Frequency of Cranial Nerve Involvement in Patients with Guillain-Barre’ Syndrome

ABSTRACT

Objective: To determine the frequency of cranial nerves involvement in patients with Guillain-Barre’ Syndrome

Study design: Cross sectional/observational study

Place and duration: This study was done in neurology ward of PIMS Islamabad and medical wards of Bahawal Victoria Hospital Bahawalpur from March 2014 to April 2015.

Materials and methods: Patients of either gender presenting with the primary diagnosis of Guillain Barre Syndrome having age of 15 years or above and admitted to neurology ward of Pakistan Institute of Medical Sciences Islamabad and medical wards of Bahawal Victoria Hospital Bahawalpur were included in the study. Diagnosis of Guillain Barre Syndrome was made on nerve conduction studies. Facial palsy (unilateral or bilateral facial weakness), bulbar palsy (dysphagia, nasal regurgitation, slurring of speech, weakness of the soft palate and/or absent gag reflex) and need of mechanical ventilation within first two weeks of start of disease were recorded on preformed proforma.

Results: 106 patients were included in the study. The mean age of patients was 34.75 years with standard deviation of 16.590 years. Out of 106 patients, 75 (70.75%) patients were male and 31 (29.25%) were female. 32 (30.19%) patients had facial nerve palsy, 42 (39.62%) patients had bulbar palsy (dysphagia, nasal regurgitation, slurring of speech, weakness of the soft palate and/or absent gag reflex) and need of mechanical ventilation within first two weeks of start of disease were recorded on preformed proforma.

Conclusion: About one third of GBS patients have some degree of cranial nerve dysfunction during their illness. Bulbar palsy is more common followed by facial weakness.

Key words: Guillain-Barre’ Syndrome, GBS, Bulbar palsy, facial palsy, respiratory weakness, mechanical ventilation

Introduction

Guillain-Barre’ Syndrome (GBS) is a common cause of acute onset weakness. It is an immune-mediated disease that is usually preceded by gastrointestinal or respiratory tract infection in most of the cases. Various pathogens like Mycoplasma pneumonia, Campylobacter jejuni, Epstein Bar virus and Cytomegalovirus are involved in its pathogenesis and immune response directed against these organisms may cross react with myelin sheath and other neural tissues causing weakness. GBS occurs in whole world with various proportions in all seasons. The incidence varies between 0.4-1.7 cases per 100,000 persons in one year. Core clinical feature of GBS is rapidly progressive ascending weakness starting from lower limbs in the most of cases. Maximum weakness is reached within 2-4 weeks from the onset of illness. After reaching maximum weakness, patients may have a plateau phase of disease lasting from several days to weeks or many months. This phase is followed by slower recovery phase of illness of varying duration of several months to years. Half of patients may have different cranial nerves involvement. In Europe, about a third of patients with GBS are mildly affected and they keep on walking with mild difficulty. Combination of rapidly progressive...
weakness in legs and arms, absent reflexes and protein
cell dissociation on CSF examination, is a hallmark for
the diagnosis of GBS.\textsuperscript{4}

Facial nerve is commonly involved in GBS, occurring in
at least half of patients. Other cranial nerves like bulbar
nerves, abducent and oculomotor nerves are less often
affected.\textsuperscript{5} Miller Fisher Syndrome is a clinical variant of
GBS in which patients may have a triad of ataxia,
areflexia and ophthalmoplegia.\textsuperscript{6,7} Brainstem (Bickerstaff)
encephalitis is another overlapping syndrome that
presents with sudden onset of weakness having multiple
cranial or peripheral nerve involvement. Later on
disturbances of consciousness and even coma may
occur in the course of this disease. Recognition of this
disease is important, because this illness may improve
after the start of treatment.\textsuperscript{8}

