Case Report

Guillain–Barré syndrome in pregnancy: A conservatively managed case

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Abstract

Guillain–Barré syndrome (GBS) is an autoimmune disease. Estimated population incidence ranges from 0.62 to 2.66 cases per 100,000 person-years across all age groups. We report a case of GBS in a 22-year-old primigravida who presented at 36 weeks of the period of gestation (POG), with complaints of bilateral progressive lower limb numbness and weakness for 2 weeks duration. Magnetic resonance imaging of the brain was done to exclude other possible causes. Diagnosis of GBS was made according to the Brighton criteria, which our patient falls into Level 2. She received intensive care management. The patient improved rapidly without any specific management. She went to labor spontaneously and delivered a healthy baby with a birth weight of 2.8 kg at 38 weeks of POG. She continued to receive supportive therapy and improved significantly.

Keywords: Conservative management, Guillain–Barré syndrome, intravenous immunoglobulin, pregnancy

Introduction

Guillain–Barré syndrome (GBS) can be described as a collection of clinical syndromes that manifest as an acute inflammatory polyneuropathy, with resultant symmetrical rapidly progressive muscle weakness, areflexia, and raised cerebrospinal fluid (CSF) protein level, but normal cell count in CSF. Two-thirds of cases are preceded by symptoms of upper respiratory tract infection or diarrhea. The most frequently identified infectious agent associated with subsequent development of the GBS is *Campylobacter jejuni*.[1] Miller-Fisher syndrome (MFS) is recognized as a variant of GBS. MFS is a rare disorder that is characterized by the acute onset of ophthalmoplegia, ataxia, and areflexia/hyporeflexia.[2] GBS can occur in any trimester and in the postpartum period. However, it is more common in the third trimester and the first 2 weeks of postpartum. GBS is known to worsen in postpartum period due to an increase in delayed type of hypersensitivity. Delayed diagnosis is common in pregnancy or immediate postpartum period because the initial nonspecific symptoms may mimic changes in pregnancy. GBS should be considered in any pregnant patient complaining of muscle weakness, general malaise, tingling of the fingers, and respiratory difficulty.[3,4]

We report this case to review management options of GBS in pregnancy.

Case Report

A 22-year-old primigravida at 36 weeks period of gestation (POG) presented with bilateral progressive lower limb weakness for 2 weeks duration with difficulty in walking and difficulty in sitting and standing from a supine position. Weakness was associated with mild upper limb numbness but no associated respiratory or cardiovascular symptoms.

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At presentation, examination revealed sinus tachycardia, normal blood pressure, power of 3/5 in bilateral lower limbs and 5/5 in bilateral upper limbs, hypotonia, diminished bilateral ankle reflexes with preserved knee joint reflexes, flexor plantar response, and no involvement of all modalities of sensation.

A clinical diagnosis of GBS was made, and further investigations were arranged to rule out other possibilities.

Blood investigations including full blood count, serum electrolytes, serum creatinine, serum calcium, serum magnesium, and creatinine phosphokinase-MB remained within normal limits. Electrocardiogram revealed sinus tachycardia.

She developed difficulty in swallowing, without difficulty in breathing. The patient was transferred to the Neurology Intensive Care Unit for further care.

Nerve conduction studies show feature of acute inflammatory demyelinating polyneuropathy.

Magnetic resonance scanning of the brain and spinal cord was arranged on day 3 after presentation and found to have normal.

There was no further deterioration of her symptoms. Fetal assessment with cardiotocography and ultrasound scanning confirmed expected fetal growth and well-being.

Her muscle tone, power, and reflexes gradually improved within a short period and decided not to do lumbar puncture and manage conservatively without intravenous immunoglobulin (IVIG) treatment. Thromboprophylaxis was started from day 1. Physiotherapy was initiated. Following spontaneous onset of labor, she delivered a healthy baby with a birth weight of 2.8 kg (Apgar 1–10, 5–10, 10–10) after 17 days of presentation (at 38 weeks of POG).

Discussion

GBS has a very low incidence during pregnancy, estimated population incidence ranged from 0.62 to 2.66 cases per 100,000 person-years across all age groups.[5] It is usually preceded by a bacterial or viral infection. It has also been linked to underlying systemic diseases, certain malignancies, surgery, pregnancy, trauma, severe infection, and tissue transplantation; acute cytomegalovirus infection complicating twin pregnancy has been reported. Our patient gave a history of acute gastroenteritis. High maternal and perinatal mortality rate (>10%) is associated with GBS. Maternal mortality is usually due to respiratory complications and perinatal due to preterm labor and delivery.[6]

Delayed diagnosis during pregnancy and immediate postpartum period is possible due to initial nonspecific symptoms which may mimic changes in pregnancy.[6]

The diagnosis of GBS is based on descriptive clinical, laboratory, and electrodiagnostic criteria. There are no pathognomonic, clinical features, and at present, no biomarkers are available to discriminate from disorders resembling GBS.

