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VARIANTS AND VARIATIONS AMONG GULLIAN BARRE SYNDROME PRESENTING AS ACUTE FLACCID PARALYSIS

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ABSTRACT

Objectives: Acute flaccid paralysis (AFP) is a clinical syndrome characterized by rapid onset of weakness that frequently includes respiratory and bulbar weakness. An accurate and early diagnosis of the cause has important bearing on the management and prognosis. Guillian Barre Syndrome (GBS) is a post infectious polyradiculoneuropathy involving mainly motor but sometimes sensory and autonomic nerves. **Methods:** Retrospective and hospital based study was carried out at Department of Neurology CMC Hospital Larkana to find out the clinical features of GBS including existing treatment modalities and its outcome, prognosis of the disease in relation to its severity. Files of the patients aged (8-60) years from Jan-2013 to Dec 2013 fulfilling AFP criteria were included. Diagnostic features included weakness or paresis of limb or flaccid paralysis with or without sensory symptoms or autonomic symptom, cranio-bulbar symptoms, together with laboratory features like albumino-cytological dissociation, nerve conduction velocity (NCV), ECG, Serum electrolytes and MRI. **Results:** 55 patients were included in the study during 12 months of study period. Among them 29(53%) were diagnosed as GBS, 21(38%) patients were associated with hypokalemic periodic paralysis and 5(9%) patients as idiopathic neuropathy. All of them had undergone NCV test and classified further as AIDP (Acute inflammatory demyelinating polyneuropathy)-18 patients, AMAN (Acute motor axonal neuropathy) – 8 patients and AMASAN (Acute motor and sensory axonal neuropathy) -3 patients. Majority patients presented with symmetrically ascending paralysis with gradual onset involving all limbs. Males were more affected in our study with male female ratio as 1.62:1 and 23 patients were seen during winter and spring season. There was slightly higher age range during 2nd and 3rd decade and second peak in 5th decade. 10% patients had relapse within 5 year. Associated diseases were URTI, pneumonia, sore throat, diarrhea in majority. Facial Nerve palsy was commonest cranial nerve involvement. Majority improved with supportive treatment alone, 11% patient required ventilator support and 22% referred for plasmapheresis or IVIG. At 3 months follow-up almost half patients fully recovered and improved upon further follow up. **Conclusion:** Timely diagnosis and prompt treatment is key to manage and support these treatable diseases. Proper education, physiotherapy and psychological support is required.

Keywords: Acute flaccid paralysis, GBS, Hypokalemic periodic paralysis, AIDP, AMAN, AMASAN.

INTRODUCTION

Acute flaccid paralysis (AFP) is a clinical manifestation characterized by weakness or paralysis and reduced muscle tone. This condition can become fatal if it affects the respiratory muscles, posing the threat of suffocation and mortality. An accurate and early diagnosis of the cause has important bearing on the management and prognosis. In 1916, 3 French physicians (Guillain, Barré, and Strohl) described 2 French soldiers with motor weakness, areflexia, cerebrospinal fluid (CSF) albuminocytologic dissociation, and diminished deep tendon reflexes. Guillain-Barré syndrome (GBS) can be described as a collection of clinical syndromes that manifests as an acute inflammatory polyradiculoneuropathy with resultant weakness and diminished reflexes. Historically, GBS was a single disorder; however, current practice acknowl-

edges several variant forms Like AIDP, AMAN, AMSAN and Miller Fisher Syndrome. Among 2 predominant subtypes, a demyelinating subtype (AIDP) predominates in the United States and Europe, and axonal subtype (AMAN) is the predominant form in China⁽⁷⁾. Previous clinical studies suggested that AMAN also occurs in Mexican children⁽¹⁾. In the USA, the incidence is 1.2-3 per 100,000 inhabitants making it the most common cause of Acute Flaccid Paralysis⁽⁵⁾. The disease is assumed to be autoimmune and operated by a preceding infection, most of the time respiratory or gastrointestinal infections. Generally infections by microorganisms such as Campylobacter jejuni, CMV, Mycoplasma pneumonia, or influenza virus exist several weeks prior to approximately two thirds of GBS cases⁽⁴⁾. As Acute Flaccid Paralysis carries high mortality if not treated early so timely diagnosis and prompt management can save the life by preventing the complica-

