Original Research Paper

Volume-9 | Issue-4 | April-2019 | PRINT ISSN No 2249-555X

Paediatrics

GUILLAIN-BARRE SYNDROME - AN ATYPICAL PRESENTATION

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ABSTRACT Guillain-Barre Syndrome is a post-infectious polyneuropathy involving mainly the motor, but sometimes the sensory and autonomic nerves as well. We present an atypical case of Guillain-Barre Syndrome in a 12 year old boy.

Male child presented with loose stool 3 days back for one day followed by abrupt onset of numbness in the palms and fingers on the second day and weakness with pain in the calf and ankle on the third day. On central nervous system examination, power in upper and lower limbs was 3/5 with normal tone and intact superficial reflexes. Elicitation of deep reflexes revealed absent ankle jerk, while knee jerk, biceps and triceps reflexes were normal. Laboratory parameters were normal and EMG-NCV was suggestive of acute demyelinating polyneuropathy, likely Guillain Barre Syndrome. Patient was started on intravenous immunoglobulin infusion @ 1gm/kg/day and monitored for progression of symptoms.

KEYWORDS : Guillain-Barre syndrome, acute demyelinating polyneuropathy, immunoglobulin.

1. Introduction:

Guillain–Barré syndrome (GBS) constitutes an important proportion of acute flaccid paralysis cases world-wide. It is a condition characterized by various degrees of weakness, sensory abnormalities, and autonomic dysfunction due to damage to peripheral nerves and nerve roots.[1] Although the underlying etiology and pathophysiology of GBS are not completely understood, it is believed that immune stimulation plays a central role in its pathogenesis.[2] It is considered to be an immune-mediated disorder resulting from generation of autoimmune antibodies and/or inflammatory cells which cross-react with epitopes on peripheral nerves and roots, leading to demyelination or axonal damage or both.[3] First described by French neurologists Guillain, Barré, and Stohl in 1916, understanding of the disorder has increased tremendously in the past 2 decades.[4] Guillain-Barre Syndrome is a post-infectious polyneuropathy involving mainly the motor, but sometimes the sensory and autonomic nerves as well. The annual incidence of GBS has been estimated at between 0.4 and 4.0 cases per 100,000 populations per year. [5-7] Many studies have suggested that men are more likely to be affected than women. Most cases are sporadic and there does not appear to be a seasonal pattern, with some exceptions.[8] Clinically, GBS is characterized by the acute or subacute onset of varying degrees of weakness in limbs or cranial nerve-innervated muscles, associated with hypo- or areflexia, and a characteristic profile in the cerebrospinal fluid (CSF).[1,9] We present an atypical case of Guillain-Barre Syndrome in a 12 year old boy.

2. Case Report:

12 year old male child born of non-consanguineous marriage, hailing from Uttar Pradesh, presented with complains of loose stools 3 days back for one day, followed by abrupt onset of numbness in the palms and fingers on the second day and weakness with pain in the calf and ankle on the third day. Loose stools were watery in consistency, 4-5 episodes/day, non- blood tinged. Numbness began on the second day of illness, initially in the fingers and palms, was acute in onset, progressive, and associated with difficulty in distal movements (buttoning/unbuttoning, writing). This progressed to weakness in calf and feet the following day, progressive, associated with pain and difficulty in range of movement, and not associated with any diurnal variation. There was history of difficulty in walking. No history of fever, upper respiratory tract infection, trauma, joint pain. No history of bladder or bowel involvement, convulsions, cranial nerve involvement, breathing difficulty. He was the first order child with no significant family history. Birth and developmental history was normal. Child was immunized till 5 years of age as per the national immunization schedule. Socio-economically, he belonged to the lower middle class.

On examination, patient was conscious, cooperative, with heart rate of 98/min and respiratory rate of 26/min. There was no wasting or stunting. On central nervous system examination, power in upper and lower limbs was 3/5 with normal tone and intact superficial reflexes. Elicitation of deep reflexes revealed absent ankle jerk, while knee jerk, biceps and triceps reflexes were normal. Blood haemogram was normal with haemoglobin of 12.5 gm/dl, total leucocyte count of 8000/cmm and platelets of 3,66,000/cmm. Electrolytes and serum calcium levels were normal (sodium: 140, potassium: 4, chloride: 103, calcium: 9.7 mg/dl). Cerebrospinal fluid analysis revealed albumin-cytological dissociation. An EMG-NCV was done which revealed acute demyelinating polyradiculoneuropathy, likely Guillain-Barre Syndrome.

Patient was started on intravenous immunoglobulin @ 1gm/kg/day for 3 days and was monitored for symptoms. He tolerated the treatment well with no progression of symptoms and was discharged on the 7th day of admission. Currently, as per the Guillain-Barre disability scale, patients has grade 2 disability (i.e. - Able to walk without support of a stick (5 m across an open space) but incapable of manual work/running). He is on regular physiotherapy sessions and a close follow-up.

Table: 1- Guillain–Barré syndrome disability scale[10]

0	Healthy
1	Minor symptoms or signs of neuropathy but capable of manual work/capable of running
2	Able to walk without support of a stick (5 m across an open space) but incapable of manual work/running
3	Able to walk with a stick, appliance, or support (5 m across an open space)
4	Confined to bed or chair bound
5	Requiring assisted ventilation (for any part of the day or night)
6	Death

3. Discussion:

Progression of weakness in Guillain - barre syndrome is usually ascending, known as Landry Ascending Paralysis, gradual in onset and appearing approximately 10 days after an initial infection. Our patient presented with an atypical presentation of rapidly progressing descending paralysis within hours of a gastrointestinal infection.

Early diagnosis of the disease on the basis of clinical and electrophysiological findings, prompt initiation of immunoglobulin infusion and close monitoring of the patient is essential in halting the progression of the condition.

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