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The precipitating factors and outcome of a thirty nine Patients with Guillain-Barre Syndrome admitted in the ICU of Al-Mawani General Hospital in the Governorate of Al- Basrah'

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Abstract

Objectives: To study the possible precipitating factors and the variability of the outcome in patients with severe Guillain- bare syndrome according to age, sex and types of treatment.

Methods: A single center, retrospective, descriptive study of 39 Patients with Guillain-Barré syndrome were studied between April 2007 to October 2010, their ages were between 3-63 year. They were 21 males and 18 females. Diagnosis was confirmed by typical clinical feature and ancillary investigations like electro diagnosis plus exclusion of other diagnoses. The history of any preceded or associated diseases before one month were clarified, treatment strategies and outcomes of the diseases were also analyzed. The study was conducted in the intensive care unit in AlMawani general hospital Basrah, Iraq.

Results: 23 patients had respiratory tract infection precede the neurological deficit, 7 patients had diarrhea and 22 patients occur at winter season. All the 39 patients develop limbs weakness, 30 patients develop shortness of breath and 24 of them needed mechanical ventilation. From the 26 patients with intravenous immunoglobulin therapy (IVIG) 17 had complete recovery, 5 left with neurological deficit and 4 of them died.

Conclusions: respiratory tract infections are the commonest precipitating diseases preceding GBS in this study, most cases occur in winter. Early immunoglobulin therapies result in a high percentage of complete recovery and a less percentage of death.

Key Words: Guillain- Barre Syndrome. Precipitating diseases. Immunoglobulin therapy.

الخلاصة

تم دراسة الامراض المصاحبة والمسببة في المرضى الذين يعانون من الشلل الرخوي، حيث تم دراسة تسعة وثلاثون مريض يعانون من أعراض الشلل الرخوي مع تحديد أصابة المريض بالتهاب الجهاز التنفسي الحاد، الاسهال الحاد، التهاب فايروسي، لقاحات أو أي أدوية ومواد كيميائية يمكن أن يكون تعاطها المريض. جميع المرضى كانوا يرفدون في العناية المركزة لمستشفى الموائ العام في البصرة جنوب العراق. تمت الدراسة للفترة من أبريل 2007 ولغاية أكتوبر 2010، تتراوح أعمار المرضى بين 3-63 سنة، 21 من الذكور و 18 من الاناث، 23 يعانون من التهاب الجهاز التنفسي و 7 من الاسهال الحاد. كل المرضى ظهرت لديهم علامات الشلل في الاطراف، 30 مريضا كانوا يعانون من ضيق النفس، احتاج 24 منهم الى التنفس الميكانيكي. توفر علاج المناعي الامينو غلوبولين ل 26 من مرضى الشلل الرخوي وأظهرت النتائج ان الاستجابة له كانت افضل عند المرضى اللذين أخذوا الجرعة مبكرا، حيث أن تحسن المريض كان بنسب عالية مع قلة الوفيات.

Introduction

Guillain-barré syndrome (GBS) is a common challenging clinical problem affecting significant number of population of different age and gender.

Historically in 1859 Landry published a report on 10 patients with ascending paralysis and in 1913, 3 French physicians ,Guillain, Barré and Strohl described 2 French soldier patients with motor weakness and cerebrospinal fluid showing albumin cell dissociation, the identified syndrome was later named Guillain-Barré Syndrome ^(1,2) .Guillain-Barré syndrome; is an acute rapidly progressive demyelinating polyneuropathy of auto immune etiology ,though the exact cause is not yet identified but it has been shown an increase incidence following gastrointestinal diarrheal illness and respiratory infections as supported by the increase incidence in winter .^(2,3,4) Some other rare GBS associated antecedent events have been reported such as surgery, cancer, pregnancy, autoimmune diseases, use of drugs, spinal anesthesia, non-Hodgkin's lymphoma, insect stings, epidural-general anesthesia, surgery for obesity, olanzapine administration and transplantation operations. Several cases have been found to develop GBS after therapeutic injection of bovine brain ganglioside preparations: ^(5,6,7) There are numerous reports of incidence rates that range from 0.6 to 4.0/100,000 population. Data from the mayo clinic, based on national institute of neurological disorders and stroke diagnostic criteria, and thorough ascertainment methods gave a crude incidence of 1.7 per 100 000 per year.^(6,7,8) .Early hospitalization, diagnosis and early treatment is mandatory and may decrease the mortality and morbidity with long term disability, despite the use of different therapeutic regimen, still the mortality and residual disability are very significant. Paralysis may progress rapidly within hours to days to cause respiratory muscle paralysis and cardiac decompensation which

