar weakness and respiratory failure, she was intubated and started on ventilatory support, IVIG, thromboprophylaxis for prevention of deep venous thrombosis and physiotherapy. Periodic fetal assessment with cardiotocography and ultrasound scanning confirmed expected fetal growth and well-being. She had features of autonomic dysfunction in the form of excessive sweating, tachycardia and hypertension which was managed with oral labetalol. Daily urine examination for albuminuria, twice a week liver function test and uric acid was done to rule out the development of preeclampsia. She also received intravenous betamethasone for fetal lung maturity as GBS in pregnancy is associated with a high risk of preterm spontaneous labour. In view of the expected prolonged duration of ventilatory requirement, tracheostomy was done on day 11 of hospitalisation. Physiotherapy was done regularly with care for prevention of bed sore, deep venous thrombosis, joint range of motion to prevent contracture and splinting was done to prevent wrist and foot drop. She had spontaneous onset of labour at 34 weeks of gestation when she was on ventilatory support and delivered a normal preterm girl baby with a birth weight of 2 kg in neurology ICU. The baby did not require any resuscitation at birth and was started on direct breast feeds. Weaning from the ventilator was done by daily monitoring negative inspiratory force. She was successfully weaned from the ventilator on day 32 of hospitalisation and tracheostomy closure was done on day 42.

OUTCOME AND FOLLOW-UP

At the time of discharge, she was able to sit with support and taking feeds orally, and power of grade II in both upper and lower limbs. Her GBS disability score at the time of discharge was 4. Her postpartum course was uncomplicated and did not had relapse of symptoms. Her baby was

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healthy and on breast feed at the time of discharge without any complications.

DISCUSSION

GBS can occur in any trimester of pregnancy and postpartum period but particularly in the third trimester and first 2 weeks postpartum.⁴ It was reported that as many as 34.5% of women suffering from GBS during pregnancy required ventilator support and the most common cause of maternal mortality is a respiratory failure.⁵ Our patient presented in the third trimester and had a stormy intensive care unit course complicated with autonomic dysfunction.

This could be compared with the work of Vasudev *et al*, who managed a similar case in the third trimester of pregnancy with plasmapheresis with good outcome.⁶ The management of pregnancy with GBS does not differ much from that in non-pregnant patients with this disease. Management includes IVIG, plasmapheresis and ventilatory support, identification and treatment of infections, prophylaxis for venous thromboembolism, pain management and management of the psychosocial distress resulting from the disease wherever required.⁷ The efficacy of IVIG is well established and can be safely given during pregnancy. IVIG and plasmapheresis are the treatment of choice for GBS. IVIG is preferred due to lesser complications compared with plasmapheresis.⁸ Those with autonomic instability, usually manifesting as tachycardia and hypertension, are managed on labetalol because it preserves uteroplacental blood flow.⁹ Our patient had autonomic instability from day 1 of admission manifested with excessive sweating, tachycardia and hypertension. It was a very close differential diagnosis of preeclampsia and we monitored her for both the development of preeclampsia and managed autonomic instability with labetalol. Despite neurological deficits, impairment of uterine contraction activity was not observed and the ability of normal vaginal delivery was demonstrated. Therefore, maternal GBS is not an indication for caesarean section and operative delivery should be reserved for obstetric indications only.¹⁰ In a retrospective study of 47 patients with GBS complicating pregnancy, only 2 patients required caesarean section.¹¹ In case of ventilator support is required in pregnancy, the risk of premature birth has been noted to be greatly increased.¹²

Our patient also had a spontaneous premature normal delivery while she had quadriplegia and was ventilator dependent. GBS is known to worsen in postpartum period due to an increase in delayed type of hypersensitivity.¹³ Our patient did not had relapse of symptoms. She required multidisciplinary approach involving neurologist, obstetrician, intensivist, radiologist and physiotherapist and paediatrician. In conclusion, GBS rarely complicates pregnancy. Early diagnosis, multidisciplinary input and prompt immune modulatory therapy are the cornerstones in the management of GBS during pregnancy and postpartum to improve outcomes for the mother. This case further highlights

the importance of multidisciplinary efforts for successful outcome in such cases.

Learning points

- Despite neurological deficits, impairment of uterine contraction activity is not observed and patient can have normal vaginal delivery.
- Termination of pregnancy did not shorten disease duration or improve maternal outcome and is not indicated for Guillain-Barre syndrome (GBS) complicating pregnancy.
- Early diagnosis and active treatment with the use of intravenous immunoglobulin together with prevention of complications is the key to success in management of pregnant women with GBS.

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Contributors RJ was involved in acute management of the patient and outpatient follow-up. PSR actively participated in literature review, writing of the case report, proofreading and submitting. KT was the intensivist in-charge for managing this case. AZ was involved in obstetric management of this patient.

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