tions like Respiratory failure, Autonomic dysfunction, Infections, residual disability and pulmonary embolism. A 2008 epidemiologic study reported a 2-12% mortality rate despite ICU management⁽⁵⁾. Causes of GBS-related death include acute respiratory distress syndrome (ARDS), sepsis, pneumonia, venous thromboembolic disease, and cardiac arrest. Most cases of mortality are due to severe autonomic instability or from the complications of prolonged intubation and paralysis⁽⁶⁾. The Aim of our study is to know the burden of different causes of acute flaccid paralysis, variants among the GB syndrome, seasonal variations and importance of accurate and timely management to allow health care personnel to identify and report such cases to avoid disability and death.

OBJECTIVE

To determine the clinical presentations of acute flaccid paralysis, electrophysiologic subtypes of GBS, seasonal variations and their outcome in our setup.

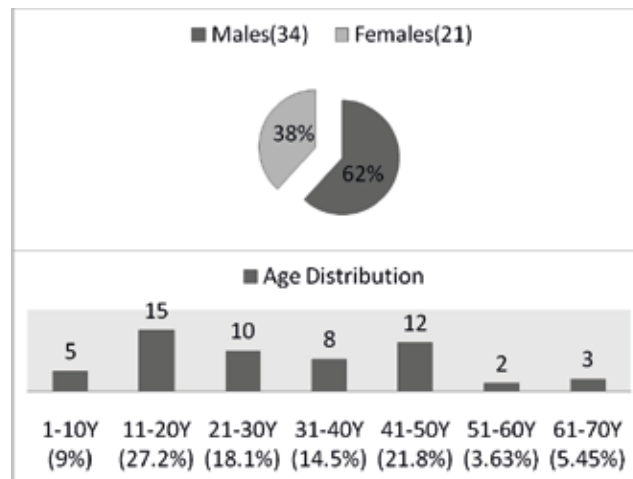
METHODOLOGY

This was a retrospective, cross sectional study conducted at the Department of Neurology, Chandka Medical College Hospital, Shaheed Mohtarma Benazir Bhutto Medical University Larkana from Jan 2013 to Dec 2013. Cases fulfilling Acute flaccid paralysis criteria were reviewed according to WHO Rapid onset of weakness of an individual's extremities, often including weakness of the muscles of respiration and swallowing, progressing to maximum severity within 1-10 days. The term "flaccid" indicates the absence of spasticity or other signs of disordered central nervous system (CNS) motor tracts such as hyperreflexia, clonus, or extensor plantar responses⁽⁸⁾. Data was collected for Age, Sex, Area of distribution, seasonal variation, Cause of Acute flaccid paralysis, Electrophysiologic subtypes of GBS and their Management. The cases were followed in outpatient. Data was collected by researcher himself and analysis was done on SPSS version 17. Patients with symmetrical flaccid paralysis with or without sensory symptoms or autonomic symptom and progressive motor weakness of more than one limb which cease to progress by four weeks were included in the study. While suspecting cases of Myopathies and Spinal lesions were excluded from the study. Diagnosis was made upon history, Clinical examination, CSF Analysis, Serum electrolytes and Nerve conduction studies while MRI was performed where ever needed.