required ventilatory support. ^(7,9) . Subcutaneous heparin should be given to reduce the risk of venous thrombosis. Immunoglobulin given intravenously (IVIG) within the first two weeks reduces duration and severity of paralysis. Plasma exchange is also of proven benefit in shortening disability, though its uses are limited by the general condition of patient and by the presence or absence of autonomic stability of patients. ⁽⁹⁾ Corticosteroids are not proved to be useful, though are widely used to treat many autoimmune disorders ^(8,9,10) . unfortunately not useful in GBS apart from their benefit in hastening recovery if given as methyl prednisone in combinations with intravenous immunoglobulin (IVIG) and reducing pain from GBS as shown by a Dutch trial. ^(10,11) and further clinical trials were suggested. Recovery begins, with or without treatment, between several days and 6 weeks from the outset. Prolonged ventilation may be necessary. The most feared complication is the respiratory system involvement, respiratory muscle paralysis and the cardiac decompensaions that may cause deranged cardiac function. Infections are another problem that may complicate GBS cases. ^(12,13,14) .Improvement towards independent mobility is gradual over many months but may be incomplete. Some 15% either die or are left disabled...^(14,15) The aim of this paper is to study the diseases that are possibly precipitating GBS and to study the outcome of GBS and its variability according to age, sex and treatment and factors affect the prognosis.

Patients and Methods

This descriptive, retrospective study was carried out in Almawani General Hospital Basrah, Southern Iraq; data

were collected from records of 39 patients with Guillain-Barré syndrome. They were admitted to the intensive care unit at the period from April 2007 to October 2010. Twenty one (53.8%) were males and eighteen (46.2%) were females, their ages ranged from 3 to 63 years with a mean of 39.44 years. The diagnosis based on criteria of the national institute of neurological and communicative disorders and stroke (NINCDS).^(15,16,17). Data collected from records of patients admitted to ICU which involve the age, gender, date of arrival to ICU, precipitating diseases of GBS which include: respiratory tract infections, flu illness, vaccinations, acute gastroenteritis, any malignant disease as lymphoma or chemotherapy and recent surgery. The given treatments were analyzed; the months of admission were also recorded to evaluate the seasonal variations if any. The Hughes or GBS disability scales was used to evaluate the clinical status of the patients and their outcomes. The time of initiation of ancillary and specific treatments, the use of invasive interventions (tracheostomy) or ventilators, date of discharge, outcome of patients and their outcome were also recorded and analyzed. The consent to be involved in the study, from the patient or his relatives, was obtained for each patient.

The Hughes or GBS disability scale was used to evaluate the clinical status of patients;

Grades:

Grade 0=healthy;

Grade 1=minor signs or symptoms of neuropathy but capable of manual work.

Grade 2=able to walk without support of a stick but incapable of manual work;

Grade 3=able to walk with a stick, appliance, or support;

Grade 4=confined to bed or chair bound;

Grade 5=requiring assisted ventilation;

Grade 6=dead

Short-term prognosis was defined as mortality and morbidity in the acute phase of the disease (first 30 days). Data were processed using the SPSS statistical package (version 19). Data were analyzed and cross tabulated by chi-square test for paired data or Fisher's exact test if frequencies were small, Continuous variables were analyzed using Student's test, with $P < 0.05$ considered significant.

Results

The age and sex distribution of the patients in this study were: 53.8% males and 46.2% females. Most of them (64.12%) were less than 20 years old. There is no significant difference between males and females when the age factor is affixed. Table-1 Respiratory tract infection (RTI) were present in 23(58.9%) of patients, males were 11(47.8%) as compared to 12(52.2%) of female patients. RTI were the most frequent precipitating factor in this study. Diarrhea was the second frequent factor incriminated to precipitate GBS. It was registered in 7(17.9%). They were 5(71.4%) males patients, as compared to 2 (28.6%) female patients. Table-2 The incidence of GBS was more frequent in winter. It was recorded in 22(56.5%) patients. Summer was the second most frequent season for the disease. 11(12.8%) case was reported 8 in this season. Table 3 weakness of the limbs was present in all the patients 39(100%), males 18(46.2%) as compare to females 21(53.8%). Shortness of breath was present in 30(76.9%) of patients, the males 18(60%) were more frequently affected than females 12(40%). Parasthesia and pain were present in 28 (71.7%) and 22 (56.4%) of patients

respectively. also cranial nerve palsy and dysautonomia were present in 15 (38.4%) and 8 (20%) respectively. Table-4 65.3% of the patients who received (IV IG) therapy had complete recovery,19.2% developed neurological deficit and 15.5% dead. While no patients treated by steroid had complete recovery, 15.4% had neurological deficit and 84.6% of them died .There is significant difference

between the mode of therapy and the outcome of patients (p-value <0.001). Table 5 80.8% of patients treated by (IVIG) not need mechanical ventilation, while 76.9% of patients treated by steroid need a mechanical ventilator. There is significant association between therapy type and the future need for mechanical ventilation. Table6

