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Original Article

Clinical Profile of Guillain-Barre Syndrome in a Tertiary Care Center

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Abstract

Background: Guillain-Barre syndrome (GBS) is an acute, mostly demyelinating polyradiculoneuropathy of an autoimmune etiology. It is no longer called a pure demyelinating disorder and there are axonal variants of the same described as acute motor axonal neuropathy, acute sensory motor axonal neuropathy, and Miller Fisher variants.

Materials and Methods: All patients above the age of 18 presenting with acute flaccid paralysis were evaluated. Asbury's criteria was used to diagnose GBS. They were subjected to nerve conduction study and cerebrospinal fluid analysis. GBS disability scoring system (Erasmus GBS outcome score) was also assessed. Events occurring during the period of hospitalization were noted.

Results: A total of 50 patients with GBS were evaluated. Of these 76% were males and 24% were females. There were two peaks in the age wise distribution, one at 20-30 years and another at 40-60 years. The most common antecedent event was fever. The most common presenting signs and symptoms were motor weakness, followed by sensory symptoms such as tingling or numbness of the affected limbs. Respiratory difficulty as the presenting symptom was seen in 10 patients. Classical GBS was the most common presentation. A majority of the patients fulfilled 5-7 of Asbury's criteria. Evidence of protein cytological dissociation was seen in 88%. The majority of patients had demyelinating motor neuropathy with prolonged or absent F waves. 19 (38%) patients required intubation. Of these intubated patients, 26% died, 26% recovered, and 48% of them required tracheostomy. 42 patients received immunoglobulin therapy and eight patients underwent plasmapheresis. Six patients died in this study. Five of these patients died due to sepsis, predominantly respiratory. One person died due to intractable ventricular tachycardia.

Conclusion: Atypical GBS was uncommon. Most of the patients were managed with immunoglobulin. Mortality rate was higher and occurred mostly with a secondary sepsis.

Key words: Cerebrospinal fluid, Guillain-Barre syndrome, Immunoglobulin, Nerve conduction study, Plasmapheresis

INTRODUCTION

Guillain-Barre (pronounced as Ghee-Yan-Bar-Ray) syndrome (GBS) is an acute and more often than not, demyelinating polyradiculoneuropathy of an autoimmune etiology. This disease has forever perplexed the minds of neurologists and physicians alike for its florid presentation and equally good recovery. With polio declared as being eradicated from this country, GBS has now become the foremost diagnosis in patients presenting with acute flaccid paralysis. Although this disease has been identified a century ago, there are still many unanswered questions about it. For instance, though GBS has been classically described as an autoimmune disease, the molecular mimicry has never been attributed to any specific cause. The antecedent events that trigger this neurological disease are so vast and wide that it is difficult to pinpoint any specific event that serves as a hallmark for this disease. Over the last few decades our understanding of GBS has also changed, and we currently know that it is not a pure demyelinating disorder and there are axonal variants of the same described as acute motor axonal neuropathy, acute sensory motor axonal neuropathy, and Miller Fisher variants. Other variants of GBS such

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as pandysautonomia and polyneuritis cranialis occur so infrequently that no exact figure on their incidence or prevalence can be given. Even today the disease is best diagnosed by the clinical profile of patients who present with flaccid paralysis with areflexia, cerebrospinal fluid (CSF) analysis that shows protein cytological dissociation and nerve conduction studies (NCS) that show evidence of demyelination, with abnormal F waves. There is no single diagnostic test that confirms or rules out the possibility of GBS. In general, this disease carries good prognosis with mortality rates around 6% worldwide. Studies have been done to try and identify factors that could be considered as bad prognostic signs, but they have never been consistent.[1,2]

In this study, we look at the clinical profile of patients who presented with features of GBS to a tertiary care center and their clinical events in the hospital are recorded until the time of discharge.

MATERIALS AND METHODS

All patients above the age of 18 presenting with acute flaccid paralysis were evaluated from June 2011 to June 2013. The patients were then subjected to Asbury's criteria for diagnosing GBS (Asbury Criteria features required for the diagnosis - symmetrical weakness in 4 limbs: Features strongly supporting the diagnosis - progression of symptoms over days to 4 weeks, relative symmetry of symptoms, mild sensory symptoms or signs, cranial nerve involvement especially facial nerve, recovery beginning 2-4 weeks after progression ceases, autonomic dysfunction, absence of fever at onset, high concentration of protein in CSF, and typical electrodiagnostic features). Laboratory investigations such as complete blood count, random blood sugar, urine analysis, renal and liver function tests, and serum electrolytes were done in all cases. Two-dimensional echocardiography was done in specific patients as required. The patients were subjected to NCS and CSF analysis. An account of the antecedent events of clinical importance was recorded. The patients were then subjected to GBS disability scoring system (Erasmus GBS outcome score [EGOS]). The patients were monitored daily. Their disability score was reassessed every week till discharge. The need for ventilation was assessed on a day to day basis and if required tracheostomy was performed. The time and type of therapeutic intervention (including intravenous immunoglobulin [IVIg] and plasmapheresis) were recorded. This was done till the patient was discharged or a terminal event occurred.

RESULTS

The patients presenting with acute flaccid paralysis from June 2011 to July 2013 were identified and among them 50 patients had GBS. Of these 76% were males and 24% were females. There were two peaks in the age wise distribution of this disease, one at 20-30 years and another at 40-60 years (Table 1).

Eight patients each had diabetes and hypertension, three had coronary artery disease. GBS patients presented throughout the year. There was a sharp rise in the incidence of GBS in the months of October (10 patients) and November (six patients). The majority of patients had no antecedent events (23 patients), and the most common antecedent event that was found was fever (10 patients) followed by loose stools (eight patients).

