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To Evaluate The Clinical And Electrophysiological Aspects of Guillain Barre Syndrome In PBM Hospital Bikaner In Children Age Group.

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Abstract

Background: Guillian Barre Syndrome (GBS) is an acute, frequently severe and fulminant polyradiculoneuropathy that is autoimmune in nature, involving mainly motor but also sensory and sometimes autonomic nerves.

Methods: The study was carried out in the Department of Paediatrics, S.P. Medical College & Associated Group of Hospitals, Bikaner, Rajasthan. The study subjects included all patients aged 15 years and below with clinical presentation of Guillain Barre syndrome.

Results: Clinically 19 patients (61.29%) were admitted with quadriparesis and 12 patients (38.71%) with paraparesis. In maximum cases the proximal and distal muscles in each limb were equally affected. In 12 patients (38.70%) bulbar muscles were involved. 32.26% of them required ventilatory support. Areflexia was present in all the patients. In our study the mixed subtype is the commonest electrophysiological pattern of that is equally distributed in all the age groups of the Guillain Barre syndrome patients in our zone present in (45.16%) cases followed by axonal (29.03%), demyelinating (16.12%), inexcitable (6.45%) and normal pattern.

Conclusion: Thus by our study we conclude that GBS should be suspected in all the patients presenting with acute flaccid paralysis in different age groups. Mixed type is the predominant electrophysiogical pattern seen in almost all age group.

Keywords: Guillain Barre Syndrome, Paraparesis, Quadriparesis.

Introduction

Guillian Barre Syndrome (GBS) is an acute, frequently severe and fulminant polyradiculoneuropathy that is autoimmune in nature, involving mainly motor but also sensory and sometimes autonomic nerves. The Guillain Barre Syndrome is the commonest cause of acute flaccid paralysis in much of the world, after the introduction of vaccines for Poliomyelitis[1].

Although there are no universally accepted criteria for electrophysiological identification of various subtypes in the developed countries. The electrophysiological study may play an important role in further investigation of the pathogenesis and assessment of prognosis[2]. Based on electrophysiology and pathology, GBS can be divided into

either predominantly demyelinating or predominantly axonal pattern and can be also in mixed pattern[3].

Several studies in past years showed that clinical and electrophysiological feature of GBS in China and India were different in some way from those in developed countries[4-6]suggesting predominantly AMAN pattern.

We planned this study to see the electrophysiological pattern in patients with GBS at the time of admission along with its relation to disability pattern. The same parameters were followed at one month ±7 days and the data was analyzed. Any relation to prognosis in different variants was also studied.

Material And Methods

The study was carried out in the Department of Paediatrics, S.P. Medical College & Associated Group of Hospitals, Bikaner, Rajasthan.

The study subjects included all patients aged 15 years and below with clinical presentation of Guillain Barre syndrome admitted in Department of Paediatrics, PBM & Associated Group of Hospitals, during January 2018 to December 2018. The clinical diagnosis of Guillain Barre Syndrome was based on criteria given by Asbury and Cornblath (1990)

We recorded data on age, sex, preceding events, date of onset of disease, clinical manifestations including initial symptoms and neurological findings as per proforma during the course, result of CSF study and serial electrophysiological changes including specific treatment given.

Essential Criteria

1. Progressive weakness of >1 limb due to neuropathy
2. Total areflexia
3. Absence of other causes of acute polyneuropathy like porphyria, toxin exposure (lead and botulinum) and diphtheria.

Supportive Criteria

1. Progression of symptoms over 4 days to 4 weeks.
2. Relative symmetry of symptoms and relatively symmetrical weakness.
3. Mild sensory symptoms or signs.
4. Cranial nerve involvement especially bilateral weakness of facial muscles.
5. Autonomic dysfunction and vasomotor instability.
6. Absence of fever at the onset
7. Albumino-cytological dissociation in CSF (raised proteins and <10/mm^3 cells) after one week.
8. Neurophysiologically marked slowing of nerve conduction velocity.
9. Recovery beginning 2-4 weeks after cessation of progression.

Exclusion Criteria

1. Doubtful diagnosis

a. Sensory level

b. Marked persistent asymmetry of symptoms or signs.

c. Severe and persistent bladder or bowel dysfunction.

d. More than 50 cells per cubic millimetre in CSF.

e. Diagnosis of Botulism, Myasthenia gravis, Poliomyelitis or Toxic neuropathy

2. Abnormal porphyrin metabolism
3. Recent Diphtheria
4. Purely sensory syndrome without weakness
5. No Past history of Guillain Barre Syndrome
6. Severe concurrent medical illness

Observations

Table 1: Age and sex distribution of patients with Guillain Barre Syndrome

Age Group (years)	Gender		Total
	Female	Male	
	No. (%)	No. (%)	No. (%)

0-5	5 (45.5%)	6 (54.5%)	11(35.5%)
6-10	4 (40%)	6 (60%)	10(32.25%)
11-15	2 (20%)	8 (80%)	10(32.25%)
Total	11 (35.48%)	20 (64.51%)	31(100%)

Maximum cases (35.50%) were from the age group 0-5 years followed by 6-10 years and 11-15 years age groups (32.25% in each).

