ORIGINAL ARTICLES

FREQUENCY OF GUILLIAN-BARRE SYNDROME AND ITS VARIANTS AT MILITARY HOSPITAL RAWALPINDI

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ABSTRACT

Objective: To assess the frequency, variants, clinical profile and outcome of patients admitted with the diagnosis of Guillian-Barre Syndrome at neurology unit of a tertiary care teaching hospital of Pakistan.

Study Design: Cross-sectional study.

Place and Duration of Study: Neurology department, Pak Emirates Military Hospital Rawalpindi, from Jan to Dec 2018.

Methodology: This study was conducted on 42 patients of Guillian-Barre Syndrome admitted in hospital. Demographic profile included age, gender, and the symptoms with which the patient presented. Medical and neurological complications were also documented among the target population. Outcomes included recovery, shifting to intensive care unit and death.

Results: Out of 42 patients included in the final analysis 25 were male and 17 were female. Most of the patients presented with lower limb weakness followed by numbness. Acute inflammatory demyelinating polyneuropathy was the commonest while cranial nerve variant was least reported. Motor deficit were the commonest complication faced by the patients followed by respiratory involvement. Out of 42 patients, 34 (80.9%) recovered, 4 (9.5%) were shifted to the intensive care unit and 4 (9.5%) died.

Conclusion: This study gives an insight into the pattern of a fairly common neurological illness which if diagnosed and managed in time has a good outcome in our set up. Management of critically ill Guillian-Barre Syndrome patients involves a multidisciplinary team with a need of intensive care unit during the course of management. Variety of types and presentations should be part of training of neurology doctors as well as internal medicine doctors.

Keywords: Demographic profile, Frequency, Guillian-Barre Syndrome.

INTRODUCTION

Guillian-barre syndrome (GBS), previously considered a rare diagnosis is a fairly common neurological diagnosis in today's era[1]. First case was reported in 1916 and was named after the scientist who reported the first case of this neuromuscular disease[2]. GBS can present in number of ways but usually acute flaccid paralysis of lower limbs is the presentation in most of the patients. With recent advances and different patterns seen in clinical practice this disease has emerged as a syndrome more than just a demyelinating condition[3].

Neurology is an emerging specialty in our part of the world with less than 200 qualified neurologist available to counter the burden of neurological illnesses in our population[4]. On the other hand neurological illnesses are on a rise and made part of the global mental health gap campaign for the low and middle income countries which states there is a big gap on the demand and supply of the treatment options available in the community[5,6]. GBS can present with a variety of symptoms so exact diagnosis and a specially tailored management plan is required to save the patient from death or other untoward complications[7].

A lot of work has been done all over the world on accurate diagnosis and management of GBS in all parts of the world. A study done in a tertiary care hospital of Iran reported 388 cases in the five years with a clear male predominance and most cases occurring in the months of spring and winters[8]. Another study done in China revealed that acute inflammatory demyelinating polyneuropathy was the commonest sub type reported and dysautonomias were predictor of poor prognosis at six month time[9]. Another study done in another neighboring country of ours concluded that GBS was least prevalent in the children with age less than two years and axon involvement emerged as a sign of poor prognosis among the patients[10]. Therefore clinical presentation, epidemiology of subtypes and treatment response vary from one population to the other, which demands a thorough understanding of the overall spectrum of illness inorder to diagnose and treat effectively.

Various etiologies have been reported to be linked with this condition. Immunological basis has

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been the most agreed basis on GBS. It has been observed that this immune reaction has been linked with infection of gastrointestinal tract or urinary tract. Winter season has also been linked significantly with the onset of GBS in various studies done in different parts of the world.

Being from a developing nation with limited resources this topic is of utmost relevance in good clinical practice. Early recognition and management not only reduces the mortality but also limit the ICU admission and disability in the patient. Very limited local data is available on this aspect and that too comprise of few case reports and reviews but no study has so far been conducted at a tertiary care military hospital receiving patients from all over Pakistan including the public sector tertiary care hospitals. This study was planned with the rationale to look for the frequency, clinical profile and outcome among the adolescence and adult patients admitted in the neurology unit with the diagnosis of GBS in a tertiary care teaching hospital of Pakistan.

