Segmental and suprasegmental speech processing in a child with Landau-Kleffner Syndrome

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Abstract

In the present study we examined the speech processing skills of a child having Landau-Kleffer syndrome (LKS). We assessed two kind of language processing skills using event related brain potentials (ERPs): the discrimination of segmental and that of suprasegmental cues. Results indicate that the two kind of cues were processed asymmetrically: the mismatch negativity component (MMN) was obtained for the phoneme difference, however no difference in the ERPs was obtained for the stress pattern difference. At the same time, another patient having Benign epilepsy of childhood with centrotemporal spikes (BECT), a childhood epilepsy similar to LKS to some extent, with no language problems showed MMN to both kind of difference. Beside the ERP study several other assessment were carried out so as to obtain a more complex picture on the language deficit, including the examination of the source of epileptic activity using electric source localizations method and PET, and the neuropsychological assessment of cognitive and behavioral skills.

As a conclusion we believe that LKS is language disorder that can be used to study what happens to the language system if in the course of development the essential neural circuits are severely disturbed. We believe also that the tools we use, that is the electrophysiological measurement of pre-attentive language processing is a fruitful approach since it makes possible the assessment of subtle processing skills while it does not require the patient / child to be responsive.

1. Introduction

Understanding spoken language is based on the processing of two kind of acoustical features: segmental and suprasegmental. Processing of segmental information (acoustical information of individual phonemes) received a much higher attention in the field of speech perception than the prosodic information (acoustic properties that are not the properties of individual segments and can apply to a string of several units). However it is evident that prosodic information (intonation, speech rhythm, stress. etc.) is present in the speech-flow and its role extends to many levels of language processing. Thus, the segmentation of speech stream into words is partly accomplished by the use of stress information, [1], syntactic parsing is sometimes helped by intonation [2], and the pragmatic and emotion-expressive role of prosody is undoubted. Therefore it may be presumed, that proper processing of prosodic cues is essential for our understanding of language.

In the present paper we present the case of a child having Landau-Kleffner Syndrome (LKS), as a clinical model for speech perception. LKS is a rare childhood epilepsy accompanied by acquired aphasia [3;4]. It is mainly characterized by an abnormal EEG pattern (focal spike-and-wave activity) and progressive language loss. The onset of the disorder is usually at the age of 3-8 years, and it does not always manifest in clinical seizures which makes the diagnoses of the syndrome problematic. The prognosis is relatively good, as a spontaneous recovery from the epilepsy is likely at the beginning of adolescence, but this is not the case for the acquired language disorder. A severe residual language deficit may remain, and the onset of the syndrome is of a major effect on the outcome: the younger the child at the onset, the worse is the chance of recovery [5].

It is agreed that in LKS the epileptic activity leads to a severe language deficit. The question is whether the language problem is broad or specific, and if specific, which level(s) of processing are possibly affected. Accordingly, we wanted to see whether the processing of both segmental and suprasegmental cues is working well in LKS. As a clinical control, we examined a child having Benign epilepsy of childhood with centrotemporal spikes (BECT), which is a similar childhood epilepsy, but without language or other serious cognitive problems [6].

2. Methods

2.1. Medical history

The patient we examined was a 6 years old boy having Landau-Kleffner Syndrome. His medical history commenced at the age of 4.5, when he had a single episode of tonic-clonic seizure after waking up. Previously, he did not have any other medical problems, and the general examination did not reveal any physical or neurological problems. The EEG examination revealed continuous slow spike-and-wave (SSW) activity in sleep, and sometimes even in awake state. The activity was focal and could be localized to the left hemisphere of the brain, mainly in the centro-temporal region. During the following 1.5 year, he received continuous medical treatment, and although the epileptic activity could be controlled by using ACTH, his understanding and production of speech became very low.

The BECT patient was a 6 years old girl, who also had SSWs in sleep and in awake, but she demonstrated no language problems, but mainly behavioral.

2.2. ERP study

Processing of segmental and suprasegmental speech cues were assessed using a passive oddball paradigm. The setting was identical to the one used in a previous study with normal adults [7]. The segmental condition consisted of the discrimination of word initial phonemes, where a two syllable meaningful word /bɔnaːn/ was contrasted with its meaningless pair differing only in the initial phoneme (/p/ vs. /b/). The suprasegmental condition used differing stress patterns: the obligatory Hungarian trochaic stress pattern (stressed followed by an unstressed syllable) was contrasted with a non-existing iambic pattern (unstressed followed by a stressed syllable). For that purpose the same word /bɔnaːn/ was used, and the iambic pattern was created by pronouncing the word with this different stress pattern.

Stimuli were presented in a passive oddball paradigm. In general, the passive oddball paradigm involves presenting frequent (standard) and rare (deviant) stimuli differing in some discriminable way. The deviant stimulus usually elicits a frontocentrally negative ERP component 100-250 ms after the onset of change, the so-called Mismatch Negativity (MMN) [8]. This component is elicited irrespective of whether the stimuli are attended or not, and provides a stable measure for electrophysiological assessment of auditory perception, even in developmental or clinical populations (see for instance [9]). Here, the word /bɔnaːn/ was used as standard, and the two contrasted stimuli (phoneme and stress) as deviants.

