Guillain Barre Syndrome in Children at Tikur Anbessa Specialized Hospital
Tigist Bacha, MD

Abstract

This is a six years retrospective descriptive study conducted in the pediatric and child health department of Tikur Anbessa Hospital from September, 2001 – September, 2006 G.C to assess the pattern of acute flaccid paralysis (AFP) and the clinical and epidemiologic features of Guillain Barre syndrome (GBS).

Data was collected from medical records of all patients admitted with diagnosis of AFP, and analyzed using standard statistical tests with SPSS version 14 software. Out of 70 admitted cases of AFP, forty six (65.7%) were males and 24 (34.3%) were females. Sixty seven cases (95.7%) were diagnosed to have GBS and the rest three were compatible with poliomyelitis, transverse myelitis and post injection neuritis. Out of the 70 cases 66 (94.3%) have received at least one dose of polio vaccination and the rest 4 (5.7%) were never vaccinated.

Out of the cases diagnosed to have GBS, 44 (65.7%) met NNCDS diagnostic criteria. History of antecedent event was obtained in 31/67 (46.3%) patients. Majority of the patients 45 (67.17%) presented with ascending reflexes quadriparesis, 2 (2.98%) patients with descending areflexic quadriaparesis, 19(28.35%) only with lower limb involvement and 1 (1.5%) with typical miller-fisher type. Sensation was affected in 4 patients.

Cytoalbuminological dissociation was found in 27(40.3%). There were 11 deaths (16.4%) of whom five were admitted to ICU the rest six didn’t. This study showed that the commonest cause of AFP is GBS which is associated with high mortality. This high mortality rate 11/67 (16.42%) is attributed to absence of pediatrics ICU, late arrival to Hospital after onset of illness, and poor supportive care.
INTRODUCTION

Acute flaccid paralysis (AFP) is the clinical condition diagnosed in any child under 15 years of age with acute floppiness of one or more limbs or any age in whom clinician suspect polio (1,2). It is caused by many conditions including Guillain Barre syndrome (GBS), poliomyelitis, transverse myelitis and metabolic neuropathy like hypokalemia (3,4). The most common cause of AFP is GBS followed by transverse myelitis (2,5,6,7).

In Ethiopia, the polio eradication initiative (PEI) was started in 1996. Surveillance for acute flaccid paralysis (AFP) was initiated in May, 1997. AFP surveillance is the detection of at least one AFP case per 100,000 children under 15 years of age. AFP surveillance depends on immediate reporting, investigation of AFP cases, routine monthly reporting of cases including zero reporting (1, 9).

GBS is an acute inflammatory polyneuropathy. The cause is unknown but autoimmunity is incriminated. Unlike polio, GBS is usually symmetrical (asymmetrical in 9%), and fever at presentation is not present. Paralysis develops acutely over days, or at most weeks. After brief plateau the patients' improvement begins with gradual resolution that lasts from weeks to months. 50% has bulbar involvement, 33% of them require ICU admission 25% required mechanical ventilation 5-10% mortality rate (2-3% in best ICU) and 80% complete recovery. Treatment is intensive care support; early intravenous immunoglobulin (IVlg) therapy and plasma exchange hasten early recovery. Corticosteroid alone outcome is controversial (3,8,10).

The objectives of the study are to describe pattern of AFP, determine the most common cause of AFP, and determine the clinical and epidemiological feature of GBS.

Material and method

Retrospective descriptive study was done in pediatric and child health department of Tikur Anbessa Hospital, Addis Ababa. Study groups were all children admitted with the case definition of AFP from September 2001 – September 2006. The medical report of Statistics Office registered for AFP surveillance, Pediatric wards, surgical and medical ICU and neurology clinic registration books were reviewed to trace all cases. This hospital is the only pediatric tertiary Hospital serving for all the country.

In this study:

Antecedent triggering event was defined as the presence of respiratory, gastrointestinal, febrile illness or vaccination for the previous 4 weeks.

