Case Report

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Guillain-Barré syndrome in pregnancy: A case report

Misai Hukuimwe, Tawanda T Matsa and Muchabayiwa F Gidiri

Abstract

Guillain-Barré syndrome is a rare condition in pregnancy which is characterised by symmetrical progressive ascending polyneuropathy. A case of a 16-year-old nulliparous woman who presented with rapidly progressive limb paralysis following an upper respiratory tract infection a week prior to presentation is discussed. She was intubated as she had developed respiratory failure and managed in the intensive care unit by a multidisciplinary team. Plasma exchange and intravenous immunoglobulin were not readily available so she was managed conservatively. The management of Guillain-Barré syndrome, maternal and foetal outcomes have been discussed.

Keywords

Guillain-Barré syndrome, intravenous immunoglobulin, plasma exchange, pregnancy

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Introduction

Guillain-Barré syndrome (GBS) is a rare condition, and it is even rarer during pregnancy with incidences of 1.2–1.9 cases per 100,000 annually.[1] It is the most common cause of acute flaccid paralysis affecting all age groups.[2] GBS represents a heterogeneous group of immune-mediated peripheral neuropathies, resulting in muscle paralysis which in most cases is symmetrical. The most predominant form of GBS is acute inflammatory demyelinating polyneuropathy, with 10% of patients having bifacial weakness.[1]

The pathology of GBS is unknown, but it is thought to be due to molecular mimicry between epitopes found on cell walls of microorganisms and gangliosides found on Schwann cell membrane.[2] In a study exploring the relationship between pregnancy and GBS, the risk of GBS is lower during pregnancy and increases post-delivery.[3] This is due to return of cellular immunity, with an increase in delayed type hypersensitivity, which is depressed during pregnancy.

Presentation of case

We report a case of a 16-year-old nulliparous woman at 31 weeks gestation who was referred to us for rapidly progressive paralysis of limbs with subsequent development of respiratory failure within 48 h of the onset of the symptoms. This case was managed at Parirenyatwa Group of Hospitals, one of the two central hospitals in Harare, Zimbabwe. The patient had presented to a local provincial hospital about 260 km away, with a history of sudden collapse at home because of limb weakness. This was on the background of an upper respiratory tract infection a week prior to presentation of which she did not seek medical treatment for. She did not have any comorbidities and was well prior to this illness. HIV test was negative, and she had no history of trauma. She was put on ventilatory support as she was now saturating at 70% on free air.

Initial diagnosis/assessment

Examination revealed notable areflexia, flaccid paralysis of all limbs with power of 0/5, on ventilatory support with preserved mentation. She was apyrexial, with a tachycardia of 112 beats/min, normotensive, saturating 98% on ventilatory support. Abdomen was gravid, soft and

Department of Obstetrics and Gynaecology, College of Health Sciences, University of Zimbabwe, Avondale, Zimbabwe

Corresponding author:
Misai Hukuimwe, Department of Obstetrics and Gynaecology, College of Health Sciences, University of Zimbabwe, B Floor Old Health Sciences Building, Mazowe Street, Avondale, P.O. Box A178 Harare +263, Zimbabwe.
Email: mhukuimwe@gmail.com

nontender with a height of fundus corresponding to 32 weeks gestation. Foetal heart was present. Cerebrospinal fluid (CSF) was initially unremarkable but serial examinations showed cytoalbuminogenic dissociation with a white cell count $<1/mm^3$ and total protein of 0.81 g/dL (Normal Range, 0.15–0.45). Clinical picture and CSF examination were consistent with acute inflammatory demyelinating polyneuropathy (GBS in pregnancy).

Treatment/management

The patient was admitted to intensive care unit (ICU) for ventilatory support and was managed by a multidisciplinary team. She was put on prophylactic enoxaparin and received a course of antenatal corticosteroids for foetal lung maturity in anticipation of preterm delivery. The planned definitive management for the GBS was intravenous immunoglobulin (IVIG) or plasmapheresis, but these were not readily available. Serological tests for *Campylobacter jejuni*, cytomegalovirus (CMV), varicella zoster virus, Epstein–Barr virus and *Mycoplasma pneumoniae* were not readily available but were considered as there is often an association. An ultrasound scan was done which confirmed a viable foetus at around 30 weeks gestation. The patient was then managed conservatively. On day 9 post-admission she went into labour and delivered a fresh stillbirth with a birthweight of 1200 g. On day 19 in ICU the patient was taken to theatre for tracheostomy because of prolonged intubation.

Outcome

The patient was discharged from ICU after 66 days to high dependency unit (HDU) where she stayed for 37 days. She was then discharged to the ward when her power was 3/5 on the right side and 1/5 on left side. She continued to receive physiotherapy until she was discharged back to the referring institution on day 115. Her condition deteriorated and became oxygen dependent and unfortunately died on day 117.

The patient provided verbal consent for the publication of her case while she was recovering in the high-dependency unit, following extubation and before being discharged to the medical ward. The Joint Research Ethics Committee and the Medical Research Council of Zimbabwe do not require ethics approval for a case report. In Zimbabwe, if a patient below the age of consent becomes pregnant, she becomes an `emancipated adult' because she is carrying a life. She can give consent despite her age.