RESULTS

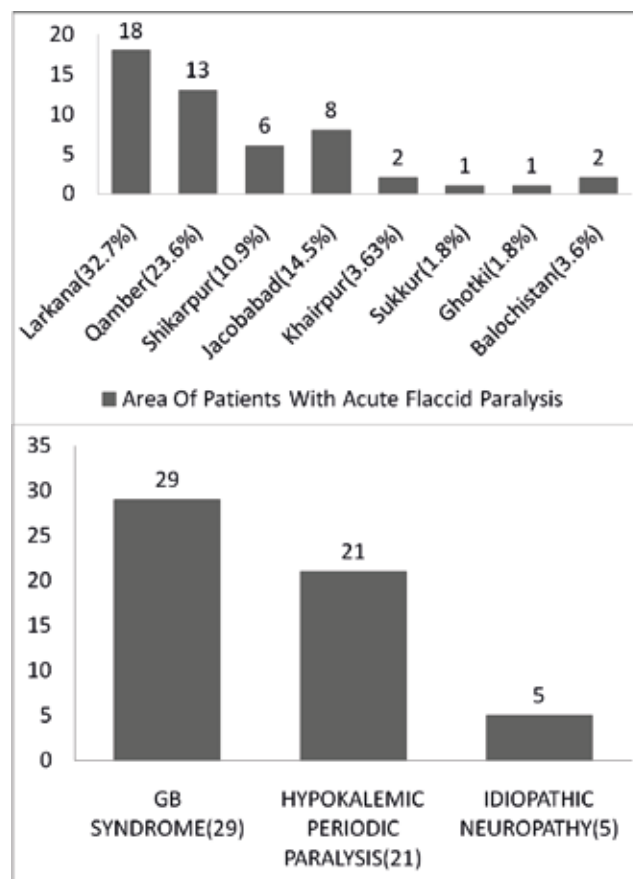
There were total 55 patients who were included in our study. Out of them males were in predominance with a total number of 34(61.8%) and females were 21(38%),

with the ratio among male and female of 62:1.

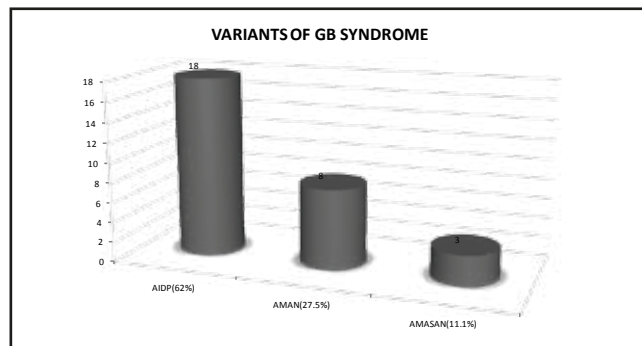


Mean age of the patients was 36 years. Majority 15(27.2%) were in 2nd decade (11-20 years), 10(18.1%) were in 3rd decade and 12(21.8%) were in 5th Decade of age.

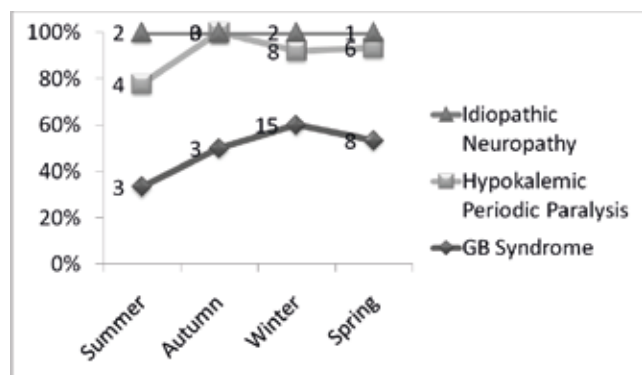
Most of them belong to district larkana n=18(32.7%) while rest belong to Qamber n=13(23.6%), Shikarpur 6(10.9%), Jacobabad 8(14.5%), Khairpur 2(3.63%), Sukkur 1 (1.8%), Ghotki 1(1.8%) and parts of Balochistan 2(3.6%).



Among them 29(53%) were diagnosed as GB syndrome , 21(38%) as Hypokalemic periodic Paralysis and 5(9%) as Idiopathic Neuropathy where no cause was found..

