Case Report

Guillain Barre Syndrome (GBS) with respiratory and autonomic involvement in pregnancy. A Rare Case Report.

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Abstract: Guillain-Barré syndrome (GBS) represents a heterogeneous group of immune mediated peripheral neuropathies. A feature common to all GBS variants is a rapidly evolving polyradiculoneuropathy which presents as rapidly progressive ascending weakness because of a previous triggering event, most often an infection of gastrointestinal or respiratory tract. GBS rarely complicates pregnancy. If at all it occurs in pregnancy , then it is usually severe. We report a 26-year-old primigravida who developed progressive heaviness of both lower limbs in her third trimester of pregnancy followed by dyspnea , later on diagnosed to have severe form of GBS with respiratory and autonomic failure.

Keywords: GBS, peripheral neuropathy, immune, trimester, autonomic

INTRODUCTION

Guillain-Barré syndrome (GBS) also known as acute demyelinating polyradiculoneuropathy is characterised by the acute or subacute onset of varying degrees of weakness in limbs or cranial nerve-innervated muscles, associated with decreased or absent deep tendon reflexes, sometimes autonomic failure, albumin-cytological dissociation in the cerebrospinal fluid and characteristic electrodiagnostic studies. French physician Jean Baptiste Octave Landry first described the guillain barre syndrome in 1859.

The aetiology and pathophysiology of GBS are not completely understood, but it is believed to be an immune-mediated process, resulting from fostering eventually autoimmune antibodies and inflammatory cells that cross-react with epitopes on peripheral nerves and roots, leading to demyelination, axonal damage or both. This immune response is thought to be initiated in response to a variety of antigenic stimuli, such as viral or bacterial infection, particularly Campylobacter jejuni, which cause diarrhoea.

Guillain-Barré syndrome (GBS) is rare in pregnancy with an estimated incidence between 1.2 and 1.9 cases per 100,000 people annually, and it carries a high maternal risk. Relapse in GBS has been reported to occur in 5.5-6.8% of patients. [1,2] Under blanket term Guillain-Barré syndrome are several recognisable variants with distinct clinical and pathological features. All patients with Guillain-Barré syndrome need meticulous monitoring and supportive care.

Early initiation of intravenous immunoglobulins (IVIg) or plasma exchange is of proven benefit and crucial, especially in patients with rapidly progressive weakness. Because a quarter of patients need artificial ventilation and many develop autonomic disturbances, many patients need admission in the high or intensive care setting.

CASE REPORT

A 26 year old primigravida with 32 weeks of gestation presented to us with two days history of heaviness in both lower limbs associated with mild tingling and weakness in both upper limbs. She was apparently alright two days back but on asking lead questions she reported to have febrile illness 15 days prior to this episode. The fever was intermittent type associated with myalgia and mild diarrhoea. She recovered from fever within two days, there was no history of any sensory level, sensory ataxia, urinary incontinence, excessive vomiting, blurring of vision or seizures.

On General examination: pulse was 122 beats per minute with regular rhythm, BP was 210/120 mm of Hg in right arm in supine position and on standing for 3 mins the blood pressure
was 186/92 mm of Hg suggestive of postural hypotension and autonomic involvement. Mild pallor was present and bilateral oedema feet was present. On neurological examination higher functions were normal. Cranial nerve examination revealed a right infra nuclear facial palsy. Motor system examination revealed power grade of 3/5 in proximal as well as distal group of muscles of left lower limb, 4/5 in right lower, 3/5 right upper limb and 4/5 left upper limb. Hypotonia in all four limbs was present and all the deep tendon reflexes were absent even with reenforcement. Sensory system examination was normal.

Investigations : Her complete blood count, urine analysis, Liver and renal function tests, and serum electrolytes were normal. In view of the above findings the clinical diagnosis of GBS was entertained and the patient was admitted in the medicine ward.