The EEG registration was performed using a 32 channel recording system (BrainAmp amplifier, BrainVision Recorder recording software and BrainVision Analyzer analysis software, BrainProducts GmbH) with electrodes mounted according to the 10-20 electrode placement system by means of an elastic electrode cap, on sites Fp1, Fp2, F9, F7, F3, Fz, F4, F8, F10, FC5, FC1, FC2, FC6, T9, T7, C3, Cz, C4, T8, T10, CP5, CP1, CP2, CP6, P7, P3, P4, P8, O1, O2, P9, P10, and with Pz as reference and with a ground placed between Fz and Fpz in the midline. The sampling rate of the registration was 500 Hz, and an online bandwidth filter of 0.15-70 Hz was applied.

2.3. Localization of epileptic source

Several methods were applied in order to obtain a comprehensive view on the location of possible brain generators of epileptic activity. Localization of SSW activity based on awake EEG and a PET measurement was applied.

For inferring the possible localization of SSW source, several SSWs were averaged. This was performed using the automatic pattern search feature of BESA 5.0 software (MEGIS Software GmbH). The first SSW was identified visually, then an automatic search was applied in order to trace all SSWs with similar spatio-temporal patterns (with a correlation threshold of r>0.75). Consequently, 53 (LKS) and 25 (BECT) SSWs were obtained, the average of which served for analysis of the epileptic activity.

The source analysis was performed with BESA 5.0, and followed the sequential dipole fitting strategy described by Scherg, Bast and Berg [10]. First, a regional source was fitted to the spike peak, and then other regional sources were fitted to the onset phase so as to minimize the unexplained variance, until the unexplained variance reached a level below 5%.

The PET study was performed in the PET laboratory of the Debrecen University and was part of the neurological examination. As tracer 2-[18F]Fluorodeoxyglucose (FDG) was used in a GE 4096 PLUS PET scanner (axial FOV: 105 mm). Data acquisition was started 40 minutes after the injection of 3.7 mCi FDG and 5 min long successive exposure times were chosen. During the scans the child was laying still in his back with the eyes open and had no task. Reconstructed PET data were registered with T1 weighted MR scans. PET was only available for the LKS child.

2.4. Neuropsychological assessment

The neuropsychological assessment was performed by means of a comprehensive neuropsychological test battery, NEPSY® [11]. The test includes several different tasks intended to evaluate various aspects of the child's cognitive profile. The main cognitive functions assessed are attention and executive functions, language, memory, visuo-spatial functions and sensorimotor functions. All tests were translated into Hungarian. The language tasks were established according to the original aims of the NEPSY test, and taking into account the specific features of Hungarian language. Neuropsychological assessment was performed only with the LKS child.

3. Results

3.1. ERP results

ERPs of both children to both deviant conditions are shown in Figure 1. It must be noted that for the BECT patient ERPs were calculated from a smaller number of responses, because her EEG was highly contaminated by spike-wave activity. The difference waves demonstrate a negative deflection at about 200 ms for the phoneme deviant for both patients. This negative deflection can be considered as an MMN component indicating the automatic processing of /b/-/p/ phoneme contrast. At the same time, the stress deviant elicited the same MMN only for the BECT patient but not for the LKS patient.

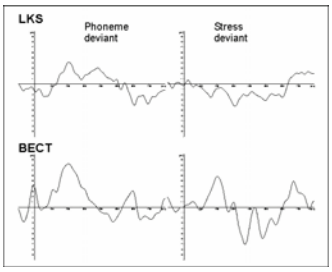

Figure1: Difference waves (ERPs to deviant minus standard stimuli) for both LKS and BECT patients.

3.2. Results of source localization

The results of source localization are shown on Figure 2. As it can be seen, the SSW activity could be modeled by three regional sources for the LKS patient and with only one for the BECT patient. The location of the main source was in different cortical areas: for the LKS patient, it could be localized in the posterior part of the left superior temporal lobe, while for the BECT patient it was in the left fronto-central lobe.

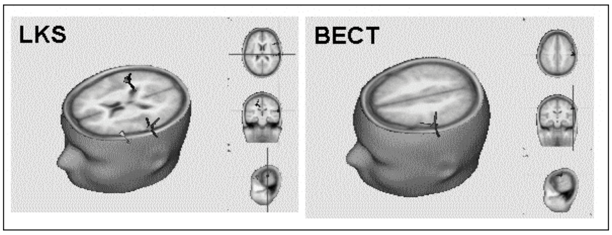

Figure2: Results of source localization for both patients

PET measurement was obtained only for the LKS patient. The MR and registered FDG-PET images are overlayed and shown in Figure 3. Comparing the regional radioactivity concentration of the different anatomical regions a higher FDG uptake was observed in the left temporo-parietal cortex relative to the rest of the brain, indicating a pronounced hyper-metabolism.