The Brighton Collaboration is an international collaboration sponsored by the World Health Organization to facilitate the development, evaluation, and dissemination of high quality internationally standardized case definitions for various illnesses, with the aim of improving vaccine safety. These innovatory "Brighton criteria" also account for the level of diagnostic certainty based on the presenting findings at clinical and additional examinations, ranging from Level 1 (highest level of diagnostic certainty) to Level 4 (reported as GBS, possibly due to insufficient data for further classification).[7]

Table 1; key diagnostic criteria and Brighton case definitions for GBS (diagnosis of GBS and validation of Brighton criteria).[7]

According to the Brighton criteria, our patient falls into Level 2 of diagnostic certainty for confirmation of GBS.

GBS can occur in any trimester of pregnancy and postpartum period but specifically in the third trimester and the first 2 weeks postpartum. Our patient develops GBS at 36 weeks of POG. GBS is known to worsen in postpartum period due to an increase in delayed type of hypersensitivity.[6] A maternal mortality of 7% has been quoted (nonpregnant GBS has mortality <5%.[7]

The management of GBS in pregnancy is somewhat similar to that in the nonpregnant population and includes IVIG, plasmapheresis, and ventilator support wherever required. Immunomodulation with plasmapheresis and IVIG has been found to improve treatment outcomes with full recovery in 70–80% of patients.[8] Even though IVIG is a proven effective treatment for GBS, not all patients recover enough after a standard IVIG dose. In one case series, IVIG was needed only for two patients out of 47 cases of GBS in pregnancy.[9] It is an expensive treatment that may induce (generally minor) side effects and is currently not indicated (proven to be effective) in mildly affected GBS patients.[10] Patients may experience mild side effects including headaches, stiffness of the neck, nausea, dizziness, vomiting, chills, fever, low blood pressure, and arrhythmia for up to 48 h after being treated

Table 1: Level of diagnostic certainty

Diagnostic criteria	1	2	3	4
Bilateral and flaccid weakness of limbs	+	+	+	+/−
Decreased or absent deep tendon reflexes in weak limbs	+	+	+	+/−
Monophasic course and time between onset and nadir 12 h to 28 days	+	+	+	+/−
CSF cell count <50/μl	+	+[a]	−	+/−
CSF protein concentration > normal value	+	+/−[a]	−	+/−
NCS findings consistent with one of the subtypes of GBS	+	+/−	−	+/−
Absence of alternative diagnosis for weakness	+	+	+	+

[a]If CSF is not collected or results not available, nerve electrophysiology results must be consistent with the diagnosis GBS. +: Present; −: Absent; +/−: Present or absent; NCS: Nerve conduction studies; GBS: Guillain–Barré syndrome; CSF: Cerebrospinal fluid

or in the initial stages of the treatment. These symptoms disappear after a few hours or days. If they occur during the treatment, they can be minimized by reducing the rate of IVIG infusion. The treatment can also trigger allergic reactions such as rash on the palms of the hands. Our patient received care at the Intensive Care Unit and did not require any specific therapy including IVIG or plasmapheresis due to rapid clinical improvement.

Physiotherapy is a mainstay in management of GBS patients and 58% of the patients will receive complete responses following physiotherapy.[11] Satisfaction with physiotherapy is about 87%.[12] Our patient received physiotherapy from day 1.

Limitation of movements, pregnancy, and gravid uterus increases the risk of thromboembolism in these patients. Thromboprophylaxis with thromboembolic deterrent stockings and low molecular weight heparin should be started as early as possible and need to continue until the patient regain full functionality.

GBS complicating pregnancy has a good fetal outcome. Unnecessary obstetric intervention must be strongly resisted.[13] Early delivery is indicated only if there is a respiratory compromise and have problems with artificial ventilation. Cesarean section should be for obstetrics indications.[14] Assisted vaginal deliveries can be considered to reduce prolonged second stage of labor. Our patient had a spontaneous vaginal delivery with birth weight 2.8 kg.

A high index of suspicion for early diagnosis and involvement of multidisciplinary supportive care in a GBS-complicated pregnancy improve the prognosis for both mother and fetus.

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Conflicts of interest

There are no conflicts of interest.

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