Case Report

A Rare case of Guillain-Barré syndrome in pregnancy treated with plasma exchange

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Abstract

Guillain-Barre syndromé (GBS) is an autoimmune disorder. It is rare in pregnancy as there is a decrease in cell-mediated immunity. A case of 28-year-old pregnant woman who presented with acute flaccid quadriplegia suffering from GBS is discussed in this study. She was treated with plasma exchange in her immediate post-partum period. The management of GBS in pregnancy has been discussed.

Key words:

Guillain-Barré Syndrome in pregnancy, plasma exchange, therapeutic plasma exchange

Access this article online

Website: www.ajts.org

DOI: 10.4103/0973-6247.126695

Quick Response Code:

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Introduction

Guillain-Barré Syndrome (GBS) is an acute demyelating polyneuropathy, characterized by progressive, ascending paralysis and areflexia with or without abnormal sensory function. Symptoms are preceded by an antecedent event in about two-thirds of patients.[1,2] The disease has been linked to bacterial and viral infections, systemic diseases, neoplasia, pregnancy, traumatic injury, and organ transplant. GBS has been considered a devastating disease because of its unusual and sudden onset. About one-third of the patients with GBS will require mechanical ventilation and most GBS-related deaths occur as a result of respiratory failure. In the developing world, where health facilities are limited, GBS has been shown to be an important cause of flaccid paralysis. Population-based surveys attempting to document the annual incidence of GBS have been conducted in various countries worldwide and generally are in agreement on a rate of 1-3 per 100,000 individuals annually.[3,4] In a cohort study, age-adjusted relative risks indicate that the risk for GBS is lower during pregnancy and increases after delivery.[5] It is known to worsen during the post-partum period due to a rapid increase in delayed-type of hypersensitivity during this period. Relapse during successive pregnancies has been reported.[6] The occurrence of disease in the third trimester carries the risk of respiratory complication and prematurity. There is no specific therapy for GBS; however, plasma exchange and intra-venous immunoglobulin (IVIG) administration have been shown to reduce the progression and severity of disease; in fact, it has been found to accelerate the recovery of these patients. Reports of treatment of GBS complicating pregnancy with IVIG and plasma exchange are available in literature. Our case is unique as we treated this critically ill patient admitted in intensive care unit (ICU) with plasma exchange in her immediate post-partum period.

The present case is of a 28-year-old pregnant female who presented with severe form of GBS with incipient respiratory failure and was successfully managed with plasma exchange and ventilator support. The report of the case under reference will add to the very limited literature of GBS in pregnancy and treatment of the same in immediate post-partum period with plasma exchange.

Case Report

A 28 years old female, Gravida 1 Para1was referred to Gynecology and Obstetrics Department of our hospital at 36 weeks of pregnancy with 4 days history of progressive weakness of limbs and difficulty in walking, which culminated in loss of ability to walk; the weakness progressed gradually to upper limbs, with a 1 day history of difficulty in swallowing and breathing.

The patient was in her usual state of health 4 days back when she noticed weakness in both lower limbs after she got up in the morning. The weakness worsened the following day and she was unable to walk. By the end of 3rd day, weakness was also noted in upper limbs and had difficulty in lifting her arms. On the 4th day of her illness, she had difficulty in swallowing and breathing and was referred to the hospital.

On examination, the patient was found acutely ill, pale, afebrile, acyanosed, anicteric, and hydration status was satisfactory. She was dyspneic with shallow breathing. Her breathing was vesicular with no added sounds. Cardiovascular examination showed no

abnormality. On abdominal examination, she was almost in full — term pregnancy (36weeks) and was not in labor. On neurological examination, she was found to be conscious but restless, and apprehensive. She was aphonic and had weak cough reflex and gag reflex; all her cranial nerves were normal. She presented with all the features of flaccid quadriplegia with grade zero power in both lower limbs and grade three in upper limbs. Muscle tone was decreased and deep tendon reflexes were lost. There was no sensory impairment or bladder and bowel involvement. Nerve conduction tests and cerebrospinal fluid analysis suggested diagnosis of GBS.

The patient was taken up for Cesarean Section, surgery was conducted under general anesthesia, and a 2.5 kg healthy baby was delivered. The patient could not be extubated following surgery as she was not able to maintain spontaneous breathing. She was then shifted to ICU and kept on ventilator and the next day plasma exchange was started; this was the 5th day of illness. Therapeutic plasma exchange was performed as first line treatment and IVIG or steroids were not tried before, after, or simultaneously with plasma exchange. Plasma exchange was done on alternate days, with a total of five sessions and in each session; 2308.8 ± 182.73 ml (range: 2105-2562 ml) of plasma was removed. This was in accordance with the guidelines of removing 1-1.5 times plasma in each session. Five percent albumin was used as replacement fluid. No plasma was used as the patient's coagulation profile was within normal limits. Peripheral femoral access was used to carry out the procedure; proper catheter care was maintained and lines were flushed with heparin before and after the exchange. The patient had no complications during these sessions. Complete coagulation profile including: prothrombin time (PT), prothrombin index (PTI), activated partial thromboplastin time (APTT); complete blood count (CBC) including haemoglobin, platelet count; electrolytes (Na^+, K^+, Ca^{++}); s. protein levels were checked before and after the procedure and remained within normal limits throughout the treatment period. After three sessions of plasma exchange, i.e., on the 9th day of illness, the patient was weaned off the ventilator and physiotherapy started. The patient was discharged on 14th day of illness. At the time of discharge, the power was 5/5 in upper limbs and 3/5 in the lower limbs. Follow-up of the patient after 2 months showed no residual weakness.

Discussion

GBS is a neurological disorder resulting primarily in muscle paralysis, which in most cases is symmetrical. GBS occurring in pregnancy is associated with an increased need for ventilator support and an increase in maternal mortality.[7,8] Pregnancy is associated with a decrease in cellular immunity and increase in humoral immunity; this shift is because of production of interleukin 10. After pregnancy is terminated, this is reversed and this accounts for the increased incidence and worsening of symptoms in the post-partum period.

Immunomodulation with plasma exchange and IVIG have been found to improve treatment outcome; 70-80% of patients recover fully. Mehndiratta *et al.* described the efficacy and cost-effectiveness of current therapies in GBS.[9] Clinical trials indicate that plasma exchange is more effective than supportive treatment alone in reducing the median time taken for patients to recover. IVIG and plasma exchange are the treatment of choice for GBS. IVIG is preferred due to lesser complications compared with plasma exchange.[10] However, in a set up like ours, cost of treatment is also a very important factor. Cost of plasma exchange is very less compared with IVIG with similar results. Goyal *et al.* have described the management of a primigravida presenting at 26 weeks' gestation with plasmapheresis.[11] Reports of treatment of Acute inflammatory demyelinating polyneuropathy (AIDP) in pregnancy with IVIG[12,13] and plasma exchange[14,15] are available in the literature. Our case was unique as we treated this case with plasma exchange in the immediate post-partum period of pregnancy. High-quality intensive care remains the most important aspect in the management of severe cases of GBS.

To conclude, GBS is rare in pregnancy but can be managed with intensive care involving ventilator support, plasma exchange or IVIG, proper nutrition, infection control and psychological support.

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Cite this article as: Vasudev R, Raina TR. A Rare case of Guillain-Barrı syndrome in pregnancy treated with plasma exchange. Asian J Transfus Sci 2014;8:59-60.

Source of Support: Nil, **Conflicting Interest:** None declared.