Diagnosis of GBS is generally made on history and
clinical examination. Nerve conduction studies (NCS)
and electromyography (EMG) studies can be useful in
the diagnosis of GBS. Delayed distal latencies, temporal
dispersion of waveforms, slowed nerve conduction
velocities, prolonged or absent F waves and H-reflexes
are characteristic findings for demyelination GBS. Low
amplitude of compound muscle action potential (CMAP)
may support axonal GBS. Needle EMG examination
shows neuropathic pattern and it may normal in acute
nerve lesions. Lumbar puncture for cerebrospinal fluid
(CSF) studies is recommended in acute phase of GBS
and it shows albuminocytologic dissociation.\textsuperscript{9} CSF
protein concentrations in patients with GBS are often
normal in the first week, but may increase in more of the
patients at the end of the second week.\textsuperscript{9}

The aim of this study was to determine the frequency of
cranial nerve involvement in patients with GBS in our
local clinical setting which would help in early
management of these patients and reduce the
associated morbidity and mortality.

Materials and Methods
This is a cross sectional/observational study in the
patients presenting with the primary diagnosis of Guillain
Barre Syndrome (diagnosed on nerve conduction study)
having age of 15 years or above and admitted to
neurology ward of Pakistan Institute of Medical Sciences
Islamabad and medical ward-I of Bahawal Victoria
Hospital Bahawalpur during the period of March 2014 to
April 2015. Patients of both genders were included.
Approval from the Hospital Ethics Committees of both
hospitals was taken prior to conducting the study.
Patients having a sensory level on clinical examination,
pregnancy, systemic illness like diabetes, chronic renal
failure, hereditary neuropathies and toxin exposure,
hypokalemic periodic paralysis (serum potassium
<3.5mEq/L) and presence of pulmonary comorbidity on
history &/or chest X-ray were excluded from the study.
The final diagnosis of Guillain Barre Syndrome in these
patients was made on the basis of clinical presentation
(progressive symmetrical weakness in the arms and
legs with or without sensory disturbances,
areflexia/areflexia, in the absence of a CSF cellular
reaction) and nerve conduction studies. Already
designed proforma was used to record the detailed
medical history, facial palsy (unilateral or bilateral facial
weakness), bulbar palsy (dysphagia, nasal regurgitation,
slurring of speech, weakness of the soft palate and/or
absent gag reflex) and need of mechanical ventilation
within first two weeks of start of disease. Data was
entered and analyzed by using SPSS 15.0

Results
106 patients were included in the study. The mean age
of patients was 34.75 years with standard deviation of
16.590 years. The minimum age of patients was 13
years, maximum age was 75 years, median age was 30
years and mode age was 30 years. Out of 106 patients,
75 (70.75%) patients were male and 31 (29.25%) were
female. The frequency of facial palsy, bulbar palsy and
need of mechanical ventilation was shown in figure no:
1.

Figure no 1. Frequency of facial palsy, bulbar
palsy and mechanical ventilation

Out of 106 patients, 22 (62.9%) male and 13 (37.1%)
female patients had need of mechanical ventilation with
insignificant p value of 0.209. Out of 106 patients, 29 (69%) male patients and 13 (31%) female patients had bulbar weakness with insignificant p value of 0.754. Facial palsy in different gender of patients was shown in figure no: 2. Frequency of facial palsy, bulbar palsy and need of mechanical ventilation in different age groups was shown in table no: 1

Table no I. Facial palsy, bulbar palsy and mechanical ventilation in different age groups

<table>
<thead>
<tr>
<th>Age group of patients</th>
<th>Total n=106</th>
<th>p-value</th>
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<tr>
<td>13-35 years</td>
<td>32</td>
<td>0.113</td>
</tr>
<tr>
<td>36-55 years</td>
<td>24</td>
<td></td>
</tr>
<tr>
<td>56-75 years</td>
<td>5</td>
<td></td>
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<tr>
<td>Facial palsy n=106</td>
<td>15</td>
<td>14.2%</td>
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<tr>
<td></td>
<td>12</td>
<td>11.3%</td>
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<tr>
<td></td>
<td>5</td>
<td>4.7%</td>
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<td>Bulbar palsy n=106</td>
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<td>22.6%</td>
</tr>
<tr>
<td></td>
<td>12</td>
<td>11.3%</td>
</tr>
<tr>
<td></td>
<td>6</td>
<td>5.7%</td>
</tr>
<tr>
<td>Mechanical ventilation n=106</td>
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<tr>
<td></td>
<td>10</td>
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</tr>
<tr>
<td></td>
<td>6</td>
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</table>