All age groups had near similar number of cases. **The male to female ratio was 1.8:1 (64.5% males).**

Table 2:Preceding illness before onset of neurological symptoms

Preceding Illness	No. of Cases	Percentage
Respiratory	11	35.48
Gastrointestinal	4	12.90
Other	10	32.25
None	6	19.35

Most of the preceding illness occurs 1-4 weeks before the neurological symptoms.

Preceding illness before the onset of neurological symptoms was present in 25 patients (80.64%). The most common preceding illness was respiratory (35.48%) followed by fever without definitive diagnosis (32.25%) and gastrointestinal infection (12.90%).

Table 3:Neurological features at the time of Presentation

Presentation	No. of Cases	Percentage
Quadriparesis: • Alone • Global motor weakness • With bulbar involvement • With Respiratory Insufficiency	 19 7 10 10	 61.29 22.58 32.26 32.26
Paraparesis Only	12	38.70
Cranial Nerve Involvement VII IX, X, XI	12 7 10	38.70 22.58 32.26
Sensory symptoms (Pain or numbness)	10	32.26
Autonomic dysfunction (bladder dysfunction, hypotension, hypertension, sinus tachycardia, arrhythmia)	11	35.48

Total 19 (61.29%) patients were admitted with quadriparesis alone, 7(22.58%) patients had global weakness, 10(32.26%) had bulbar involvement and 10 patients (32.26%) had quadriparesis along with respiratory muscle involvement.

12 patients (38.70%) presented with Paraparesis only without any progression of disease with equal distal and proximal muscle weakness was seen.

12 patients (38.70%) presented with cranial nerve involvement along with muscular weakness .Bulbar palsy was most commonly seen.

Sensory symptoms in the form of pain, numbness, paresthesias were present in 10(32.26%) patients with neurological weakness.

Autonomic dysfunction were present in 11(35.48%) of patients with neurological weakness, sinus tachycardia was the most common autonomic disturbance seen followed by transient hypertension.

Table 4: CSF Analysis

Protein (mg/dl)	≤50		51-75		76-100		>100	
	No.	%	No.	%	No.	%	No.	%
No. of Cases (%)	2	6.5	10	32.3	5	16.1	14	45.2
Mean	113.53mg/dl							
Cell Count (Cells/µl)	≤5		6-19		20-49		≥50	
No. of Cases (%)	22	71.0	8	25.8	1	3.2	0	-
Mean	5.70cells/µl							

We analyzed total protein concentration (mg/dl) and total cell count (cells/µl) in CSF of every patient.

The protein concentration ≤ 50 mg/dl was present in only 2 patients (6.45%) , 51-75 mg/dl in 10 patients (32.3%), 76-100 mg/dl was present in 5 patients (16.13%) and >100 mg/dl was present in 14 (45.16%) of patients. Mean CSF protein concentration was 113.53 mg/dl.

The cell count of ≤5 cells/µl was present in 22 (70.97%), cell count between 6-19 cells/µl was present in 8 (25.80%) cases.

Only 1(3.23%) patient showed cell counts between 20-49 cells/µl. The cells in all the cases were lymphocytes. Mean cell count was 5.70 cells/µl.

Table 5:Electrophysiological pattern of GBS at the time of first presentation

Pattern	No. of cases	Percentage
Demyelinating	5	16.12
Axonal	9	29.03
Mixed	14	45.16
Inexcitable	2	6.45
Normal	1	3.2
Total	31	100

Table shows the different electrophysiological subtypes of GBS patients observed by motor nerve conduction study.

Thus we conclude that mixed pattern was the most common followed by axonal pattern followed by demyelinating pattern.

Discussion

Out of the 31 patients studied, 20 (64.5%) were males while 11(35.48%) were females, the sex ratio being 1.8:1 . The age group of patients ranged from 3 years to 15 years. In our study all age groups had near equal number of cases with male predominance in all the age groups. This was consistent with all the previous studies on GBS. Similar age and sex profile has been reported by Ropper et al[7] (1991). They reported that Guillain Barre syndrome may occur in any age group, with bimodal distribution (occasionally including infancy) in either sex with male predominance.

GBS is believed to be an autoimmune disease. An otherwise unremarkable infection, such as an upper respiratory tract infection, often predates the onset of GBS by 10-14 days. Many antecedent infections have been identified including Campylobacter jejuni, Cytomegalovirus, Mycoplasma pneumonia, Epstein Bar virus and Influenza virus. Surgery and immunization have also been associated with GBS. Infection triggers an autoimmune response against the peripheral nerve myelin, due to the invading antigen having antigen similarity to the myelin antigen. In our study, preceding illness was present in 25 patients (80.64%), in the form of gastrointestinal infection, respiratory infection, fever due to unknown cause. Only 8 patients (19.35%) developed flaccid paralysis without any identified preceding illness.

The present study showed that the most common preceding illness was respiratory (35.48%) followed by fever without focus (32.25%) followed by gastrointestinal infection (12.90%).