METHODOLOGY

This observational study was conducted at the Neurology Unit of Pak Emirates Military Hospital Rawalpindi from January to December 2018. Sample was gathered by using the non-probability consecutive sampling technique. All patients between the age of 12 and 65 years admitted in the neurology department and diagnosed as GBS by the consultant neurophysician were included in the study. Patients who were referred from other military, public sector and private hospitals with the same diagnosis were also included in the analysis in addition to the referrals from the other wards of own hospital. GBS diagnosis was made by incorporating the following clinical and diagnostic modalities according to diagnostic criteria from the National Institute of Neurological Disorders and Stroke FROM 1990[11-15].

1. Acute progressive symmetric weakness of the extremities with areflexia or hyporeflexia
2. Albuminocytological dissociation in cerebrospinal fluid (raised protein and total cell count of ≤10/mm^3)
3. Demyelinating/axonal neuropathy on electrophysiological studies.

All suspected cases of GBS underwent electrophysiological studies within 48 hours of admission. Needle EMG was also performed. At least one motor and one sensory nerve wastested on the upper and lower limbs. Response was recorded in all the extremities. Additionally, routine motor conduction studies were performed on the median, ulnar and tibial nerves using conventional procedures. Sensory nerve studies were performed on the median and sural nerves. The amplitude of the negative phase was measured for compound muscle action potentials and sensory nerve action potentials.

In this study, the patients were classified into the following categories/variants based on the existing electro-diagnostic criteria: acute inflammatory demyelinating polyneuropathy (AIDP) group, acute motor axonal neuropathy (AMAN) group, acute motor and sensory axonal neuropathy (AMSAN) group, miller-Fisher syndrome (MFS) group and cranial nerve variant (CNV).

Exclusion criteria were the patients with <12 years of age or those with unclear medical diagnosis. Pregnant patients or those with CSF India ink stain positive for the fungal growth or those with suspected TBM or encephalitis were also not included in the study. Patients with post traumatic meningitis or those with post injection syndrome or poliomyelitis were also the part of exclusion criteria. Patients with diabetes, neoplasia, hypothyroidism, renal failure, vasculitis, or history of intoxication or those who did not give written informed consent were also excluded from the study.

Ethics approval was taken from the ethics review board committee of PEMH Rawalpindi. After written informed consent from the potential participants, patients with GBS in the neurology unit of PEMH Rawalpindi fulfilling the above mentioned criteria of inclusion and exclusion were included in the study. Neurology unit of PEMH Rawalpindi is a tertiary care 40 bed facility with 4 consultants and around 10 residents. Outcomes included recovery, Shift to intensive care unit and death. A specialized proforma was designed for the study filled by the neurophysician after interviewing the patient and seeing the referral form.

All statistical analysis was performed by using the SPSS-24. Mean and standard deviation for the age of study participants was calculated. Frequency and percentages for gender, variants of GBS, symptoms with which the patient presented, complications and outcome was calculated.

RESULTS

A total of 48 patients admitted in the neurology unit with confirmed diagnosis of GBS were initially

approached to get them included in the analysis. Two were pregnant, two were either over or under age, one had no clear diagnosis of the underlying medical condition one patient did not give consent to get included in the study. final analysis 25 were male and 17 were female. Male to female ratio was 1.5:1. Mean age of patients diagnosed with GBS in our study was 32.63 ± 4.271. Other characteristics of study population have been summarized in table-I. AIDP was the commonest while cranial nerve variant was least reported. Most of the patients presented with lower limb weakness followed by numbness. Motor deficit was the commonest complication faced by the patients followed by respiratory involvement (table-II). Out of 42 patients, 34 (80.9%) recovered, 4 (9.5%) were shifted to the intensive care unit and 4 (9.5%) died (talbe-III).

Table-I: Characteristics of patients admitted with Guillian-Barre Syndrome (n=42).

Age (Years)	
Mean ± SD	32.63 ± 4.271
Range (min-max)	12-59 years
Gender	
Male	25 (59.5%)
Female	17 (40.5%)
Clinical Features at Presentation	
Lower limb weakness	19
Numbness	12
Depressed reflexes	11
Facial nerve palsy	06
dysarthria	06
Confusion	01
Others	03
Variants of Guillian-Barre Syndrome	
AIDP	22
AMAN	08
AMSAN	06
MFS	05
CNV	01

Table-II: Complications among the patients admitted with Guillian-Barre syndrome at neurology department (n=42).

Medical Conditions	Frequency
Motor deficit	10
Cranial nerve palsies	06
Respiratory involvement	07
Dysautonomias	05
Bulbar palsy	03
Arrhythmias	02
Urinary retention	02
Persistent pain	04
Others	02

Table-III: Outcome of patients diagnosed and treated as Guillian-Barre syndrome.