Twenty-four hours after admission (4th day of weakness) the patient complained of palpitations and breathlessness. Examination at that time revealed a single breath count of 11. Bilateral lungs were clear on auscultation, BP was 170/90 mm Hg. Pulse oximetry revealed SPO2 of 82% while the patient was breathing ambient air. The RR was 32 cycles per minute. in view of impending respiratory failure failure was shifted to intensive care unit and was put on mechanical ventilator. Additional serological investigations in the form of ANA, HIV, RA factor, Australia antigen, HCV, Ig G and IgM antiphospholipid antibodies were negative. Serologic tests for C. jejuni, Mycoplasma were negative. CSF was done which revealed 3 cells (all were lymphocytes) protein =140mg%,sugar= 65mg% (albuminocytological dissociation). Apart from mechanical ventilation patient was treated with IVIG 400mg/ kg per day for next 5 days, tab. metoprolol 50mg, antibiotics and IV fluids.

Her condition improved 3 days into Intra venous immune globulin therapy and she was extubated on the 9th day. A nerve conduction study was done which revealed; absent CMAP amplitude in bilateral tibial and peronial nerve, CMAP amplitude reduced with normal onset latency and CV in bilateral median nerve. SNAP amplitude could not be elicited in bilateral sural nerve, features suggestive of generalised sensory polyneuropathy. (figure 1)

The patient was discharged after 14 days of hospital stay with 4+/5 power in upper limbs and 5/5 power in right lower limb and 4+/5 in left lower limb with persistence of areflexia.

**DISCUSSION:**

GBS is an immune mediated disorder. Occurrence of GBS is rare in pregnancy. The incidence of GBS has been reported to be very low during pregnancy.[1,4] The understanding of the pathogenesis revolves around cellular immunity. There are three sets of helper T cells:

1. Th1 cells secrete pro-inflammatory cytotoxic cytokines
2. Th2 cells secrete anti-inflammatory non-cytotoxic cytokines
3. Treg (regulatory T cells) cells play a role in induction of tolerance to fetal alloantigen.

During pregnancy because of altered physiology of hormones, the Th2 cytokines predominate over Th1 cytokines and Treg activity increases during the first and second trimester. This physiologic alteration explains the rarity of GBS in first and second trimester and increase in incidence in the third trimester and post partum.[1,6] About two thirds of patients have an infection within the previous 4-6 weeks. The implicated organisms which may trigger an attack of GBS are Mycoplasma pneumoniae, Campylobacter jejuni, Cytomegalovirus, and Ebstein-Bar virus and the usual presentation is gastroenteritis.[7] Molecular mimicry plays an
important role where the preceding infection causes an autoimmune response against the various components of the peripheral nerve myelin and sometimes the axon. The classical presentation is with pain, numbness, paraesthesia, or weakness of the limbs. The management of GBS in pregnancy is similar to that in the non-pregnant population and includes intravenous immunoglobulins (IVIG), plasmapheresis, and ventilator support wherever required. The usual indication of mechanical ventilation are, decrease single breath count less than 20, bulbar palsy with aspiration pneumonias., use of accessory muscles or arterial blood gases showing pO2 45mmHg. Immunomodulation with plasmapheresis and IVIG has been found to improve treatment outcomes with full recovery in 70-80% of patients.[8] Ventilatory support is required in 25-30% of non-pregnant patients, but respiratory problems may be worse in pregnancy because of splinting of the diaphragm. In cases requiring ventilatory support in pregnancy, the risk of premature birth has been noted to be greatly increased.[9]

CONCLUSION:
To conclude, though, GBS is a rare occurrence in pregnancy it can be associated with severe co-morbidities like respiratory failure and autonomic dysfunction. A high index of suspicion should be maintained by the treating physician. Postpartum relapses may occur. The efficacy of IVIG is well-established and can be safely given during pregnancy. Early diagnosis, multidisciplinary input and prompt immune-modulatory therapy are the cornerstones in management of GBS during pregnancy and improve outcomes for the mother and foetus.

REFERENCES: