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ORIGINAL ARTICLE

Clinical and electrophysiologic features of childhood Guillain-Barré syndrome in Northeast China

Yu-Qin Ye [a,*], Ke-Ren Wang [b], Li Sun [a], Zan Wang [a]

[a] *Department of Neurology, First Hospital, Jilin University, Changchun, PR China*
[b] *Breast Surgery, China-Japan Union Hospital, Jilin University, Changchun, PR China*

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Background/Purpose: Since little has been reported in previous studies, we aimed to find the clinical and electrophysiologic characteristics associated with childhood Guillain-Barré Syndrome (GBS) in Northeast China.
Methods: The clinical and electrophysiologic data were collected and reviewed retrospectively in 33 children and 105 adults with GBS during the period between 2006 and 2010 from the First Hospital of Jilin University.
Results: Most of the children with GBS were older than 8 years of age and symptoms were severe at GBS onset. Simultaneous involvement of four limbs was the most common clinical feature, and cranial nerve involvement was common; however, previous infection, sensory nerve involvement and elevated proteins in cerebrospinal fluid occurred much less in the children with GBS than those in adult patients. Recruited children were classified as having acute inflammatory demyelinating polyneuropathy (AIDP; 41%), acute motor axonal neuropathy (AMAN; 38%), and were unclassified (21%). Electrophysiologic features and prognosis in these children were not different from those in adults. For children with AMAN, the efficacy of intravenous immunoglobulin (IVIg) was not different from that in adults; however, IVIg was not significantly effective for AIDP in these children.
Conclusion: Childhood GBS in Northeast China exhibits characteristics of clinical and electrophysiologic alternations; early diagnosis and appropriate treatments should be provided accordingly.

Conflicts of interest: The authors have no conflicts of interest relevant to this article.

* Corresponding author. Department of Neurology, First Hospital, Jilin University, Number 71, Xinmin Street, Changchun, PR China.
E-mail address: dearlea@live.cn (Y.-Q. Ye).

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Introduction

Guillain-Barré Syndrome (GBS) is the most common cause of acute and severe generalized peripheral neuropathic weakness.[1] Studies have found that one-third of childhood cases occur before the age of 3 years, and 50%—70% of the cases have an antecedent infection 2—4 weeks prior to GBS onset.[2,3] Weakness usually starts in the lower extremities, and sensory abnormalities and cranial nerve involvements are common in childhood GBS. Some reports have emphasized that in China, acute motor axonal neuropathy (AMAN) primarily affects children, and it is controversial as to whether AMAN is associated with a poor prognosis. Additionally, some studies indicate that prognosis is better in children than that in adults.[4,5] However, little has been reported regarding the clinical and electrophysiological features present in children with GBS in Northeast China. Here we aimed to determine whether there were identifiable clinical and electrophysiologic characteristics of childhood GBS in order to make early diagnosis and appropriate treatments.

Materials and methods

Patients

A retrospective study was performed for this study. Thirty-three children (age younger than 15 years) and 105 adults with GBS who were admitted to the First Hospital of Jilin University between 2006 and 2010 were recruited. All patients came from Jilin province, Northeast China and fulfilled the clinical criteria for GBS.[6] Patient criteria were evaluated using the functional grading scale produced by Hughes et al.[7] The patients were classified as severely affected (inability to walk when unaided, GBS disability scale $\geq$3) or mildly affected. Lumbar puncture (normal cell count value: 5/μL, normal protein: 45 mg/dL) was performed in children between 10 and 16 days (mean 12.7 days) after GBS onset. In adults the lumbar puncture was performed between 10 and 17 days (mean 12.5 days) after GBS onset. Intravenous immunoglobulin (IVIg) was administered when GBS was diagnosed and efficacy of IVIg was measured approximately 2 weeks after IVIg treatment. The efficacy was considered as improved if there was an increase in one or more points in Hughes score.

Electrophysiology

Nerve conduction studies (NCSs) were performed using conventional procedures. Motor NCS was performed on the median, ulnar, posterior tibial, and deep peroneal nerves. Stimulus sites for the median and the ulnar nerves were defined at the wrist and elbow and electrophysiologic recordings were performed in the abductor pollicis brevis and the abductor digit minimi. The stimulus site for the tibial nerve was at the ankle and popliteal fossa and the recording was conducted in the fiexor hallucis brevis. For the deep peroneal nerve at the ankle and fibular head, the recording was in the extensor digitorum brevis muscle. Sensory NCS included stimulation of the median nerve, ulnar nerve, superficial peroneal nerve, and sural nerve. F waves of the median and ulnar nerves were measured after compound muscle action potentials (CMAPs) were obtained in the motor NCS. The H reflex of the posterior tibial nerve was recorded from the gastrocnemius muscle. CMAP amplitude, distal motor latency, motor nerve conductive velocities (MCVs), conduction block, sensory nerve conductive velocities (SCVs), sensory nerve amplitude, average F response latencies, F wave incidence, and H reflex amplitude and latency were measured. Needle electrode examination (NEE) of the extensor digitorum communis and tibialis anterior muscles was performed. Values were defined as abnormal if they were outside of the age corrected normal range. Patients were classified as having AMAN or acute inflammatory demyelinating polyneuropathy (AIDP) on the basis of the electrodiagnostic criteria reported by Hughes and colleagues.[8] A diagnosis of AIDP was made if at least one of the following parameters were present in each of at least two nerves, or at least two of the following parameters in one nerve if all others were unexcitable and the CMAP amplitude after distal stimulation (dCMAP) $>$10% lower limit of normal (LLN). These parameters included MCV $<$90% LLN (85% if dCMAP $<$50% LLN), distal motor latency $>$110% upper limit of normal (ULN; $>$120% if dCMAP $<$100% LLN), CMAP amplitude after proximal stimulation (pCMAP)/dCMAP ratio $<$0.5, and dCMAP $>$20% LLN; F wave latency $>$120% ULN). AMAN was diagnosed as none of the features of AIDP except one demyelinating feature allowed in one nerve if dCMAP $<$10% LLN, and sensory action potential amplitudes are normal. The study protocol was approved by the Human Ethics Committee of Jilin province, P. R. China, and informed consent was obtained from all patients.

Statistical analysis

Differences in proportions were tested with the Chi-square test. A p value of $<$0.05 was considered significant.

Results

Clinical features of childhood GBS

The clinical data were summarized in Table 1. Twenty-seven patients of childhood GBS were older than 8 years of age. Simultaneous involvement of four limbs was one of the most common clinical features. Cranial nerve involvement (mainly facial nerve palsy and glossopharyngeal and vagal nerve involvements with dysphagia, choking cough, and dyspnea) was also common. Children with GBS had fewer previous infections (mainly gastroenteritis and respiratory infection), sensory abnormalities (mainly symmetrical numbness of the distal limbs and tingling), or elevated proteins in the cerebrospinal fluid when compared with adult patients. Clinical presentations in children with GBS were more severe than adult patients, six (18%) and 33 (31%) were mild patients in children and adults, respectively, of which two children and two adults deteriorated within 7 days after admitted to hospital. However, we found that treatment of IVIg in children was not as effective as in adults.