Guillain Barre Syndrome: A Rare Cause of Floppy Neonate

Sir,
The floppy infant represents a diagnostic challenge with a wide range of possible diagnoses including central or peripheral nervous system abnormalities, myopathies, genetic disorder, endocrinopathies, and metabolic diseases.[1] Guillain Barre Syndrome (GBS) is an acute inflammatory demyelinating polyneuropathy usually triggered by antecedent factors like infections, maternal diseases, perinatal infections, and recent vaccination.[2] GBS is rare in neonatal period and should be suspected when motor conduction velocity is slowed in floppy infant.

We had a 25-day-old male neonate with history of inability to move limbs. Initially, both lower limbs were affected and later progressed to upper limbs in 4–5 days with respiratory distress. On further inquiry, history of fever, sneezing, and cough was present 1 week prior to admission but there was no history of diarrhea, top feed, recent vaccination, or drugs prescription such as aminoglycoside. He was born at 38 weeks of gestation to a 23-year-old primi mother, through vaginal delivery with birth weight of 2.7 kg and Apgar score of 8 and 10 at 1 and 5 min, respectively. Mother had received adequate antenatal care with no history of maternal diseases like SLE, inflammatory bowel disease, hypo- or hyperthyroidism, diabetes, myasthenia gravis, genetic diseases, drug exposure such as magnesium sulfate, immunomodulator therapy, or infection such as CMV or radiation exposure. Mother had an uncomplicated perinatal period. Family history was not significant.

On physical examination, baby was alert with respiratory distress (SpO2 80% off oxygen), with normal physical growth without any obvious congenital malformations or dysmorphism. The pupillary responses were normal and symmetric. Ocular movements were full and no ptosis was present. Facial strength was normal. There were no tongue fasciculation and lingual papillae were present. Suck and swallow co-ordination was poor. The withdrawal of limbs to light touch and painful stimuli was absent and there was no tremor. Serial neurological examinations documented ascending paralysis culminating in absent spontaneous limb movement, severe generalized axial, and appendicular hypotonia and areflexia. Autonomic instability marked by urinary retention and heart rate variability was also present.

Cerebrospinal fluid analysis showed protein 98 mg/dL, sugar 64 mg/dL, and microscopy revealed 3–4 lymphocytes per high-power field suggestive of albuminocytological dissociation. Motor nerve conduction studies on surface electrodes showed no response to stimulus from bilateral median, ulnar, femoral, tibial, and peroneal nerves while sensory nerve conduction showed reduced SNAPs amplitude in bilateral median and ulnar nerves and no response elicited in bilateral sural nerves. F wave could not be elicited since “M” response was absent. Needle examination showed increased insertion activity but no spontaneous activity was noted. Test for serum antibodies for Cytomegalovirus and mycoplasma were negative. Blood investigations including serum electrolytes, renal function test, liver function test, thyroid function test, creatinine phosphokinase, and C-reactive protein were normal. Blood culture was negative. Polymorphonuclear leucocytosis was reported on peripheral smear.

The diagnosis of GBS was made on the basis of clinical finding, nerve conduction study and CSF finding. Baby was treated with intravenous immunoglobulin, 0.4 mg/kg/day for 5 days with supportive therapy. Improvement was seen after 48 h with resolution of autonomic instability and gradual recovery.
of muscle strength. Baby was discharged after 2 weeks and followed up for 6 months. There was appropriate growth with no sequelae or recurrence. Since the poliomyelitis has been nearly eliminated, the GBS is currently the most frequent cause of acute flaccid paralysis. The syndrome was named after the French physician Guillain, Barre, and Strohl (1916) described a benign polynuropathy in allergic polynuropathy in cerebrospinal fluid.(3) GBS is extremely rare in neonatal period/early infancy and it is usually occurs after the age of 3 years. (4) A previous infection should always be searched particularly when trying to define the presence of some microbiological agents more frequently related to GBS. Among these events the most frequent was unspecific upper respiratory tract infection followed by diarrhea. Our patient had upper respiratory infection one week prior to illness like most of the studies in literature noted that antecedent factors associated with occurrence of GBS. (5) In relation with clinical presentation, the main feature is progressive bilateral and relatively symmetric weakness that progresses over a period of 12 h to 28 days before a plateau is reached with hyporeflexia or areflexia. Our patient had classical clinical presentation of symmetrical bilateral lower limb weakness with areflexia. We did not find cranial nerve involvement but dysautonomic signs and symptoms in form of urinary retention and heart rate variability.

Al-Quadah AA et al. reported a similar case of neonatal GBS at birth. She had generalized hypotonia, paucity of limb movement, diminished stretch reflex and at 3 weeks of age motor nerve conduction study demonstrated evidence of demyelination and axonal involvement. (6) Luijckx GL et al. revealed neonatal GBS whose mother was suffering from GBS at 29th weeks of pregnancy, (7) while Jackson et al. published a case of in utero-acquired congenital Guillain Barre syndrome with maternal autoimmune disorder. (8) Similar to our case, Anastasopoulou et al. and Sharma et al. recently reported a case of acute inflammatory demyelinating polyneuropathy in newborn with healthy mother. (9,10) Buchwald et al. reported neonatal GBS due to transplacental transfer blocking antibodies from mother to child. They performed experiment on the hemidiaphragms of adult mice and neonatal and juvenile rats and demonstrated depressed quantal content by approximately 90% from serum of mother and infant and concluded transplacentally transferred blocking antibodies may be specifically directed at epitopes of the mature but not the fetal neuromuscular junction. (11) The pathophysiology which links demyelinating polyneuropathy and maternal inflammatory bowel disease is not well understood. However, Bamford et al. reported congenital GBS associated with maternal inflammatory bowel disease and responsive to intravenous immunoglobulin. (11) GBS though extremely rare in neonatal age group, it should be considered differential diagnosis of floppy neonate.

Declaration of patient consent
The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest
There are no conflicts of interest.

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