Long-term course of Landau-Kleffner syndrome: visuo-semantic and auditory aspects of comprehension

Evolução da síndrome de Landau-Kleffner: aspectos visuo-semânticos e auditivos da compreensão

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SUMMARY

Introduction. Landau-Kleffner Syndrome is characterized by normal speech acquisition followed by epileptic seizures, receptive and expressive language deterioration coupled with agnosia for non-verbal sounds, having variable long-term evolution. Case Report. It is described neurophysiologic and acoustic findings in a patient with Landau-Kleffner Syndrome, and correlate these with the results of a language evaluation carried out 7 years after the acute phase. It is performed Electroencephalography, Immitance Measurements, Basic Audiometry, Auditory Brainstem Response, Middle Latency Response, and P300. Language was evaluated by Boston Diagnostic Aphasia Examination. Electroencephalography was normal and audiological evaluation revealed normal Immitance Measurements, Basic Audiometry and Auditory Brainstem Response values. An electrode effect was present in the left hemisphere in Middle Latency Response, and bilateral P300 latencies delayed on the right. Language evaluation showed severe receptive and expressive impairment, severe phonemic substitutions, which had an impact on social and academic levels. There were contextual and gestual non-verbal compensations, evidencing intellectual and cognitive domain preservation. Conclusion. This case illustrates the specific cerebral areas that can be damaged in patients with Landau-Kleffner Syndrome and which are demonstrable by clinical evaluation and proper neurophysiology studies, showing the importance of neurological, audiological, electrophysiological and language exams in a longitudinal follow up. Keywords: Auditory evoked potentials. Electroencephalography. Hearing disorders. Landau-Kleffner Syndrome. Language. P300 event-related potentials.

RESUMO


INTRODUCTION

Landau-Kleffner Syndrome (LKS) is characterized by normal speech acquisition followed by epileptic seizures, receptive and expressive language deterioration coupled with agnosia for non-verbal sounds, having variable long-term evolution. Electroencephalography (EEG) abnormalities are seen in temporal areas, especially during sleep, where verbal language and acoustic information is processed.

The clinical picture fluctuates and presents differences over its several phases. Some improvements during development, and after the use of antiepileptic drugs and corticosteroid, are observed. Speech-language therapy, special schools and psychopedagogic management are also indicated. We describe the neuropsychiologic and acoustic findings of a patient with LKS, and correlate these with the results of language evaluation 7 years after the acute phase.

METHOD

A male right-handed patient presented epileptic seizures, speech difficulties and hyperactivity at the age of 3 years. EEG showed epileptiform discharges in the parietotemporal area. He received antiepileptic drugs and corticosteroids leading to control of seizures, but minimal speech improvement. The subject has been attending regular school since age 3, experiencing learning difficulties, and is currently in the first grade. Improvements in behavior and non-verbal communication in parallel with medical, speech-language and psycho-pedagogic treatment were observed. According to parent information, he reacts to and identifies environment sounds, and is learning Brazilian sign language.

We performed EEG, Immitance Measurements, Basic Audiometry and Auditory Evoked Responses such as brainstem, middle and long latency studies (Auditory Brainstem Response, Middle Latency Response, Cognitive Potential – P300) (Biologic equipment Traveler-Express). Language was evaluated by a number of stimuli from the Boston Diagnostic Aphasia Examination.

RESULTS

Behavioral, Electroacoustic and Electrophysiological Assessments

a) EEG: at onset of the disease, paroxystic activity in the parietotemporal areas; at the latest evaluation, 7 years after, normal in wakefulness and sleep.

b) Immitance measurements: type A tympanometric curve with presence of acoustic reflexes in both ears.

c) Tonal audiometry: normal hearing thresholds in both ears.

d) Vocal audiometry: not performed because of poor cooperation.

e) Auditory Brainstem Response (ABR): presence of waves I, III, V at 80 decibels hearing level for clicks, with absolute and interpeak I-III, III-V latencies within normal limits in both ears.

f) Middle Latency Response (MLR): absolute latency of the positive wave Pa within normal limits in both ears; significant difference between the amplitude of the wave Pa for the electrode located in C3 (condition C3/A2=2.13 microvolts) when compared with the electrode located in C4 (C4/A2=6.54 microvolts), revealing an electrode effect in the left hemisphere; significant difference between the amplitudes of the wave Pa for the electrode located in C4 ipsilateral (C4/A2=6.54 microvolts) and contralateral (C4/A1=0.86 microvolts), revealing a left ear effect (figure 1).

g) Cognitive Potential (P300): present in both ears with prolonged absolute latency (P300=410 milliseconds) in the right ear (figure 2).

