Original article

Guillain-Barré Syndrome in Nepalese Children

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Background: Guillain-Barré Syndrome (GBS) is a group of autoimmune syndromes consisting of demyelinating and acute axonal degenerating forms of the disease. This study gives the profile of 30 patients who were admitted with the diagnosis of GBS in a pediatric hospital.

Methods and material: We reviewed the admitted case notes of children admitted with the diagnosis of GB syndrome from January 2005 to December 2007. The data were analyzed to find out the demographic features, preceding illness, clinical features and CSF findings.

Result: The age ranged from 3.6 years to 14 years. The mean (SD) age was 4.9 (9.7) years. The most common symptoms were inability to walk after awaking in the morning.

The median evolving time from the onset of illness to nadir was 12 days. The CSF protein concentration ranged from 60 to <100mg% and the CSF cell counts were less than 10/cmm in 23 patients and 11-19 in 7 patients.

Conclusion: We could not find any previous study done in such a large paediatric population of this country. Our results show the similarity in pattern of age distribution, preceding events, and cell protein dissociation similar to that of other studies.

Keywords: Guillain-Barré Syndrome, diagnostic criteria, children, clinical profile, hospital.

Introduction

Guillain-Barré Syndrome (GBS) is a world wide disease[1]. It is an acute peripheral neuropathy causing limb weakness that progresses over a time period of days or at the most, up to 4 weeks. The median annual incidence of 1.3 cases per population of 100,000[2]. Based on pathological and electrophysiological studies it is of three types [3]. These are acute inflammatory demyelinating polyradiculoneuropathy (AIDP), acute motor and sensory axonal neuropathy (AMSAN), and acute axonal neuropathy (AMAN). These different types of GBS have prediction for different geographical areas. AIDP predominates in the United States and AMAN is the predominant form in China [4]. GBS has been associated with antecedent bacterial and viral infections [5], administration of certain vaccinations [6], and other systemic illnesses. There has been no systematic study of this syndrome in Nepalese children. We have collected these cases to see the clinical symptomalogy of GBS in Nepalese children admitted in a paediatric hospital based on the accepted criteria [7].

Diagnostic Criteria for Typical Guillain-Barré Syndrome [7,8]

Features required for diagnosis

Progressive weakness in both legs and arms
Areflexia

Features strongly supporting diagnosis

Progression of symptoms over days, up to four weeks
Relative symmetry of symptoms
Mild sensory symptoms or signs

Cranial nerve involvement, especially bilateral weakness of facial muscles
Recovery beginning two to four weeks after progression ceases
Autonomic dysfunction
Absence of fever at onset
High concentration of protein in cerebrospinal fluid, with fewer than 10 cells per cubic millimeter
Typical electrodiagnostic features

Features excluding diagnosis

Diagnosis of botulism, myasthenia, poliomyelitis, or toxic neuropathy
Abnormal porphyrin metabolism
Recent diphtheria
Purely sensory syndrome, without weakness

Patients and methods:

We reviewed the medical case history of children admitted with the diagnosis of GB syndrome from January 2005 to December 2007. We analyzed the age, sex, clinical manifestations which included the initial symptoms and signs, progression of the disease while in hospital, duration of hospital stay, CSF findings. Only those patients with the GB syndrome showing CSF cell protein dissociation was included in the study. The electrophysiological studies were not done in any of these patients. At the time of their maximal deficits during admission to hospital, patients were graded using a disability scale modified from Hughes et al.[9] Grade 0 is defined by normal functional state without neurological deficits; grade 1, minor symptoms or signs but being able to do manual work; grades 2 and 3, ambulation without or with assistance respectively; grade 4, chair or bed bound; grade 5, requiring mechanical ventilator; and grade 6, death.