Table1: The age and sex distribution of patients with GBS

Age group	Male	Female	Total
1 – 19	11	14	25(64.12%)
20- 39	3	3	6 (15.38%)
40 – 60	7	1	8 (20.5%)
Total	21(53.8%)	18(46.2%)	39

Table2: The distribution of precipitating diseases among the studied patients

Disease	Male	Female	Total
Respiratory infection	11	12	23(59 %)
Diarrhea	5	2	7(18 %)
Others	2	0	2
No associated illness	3	4	7

Table3: The distribution of GBS according to the seasons

The season	Number of cases
Winter	22 (56.5%)
Spring	1 (2.5%)
Summer	11 (28.2%)
Autumn	5 (12.8%)
Total	39

Table4: The distribution of clinical features of GBS according to sex

The Clinical feature	Male	Female	Total
Parasthesia	15(53.5%)	13(46.5%)	28(71.7%)
Pain	12(54.5%)	10(45.5%)	22(56.4%)
Weakness of the limbs	21(53.8%)	18(46.2%)	39(100%)
Shortness of breath	18(60%)	12(40%)	30(76.9%)
Cranial nerve palsy	9(60%)	6(40%)	15(38.4%)
Dysautonomia	5(62.5%)	3 (37.5%)	8 (20%)

Table5: The distribution of outcome of patients according the therapies

Type of therapy	Complete recovery	Neurologic deficit	Dead	Total	p-value
Immunoglobulin	17(65.3%)	5(19.2%)	4(15.5%)	26(66.7%)	< 0.001
Steroid	0	2(15.4%)	11(84.6%)	13(33.3%)	
Total	17(43.6%)	7 (17.9%)	15(38.5%)	39	

Table6: The distribution of patients on mechanical ventilation according to the type of treatment.

Type of therapy	need ventilator	Not need ventilator	Total	P value
IG therapy	5(19.2%)	21(80.8%)	26(66.7%)	0.001
Steroid therapy	10(76.9%)	3(23.1%)	13(33.3%)	
Total	15(38.5%)	24(61.5%)	39	

Discussions

Little is known about the incidence of GBS in our area. In this retrospective study, it was observed that most of the cases of GBS were in childhood group (1-19 years) as compared to young adult and middle age group, with no sex difference in younger age group. This result is similar to other studies.^(19,20) This study shows an increasing occurrence of GBS in winter season and this result is different from other countries where the attacks reported to occur mainly in spring)^(20,21). This difference can be explained by increase in the incidence of infections in winter as for example viral or bacterial triggering factors and in accordance with other studies that is in favor of infectious associations^(5,23). The outcome of Guillain-Barré syndrome depends on early diagnosis, admission to intensive care unit and administration of immunoglobulin therapy. This approach apparently reduces the need for mechanical ventilation and decrease the incidence of deaths as proved in our study. The delay of initiation of specific immunoglobulin treatments and the use of corticosteroid instead, may attribute to the increase of deaths.^(22,23) Using of steroid or delay of IVIG occurs sometimes due to shortage of supply or because of very high price that cannot be offered by some patients and this happen in other study. The choice of treatment used was greatly affected by the availability of drugs as

in case of (immunoglobulin), and the general condition of patients (presence of autonomic neuropathy) as severe postural hypotension. The exchange transfusion and steroids were used mainly in the year 2007 because the immunoglobulin was not readily available and its high price in the markets and this problem led to high incidence of mortality and neurological deficit, while in the following years 2008 -2010, immunoglobulin given to all patients in the recommended doses for the patients in the ICU. The most important variable in prognosis was the GBS scale and this result was comparable to many previous reports^(18,19, 20). In conclusion Infectious disease is the most common precipitating condition for guilain-baree syndrome; respiratory tract infection was on the top of the list, followed by diarrheal disease. Immunoglobulin therapy is the best therapy for better outcome in GBS.

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