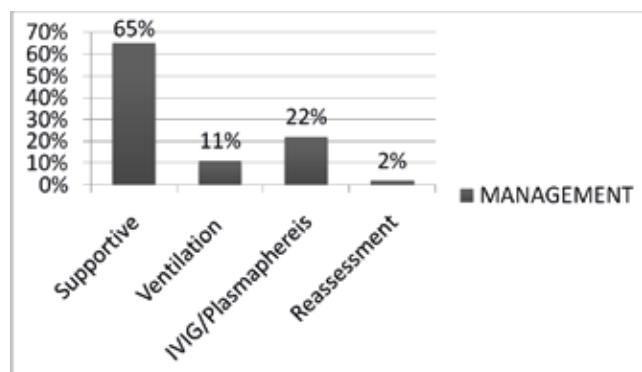


Variants of GB syndrome as identified after Nerve Conduction Studies were AIDP 18(62%) which is classic GB syndrome, Acute motor axonal neuropathy 8(27.5%) and Acute motor and sensory axonal neuropathy 3(11.1%) . The Spectrum of Seasonal variation in our study is shown below



Most of the cases were seen during winter and spring season.15 cases among the GB syndrome and 8 among Hypokalemic Periodic Paralysis were seen during winter season. Less number of cases seen during summer and autumn.

Associated diseases were upper respiratory infections in 3 patients, Pneumonia in 1 and Diarrhea in 6 patients while 14(25.4%) patients had associated facial nerve palsy.



During the treatment 65 percent patients Improved with supportive treatment and 11% required Intubation and need for Ventilation while 22% referred for Immune modulation therapy (IVIG/Plasmapheresis).

DISCUSSION

Among the 55 cases of Acute Flaccid Paralysis mostly were in 11-20 years of age group, belong to district Larkana .GB syndrome was the most common cause of Acute Flaccid Paralysis with AIDP as commonest variant. Seasonal variation was observed with more cases seen during winter and spring .Majority Improved with Supportive care. The underlying mechanism could be the explanation of pathophysiological basis in GB syndrome peak and halt of progression of disease as its natural course. The higher number of cases reported in winter and spring in our study is similar to reports from Southern Iran and Kuwait .The cases were reported throughout the year with the highest number being in January (261) and the lowest in August (173)2 . There is significant monthly and seasonal variation in the admission rate of patients with GBS in Shiraz.(IRAN) .From 389 cases of GBS, 232 (59.6%) were males and 157 (40.4%) were females. There was seasonal (P=0.004) and monthly (P=0.046) variation. Spring and winter had the most number of patients, with admissions from the month of February through June inclusive accounting for 50% of all cases (3). Immunomodulatory therapy like IVIG or plasmapheresis give the best output. Hypokalemic periodic paralysis on the contrary require bedside ECG and urgent potassium level and provide dramatic response to oral/IV potassium. At 3 months follow-up in outpatient almost half patients fully recovered and remaining improved upon further follow up. 10% patients had relapse within 5 year while. No Death or Residual deficit noticed during the course of disease.

CONCLUSION

GB Syndrome carries high mortality and is major public health problem if not diagnosed properly. Supportive treatment with observation for the progression and timely diagnosis and prompt treatment is key to raise the flaccid upon his feet. Proper education, physiotherapy and psychological support is required to treat these treatable diseases.

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Author's Contribution:

Dr Alam Ibrahim Siddiqu: Study concept and design, protocol writing, data collection, data analysis, manuscript writing, manuscript review

Dr. Ghulam Rasool Bouk: Data collection, data analysis, manuscript writing, manuscript review

Dr. Lal Chand: Data collection, data analysis, manuscript writing, manuscript review

Dr. Sarfraz Ahmed Mahesar: Data collection, data analysis, manuscript writing, manuscript review

Dr. Ghulam Mustafa Tunio: Data collection, data analysis, manuscript writing, manuscript review