Discussion

GBS is an acute demyelinating polyneuropathy with rapid onset (hours to days) of weakness. This usually produces symmetrical weakness of lower and upper limbs and axial musculature, which may progress to respiratory failure and swallowing problems. Autonomic involvement may occur to cause blood pressure fluctuations and cardiac arrhythmias. With the progression of disease, sensory symptoms like numbness, burning and tingling sensation may appear, but the picture is usually dominated by motor weakness. Although the most common presentation is an ascending weakness, but the patients can presents with cranial muscles paralysis.10

In our study, the mean age of patients was 34.75 years with standard deviation of 16.590 years. 75 (70.75%) patients were male and 31 (29.25%) were female. Cranial nerve abnormalities were present in about 30-39% patients. These results were comparable to the results of other studies. In a study conducted by Areeyapinan P et al11 showed that the most common presentation was motor weakness in 87% of patients, sensory symptoms were present in 78% of patients and facial weakness was present in 18% of patients. Electrophysiological studies showed demyelinating process in most of cases as compared to axonopathy. In another study conducted by Dhadke SV et al12 showed that younger age group was commonly affected by GBS. In this study, male to female ratio was high. Motor weakness was present in 39 out of 40 patients. Cranial nerve weakness was present in 25 out of 40 patients. Facial and bulbar muscles were equally affected in 30% of patients. Reflexes were absent in all studied patients. Some studies were showing higher frequency of cranial nerve involvement than the results of our study. In a study conducted by Netto AB et al13 showed that respiratory involvement was present in 273 patients (190 patients were men and 83 patients women). Out of 273 patients with respiratory failure, bulbar palsy, sensory involvement and symptomatic autonomic dysfunction were found in 68.1%, 32.2% and 26.4% respectively.

In another study conducted by Azim A et al14 showed that the mean age of patients was 32.4±18.12 years. Majority of patients were male. Autonomic dysfunction was present in 35% of patients and 64% patients needed mechanical support. Almost similar results were found in a study conducted by Yakoob MY et al15 with cranial nerve abnormalities in 88.2% patients, autonomic dysfunction in 61.8% patients and respiratory failure requiring mechanical ventilation in 56% of patients.

Durand MC et al16 included one hundred and fifty four patients with GBS in the study and 22% patients were subsequently required ventilation. Demyelinating GBS
was more common as compared to axonal GBS in patients who required mechanical ventilation. Facial palsy and bulbar palsy were presumed clinical factors causing the need of mechanical ventilation in patients of Guillain-Barre Syndrome. However, some studies were showing minimal cranial nerves involvement like a study conducted by Yuan CL et al who studied 49 cases of GBS, with non-specific age distribution. Motor weakness was the most common presentation. Cranial nerve abnormalities, autonomic dysfunction and respiratory failure were minimal in their patients. Acute multiple cranial neuropathies without limb weakness may be considered as variant of Guillain-Barre syndrome, which are immune-mediated processes that are triggered by various pathogens. Isolated Bell's palsy may be present as a variant of Guillain-Barré syndrome with recognized cell-mediated immunity against peripheral nerve myelin antigens. In Bell's palsy and GBS, a viral infection or the reactivation of a latent virus may provoke an immune reaction against peripheral nerve myelin sheath, leading to the demyelination of cranial nerves, especially the facial nerve. The prognosis of GBS is generally favorable with a mortality of ten percent and significant number of patients is left with severe disability.

Conclusion

In conclusion, GBS is seen in all age groups but more common in younger age group. About one third of GBS patients have some degree of cranial nerve dysfunction during their illness. Bulbar palsy is more common followed by facial weakness. Mechanical ventilation, which is the final consequence of several clinical factors is required in many patients of Guillain-Barre Syndrome.

References