Polio-compatible case is defined as a case in which one adequate stool specimen was not collected from a probable case within 2 weeks of the onset of paralysis, and there is either an acute paralytic illness with polio-compatible residual paralysis at 60 days, or death takes place within 60 days, or the case is lost to follow-up.

The diagnosis of GBS in this study is based on National Institute of Neurological and Communicative Disorders and stroke (NNCDS) diagnostic criteria. (11)
1. Progressive weakness of more than one limb due to neuropathy,
2. Areflexia or hyporeflexia
3. Duration of progress less than 4 weeks,
4. The absence of a sharp sensory level on the trunk,
5. The absence of other causes of acute neuropathy
6. Less than 50 mononuclear leukocytes per mm³ of CSF.

The data were transferred to a structured form which was then entered into computer database. SPSS Version 14, 2005 was used to process the statistical data.

Results

In this six year period 70 AFP cases (46 male and 24 female) were admitted and majority 67 (95.7%) were GBS. Three cases were suspected to have a compatible poliomyelitis, transverse myelitis and post injection neuritis each accounting 1.4%. The mean age of AFP is 5.82 year with a range of 1-12 year. Cases were reported from different regions of Addis Ababa 44, Oromia 12, Amhara 11 and South Nations Nationalities and Peoples region (SNPPR) 3 (Fig 1). The distribution over the years is shown in Fig 2 from 2001-06 (1993-1998 Eth C). More cases were seen in 1996 Eth C. Stool sample was taken for polio in 65 cases and was not taken in 5 of them. Out of the 70 cases 66 (94.3%) of the cases got at least one dose of polio vaccination and the rest 4 (5.7%) never vaccinated (table 1). Prior injection history is found in 7 (1%) of the patients.
Table 1. Cranial nerve involvement

<table>
<thead>
<tr>
<th>Cranial nerve involved</th>
<th>Frequency</th>
<th>Percentage</th>
</tr>
</thead>
<tbody>
<tr>
<td>Facial nerve</td>
<td>3</td>
<td>27.2</td>
</tr>
<tr>
<td>Optalmolopelgia</td>
<td>1</td>
<td>9.1</td>
</tr>
<tr>
<td>Glossopharyngeal nerve</td>
<td>5</td>
<td>45.4</td>
</tr>
<tr>
<td>Vagus nerve</td>
<td>1</td>
<td>9.1</td>
</tr>
<tr>
<td>Multiple cranial nerve</td>
<td>1</td>
<td>9.1</td>
</tr>
<tr>
<td>Total</td>
<td>11</td>
<td>100</td>
</tr>
</tbody>
</table>
Table 2: CSF analysis Result

<table>
<thead>
<tr>
<th></th>
<th>Frequency</th>
<th>Percent</th>
</tr>
</thead>
<tbody>
<tr>
<td>CSF cell count &gt;10/cumm</td>
<td>3</td>
<td>4.5</td>
</tr>
<tr>
<td>and protein &gt;45mg</td>
<td></td>
<td></td>
</tr>
<tr>
<td>CSF cell count &gt;10/cumm</td>
<td>7</td>
<td>10.4</td>
</tr>
<tr>
<td>and protein &lt;45mg</td>
<td></td>
<td></td>
</tr>
<tr>
<td>CSF cell count &lt;10/cumm</td>
<td>27</td>
<td>40.3</td>
</tr>
<tr>
<td>and protein &gt;45mg</td>
<td></td>
<td></td>
</tr>
<tr>
<td>CSF cell count &lt;10/cumm</td>
<td>21</td>
<td>31.3</td>
</tr>
<tr>
<td>and protein &lt;45mg</td>
<td></td>
<td></td>
</tr>
<tr>
<td>Not Documented</td>
<td>9</td>
<td>13.4</td>
</tr>
<tr>
<td>Total</td>
<td>67</td>
<td>100.0</td>
</tr>
</tbody>
</table>