The most common presenting signs and symptoms were motor weakness, which was followed by sensory symptoms such as tingling and numbness of the affected limbs. Respiratory difficulty as the presenting symptom was seen in 10 patients (Table 2).

Asbury's scoring system for diagnosis was graded from 1 to 9. Most of the patients scored around 5-7 (Table 3).

Most of the patients presented with EGOS scale score of 2 (Table 4).

88% of people had evidence of protein cytological dissociation. NCS was done for all patients admitted with GBS. Most of the patients had demyelinating motor neuropathy with prolonged or absent F waves. Some

Table 1: The study profile

Age	Male	Female	Total
<20	0	3	3
20-29	6	3	9
30-39	4	0	4
40-49	10	1	11
50-59	9	2	11
60-69	6	2	8
>70	3	1	4

Table 2: The presenting symptoms and signs

Presenting signs and symptoms	Symptoms
Weakness of limbs	48
Sensory symptoms	20
Respiratory difficulty	10
Facial nerve involvement	7
Ptosis/ophthalmoplegia	3
Bulbar symptoms	1

Table 3: Asbury score

Score	Number of patients
1	0
2	0
3	0
4	6
5	10
6	18
7	15
8	1
9	0

Table 4: EGOS disability score on admission

Score	Number of people
1	I
2	22
3	6
4	13
5	8

EGOS: Erasmus Guillain-Barre syndrome outcome score

had more than one type of conduction abnormalities (Table 5).

19 (38%) patients required intubation. Of these intubated patients, 26% died, 26% recovered, and 48% of them required tracheostomy. 42 patients received immunoglobulin therapy and eight patients underwent plasmapheresis. Six patients died in this study. Five of these patients died due to sepsis, predominantly respiratory. One person died due to intractable ventricular tachycardia. Patients with low Asbury score had a poor prognosis than with a higher score. The EGOS scoring of patients at the time of discharge was also documented. Most of them scored 0 or 1 at the time of discharge meaning normal or near normal recovery (Table 6).

DISCUSSION

GBS has always been considered a disease with a very low incidence which is estimated at around 0.7-1.5/100,000 population.[3] This being an observational study at a tertiary care center, the incidence or prevalence of this disease could not be estimated. Most of the studies that have looked at the epidemiology of GBS have noticed that this disease seems to occur more often in males than in females.[4,5] This has been true with this study also; 76% of our GBS patients were males and 24% were females. The disease was seen in all age groups, but there were two peaks of distribution that was seen, one at 20-30 years and another at 40-60. This was consistent with previous studies which had also described similar peaks.[6,7] However, the second peak seemed to occur much earlier in this study. A very interesting observation that was made in this study was the pattern of distribution

Table 5: NCS findings

NCS	Number of patients
Demyelinating motor neuropathy with prolonged or absent F waves	48
Demyelinating sensory neuropathy	4
Facial NCS abnormalities	2
Axonopathy	1

NCS: Nerve conduction study

Table 6: EGOS disability score on discharge

Score	Number of patients
0	16
1	21
2	3
3	1
4	3
5	0
6	6

EGOS: Erasmus Guillain-Barre syndrome outcome score

of the incidence of the disease during the calendar months of a year. There was a significant peak in the occurrence of this disease in the months of October and November. This also corresponded to the distribution of rainfall in Chennai during the period of study.

Most of the previous studies have shown that patients present with an antecedent event even up to 6 weeks before the neurological symptoms. In this study, 55% of them had antecedent event before getting admitted to the hospital. This was consistent with studies that had been published earlier.[8] Among the antecedent events that were reported by the patients, fever was the most common one (19%) followed by loose stools (16%). After motor weakness, the next common presentation was sensory disturbances which were seen in 20 patients. Such sensory disturbances have also been reported in previous studies.[9] The most common cranial nerve that was involved was the facial nerve which was also consistent with the previous studies.[9] Respiratory failure was seen as a presenting complaint in 20% of the patients. This correlated with the incidence of respiratory failure that has been previously documented with GBS (10-30%).[10]

Protein cytological dissociation was seen in 88% of the patients. This was consistent with previous studies.[11] Previous studies had shown that demyelinating form of GBS was the most common in European countries and in the US,[12] and the axonopathy variant was more common in China and Japan.[13] However, in this study, the demyelinating variant was the most common (96%). Most of the patients were administered IVIg (84%). The prognosis of the patient based on IVIg treatment could not be assessed because many patients had received treatment elsewhere

before being admitted in this hospital, which served as a confounding factor. Moreover, the decision as to whether IVIg needed to be administered was based solely on the treating physician and Neurologist's discretion. Of the types of GBS that were seen in this study, demyelinating variety was the most common, followed by Miller Fisher variant, and then, axonopathy.

In this study patients with poor prognosis were distributed throughout all ages. This was not correlating with previous studies which had quoted older age to be a marker of poor prognosis of patients.[14] In this study, it was found that patients with a lower score in Asbury's scale had worse prognosis than patients with a higher score. At the time of discharge most of the patients had no or minimal residual neurological deficit (EGOS Scale - 0 or 1) - 74%. Mortality rate was 12% which was higher than the ones shown in other studies worldwide (4%).[15] This was mostly due to sepsis.

CONCLUSION

Our study showed GBS to be more frequent in males, with classical GBS being the predominant type. NCS and CSF findings were the most specific for diagnosis of GBS. Asbury criteria on admission had no correlation with the prognosis. We were not able to ascertain which line of treatment was superior as 84% were given immunoglobulin therapy and only 16% underwent plasmapheresis. Mortality was seen only in patients who developed secondary complications such as respiratory sepsis.

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