IOSR Journal of Dental and Medical Sciences (IOSR-JDMS)
e-ISSN: 2279-0853, p-ISSN: 2279-0861.Volume 23, Issue 5 Ser. 4 (May. 2024), PP 07-09
www.iosrjournals.org

Sleep Eeg Is Useful In The Diagnosis Of Landau-Kleffner Syndrome

Abdellaoui Walid[1], Louanchi Malika[2], Ait-Oukaci Wassila[3], Toubal Nadia[2], Sadibelouiz Mustapha[4], Ait-Kaci-Ahmed Mahmoud[4]

[1]*neurology Department, Mostaganem Hospital, University Of Mostaganem*
[2]*neurology Department, Ibn Sina Hospital, University Of Annaba*
[3]*neurophysiology Department, Ait Idir Hospital*
[4]*neurology Department, Ait Idir Hospital, University Of Algiers*

Abstract:
Background*: Landau-Kleffner syndrome (LKS), also known as acquired aphasia with epilepsy, is a rare age-related epileptic encephalopathy characterized by developmental regression in the area of language and electroencephalogram (EEG) abnormalities located mainly around the temporoparietal areas. When present, seizures occur more frequently during sleep. Behavioral disorders can be part of the clinical picture.*
Clinical case report: *Landau-Kleffner Syndrome LKS is a rare childhood epileptic syndrome characterized by acquired aphasia, epileptic abnormalities and epileptic seizures. In our case report on two children with LKS we compared waking EEG and sleep EEG, and which one is better in the diagnosis of LKS.*
Conclusion: *Sleep EEG is better then waking EEG in the diagnosis of Landau-Kleffner Syndrome LKS.*
Key Words*: Landau Kleffner syndrome, Acquired aphasia with epilepsy, Sleep EEG*

Date of Submission: 01-05-2024 Date of Acceptance: 10-05-2024

I. Introduction

The syndrome usually occurs in children aged 3 to 8 years with previously normal development. Aphasia, EEG alterations, seizures and behavioral disturbances are the main features of the syndrome.

Regarding aphasia, the typical presentation of the syndrome is characterized by a child who, despite apparently normal neurological development, begins to have difficulty understanding written and spoken language. Subsequently, over the days, weeks or months, the young patient manifests difficulties of expression accompanied by an alteration of spontaneous language which, in turn, becomes limited and altered.

You should know that aphasia initially manifests itself as auditory verbal aphasia which precedes expressive aphasia characterized by difficulties in processing or interpreting verbal and/or non-verbal sounds. Children may also express babbling, neologisms, verbal perseverance or even mutism.

Epileptic seizures occur in approximately two thirds of patients. Epileptic seizures are usually partial motor seizures (more common), generalized clonic seizures, and atypical absence seizures (e.g., blinking, chewing gestures, as well as lip smacking or slight jerking lip movements).

Linguistic regression is often associated with social cognitive deficits and behavioral disorders, such as attention deficit, hyperactivity, impulsivity, and distractibility. Emotional lability, anxiety and depression, sleep disorders, working memory disorders (but not long-term memory) and hypersensitivity to sound can complete this sometimes particularly complex clinical picture. [1] [2].

EEG examination during wakefulness and sleep is fundamental for diagnosis. The EEG shows more pronounced unilateral or bilateral activity in the posterior temporal regions around the sylvian fissure. This EEG activity becomes much more widespread and intense during sleep (non-REM sleep) when it is characterized by an almost continuous spike-wave pattern with a frequency of 1.5 to 2.5 spikes/s. Background EEG activity and macro sleep architecture are normal. However, during REM sleep, epileptic activity can partially interrupt, decrease or cease.

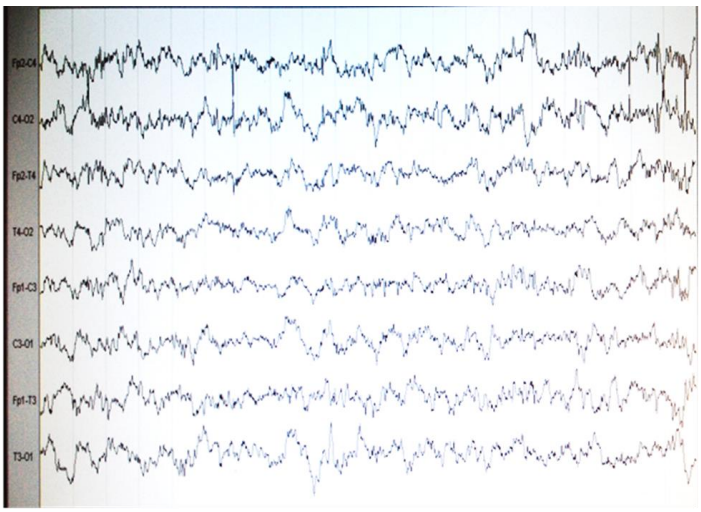
It should be noted that there is no international consensus on the most appropriate therapy. Treatment should begin as early as possible [3].