Discussion

Most patients with GBS present with a history of progressive ascending symmetrical muscle weakness, usually preceded by a respiratory or gastrointestinal infection within 6 weeks in about 60% of patients.[4] Our patient had a prior history of a respiratory tract infection before the sudden collapse due to muscle weakness. The organisms that have been implicated are *C. jejuni*, varicella zoster virus, CMV, Epstein–Barr virus and *M. pneumoniae.* In Brazil, where the prevalence of Zika virus infection is high, there has also been an association with high incidence of GBS in pregnancy.[5] While it is possible that the Zika virus may be carried worldwide by international travel far beyond of its mosquito vectors, its reported incidence in Africa is low.[6] However, there has been no reported case of Zika virus in Zimbabwe. GBS in pregnancy has been reported after trivalent influenza vaccine administration; however, our patient didn't receive any vaccination.[7] However, due to the overlap of early symptoms of GBS with symptoms of pregnancy, there is usually a delay in diagnosis resulting in increased maternal and perinatal morbidity and mortality. However, there was no delay in presentation to hospital by our patient.

The diagnosis is usually clinical with support of serological, nerve studies and CSF investigations. In resource-limited settings, the diagnosis is usually made clinically with the support of a few laboratory investigations. The most commonly used investigation is CSF examination which typically shows elevated protein with normal cell and glucose level.[8] However, the diagnosis is feasible in resource-limited settings using criteria by the GBS classification group which is based solely on clinical grounds.[5] Our patient's diagnosis was based on clinical findings and a high protein and normal white cell count and glucose.

The management of GBS in pregnancy is mainly supportive, with plasmapheresis or IVIG. About a third of patients with GBS will require mechanical ventilation and most common cause of maternal mortality is respiratory failure.[9] In addition to mechanical ventilation, supportive management also includes identification and treatment of infections, prophylaxis for venous thromboembolism, pain management and management of the psychosocial distress resulting from the disease.[10] Those with autonomic instability, usually manifesting as tachycardia and hypertension, are managed on labetalol because it preserves uteroplacental blood flow. Rehabilitation should be started during pregnancy focusing on pregnancy-associated change in figure and weight gain and post-delivery with training in movements needed for child care.[11] Plasmapheresis or IVIG are the mainstay of definitive treatment of GBS.[7] A systemic review has shown that plasma exchange is better than supportive treatment alone without a significant increase in adverse events.[12] In the same review, plasma exchange was however associated with a small but significant rate of relapse within the 6–12 months post-treatment with full recovery after 1 year of treatment. In another systematic review, it was shown that IVIG hastens recovery as much as plasma exchange with no increased adverse events with both

treatments, but there was a higher likelihood of treatment continuation with IVIG.[13] There is no added significant benefit in giving IVIG after plasma exchange. Our patient was managed on mechanical ventilation, prophylactic enoxaparin, antibiotics, analgesics and physiotherapy. She did not have any autonomic instability so she was not put on labetalol. She did well on that supportive treatment as evidenced by the clinical improvement with eventual discharge back to the referring hospital. Immunotherapy has not been shown to reduce mortality in GBS and mortality is due to disease-related issues and secondary complications developed in hospital due to prolonged disease course.[14] We think this patient did not have a recurrence but she still needed good nursing care which was not provided at the referral hospital as her condition deteriorated within a few days of her arrival there. There is generally staff shortage within the public health system which is worse for peripheral hospitals as there are no incentives for staff to go and work there.

The patient went into spontaneous labour and delivered a fresh stillbirth 9 days after admission. We did not have a post-mortem done on the foetus as the family did not consent to it. It may however be possible that the maternal infection might have crossed to the foetus transplacentally and caused a fatal foetal infection. GBS following CMV infection is more severe with a higher frequency of respiratory insufficiency, cranial nerve involvement and severe sensory loss than in those with GBS following *C. jejuni* infections.[15] In utero deaths are more frequent with CMV infection than with any other aetiologies. Unfortunately, we could not do serological test in our patient, but CMV infection of both the mother and the foetus is a possibility. Even if we had been able to do CMV serology, the efficacy of CMV treatment during pregnancy is controversial. Foetal outcomes are, however, generally good.[16]

The course of recovery of patients from the disease is usually long with patients being maintained on mechanical ventilation for more than 50 days and being hospitalised for about 90 days.[10] The condition may worsen post-delivery because of changes in the immune system with return to predominantly cellular immunity which is suppressed during pregnancy. The risk of GBS increases after delivery, particularly in the first 2 weeks after delivery.[3] Relapse of GBS in pregnancy has also been reported which is managed with repeat IVIG.[17] Such relapses could be true relapses or natural disease progression or pharmacological relapses which are due to immunomodulatory drug wear off. A systematic review also found a small but significant increase in GBS relapse in those treated with plasmapheresis in the 6–12 months post initial treatment.[12] About 70%–80% of patients with GBS will recover fully.[18] In cases of GBS in pregnancy that requires ventilatory support, the risk of preterm delivery is high. Our patient continued to recover on the ward with gradual return of power in the upper limbs and she was discharged back to the referring institution where she later died. Poor prognosis in GBS has been associated with rapid onset of illness, severe degree of paralysis, muscle wasting, prolonged period of peak paralysis lasting more than 2 weeks and a delay in onset of recovery lasting more than 3 weeks as well as respiratory involvement.[19] Our patient had all these bad prognostic features.

To conclude, GBS is rare during pregnancy and one has to maintain a high index of suspicion to make a diagnosis of GBS in pregnancy. Symptoms of GBS can be confused with normal pregnancy symptoms. The diagnosis is usually made on clinical grounds supported by CSF examination, serology and nerve studies. Management of GBS is by a multidisciplinary team and is supportive including prophylaxis for venous thromboembolism, treatment of infections, pain management, management of autonomic instability and management of psychosocial distress and 60% will require mechanical ventilation. Plasma exchange and IVIG are definitive management and are better than supportive treatment alone. However, these are not readily available in resource-limited settings because of cost and the recovery course can be very long, requiring prolonged periods of intubation and despite best efforts maternal mortality occurs.

Declaration of conflicting interests

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