Speech Evaluation

a) Conversation and spontaneous speech

The subject responded to non-verbal communication cues and reacted to gestures and expressions, although he did not understand propositions and did
not respond to verbal calling. When requested to participate in direct interlocution, he produced gestures and unintelligible verbal utterances, which were key words with intense phonetic-phonological alterations, and surprising preservation of the melodic contour, accent and rhythm of his native region.

b) Word comprehension
He correctly designated colors and numbers when prompted by acoustic stimuli. Body parts, objects and letters were not recognized, and other complex tests could not be applied.

c) Oral expression
The subject repeated isolated words with phonetic-phonological approximations and descriptions of objects (marron/brown = maku; cadeira/chair = kinká – gestures, kareira; que/that = te; and named: casa/house = gaga; banco/bench = kinká – sentir = to sit).

d) Written comprehension and expression
He recognized frequently occurring words (such as house, water, João Paulo, kite and water), wrote numerical series (1–11), his own name and simple words. In addition, the subject completed overtrained sentences with errors (a piscina é azul = azul) and wrote text segments with juxtaposed pronouns and substantives but without syntax, besides unintelligible segments.

DISCUSSION
D. evolved with a profile akin to the severe cases described in the literature, in which there was a recovery of the capacity to respond to non-verbal sounds of the environment. However, this ability was not enough to achieve the phonemic discrimination that presupposes quick acoustic change perception. The severe impairment in performing auditory-verbal pairing tasks supports our hypothesis that D. presented a disorder in this aspect.

The oral language disorders were severe, where language was limited to the emission of lexical fragments. In the few repetitions, D. tended to maintain the articulation zones (visual cues support), hindering other phoneme features. Generally, intonation contour preservation is rarely mentioned in the evolution of severe cases with LKS. In the context of damage, in the MLR and P300, traces of improved right hemisphere integrity can be observed where this could explain regional melodic accent preservation in D.’s emissions.

The written language disorder presented by D. has also been verified in cases where the syndrome occurred in the period before the acquisition of this modality.

Aphasia with intense reception obliteration retains characteristics of childhood manifestation. The lack of language prevented sophisticated analysis from verifying semantic system integrity. However, reactions to situations involving visual input (gestures and objects) allow us to infer its preservation. Dissociation in the ability to identify stimuli via visual and auditory input was noted: our patient demonstrated a greater facility for identifying colors and numbers, in contrast to identifying other items. Colors and numbers are coded by predominantly visual experiences where this probably took place prior to the disease. Letter coding is similar, although this occurred later, when D. already presented the hearing impairment.

Although it is recognized that the prognostic of childhood aphasia is favorable for majority of cases, Martins et al identified the type and location of the aphasia presented by our patient as factors that could have a negative influence on recovery.

In the electrophysiological exams, we were able to verify absence of brainstem auditory pathway damage in the ABR, a finding in agreement with that described by Msall et al, along with an alteration in late auditory evoked potentials. In the present study, alterations in the MLR were found, more specifically an electrode effect in the left hemisphere and a left ear effect, as well as in the cognitive potential, with a prolonged latency of the P300 wave in the right ear. According to Musiek et al, patients with central nervous system disorder present altered MLR in the same side of the lesion. This alteration can be observed by the electrode effect, due to the fact that this electrode is closer to the lesion. On the other hand, the same author affirms that the ear effect may be observed ipsi and contralaterally to the lesion. These results are compatible with the deficits detected in the language reception and emission tasks in our patient, which, according to the specialized lit-
The electroencephalogram presented alteration over a long period, normalizing after 7 years. The abnormalities observed were seen in the posterior region of the hemispheres (parietotemporal area), which agrees with the other findings.

The results of the electrophysiological and language evaluations are coherent, given that D. presents left hemisphere dysfunction – evidenced by results obtained in the MLR and P300 (left temporal lobe lesion, one of the areas responsible for language and auditory information processing) and also presents speech comprehension difficulties, as well as speech production alteration.

**CONCLUSION**

This case demonstrates the importance of neurological, audiological, electrophysiological, and language exams in a longitudinal follow up, in order to relate semiologic data and foresee prognostics, essential requirements to enable precise definitions to be made from a clinical standpoint.

The syndromic etiology, bilateral affection, extension and duration of the lesion, allied with the absence of special education opportunities indicate an unfavorable prognosis. Indeed, this may have limited the development of compensatory language abilities in this child. Functional compensation related to the anterior regions of the brain, such as context management, and those related to posterior regions, such as use of visual cues in phoneme imitation, were also evidenced.

**REFERENCES**