The occurrence of infective antecedents during the month preceding the onset of Guillain Barre syndrome was investigated by means of a self administered questionnaire and the results were not modified by laboratory investigations. As stool culture may be negative by the time the symptoms of GBS appear, usually one to three weeks after the diarrhoel illness hence serological testing for elevated levels of serum IgA, IgM and IgG, specific for Campylobacter jejuni should be performed, in addition to stool culture. We were not able to identify the causes of preceding illness, as these tests are not available in our college.

A study done by the Italian Guillain Barre Study group[3] (1996) showed that an antecedent event was reported in

60.9% of the 297 patients and the commonest illness was Influenza (26.3%) followed by upper respiratory tract infection (15.2%) and the gastroenteritis (8.8%). Study by Tam et al[8] found positive associations between GBS and infection with Campylobacter jejuni, Epstein-Barr virus and influenza like illness in the previous two months as well as evidence of a protective effect of Influenza vaccination. The excess risks of GBS following Campylobacter enteritis was 60 fold and 20% of GBS cases were attributable to this pathogen.

In a study by Stowe et al[9] reported relative incidence of Guillain Barre syndrome within 90 days of vaccination was 0.76, in contrast, the relative incidence of GBS within 90 days of Influenza like illness was 7.35. In a study by Jacobs et al[137] reported that infections with many agents have been reported preceding GBS. Their study showed that in GBS patients infections with Campylobacter Jejuni (32%), Cytomegalovirus (13%), and Epstein-Barr virus (10%) were significantly more common than that in controls.

Guillain Barre syndrome is predominantly motor neuropathy, a feature noted reportedly from its earliest description. At the time of presentation the paralysis is usually distally accentuated and involves the legs before the arms ("ascending paralysis, Westphal 1876)[10]. In our study maximum number of patients of Guillain Barre syndrome had quadriparesis (61.29%) at the time of admission. Ten patients (32.26%) developed respiratory insufficiency who needed ventilatory support, 10 patients (32.26%) showed involvement of the bulbar muscles and had nasal regurtitation with difficulty in swallowing the food. Seven patients (22.58%) showed global weakness. Twelve patients out of 31 patients were admitted with paraparesis without any progression of the disease. The cranial nerves were involved in 12 patients along with the limb weakness. 32.26% of the patients had bulbar palsy followed by facial nerve palsy (22.58%). Every patient presented with sudden onset of weakness of lower or all four limbs, with symmetrical involvement and upward progression of the disease. Hypotonia and areflexia was present in all the cases.

Winer at al[11] also noted that muscle weakness usually starts in the leg and ascends to the arm. Proximal muscle weakness may be prominent form of onset.

Although weakness is the most predominant feature of GBS, in about 60% to 70% of cases the onset of disease is heralded by the development of distal paraesthesia and sometimes pain and muscle cramping[12]. In our study the sensory symptoms were present in 32.26% patients either in the form of paraesthesia or myalgia before the neurological illness. In a study by Shrivastava et al[13] 3% of the patients had sensory involvement.

In our study we found autonomic disturbance in 35.48% of the cases, most common manifestation was sinus tacychardia, followed by transient hypertension, arrhythmia, urinary dysfunction (transient urinary retention). In a study done by Lichtenfeld[14] 50% of Guillain Barre patients had autonomic dysfunction.

In our study the laboratory investigations of all patients showed abnormality in CSF analysis only. Other investigations were normal like haemogram, renal function test, liver function test, serum electrolytes and urine for porphobilinogen.

CSF protein concentration was raised in 29 patients out of the 31 patients (93.5%) cases. The mean CSF protein concentration was 113.53 mg/dl ranging from 26.9 mg/dl to 261 mg/dl. The CSF cell count was normal in 22 (70.96%) patients and only 9 patients (29.03%) showed slightly raised mononuclear cells. CSF cells ranged from 2-30 cells/µl. The mean CSF cell count was 5.70 cells/µl.

This study also showed the albumino-cytological dissocioation of CSF in 93.54% patients of Guillain Barre syndrome.

Our studies of CSF analysis are consistent with the study of Hung et al[15], which showed CSF protein concentration was elevated in 93.8% of cases. Results are also consistent with study of Hung et al[15] who had shown elevated CSF protein concentration in 93.8% of cases and <30 cells/µl in CSF in all patients. In a study by Gonzalez et al[141] 84% of the patients presented with albuminocytological dissociation that is consistent with our results.

In our study the mixed subtype is the commonest electrophysiological pattern of that is equally distributed in all the age groups of the Guillain Barre syndrome patients in our zone present in (45.16%) cases followed by axonal (29.03%), demyelinating (16.12%), inexcitable (6.45%) and normal pattern (3.21%) . This observation is consistent with the study made by Italian Guillain Barre Study Group[8] 1996 in which 45.2% of cases had mixed pattern and 29% cases had axonal, and 10.7% patients had demyelinating pattern. In a study conducted by Nagasawa and Kuwabara[16] in Japan out of 31 GBS children 48% had AMAN And 35% AIDP.

Conclusion

Thus by our study we conclude that GBS should be suspected in all the patients presenting with acute flaccid paralysis in different age groups. Mixed type is the predominant electrophysiogical pattern seen in almost all age group.

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