From those diagnosed as having GBS 44(62.8%) met NNCDS diagnostic criteria. The mean age was 5.9 with ranges of 15 months -12 years. Most of them were male. History of antecedent event was obtained in 31(46.3%) patients. Respiratory symptoms accounted 19(27.1%), gastrointestinal symptoms 9 (11.4%), both gastrointestinal and respiratory symptoms 1 (1.4%), 1 (1.4%) post vaccine (Rabies), and 1 (1.4%) had prior malarial attack. The pattern of progression of paralysis was ascending areflexic quadripareisis in 45 (67.2%) patients, descending areflexic quadripareisis in 2(3%), patients with only lower limb involvement were 19 (28.3%) and one (1.4%) was Miller - Fisher variant. The mean interval between onset of symptoms to hospital admission was 6 days with a range of 5 hrs-14days. Rapidity of the progression ranged from 1 day to 14.0 days the mean being 3.44 days. The mean duration of hospital stay is15.5days. Out of the 17 patients whose blood pressure was measured 12 patients had normal measurement and 5 had transient hypertension. Bladder and bowel dysfunction was reported in 44 out of 64 documented cases. Cranial nerve palsy was reported in 11 patients. As shown in table xxx the commonest were cranial IX (45.4%) followed by facial nerve (27.2%). Multiple cranial nerve involvement was found in 1(9.1%) patient. Sensation was affected in 4 (6%) cases. Ataxia was documented in 8(11.9%) patients. Cytoalbuminological dissociation was found in 27(40.3%) patients. see table 5. EMG was done in 5 patients with GBS out of which 2 were demyelinating, 2 axonal and 1 mixed axonal and demyelinating.

Fifteen15/67 (22.39%) patients required ICU care out of which 6 didn’t get the service. There were 11(16.4%) deaths. Five died in ICU and 6 patients died in the pediatric ward. Four of them came within 4 days of onset of illness and 7 of them after 4 days. Respiratory failure was considered as a cause of death in 6. Respiratory failure and infection were cause of death in 5 of them (3 had pneumonia, 1 urosepsis). Specific treatment such as plasmapheresis and Immunoglobulin was given to none of the patients. Prednisolon was given only for 4 patients (1.0%).HIV screening was done only for one patient and the result was non reactive.
Discussion

This is the first study done on pattern of AFP and clinical and epidemiology of GBS in the pediatric age groups. The predominance of male is similar to other studies (6-10). The common AFP identified was GBS which also correlates with other studies in Australia, Bangladesh, and Honduras (2,6-8).

Regarding GBS the antecedent events are lower in this study than other studies done in Kenya, Tanzania, Nigeria and including the study done in this hospital in adults (12-15). Similar to other studies respiratory symptoms were the commonest antecedent events (12). The cranial nerve involvement are common findings similar to that of Kenya and adult Ethiopian patient’s (12, 13).

In this study one patient gave history of antecedent malarial attack the species not documented. From other studies the development of Guillain–Barré syndrome was reported in 10 patients who had had acute P. falciparum malaria during its seasonal exacerbation is reported (16).

The high mortality rate is higher to the report from other studies done in Kenya (13) and Tanzania (9). Also a higher mortality was observed in adult study done in the same hospital (12). The main attributable cause is lack of good pediatric ICU in this Hospital which could be compounded by the late appearance of patients to the hospital. In addition specific therapies such as immunoglobulin and plasmapheresis are not available in the setup.

This study showed a high need of having pediatric ICU. We can see the mortality rate from GBS is high some of which could have been prevented if there was a pediatric ICU. The care of critical patients and universal precautions for infection prevention should be encouraged. In addition the use of specific treatments such as plasmaphereis and immunoglobulin should be introduced.

For five cases stool sample for polio was not taken out of whom four died. Taking specimens as soon as possible especially on critical patients, including on weekend may increase the surveillance. Improving the AFP surveillance increased by case investigation reporting cases as early as possible especially on critical patients should be encouraged.
In addition care seeking behaviour should be improved so that patients are brought to hospital as early as possible. Finally a further large scale study with more investigation modalities should be done in the near future.
References


Acknowledgments:
I extend my thanks to Dr. Ahmed Bedru for his invaluable advice in the conduct of the research. Ato Tilahun Zimita for his assistance in data entry and analysis.