

GUILLAIN-BARRÉ SYNDROME IN NORTH EASTERN IRAN; 1999-2005

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Abstract

Objective

To study the clinical presentation, hospital course and outcomes of patients admitted with Guillain-Barre Syndrome (GBS) to three tertiary care hospitals in Mashhad, Iran.

Materials and Methods

The records of all patients admitted with flaccid paralysis between April 1999 and January 2005 were reviewed and those with the diagnosis of GBS were included in the study. Standard questionnaires were used to record clinical data on was recorded on a standardized questionnaire, which included patients' age, sex, antecedent infectious history, neurological signs and symptoms and ventilation requirements. The hospital course, including therapy given and the functional status of patients, was analyzed, including therapy given and the functional status of patients.

Results

Ninety-one cases of acute flaccid paralysis were admitted to the hospitals during the study period. Eighty-three cases, age range 10 months to 11 years, were later diagnosed as GBS afterwards, with an age range of 10 months to 11 years. The mean age for disease onset was 4.2 years; there were 47 boys and 34 girls, male to female ratio 1:0.7. Upper respiratory tract infection (62.6%) was the most common antecedent event, followed by gastrointestinal infections (19%), urinary tract infection (1.2%) and chicken pox (2.4%), while the remaining cases (14.8%) had no other cases (14.8%) did not have any reliable history of any preceding antecedent infections. Most patients developed GBS within one month of the preceding infection. Cranial nerve abnormalities (19.3%), autonomic dysfunction (7.2%) and respiratory failure requiring intubation (10.8%) were also common. The in-patient mortality was 2.4% (2 of 83).

Conclusion

GBS was found to occur slightly more often in male patients, majority of whom had histories of previous infection. Despite persistent disability, in-hospital mortality was low.

Keywords: Guillain-Barré Syndrome, Acute Flaccid Paralysis, Acute Weakness, Children.

Introduction

Guillain-Barré Syndrome (GBS) is the leading cause of acute neuromuscular weakness in the developed world (1). Different terminologies e.g. acute inflammatory demyelinating polyneuropathy (AIDP), acute motor axonal neuropathy (AMAN), acute motor-sensory axonal neuropathy (AMSAN) and Miller Fisher Syndrome,

are all included under the GBS rubric (2). GBS leads to a wide variety of deficits, characterized by rapidly evolving, symmetrical and often ascending limb weakness, loss of deep tendon reflexes, variable sensory signs and autonomic dysfunction (3). Occurring worldwide, it affects people of all ages and both sexes. Studies on patients with GBS indicate that the disease is the result of aberrant immune responses against components of peripheral nerves. The exact etiology of Guillain-Barré Syndrome remains elusive and incompletely defined. It is generally considered to be a post-infectious disease, with almost two-thirds of patients reporting an infectious illness prior to the onset of GBS symptoms (4).

The purpose of this study was to review and investigate the clinical course and outcome of patients with Guillain-Barré Syndrome admitted to our hospital over a 6-year period from 1999 to 2005. Special emphasis was laid on the characteristics of clinical and laboratory conditions and the complications which developed during their hospital stay. Symptoms at time of admission and on the 60th day of presentation were compared and analyzed.

Patients and Methods

The study was carried out at the Qaem, Imam Reza and Dr. Sheikh Hospitals of Mashhad University of medical sciences over a 6-year period (from 1999 to 2005). Of ninety-one cases of acute flaccid paralysis admitted in these hospitals, 83 patients were diagnosed with Guillain-Barré Syndrome, the diagnosis being confirmed on the basis of clinical presentation, CSF findings, electromyography and nerve conduction studies. A structured questionnaire was designed which documented age of the patient, antecedent history of infection, neurologic dysfunction, admission in intensive care unit (ICU) and the duration of hospital stay. Patient's records also included complications during hospital stay, management, and the treatment given. Using the Modified Rankin Scale, patient symptoms at time of admission and again degree of disability on the 60th day following presentation by Modified Rankin Scale(5). All information was coded to retain patient confidentiality. Data collected was entered and analyzed using SPSS 11.5.

Results

The study included 83 GBS patients, mean age 4.2 years (range 10 months to 11 years); 47 (56.6%) were male, and the male to female ratio was 1: 0.7.

Seventy (84.3%) patients reported a history of infection having occurred within the month prior to onset of GBS; of infections reported, upper respiratory tract infection was the most common (62.6%), followed by gastrointestinal infections (19.0%), chicken pox (2.4%) and urinary tract infection (1.2%); remaining cases however had no histories of prior infection. Sixty nine (83.1%) patients had the initial symptoms of limb weakness and ataxia, the inability to walk; 14 (16.9%) patients had four limb weakness. Another universal finding seen during the course of illness was absence of or reduced deep tendon reflexes (DTR); 48 (57.8%) had decreased DTR, in 23(27.7%) no DTR was detected, and 12 (14.5%) patients had normal DTR. Thirty-nine (42%) patients had cranial nerve involvement including ptosis, gaze problems, nystagmus and facial nerve paralysis and gag reflex dysfunction. Twenty eight (33.7%) patients were admitted in ICU because of severe illness. Sensory disturbance (pain, and paresthesia) was seen in 54 (65.1%) of our patients (Table 1).