Results:

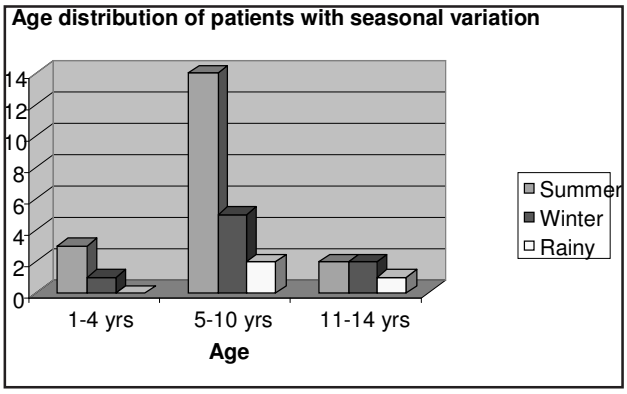
During this period 30 patients were admitted with the diagnosis of GB syndrome. All patients had the CSF findings suggestive of cell protein dissociation. None of our patients had electrophysiological studies done as these facilities were not available in our centre.

Table 1. Demographic features, preceding illness, clinical features and CSF findings

Mean age in yrs	9.7
Sex male/female	13/17
Preceeding events	
Rash only	1
Upper respiratory infection with rash	3
Fever	7
Diarrhoea	9
Clinical featrues	
Limb weakness	30
Sensory disturbances	17
Facial weakness	1
Bulbar dysfunction	1
Autonomic dysfunction	
Hypertension	12
Hypotension	2
Bladder dysfunction	3
Ataxia	8
CSF	
Protein>100 mg%	30
Cell <10/cmm	23
Cells 10-20/cmm	7

Age, sex and seasonal distribution:

The age ranged from 3.6 years to 14 years (14 years is the upper limit of patients who can attend this hospital). The mean (SD) age was 4.9 (9.7) years. 13 patients were male and 17 patients were female. 14 patients in the age group of 5-10 years were admitted during the summer (Chaitra-Ashad: April-July) season.



Preceding illness:

Nine patients had preceding history of diarrhea, three had cold and four had rash.

Clinical features:

The most common symptoms were inability to walk after awaking in the morning. Six children aged 6-12 also complained of tingling sensation in the lower limbs bilaterally. Only one patient had left sided facial palsy. One patient had ascending paralysis and was referred for ventilatory support and he was kept in ventilator in another hospital on 5th day. Six patients had gradual involvement of

upper limbs bilaterally. The median evolving time from the onset of illness to nadir was 12 days

Cerebrospinal fluid: cell and biochemistry

Lumber puncture was performed in all children suspected of GB syndrome and only those children were included in the study had cell protein dissociation. All lumber puncture were done within one month (range 6 - 30 days). The CSF protein concentration ranged from 60 to <100mg%. We could not calculate the mean as the laboratory reported up to 100 or more than 100mg%. The CSF cell counts were less than 10/cmm in 23 patients and 11-19 in 7 patients.

Discussion

We could not find any study done in paediatric population of this country. This study does not give the type of GB syndrome relating with electrophysiological study but gives the profile in a group of children admitted in a paediatric hospital. Most of the study shows the higher incidence in adults but occasional higher incidence in children has been suggested.[10, 11] We could not compare with adult population in this study as our population was children only. Our results show the similarity in pattern of age distribution similar to that of northern China[12]. The seasonal preponderance of summer months was also seen in northern china and Paraguay as ours [12,13]. In our study only one patient had bulbar involvement and another one child had cranial nerve involvement. Cranial nerves are affected in over 50% of all cases, with the facial nerves being affected the most.. Bilateral oculomotor nerve palsy has also been reported [16].

We could not correlate our patients with infectious diseases as this is the retrospective study. There are enough reports of Camphylobacter jejuni, Mycoplasma pneumoniae, Hepatitis A virus, Cytomegalovirus, Epstein-Barr virus, and Hepatitis B virus associating with GB syndrome [14]. Preceding events were identified in only 43% of our population which is close to other series [15].

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