OPEN ACCESS

Saudi Journal of Medicine

Abbreviated Key Title: Saudi J Med
ISSN 2518-3389 (Print) |ISSN 2518-3397 (Online)
Scholars Middle East Publishers, Dubai, United Arab Emirates
Journal homepage: https://saudijournals.com

Case Report

Post-Epileptic Aphasia Revealing LANDAU- KLEFNER Syndrome in a Child

Najoua Belhaj[1*], Razika Bencheikh[2], Lina EL Messouadi[1], Ahmed Ould mohammed[1], Mhammed Azedine[1], Ngouya Koumba Hernandez Vasthi[1], Mohammed Anass Benbouzid[2], Leila Essakalli Houssyni[2]

[1]Resident Physician in Otorhinolaryngology, Department of Otorhinolaryngology, Head and Neck Surgery, Ibn Sina University Hospital, Faculty of Medicine, Mohammed V University, Rabat, Morocco
[2]Professor of Otorhinolaryngology, Department of Otorhinolaryngology, Head and Neck Surgery, Ibn Sina University Hospital, Faculty of Medicine, Mohammed V University, Rabat, Morocco

DOI: 10.36348/sjm.2021.v06i06.002 | **Received:** 22.03.2021 | **Accepted:** 29.04.2021 | **Published:** 06.06.2021

***Corresponding Author:** Belhaj Najoua

Abstract

Aphasia in children is generally still little explored because it is rare. Landau-Kleffner syndrome (LKS) is rare and causes specific difficulties in children with it. Among the neuropsychological difficulties (cognitive and attentional), we find a significant initially receptive aphasia. We report in this work the observation of a 13-year-old child seen in consultation for aphasia at the age of 07 years post epileptic seizure. In this patient the diagnosis of LKS was retained after the etiological assessment and the complementary examinations carried out.

Keywords: Landau-Kleffner syndrome (LKS), KLEFNER Syndrome, aphasia, neuropsychological difficulties.

INTRODUCTION

Aphasia in children is generally still little explored because it is rare; Children with stroke or various brain injuries will often have a range of motor and neuropsychological disorders, but aphasia, which can be temporary, will most often be recovered quickly thanks to brain plasticity. Many means of re-education, compensation and rehabilitation can then be put in place to help the children. In children in the process of building their language, aphasia presents itself as an obstacle in its development. We can find infantile aphasia in Landau-Kleffner syndrome, described by Landau and Kleffner in 1957. We report in this work the observation of a 13-year-old child seen in consultation for aphasia at the age of 07 years. post epileptic seizure.

CASE REPORT

We report the case of a 13-year-old child, resulting from a continued pregnancy, brought to term.

The child is not from a consanguineous marriage. The patient presented at the age of 7 years an epileptic seizure of unknown origin; after the epileptic seizure the parents observed post-epileptic aphasia.

The patient was followed in the pediatric neuro department, the sodium valproate has been prescribed, the etiological assessment was completed by an electroencephalogram,that showed no abnormalities.

The brain magnetic resonance imaging and the brain scan were also normal. The ENT clinical examination of the child was normal, we completed the assessment with a recording of the auditory evoked potentials (Figure-3), the audiogram and the temporal bone scan were normal (Figure 1 & 2), the patient was taken care of by speech therapists .