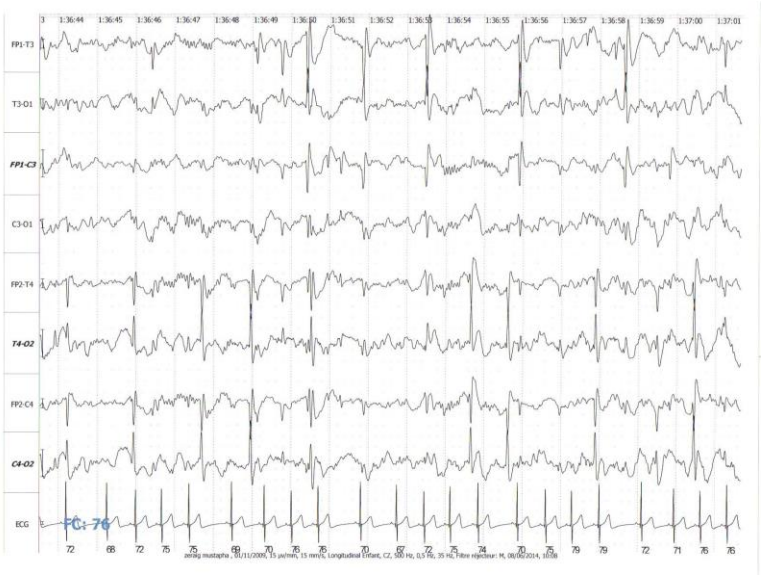
Corticosteroids (oral prednisone 1 mg/kg/day for 6 months or oral prednisolone 2 mg/kg/day for at least three months) could be useful for improving or at least stabilizing linguistic and cognitive skills. and behavioral. . [4] Their use, also in combination with benzodiazepines (BDZ), is recommended when epileptic activity and language disorders persist despite treatment. [5].

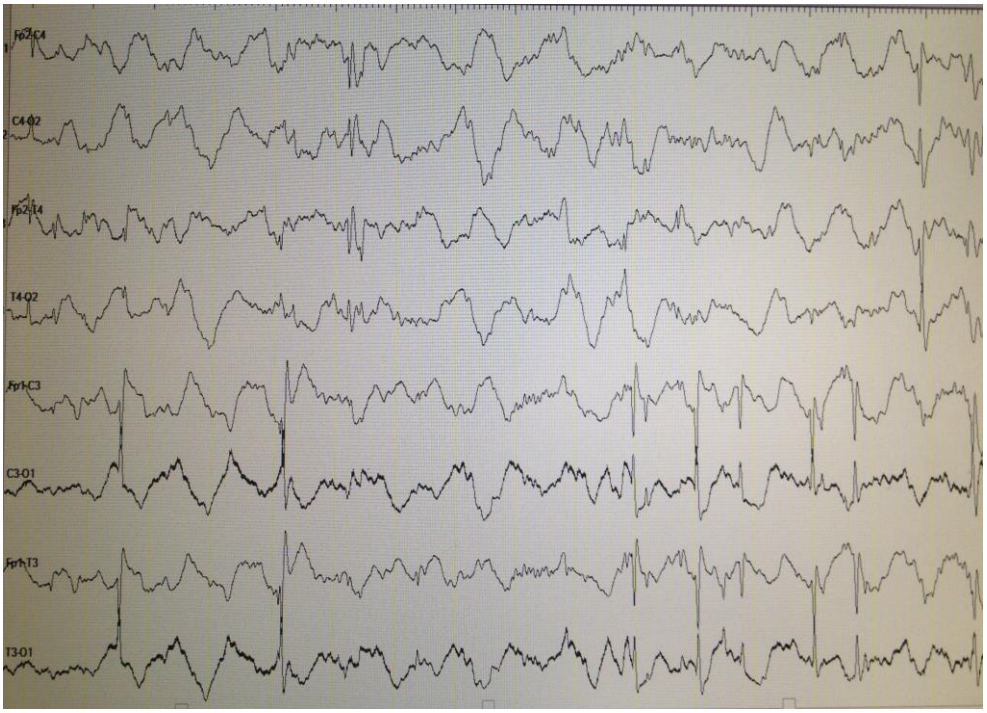
Multiple subpial transections (MSTs) in the posterior temporal lobe of the dominant hemisphere are a surgical approach that attempts to interrupt epilepsy while preserving the eloquent cortex. This involves the sectioning of the horizontal corticocortical fibers while sparing the vertical cortico-subcortical fibers. Grote et al. showed that MST could induce the restoration of linguistic abilities in children with Landau-Kleffner syndrome [6]. The use of this invasive technique should be limited to severe and drug-resistant cases.

