Case Report

Congenital Guillain-Barre Syndrome: A Rare Entity

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Abstract

A term male infant presented with generalized hypotonia, paucity of lower limb movements, and diminished muscle stretch reflexes. At 3 weeks of age, motor nerve conduction studies demonstrated evidence of demyelination and axonal involvement. These findings indicated demyelination and patient was diagnosed to have congenital Guillain-Barre syndrome. Improvement was seen after a course of IVIG. We are reporting this case because of rarity of its occurrence and should be considered in differential diagnosis in floppy neonate.

Key words

Guillain- Barre syndrome, Congenital, Floppy neonate, IVIG.

Introduction

Guillain-Barre syndrome is an acute inflammatory demyelinating polyneuropathy that is rarely diagnosed in neonatal period. The diagnosis of Guillain-Barre syndrome is suspected when motor nerve conduction velocities are slowed in a floppy neonate. We reported here a case of term male newborn who had the clinical manifestations and accompanying electrophysiologic studies to fulfil the criteria of Guillain-Barré syndrome.

Case report

A male baby was born at term gestation to 28 year gravida 2, para 1 mother. Labour and delivery were uneventful. Antenatal history was unremarkable except that mother reported decreased fetal movements 1 week before delivery. Apgar scores were 8 and 9 at 1 and 5 minutes respectively. Birth weight was 3000 gm, length 49 cm and head circumference was 34.5 cm. The baby was referred to a tertiary centre after birth as baby was hypoactive, hypotonic

and poor sucking. After admission baby was found to have generalized hypotonia, reduced limb movements and diminished moro and stretch reflexes. Baby required brief ventilator assistance. Hematological investigations, head ultrasound, MRI brain and metabolic workup was normal. Baby did not receive any vaccine (oral/parenteral) before admission and during hospital stay.

At 3 weeks of age, oxygen supplementation was required for desaturations. Serial neurological examinations showed ascending paralysis and areflexia. Autonomic instability was present, marked by unstable heart rate and increasing episodes of apnea requiring intubation.

Electromyography (EMG) revealed abnormal spontaneous activity in biceps, quadriceps and tibial muscles. Nerve conduction studies failed to elicit motor or sensory response. Creatine kinase was in normal limits.EEG was also in normal limits. CSF analysis done at 28 days of life was also normal, with protein 45mg/dl, glucose 86 mg/dl and total cell count 4/mm^3 (all lymphocytes). Repeat CSF done on 38 day of life showed a protein rise to 120 mg/dl, glucose 60 and total cells 5/mm^3 (all lymphocytes). Muscle biopsy done was normal. Clinical picture of ascending weakness and areflexia, with normal muscle biopsy suggested demyelination. IVIG 500 mg/kg/day was given over 5 days. Improvement was seen after 72 hours with rapid resolution of autonomic instability and gradual descending recovery of strength. Baby was being followed up and at the age of 10 months child recovered completely with normal tone and tendon reflexes.

Discussion

The acute and chronic form of inflammatory demyelinating polyneuropathy have been reported only rarely in the neonates [1-4]. In all cases, hypotonia is apparent at birth with weakness and hyporeflexia. Where tested, motor nerve conduction velocities were reduced or absent. In congenital acute inflammatory demyelinating polyneuropathy or Guillian-Barre syndrome, early improvement occurs within first few months with complete recovery by one year. Our patient was symptomatic at birth with diffuse hypotonicity followed by ascending flaccid quadriparesis, areflexia and autonomic instability. The differential diagnosis of such floppy neonate is broad [5], however consistent with demyelinating disorder, our electrophysiological studies revealed evidence of demyelination and axonal involvement [6-8].

There are case reports which suggest an association with maternal inflammatory bowel diseases and neonatal GBS [9, 10]. The relationship between maternal inflammatory bowel disease and congenital Guillain Barre Syndrome suggest an immune mediated process. Direct neonatal antibody synthesis by neonate against a maternal antigen can be a consideration; however in our patient we could not find any maternal cause.

These babies may be diagnosed with transient neonatal hypotonia or would not come to medical attention because of the minor nature of the symptoms. Conversely these infants may be severely affected with resulting miscarriage and early infant death.

Treatment might not be required and there is gradual improvement over first few months with almost complete recovery by 1 year of age, however rapidly progressing ascending paralysis needs treatment with IVIG. Further studies are awaited regarding epidemiology, pathogenesis, outcome of such neonates and spectrum of disease.

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