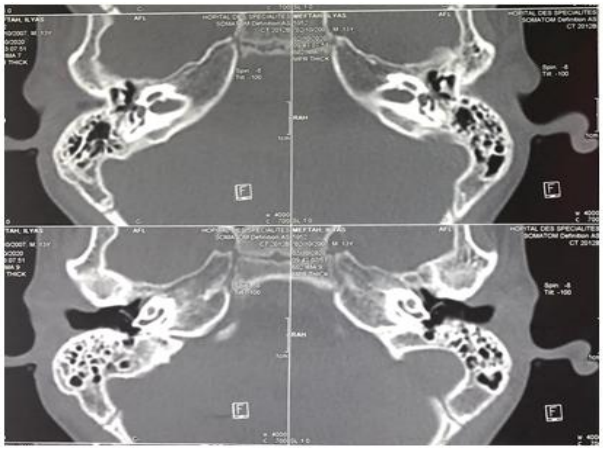

Fig-1: Axial & coranal temporal bone CT cut

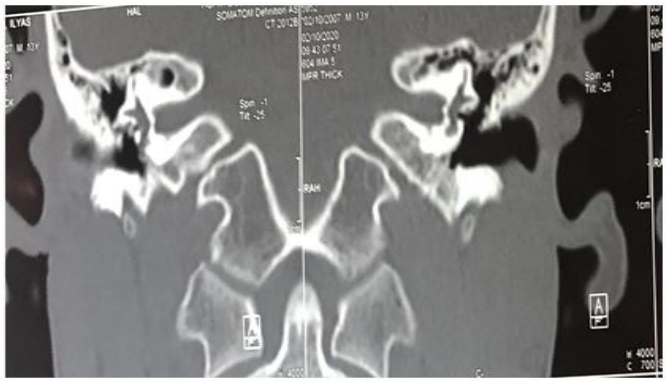

Fig-2: Axial & coranal temporal bone CT cut

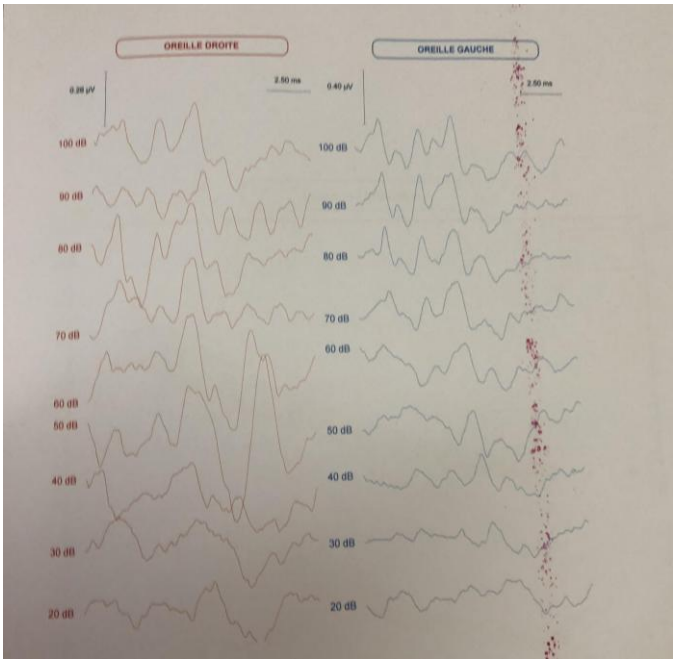

Fig-3: The auditory evoked potentials: at 20 db the 5 waves are presents in both ears

DISCUSSION

Childhood aphasia is defined by Seron [1-3] as a "language disorder resulting from objective damage to the CNS (central nervous system) and occurring in a child who has normally acquired a certain level of comprehension and oral expression". The etiologies are varied. We can thus find, in children, cases of aphasia by lesions of vascular origin, aphasia following bacterial or viral infections, tumor aphasia, traumatic aphasia, the frequency of which is increasing. higher, and epileptic aphasia in children [1-3].

For the first time in 1957, Landau and Kleffner presented the case of five children with a syndrome linking epilepsy and aphasia, mostly receptive. These children had suffered a loss of previously acquired language skills, both in expression and in comprehension. Following extensive examinations, the only abnormality found was in the tracing of electroencephalograms (EEG) in the temporal areas. According to them, the manifestations of this "convulsive disorder" were generalized, partial, myoclonic or "petit mal" convulsions, that is to say absences. Short, sometimes severe, and sudden onset paroxysmal abnormalities were found on the EEG.