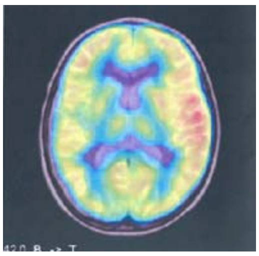

Figure3: Overlayed MR and FDG/PET slice

3.3. Results of neuropsychological assessment

Neuropsychological assessment was performed only for the LKS patient. The results showed a major dissociation of the visual and verbal functions, which was most obvious in the domain of attention, where a good visual attention performance was contrasted with a low auditory attention performance. The considerably low values of the auditory-verbal domain could also be well seen in the language and memory profiles and subtest. The memory subtest scores were very low, and the worst performance occurred when verbal recall was needed to accomplish the task. The performance in two tasks was considerably high (3 standard deviations above mean): that of the "Tower" and "Block Construction". Each of those task required manipulation of objects. So it seems that the patient had highly spared skills in both domains, which were much above the expected level, therefore they might be considered as possible compensation strategies used in every day's life of the child.

In sum, the neuropsychological assessment showed a particular dissociation of language, acoustic attention and verbal memory versus executive functions, visual attention and visuospatial functions, suggesting that the disturbed functions of the left centro-temporo-parietal areas are expressed in all related tasks, while the right parietal areas are left intact.

4. Discussion

The evaluation of segmental and suprasegmental discrimination skills using the method of ERPs demonstrated that the LKS patient had an MMN component in the phoneme task, but not in the prosodic task, while the BECT patient showed the MMN component is both tasks. Given that the MMN is believed to index pre-attentive discrimination of stimuli [8] the interpretation of these results is that the LKS patient appears to have a preserved automatic phoneme discrimination, while the discrimination of altered stress pattern is deficient. At the same time the processing of both segmental and suprasegmental cues seems to work normally in the BECT patient.

Regarding the results of SSW source localization, it can be concluded that the two patient had the major source of epileptic activity in slightly different cortical region (superior temporal lobe and fronto-central lobe).

For the LKS patient the localization of SSW activity is further supported by the results of PET measurement, which showed hypermetabolism in the left superior temporal regions. This result is in agreement with the findings of Maquet et al. [12] who also found focal hypermetabolism in all 6 patients measured during the active phase of LKS. The hypermetabolism was localized to either the left or the right hemisphere, and it was supposed to be due mainly to the focal epileptic activity, affecting mostly cortical areas.

It can be hypothesized that the epileptic activity influences the functioning of specific brain areas leading to specific deficit of those cognitive functions which are linked to these areas. Therefore it can be supposed that the different locus of SSW activity in the two patients causes the dysfunction of different elements of language network, and this is manifested in the ERP results. For the LKS patient this is further supported by the results of neuropsychological assessment, showing the dissociation of verbal and non-verbal skills, and especially the dysfunction of memory functions.

The importance of verbal working memory is emphasized by findings showing a specific deficit of working memory system [13]. The separate assessment of phonological store and auditory loop components of verbal working memory showed impairment only in the phonological store. Thus, one possible deficit contributing to the language problem in LKS might be the dysfunction of verbal working memory.

In a recent model about the functional neuroanatomy of language [14] it is suggested that the verbal working-memory system is an important part of those brain-circuits participating in the processing of speech. The verbal working memory is considered to consist of an articulatory loop and a phonological store, and a separate mechanism is supposed to exist for interfacing between the two. This interface can be possibly localized in a specific are at the conjunction of temporal and parietal lobe, called the Sylvian-parietal-temporal (Spt) area.

Taken together the Hickok and Poeppel model and the results of Metz-Lutz showing the deficit in verbal working memory in LKS children, we believe that our data can be interpreted in the light of these as follows. We suggest, that the main deficit in the LKS patient is the dysfunction of area Spt which partly coincides with the source of epileptic activity. The defected functioning of area Spt causes the dysfunction of verbal working memory and this in turn leads to the language problems. It is obvious that the phoneme discrimination does not have to be affected by working memory problems, while the processing of suprasegmental cues might be. Evidently, we can not exclude the possibility that the deficit of the processing of prosodic cues is due to some other factors. At least we can say, that the processing of those cues is not performed by the same mechanisms as the processing of segmental cues (as this was left intact in our LKS patient). What is obviously needed is an extended model of speech perception which can incorporate the processing of prosodic cues, and their possible neuroanatomical basis.

5. Conclusions

To conclude, we found a dissociation in the processing of segmental and suprasegmental speech cues in a child with Landau-Kleffner Syndrome, using the method of ERPs. Several other techniques showed converging results in that the patient presented a selective impairment of the language system, and especially the verbal working memory seems to be defected. At the same time, a child with a similar childhood epileptic disorder, BECT, demonstrated spared processing for both kind of cues. The epileptic slow spike and wave activity present in both children could be localized in different brain areas, supporting that different brain areas affected in the two syndromes lead to the dysfunction of different aspects of language comprehension.

We believe that LKS is a language disorder that can be used to study what happens to the language system if in the course of development the essential neural circuits are severely disturbed. We believe also that the tools we use, that is the electrophysiological measurement of pre-attentive language processing is a fruitful approach since it makes possible the assessment of subtle processing skills while it does not require the patient / child to be responsive.

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