Table1. Clinical features in 83 patients with GBS.

Clinical features	Frequency	%
Sensory disturbance	54	65.1
Cranial nerve involvement	39	42
Respiratory distress	16	19.3
Respiratory failure requiring intubation	9	10.8
Hypertension	6	7.2
ICU admission	28	33.7
Decreased DTR	48	57.8
No DTR	23	27.7
Electroneurography		
Axonal variant	17	20.5
Not characteristic	4	4.8
Demyelinating variant	62	74.7

The electromyographic and nerve conduction studies (60/83 patients) in the first week of the GBS (mean \pm S.D. = 5.3 ± 2 days) showed that the demyelinating type of neuropathy was the predominant form of GBS in our patients. Lumbar puncture was done an average 8 days after the beginning of paralysis. Raised protein levels in cerebrospinal fluid (CSF) were seen in 19 (22.9%) patients while 27 (35.5%) had normal values; 37 (44.6%) patients refused to undergo lumbar puncture (LP). The average duration of stay in hospital was 8.7 days (range 1 to 23 days). The most common condition seen, 16(57.1%) of 28 ICU admitted cases, requiring ICU admission was respiratory distress; 9(10.8%) patients had to be intubated for respiratory failure. A majority of patients exhibited a good recovery within sixty days following the initiation of symptoms. In addition to supportive care, 2 patients (2.4%) were managed with plasmapheresis, 46 (55.4%) received intravenous immunoglobulin (IVIg), and 1 (1.2%) received methyl prednisolone. Nine patients (10.8%) received IVIg and methyl prednisolone. Nine patients (10.8%) were treated with IVIg and Plasmapheresis. Two patients (2.4%) were treated with all the three therapies (IVIg, methyl prednisolone and plasmapheresis). Fourteen patients received supportive care per se and were not given any drugs. Six (7.2%) patients, received anti hypertensive drugs besides other drugs. Two patients (2.4%) died of cardiac arrest while they were admitted in hospital.

As all the cases were admitted with acute flaccid paralysis, their condition on day 60 was studied; the median Rankin score, which at admission was 4.5, at discharge had decreased to 3. Results showed that 59 cases (73%) had no remaining paralysis, whereas 22(27%) still had some degree of paralysis on day 60, with a median Rankin score of 1.5.

Discussion

Guillain-Barré Syndrome (GBS) is a relatively common cause of neuromuscular weakness (1, 3-10). Unfortunately, there is a paucity of published information on the local incidence of the condition.

In the current study, we collected data on GBS patients admitted to the hospitals mentioned between 1999 and 2005. The male to female ratio of 1:0.72 in our patients was similar to that cited in other studies (2, 6-10).

Most studies showed an antecedent history of infection in 50 to 71% of patients with GBS (2, 6-8). Our findings showed a higher incidence (84.3%) of antecedent history of infection. In most studies, non-specific upper respiratory tract infection was the most common preceding event (2, 6-9), similar to the findings of our study.

In our study, cranial nerve involvement was seen in 19.2% of patients, which differs from that reported by Cheng (7), Mckhann (8) and Rong-Kuo Lyu (9). None of our patients had confusion or signs and symptoms suggestive of encephalopathy.

The average duration of hospital stay in our patients was 8.7 days (range 1 to 23 days), comparable to that previously reported and relevant to pathologic type of GBS (11,12).

Occurrence of hypertension as part of autonomic dysfunction was seen in six patients, higher than that reported in another study (13).

The percentage of patients requiring ventilatory assistance was lower in our study (10.8%) as compared to those between 21% to 43.1% reported in literature (3,4,6).

Most of our patients had started recovering at the time of discharge. Our patients had a median Rankin score of 4.5 at initial admission; this may be because of late presentation, late recognition or rapid progression of disease. The disease itself may be more severe and/or rapidly progressive in our population. The mortality in our series is low (2.4%), which is comparable with rates of other series (10,11,12).

Most of our patients had demyelinating neuropathy. All our patients who had prolonged recovery had either axonal or undifferentiated Guillain-Barré syndrome (EMG had been suspicious and could not detect any form of GBS). These results indicate that Guillain-Barré syndrome is a disease with a good prognosis in patients who survive the acute stage(9,14-17).

Regarding the small number of patients treated with steroids or combination of IVIg and Plasmapheresis or IVIg and steroids, we were unable to draw any definite conclusion regarding methods of treatment.

Conclusion

In conclusion, GBS was seen in all the pediatric age groups, with a majority of patients having a history of previous infections. Rapid progression, mild disease,

lesser need for ventilation and slow recovery were seen in most patients. Inpatient mortality was low and severe disease on presentation followed by protracted recovery was not associated with poor outcome.

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