Landau-Kleffner syndrome (LKS) is also called "acquired aphasia with epilepsy or AAE" acquired epileptiform aphasia "," epileptic aphasia ". There is a consensus on the two main symptoms of SLK: on the one hand, acquired aphasia, with loss of expressive and especially receptive language skills, in a normally developing child; and on the other hand, epileptiform seizures. Language deficit is directly caused by epileptic discharges in critical areas of language. Rather, LKS is said to be a "situation", not a "syndrome", in which epilepsy produces a prolonged or permanent disturbance of neural circuits responsible for higher cortical functions. Language deficit is directly caused by epileptic discharges in critical areas of language. Rather, LKS is a "situation", not a "syndrome", in which epilepsy produces a prolonged or permanent disturbance of neural circuits responsible for higher cortical functions, beyond aphasia alone [3-5].

LKS affects children with normal development. Landau-Kleffner syndrome is a predominantly male condition with an M / F sex ratio of 2 and occurs mostly at an age between 5 and 7 years, the extremes being 2 and 12 years. Epileptic seizures are the mode of onset of the condition in 60% of cases, as is the case with our patient. Their semiology and frequency vary from child to child and within the same child. Seizures usually respond well to antiepileptic therapy and often go away before the age of 14. They are particularly sensitive to sodium valproate, ethosuximide and benzodiazepines. Phenytoin, carbamazepine and phenobarbital are ineffective and may even have a deleterious effect on seizures and thus worsen the prognosis. Language disturbances are constant during Landau-Kleffner syndrome. They are as much about comprehension as they are about expression. One of the hallmarks of Landau-Kleffner syndrome is hearing impairment, although not constant [6-9].

LKS does not cause structural damage, but a form of functional ablation "of normal language behavior. Most often, other functions including intelligence and behavior are not affected. The cases reported in the literature are very varied. Aphasia can last from two weeks to several years. The onset of LKS can be sudden or gradual and in 90% of children occurs as receptive aphasia, followed by rapid and severe reduction in speech. Auditory-verbal agnosia can be severe in normal hearing. Even familiar noises are not recognized or distinguished, leading to severe impairment of oral language, or even its abolition [1, 2]. Consequently, one finds in the children reached an expressive deficit with articulation, phonological, lexical, morphosyntactic disorders, even a jargon or a total mutism. Behavioral disturbances are seen in Landau-Kleffner syndrome at a frequency of 75% and are most often moderate in intensity. Usually it is hyperkinesia, outbursts of aggressive behavior or oppositional behavior.

LKS is indeed one of the epileptic syndromes. Over 75% of children with SLK experience simple partial, infrequent, often nocturnal seizures, especially when the syndrome develops. Despite the severe EEG abnormalities, 20-30% of children with LKS do not have "clinical" seizures but only spike waves in the temporal lobe. These "subclinical" seizures are characterized by unilateral or bilateral focal point waves, mainly located in the temporal and posterior temporal regions, during slow sleep [3-5].

The etiology of LKS is unknown but the genetic hypothesis is found in the literature, without direct evidence. The main goal of treatment is to influence aphasia, as "classic" seizures are easily controlled with anticonvulsant therapy. Benign epilepsy can be treated, but several drugs tested have no effect or even aggravate it. The authors mention the effectiveness of corticosteroids, allowing a significant improvement in language, with transient, non-dangerous, relatively rare side effects (weight gain, behavioral disturbance, hypertension). The benefits of this treatment appear to be long-lasting and outweigh the side effects. However, it should be noted that poor adherence to treatment is a frequent cause of non-control of symptoms. Unlike epileptic seizures which are controlled by treatment, speech and behavior disorders have a poor prognosis [10]. The latter give rise to permanent sequelae if paroxysmal brain activity has lasted for more than two years [10]. Although there are usually no morphological abnormalities on brain imaging.

Surgical therapy (multiple subpay transection) has also proven successful. A surgical technique first performed by Murrell in 1995 consists of a section of cortical fibers responsible for the connection of epileptogenic activity allowing regression of seizures and aphasia [11]. This technique was used in 1999 by Grotte et al., who noted definitive clinical improvement in 11 of the 14 patients included in their series. Likewise, Irwin et al. reported 5 cases of Landau-Kleffner syndrome with severe speech and behavioral disturbances that improved with this surgical technique [12]. Recently, the effectiveness of vagus nerve stimulation has been demonstrated on epileptic seizures. drug-resistant [13]. Speech therapy is essential because it is necessary to provide an effective communication system in order to improve behavioral difficulties. The use of Sign Language and / or integration into a school for the deaf are possible and will undoubtedly promote the emergence of oral language [6, 14].

