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Guillain Barré Syndrome in A 3-Year-Old Child: A Rarity

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Abstract

Guillain Barré syndrome (GBS) is a group of neuropathic conditions characterized by progressive limb weakness and hyporeflexia that might lead to quadriparesis and ultimately death. GBS is the most common cause of acute flaccid paralysis but is a rare disease in children (0.34-1.34/100,000). This study is a case of a three and half year-old male who presented with ascending symmetrical bilateral paralysis of lower limbs eventually involving all limbs which was diagnosed as GBS. This case signifies the importance of a high index of clinical suspicion and the start of prompt treatment for early recovery from the disease.

Keywords: NIL

INTRODUCTION

Guillain Barré syndrome (GBS) is a group of neuropathic conditions characterized by progressive limb weakness and hyporeflexia that might lead to quadriparesis and ultimately death [1]. The disease is autoimmune in nature such that the immune system mis-identifies its own nerve cell peptides with that of the peptides of foreign bacteria or viruses and facilitates action of autoreactive T and B cells. This is called molecular mimicry [2]

Worldwide prevalence of Guillain Barré syndrome in the paediatric age group is between 0.34 and 1.34/100,000 making it a very rare disease [3] [4].

CASE REPORT:

A 3 and half year-old male child from a rural background presented with complaints of pain and progressive weakness in both lower limbs since 4 days and complete loss of ability to walk. He also experienced difficulty in swallowing since 2 days. On further questioning, the parents of the patient mentioned that he had developed fever, cough and cold with abdominal pain and loss of appetite 6 days back for which they received treatment from a local doctor. The parents observed weakness in the lower limbs of the child two days after the fever resolved which progressively increased and ascended symmetrically, for which he was referred to our tertiary care centre.

There was no significant past history and family history. The parents denied any history of diarrhoea. They also stated that the patient has had no significant medical problems in the past and that he was completely immunised as per the National Immunization Schedule of India.

At the time of admission, the child was conscious, oriented, emotionally disturbed and had a poor general complexion. He was afebrile with stable vitals. His preliminary neurological examination revealed sluggish knee reflex and decreased tone in all four limbs with power of grade 0 in both lower limbs and grade 3 in both upper limbs. On the second day of admission, he had intermittent apnoea and bilateral crepitus was heard. The child suffered with two

episodes of generalized tonic clonic seizures with up rolling of eyeballs and drooling of saliva. The seizures were effectively controlled and the child was put on nasogastric tube. However, his respiration was not affected and he did not require artificial ventilation.

Complete blood count, ESR, Mantoux test, serum calcium, kidney function test, CSF examination, culture & sensitivity report, chest X-ray and MRI of the brain was advised. MRI revealed an incidental finding of a subarachnoid cyst with dimensions of 42*14*25 mm in left basi temporal lobe, CSF was of hazy appearance, had protein content of 45 mg/dl, sugar content of 37 mg/dl and cytology indicated predominantly polymorph with occasional lymphocytes.

A clinical diagnosis of Guillain Barré syndrome was made. The patient was then started on intravenous immunoglobulin (IV IG) with the dose of 90ml at the rate of 13ml per hour for the first half hour and then at the rate of 27ml per hour. On the third day, physiotherapy was started. Nasogastric tube was removed on the fifth day. Child was able to sit with support and tolerate liquid diet on the 6th day. By the 13th day, the tone of limbs improved significantly to grade 4 in all limbs, respiration was normal and the child tolerated solid food. Therefore, after 12 days of IVIG administration and physiotherapy, the patient recovered enough to be discharged. The parents were advised to follow up and continue physiotherapy.

DISCUSSION:

After the eradication of polio in most countries of the world, Guillain Barré syndrome is now the most common cause of acute flaccid paralysis in children (25.9%–51%) [5]. With supportive findings in patient history like a recent respiratory infection [6], loss of appetite, absent bowel and bladder movements, neurological examination showing bilaterally symmetrical ascending paralysis and CSF findings such as slightly raised protein levels (45 mg/dL) [7], we could make a clinical diagnosis of Guillain Barre Syndrome.

Differentials for GBS included Enterovirus D-68 infection [8], poliomyelitis, transverse myelitis, acute compressive lesions, Neuromyelitis Optica and meningitis. Viral culture from stool sample was not available, but the clinical recovery response of the patient to IVIG excluded the differential of Enterovirus D-68 infection. The patient was immunised and poliomyelitis has been eradicated in India since 2014 [9], transverse myelitis was ruled out due to inconsistency with the features of its paralysis, and Neuromyelitis Optica was ruled out due to absence of signs of optic nerve paralysis. Absence of neck rigidity and fundal changes indicated normal CSF pressure that ruled out meningitis and compressive lesions of spinal cord.

Intravenous immunoglobulin (IV IG) are immunoglobulins derived from donor blood that act against the autoreactive T and B cells that are involved in Guillain Barré syndrome and are therefore, the mainstay of treatment [10]. With IVIG, improvement was assumed because of the following events: 1) Overall improvement in general complexion; 2) Significant increase in tone and power of the limb muscles was observed, and 3) improvement in appetite such that he could tolerate solid food. Since recovery was evident as soon as the treatment started, electrophysiological studies to determine the subtype of Guillain Barré syndrome were deemed unnecessary.

Classically, in GBS, the disease progresses up to two weeks [11]. However, our patient showed significant improvement by the end of this period which could be because of early patient presentation to the tertiary care centre, prompt clinical diagnosis and immediate initiation of treatment.

CONCLUSION:

Though Guillain Barré syndrome is a rare presentation in the paediatric age group (0.34-1.34/ 100,000), with every presentation of AFP, there must be a high index of suspicion for GBS. Investigations like electrophysiological tests, MRI, viral culture may not always be available, especially in developing countries, suggesting the importance of clinical diagnosis. Paediatricians should be well versed with the varied presentation of the disease because clinical diagnosis is also crucial for administering prompt IVIG treatment for GBS proved by the early recovery of the patient.

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Volume 4, Issue 4; July-August 2021; Page No 1242-1244

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Volume 4, Issue 4; July-August 2021; Page No 1242-1244