Total	Recovery	Shifted to ICU	Death
42	34 (80.9%)	4 (9.5%)	4 (9.5%)

DISCUSSION

Pakistan is a low and middle income county with a high burden of neurological problems which have been under addressed and made part of the mental health gap in order to catch the due attention[5]. GBS is an interesting disease with an immunological basis which helps us to classify it as a non-communicable illness but on the other hand its link with the infections and post infectious immunological phenomenon also link it with the communicable diseases[11,12]. It has not been limited to some distinct part of the world but prevalent across the globe in all the ethnicities which makes it a challenge for neurologists all over the world[1,2]. Not much is known regarding primary prevention of this illness so main stay of management is early recognition and prompt treatment after the diagnosis has been made[7]. Treatment modalities include either intravenous immunoglobulin or plasmapharesis, both of which are very expensive. Therefore accurate diagnosis and knowledge of all aspects of disease is necessary for the physicians. This study was thus planned with the rationale to give an account of frequency and variants of GBS in our population.

Most of the patients in our study were male with male female ratio of 1.5:1. Though usually females have predominance in most of the immunological diseases but this has not been usually found in case of GBS. Similar results were reported in similar studies done in past in other countries which showed clear male dominance among the study population[9,10]. Reason might be either females have lesser chance of getting GBS or chances of females with GBS reaching the tertiary care facility are less as compared to males. Sample was drawn from the military hospital which has entitlement of military soldiers so sample is not representative of general population. More research is required to look into this parameter.

This study population reported lower limb weakness as the commonest presenting feature followed by numbness. Similar results have been reported in the studies done in the past on similar subject[10,11]. These two symptoms are the common finding among the patients of various outpatient departments (OPD) including the general practitioner and medical OPD. Accurate history, general physical and systemic examination, relevant laboratory and radiological investigations can give these symptoms a definite shape and enable the physician to reach the diagnosis. Sharing of

experience of various local settings can help the neurologist and immunological disease specialist to develop our own protocol for diagnosing GBS in suspected cases. Our country has also been in the poliomyelitis zone and still cases of polio have been reported till date[16]. Therefore this important differential diagnosis needs to be ruled out before labeling the patient as GBS.

Motor weakness was the commonest complication these patients reported in our analysis followed by the respiratory involvement. These complications have been seen among the patients of GBS in other settings as well[10,17,18]. This finding raises the importance of liaison with other specialties in the management of GBS. Physiotherapist and respiratory physicians need to be warranted in such cases. Critical care settings with mechanical ventilation facility may need to be involved in some cases of GBS.

Clinical findings and electrophysiology techniques were used to classify the patients of GBS in different variants. AIDP was the commonest variant in our analysis followed by the AMAN variant. Cranial nerve variant was reported only in one patient. Studies done in other parts of the world have also shown same results regarding the epidemiology of variants of GBS[10-12].

Outcome was avariable in our analysis. Most of our patients (80%) had a good recovery and were discharged from the neurology ward after adequate treatment. Four patients needed the mechanical ventilation and shift in the critical care unit which highlights the importance of managing such cases at a tertiary care hospital with lifesaving facilities. Mean age of our target population is also around thirty two years which further strengthens the fact of aggressive multidisciplinary management in such cases. Four patients died with complications in the ward. This mortality rate is comparable to the mortality rates of this disease in other parts of the world as well[10,12,19].

The major limitation of our study is the lack of generalizability as patients from one neurology unit of a tertiary care hospital of Pakistan were studied instead of all hospitals of the country. Randomization was not done for the mode of treatment for GBS and then looked for the outcome. Prognosis was also not compared among the different variants of GBS. Patients shifted to ICU were also not followed up so their outcome remains unclear. We suggest further studies on a broader based and a more representative sample size involving the hospitals of both public and private sector in order to generalize the results and set our own evidence based protocols regarding the suspicion and diagnosis of GBS in neurology units of our country.

CONCLUSION

This study gives an insight into the pattern of a fairly common neurological illness which if diagnosed and managed in time has a good outcome in our set up. Management of critically ill GBS patients involves a multidisciplinary team with a need of intensive care unit during the course of management. Timely diagnosis and adequate training of the neurophysicians in this aspect may serve as key to improve the outcome. Variety of types and presentations should be part of training of neurology doctors as well as internal medicine doctors.

CONFLICT OF INTEREST

This study has no conflict of interest to be declared by any author.

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