CONCLUSION

Landau-Kleffner syndrome (LKS) is rare and causes specific difficulties in children with it. Among the neuropsychological difficulties (cognitive and attentional), we find a significant initially receptive aphasia. It is important to introduce speech therapy quickly in order to help restore their language and keep the connection they have with the outside world, thanks to communication.

REFERENCES

1. Deonna, T. W. (1991) Acquired epileptiform aphasia in children (Landau-Kleffner Syndrome). Journal of Clinical Neurophysiology, 8(3), 288–298.
2. Deonna, T., & Roulet-Perez, E. (2010). Early-onset acquired epileptic aphasia (Landau–Kleffner syndrome, LKS) and regressive autistic disorders with epileptic EEG abnormalities: The continuing debate. Brain and Development, 32(9), 746-752.
3. Deonna, T., Prelaz- Girod, A. C., Mayor- Dubois, C., & Roulet- Perez, E. (2009). Sign language in Landau- Kleffner syndrome. Epilepsia, 50, 77-82.
4. Akinci, Ö., Işık, U., & Melek, İ. (2012). Landau kleffner syndrome, electrical status epilepticus in

sleep and autistic regression: an overview of literature / Landau kleffner sendromu, uykuda elektriksel status epileptikus ve otistik regresyon: Literatüre bir bakış. Dusenne Adam : The Journal of Psychiatrie and Neurologique Sciences, 157–169.

5. Delille, R., & Prudent, A. (2015). Effet d'un Entrainement De La Comprehension Lexicale Grace Aux Signes Et Aux Pictogrammes Dans Le Cadre d'un Syndrome De Landaukleffner Etude De Cas d'un Enfant Agé De 6 Ans Directeur de Mémoire Santaella Elisabeth.
6. Dugas, M., Masson, M., Le Heuzey, M. F., & Regnier, N. (1982). Aphasie acquise de l'enfant avec épilepsie (Syndrome deLandau-Kleffner) 12 observations. Rev Neurol, 132(10):755-80.
7. Beaumanoir, A., Bureau, M., Deonna, T. M., & Tassinari, C. A. (1995). Continous spikes and waves during slow sleep. London, John Libbey, 213-216.
8. Roger, J., & Dravet, C. (1984). Les syndromes épileptiques del'enfant et de l'adolescent. Eurotext Neurol, 185-95.
9. Zivi, A., Broussaud, G., Daymas, S., Hazard, J., & Sicard, C. (1990). Syndrome aphasie acquise-épilepsie avec psychose. A propos d'une observation. In Annales de pédiatrie (Paris) (Vol. 37, No. 6, pp. 391-394).
10. Smith, M. C., & Hoeppner, T. J. (2003). Epileptic encephalopathy of late childhood: Landau-Kleffner syndrome and the syndrome of continuous spikes and waves during slow-wave sleep. Journal of Clinical Neurophysiology, 20(6), 462-472.
11. Morrell, F., Whisler, W. W., Smith, M. C., Hoeppner, T. J., Toledo-Morrell, L. D., Pierre-Louis, S. J., ... & Hasegawa, H. (1995). Landau—Kleffner syndrome: treatment with subpial intracortical transection. Brain, 118(6), 1529-1546.
12. Irwin, K., Lees, J., Polkey, C., Alarcon, G., Binnie, C., Smedley, M., ... & Robinson, R. O. (2001). Multiple subpial transection in Landau- Kleffner syndrome. Developmental Medicine & Child Neurology, 43(4), 248-252.
13. Yong, D. P. (2003). The effects of vagus nerve stimulationtherapy on patients with intreatable seizures and either Landau-Kleffner syndrome or autism. Epilepsy & Behaviour, 4:286-290.
14. Maamar, M., El Alaoui-Faris, M., Ettahri, L., Benblaid, F., Medjel, A., & Chkili, T. (2006). Syndrome De Landau-Kleffner. Maroc Médical, 28(1).