Case Report

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A Rare Variable Presentation of Paediatric Gullain-Barre Syndrome: A Case of Acute Flaccid Paralysis

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Abstract

A 2.6 years old boy previously healthy presented with h/o mild fever and generalized weakness 10 days back following which he became irritable and lethargic and lower limb weakness for 3 days. Plain MRI brain was normal but the CSF study showed albumino-cytologic dissociation and Nerve conduction study showed absence of 'f' waves. He was started on IV Human Immunoglobulin (IV Ig) after which he improved and was discharged in stable condition.

Keywords: Gullain-Barre Syndrome; Acute Flaccid Paralysis; Autoimmune Disorder; Iv Human Immunoglobulin; India.

Introduction

Guillain Barre Syndrome is an acute, immune mediated polyneuropathy and an important cause of acute flaccid paralysis worldwide. There are antibodies directed against myelin sheath and axons of peripheral nerves are formed in response to preceding viral or bacterial illness, trauma, surgery or vaccination. GBS have heterogenous presentation, course and intensity. Diagnosis may be delayed especially in patients with atypical presentation, including pain and in young children. GBS is common in adults with slight preponderance for males, rare in children. This is a common cause of acute and subacute flaccid paralysis in children. Onset of symptoms are 2 to 4 weeks after respiratory or diarrheal illness, which includes ascending symmetric weakness or paralysis and areflexia or hyporeflexia [1,5] Incidence 0.4- 1.9 per 100,000.

Case Study

2.6 years male child came to ED with presentation generalised weakness for 7 days, irritability and ? pain in limbs for 7 days. There is history of fever for 3 days, 7- 10days prior these symptoms.

His parents noticed that he preferred to lie down most of the time and was unable to get up from lying down position since 3 days and he cried excessively when his legs were touched although he was moving his arms normally.

There was occasional cough while eating and had some change in his voice.

He was being treated as a normal viral fever with myalgia prior to coming to our ED. No fever now and history of bowel and bladder incontinence.

Physical examination revealed the child was conscious and oriented, breathing normally with vitals signs PR:98/min, BP: 100/60 mmhg, RR:18/min, regular, SpO_2: 100% on room air, Temperature: 98.4 F, RBS: 90mg/dl, Cardiac monitor: sinus rhythm. His Airway – Patent, Breathing- Spontaneously, Chest - AE B/L equal, no added sounds.

On CNS Examination – GCS 15/15, conscious

- power: upper limbs – b/l-5/5
  lower limbs – b/l -3/5
- DTR and Superficial reflexes- not elicitable
- All cranial nerves – normal,
- Gag reflex+, Hoarseness of voice +
- Cerebellar signs – could not ellicitable

He had no hypothermia or rash.

Cardiovascular, Respiratory and Per abdominal examinations were insignificant. In view of above findings, Neurology and Paediatric consultations were requested, radiological and PICU admission planned.

Course in the Hospital and Outcome: Patient investigated in hospital MRI BRAIN and SPINE SCREENING: Normal study.

CSF Study: Albumino – cytological Dissociation, Nerve conduction study: F wave absent. All other blood investigation – within normal limit. Patient was treated with IV Ig G. patient started improving in subsequent hospitalization and did not required ventilatory support. His lower limb power improved and discharged in stable condition.

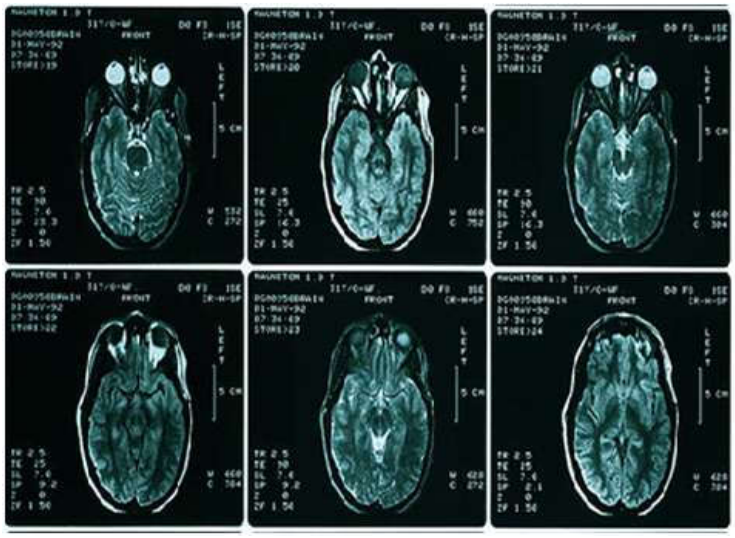
Discussion and Therapeutic considerations:

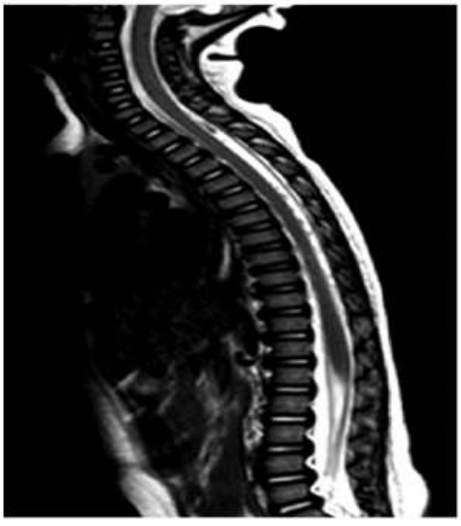
Guillain-Barré syndrome (GBS) is a disorder in which the body›s immune system attacks part of the peripheral nervous system. The first symptoms of this disorder include varying degrees of weakness or tingling sensations in the legs. In many instances the symmetrical weakness and abnormal sensations spread to the arms and upper body. If respiratory involvement is pronounced, such an individual is often put on a ventilator to assist with breathing and is watched closely for problems such as an abnormal heart beat, infections, blood clots, and high or low blood pressure. Most individuals successfully managed in ICU, although some continue to have a certain degree of weakness [1-2,5].

The pathogenesis of GBS remains unclear. Increasing data indicate that it is an autoimmune disease, often triggered by a preceding viral or bacterial infection with organisms such as *Campylobacter jejuni*, cytomegalovirus, Epstein-Barr virus, or *Mycoplasma pneumoniae*. Vaccination against the flu, rabies, and meningitis are also documented precipitating factors that have been reported [3].

When Guillain-Barré is preceded by a viral or bacterial infection, it is possible that the virus has changed the nature of cells in the nervous system so that the immune system treats them as foreign cells. It is also possible that the virus makes the immune system itself less discriminating about what cells it recognizes as its own, allowing some of the immune cells, such as certain kinds of lymphocytes and macrophages, to attack the myelin. Sensitized T lymphocytes cooperate with B lymphocytes to produce antibodies against components of the myelin sheath and may contribute to destruction of the myelin. In two forms of GBS, axons are attacked by antibodies against the bacteria Campylobacter jejuni, which react with proteins of the peripheral nerves [1-2].

The diagnosis of GBS is typically based on the presence of a progressive ascending weakness with areflexia and CSF analysis and electrophysiological study of nerve and muscles [6]. To date, treatment for GBS has been aimed primarily at immunomodulation. In pediatrics, the most effective form of therapy is generally considered to be intravenous immunoglobulin (IVIG). In general, the outcome of GBS is more favorable in children than in adults; however, the recovery period is long, often weeks to months. Rarely, it can be fatal in 5-10% of patients with respiratory failure and cardiac arrhythmia [3].





Conclusion

Diagnosing GBS in pediatric population is often delayed due to its variable presentation. This is very challenging and needs high degree of suspicion which was meticulously used in our patient who presented with variable symptoms. GBS has better prognosis in children as compared to adults and shows recovery within few weeks after being treated with IV Ig.

This case demonstrates that the physicians must be highly aware of the symptoms of paediatric GBS so as to correctly diagnose the disease early and not to confuse with other differentials like meningo-encephalitis, poliomyelitis etc.

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