



Guillain-Barre syndrome in pregnancy: A rare disease with challenging management

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Abstract

Background: Guillain-Barre syndrome is a rare autoimmune disorder in pregnancy, characterised by ascending polyneuropathy. Diagnosis is mainly clinical and treatment is mainly supportive. Poor access to plasmapheresis and intravenous immunoglobulin limits its management in low and middle income countries. This report is aimed at presenting a case of Guillain-Barre syndrome in pregnancy and challenges encountered in our setting.

Case presentation: A 32 year old multipara at 34 weeks gestation who presented with progressive limb weakness, inability to walk and difficulty in breathing. She was delivered following hypertension and respiratory distress. Baby remained in good condition. She subsequently succumbed to respiratory failure in the postoperative period.

Conclusion: Guillain-Barre syndrome is rare in pregnancy. Diagnosis is mainly clinical. Supportive care including the use of plasmapheresis and intravenous immunoglobulin are important in reducing the mortality associated with the disease. Early presentation and diagnosis is also key to its management.

Keywords: Guillain-Barre syndrome, pregnancy, neuropathy

Introduction

Guillain-Barre syndrome (GBS) in pregnancy is rare but the disease appears fatal especially in developing countries due to poor access to facilities for its management. The incidence is 1 – 2 per 100,000.¹ It is an inflammatory polyradiculoneuropathy that results in motor weakness.² The cause of maternal death is usually respiratory insufficiency and autonomic instability while neonatal mortality is usually due to preterm labour and delivery.²

Autoimmune mechanisms appear to play a role in its aetiopathogenesis. There is damage to peripheral nerves which is caused by infectious agents. GBS is preceded by nonspecific gastrointestinal or respiratory symptoms.¹ The antibodies originally

produced against infection will cross-react with native epitopes in specific gangliosides resulting in the different manifestations of the disease.^{1,3} In *Clostridium jejuni* infection, its lipopolysaccharides is similar to that found in carbohydrate moiety present in gangliosides which triggers the immune system to mistake the neuronal cells as foreign, thereby causing damage.¹

The presentation of GBS is usually ascending acute flaccid paralysis with involvement of the diaphragm resulting in respiratory failure and death. The infectious organisms implicated include *Campylobacter jejuni*, *Escherichia coli*, *Mycoplasma pneumonia*, cytomegalovirus, the Human Immunodeficiency Virus and Zika virus.³ The disease has also been linked to pregnancy and neoplasia.² About 67% of affected patients will report an infectious illness 2 – 4 weeks before the onset of symptoms.³ These symptoms include pain, paraesthesia, numbness or limb weakness.¹

Treatment of GBS is non-specific though plasmapheresis and intravenous immunoglobulin appear to be useful.⁴ Though these may be readily

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available in developed countries, this is not the case with developing countries which may increase mortality associated with the disease.

Our aim is to report a case of Guillain-Barre syndrome in pregnancy and share our challenges in its management with a view to improving care in our setting.

Case presentation

A 32-year-old housewife with two living children at 34 weeks gestation who was referred from a peripheral centre to the University of Benin Teaching Hospital with complaint of progressive lower limb weakness of 5 weeks duration and inability to walk of 4 weeks duration. Fever started a week prior to presentation. She developed loss of neck control and difficulty in breathing a day prior to presentation. There was no history suggestive of an infectious disease prior to onset of symptoms. She had no other known comorbid conditions.

Examination showed a febrile lady in respiratory distress (SpO₂ 85% in room air). She was fully conscious without any cranial nerve deficit. Reflexes were absent in both lower limbs and power was reduced in all her limbs but worse on the lower limbs. Her neck was floppy. She was dyspnoeic with respiratory rate of 30 cycles per minute and her chest was clear. Pulse rate was 120 beats per minute and blood pressure was 170/90mmHg. There were no uterine contractions and the foetal heart was 160 - 168 beats per minute.