II. Clinical Case

We are reporting the clinical case of two boys aged 6 and 8 respectively. They had normal pregnancy through a physiological delivery. The family history was negative and the psychomotor development was normal.since age five, they answering only in mono-syllables and they had a comprehension of some commonly used expressions. An aphasia occured 1 year after language disorder.In the space of just a few months later there was a severe drop in verbal comprehension. They presented idiopathic focal epilepsy with partial motor seizures since age five.Neuroimaging (CT scan, MRI) did not show any structural lesions. The first neurophysiological (EEG during wakefulness) evaluation were performed after epilepsy onset.Waking EEG showed dipahasic spike and spike-and-wave predominating over centro-temporal regions. The diagnosis of benign epilepsy with centro-temporal spike BECTS was established. Therapy with valproic acid VPA 20 mg/kg/day was begun and the clinical picture was characterized by disappearance of the seizures with no improvement in the language deficits. The second neurophysiological (sleep EEG) evaluation were performed one year after the first recording (waking EEG) and revealed generalized continuous repetitive high voltage spike and spike-and-wave discharges, activated in slow wave sleep, exceeding 50% of sleep cycles. On the basis of sleep EEG patterns the diagnosis of Landau -Kleffner Syndrome LKS was confirmed.Improvement in the language aspects with clonazepam CZ 0,1 mg/kg/day and lamotrigine 2.5 mg/kg/day.







III. Discussion

Both patients presented to our consultation for language delay and epilepsy. On the basis of sleep EEG patterns the diagnosis of Landau-Kleffner Syndrome LKS was confirmed. LKS is often misdiagnosed as autism, pervasive developmental disorder, hearing impairment, dyslexia, auditory/verbal processing disorder, speech deficit disorder attention, intellectual disability, childhood schizophrenia, or emotional/behavioral problems in early stages of presentation [7].

Ghosh et al. Reported the case of a 9-year-old boy with speech loss and seizures for two years [8]. The boy was developing normally before the seizures began. Examination of higher mental functions revealed excessive hyperactivity, self-destructive behavior, and reduced attention span. Audiological evaluation showed normal hearing thresholds bilaterally and he began to communicate through signs. EEG showed repetitive spikes and epileptiform activity in bilateral parieto-occipital regions. Clinical features and EEG led to a diagnosis of LKS [8].

Motwani et al. reported an 11-year-old boy who presented with fever followed by seizures [9]. The boy developed aphasia after his illness. No history of neonatal distress and he had normal growth and development of language, hearing and vision. His neurological examination was normal. CSF study and MRI were normal. However, the EEG showed abnormal exacerbation of spikes and waves during sleep, and the boy was diagnosed as a case of LKS and treated with sodium valproate, levetiracetam, and corticosteroids [9].

Raybarman reported the case of a 5-year-old boy who developed aphasia, attention deficit disorder, and hyperkinesia [10]. EEG showed generalized epileptiform activity. He was diagnosed with LKS. Computed tomography (CT) and MRI of the brain were normal. He improved on antiepileptic drugs and ACTH [10] .

Although the importance of a high SWI during sleep is agreed upon (Morrell et al, 1995[11].; Roulet Perez, 1995[12].; Rossi et al., 1999[13].; Eriksson et al., 2003[14].; Smith & Hoeppner, 2003[15].; Holmes & Lenck-Santini, 2006[16].), relatively little attention is paid to the SWI during the awake stage. Some authors claim that epileptic activity itself hampers cognitive function (Seri et al., 1998[17].; Gordon, 2000[18].; Pearl et al, 2001[19].; Holmes & Lenck-Santini, 2006[16]; Praline et al, 2006[20].; Tassinari and Rubboli, 2006[21].). Several authors developed grading scales to quantify the amount of epileptiform activity during sleep: Mikati et al (2002) [22]. used a gradation of 0–4 [no spike-waves (SW); 0–25% SW; 25–50% SW; 50–75% SW; 75–100% SW, respectively]. Beaumanoir (1995) [23]. and Beaumanoir et al. (1995) [24]. distinguished four categories: CSWS** (>85%), CSWS* (50–80%), CSWS (<50%) and CSWS°(no epileptiform activity.Aeby et al (2005) [25]. developed a grading system of 4 items containing epileptiform and non epileptiform parameters.

IV. Conclusion

Sleep EEG is better then waking EEG in the diagnosis of Landau-Kleffner Syndrome LKS specially after appearance of language deficits.