A clinical diagnosis of Guillain Barre syndrome with preeclampsia was made and stabilisation was immediately commenced. Full blood count was essentially normal except for marginally elevated white blood cell count. Urinalysis showed proteinuria of 2+. Electrolytes, urea and creatinine levels were within normal limits. Cerebrospinal fluid protein was elevated (1.31g/l). She was managed with the medical team, anaesthetist and haematologist.

She had surgery with spinal anaesthesia which was later converted to general anaesthesia following worsening respiratory distress. A live neonate was delivered in good condition and was received by the paediatrician. She was moved to the intensive care unit after surgery where mechanical ventilation was continued. Intravenous antibiotics and paracetamol were also continued. She had deep venous

thrombosis prophylaxis with subcutaneous enoxaparin. Possibility for plasmapheresis, use of immunoglobulin and exchange blood transfusion were discussed but these were not readily available. Her condition did not improve. She subsequently had respiratory failure leading to demise on the same day of admission.

Discussion

GBS typically presents with a preceding history of an infectious illness before the onset of neurological symptoms.¹ Though the index patient did not give any of such history there may have been a preceding mild illness which was unremarkable. Her late presentation (five weeks after onset of symptoms) may have also affected her ability to recall past events.

GBS can occur at any gestational age but seems to be more common in the second and third trimester of pregnancy.⁵ The index patient was in the third trimester. Diagnosis of GBS is mainly clinical.³ Criteria for diagnosis is based on clinical, laboratory and electro-diagnostic parameters.³ Common features of GBS are pain, numbness, paraesthesia and weakness of the limbs.⁶ There is no pathognomonic feature for the disease and may sometimes be misdiagnosed.⁶ As seen in this case report the patient was not immediately referred to our facility following onset of symptoms. It was only after there was affectation of the diaphragm with respiratory embarrassment that prompted her evacuation to our facility.

Autonomic dysfunction may also occur in GBS. This could result in hypotension or hypertension, tachy- or bradyarrhythmias, flushing, diaphoresis, ileus and bladder distension.¹ Tachycardia and hypertension seen in this patient may have been as a result of GBS. The presence of significant proteinuria gave the impression of a comorbid diagnosis of preeclampsia.

In GBS, cerebrospinal fluid (CSF) analysis shows elevation of protein (>0.55g/L) without a corresponding increase in white blood cells.³ As shown in this case report CSF protein was elevated. This usually occurs in the acute phase of the disease, resulting from widespread inflammation of the nerve roots. Radiological investigations would not assist in the diagnosis of GBS hence were not recommended for the index patient especially in a

resource constrained setting. They may however play a role in excluding other differentials.³ Management of GBS in pregnancy is same as outside pregnancy which is mainly supportive. Those affected should be managed in the intensive care unit with respiratory and cardiovascular support. Plasmapheresis and intravenous immunoglobulin are useful in up to 70 – 80% but this is not readily available in resource limited setting such as ours.^{4,7} Physiotherapy is also important in the management of GBS. Pregnancy and limitation of movement increases the risk of thromboembolism in GBS hence treatment should include the use of low molecular weight heparin and thromboembolic deterrent stockings.⁷ Delivery may be considered to aid resuscitation.

There is high maternal and perinatal mortality (>10%) associated with GBS.⁶ The cause of death in GBS is usually respiratory failure when the diaphragm is affected. Autonomic dysfunction may also be a cause of maternal mortality in pregnant women with GBS. The gravid uterus worsens respiratory compromise in pregnant women with GBS. Recovery period is variable and in some cases there is residual disability.⁸ Relapse could occur in 5.5 – 6.8%.²

Conclusion

Guillain-Barre syndrome is rare in pregnancy and diagnosis is mainly clinical. Supportive care including the use of plasmapheresis and intravenous immunoglobulin are important in reducing the mortality associated with the disease. Respiratory failure and autonomic dysfunction are the usual causes of death in GBS.

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