Asian Journal of Research and Reports in Neurology

2(1): 67-70, 2019; Article no.AJORRIN.55936

Landau–Kleffner Syndrome (Acquired Epileptic Aphasia) about a 13 Year Old Child: A Case Report

Ouajid El. Bakkali[1*], Sanae El. Moussaoui[1] and Yamna Kriouile[1]

[1]Department of Neuropediatrics, Neuro-Metabolic Diseases Unit, Pediatrics II, Children's Hospital, Rabat, Morocco.

Authors' contributions

This work was carried out in collaboration among all authors. Authors OEB and SEM designed the study, performed the statistical analysis, wrote the protocol and wrote the first draft of the manuscript. Author OEB managed the literature searches. All authors managed the analyses of the study, read and approved the final manuscript.

Article Information

Editor(s):
(1) Dr. Maria Emilia Cosenza Andraus, Universidade Federal do Rio de Janeiro, Brazil.
Reviewers:
(1) Steve Carlan, USA.
(2) Piradee Suwanpakdee, Phramongkutklao Hospital and College of Medicine, Thailand.
Complete Peer review History: http://www.sdiarticle4.com/review-history/55936

Case Study

Received 25 January 2020
Accepted 31 March 2020
Published 06 April 2020

ABSTRACT

Landau-Kleffner syndrome or acquired epileptic aphasia is an acquired infantile epileptic syndrome. It was identified by Landau and Kleffner in 1957, where six affected children were listed. It is characterized by an acute loss of speech and language and an overall regression of behavior in a child who had good psychomotor development with language normal. The rarity of this syndrome makes it demanding to diagnose. Multidisciplinary treatment will be required by pediatricians, neuropediatricians, child psychiatrists, psychologists, speech-language pathologists, otolaryngologists and radiologists. The final diagnosis is based on an altered EEG during sleep with a normal brain MRI, as well as clinical manifestations syndrome. we report the case of a 13-year-old child, with good psychomotor and cognitive development, from 1st degree inbred parents, there was no family history of childhood epileptic disorder, having presented six months ago 15 minute tonic-clonic straight hemicrises and a dysgraphia was installed gradually, against a background of apyrexia. These symptoms progressed six months later, towards a very significant regression of the oral language with difficulties of articulation and a total regression written. The sensory and motor examination was normal. The cranial nerves were intact. The walk was normal. The systemic examination was without abnormality. The studies on cerebrospinal fluid (CSF) were normal.

**Corresponding author: E-mail: doctor.bakkali@gmail.com;*

Magnetic resonance imaging (MRI) of the brain with contrast is normal. EEG objectified short, predominantly centro-temporal biphasic short bursts and during sleep, the presences of a central theta focus. The patient was put on sodium valproate, Levetiracetam, Clobazam, intravenous immunoglobulin and corticosteroid and speech therapy.

Keywords: Landau-kleffner syndrome; acquired epileptic; aphasia.

1. INTRODUCTION

Landau and Kleffner described this syndrome in children, initially called aphasia syndrome acquired with a convulsive disorder in 1957. This syndrome is characterized by a progressive regression of language accompanied by convulsions in children with normal psychomotor development, but it there is no causal relationship [1]. It interests children between 3 and 7 years of age, the male/female ratio being 2:1 [2]. Patients initially present language problems (deafness of words or auditory verbal agnosia (AVA)), with an inability to understand speech. A convulsive seizure occurs in 70% to 85% of affected patients around the age of 4 and 10 years and generally stops after 15 years. An abnormal peak and wave electroencephalogram (EEG) pattern during sleep often precedes language improvement. Behavioral changes may be observed to be due to language impairment. There is no standard treatment for LKS. The means of treatment include anticonvulsant drugs, steroids, adrenocorticotropic hormone (ACTH), the ketogenic diet, immunoglobulins and even surgery. Early use of corticosteroids or ACTH can relieve symptoms and normalize the EEG in LKS [3]. The EEG is a basic component for diagnosis, which will object to abnormal epileptic discharges in children, while clinical crises are observed in approximately 70% of cases. The EEG presents generalized, multifocal or changing transient and undulatory discharges, mainly in the temporal region, sometimes unilateral and particularly exacerbated during sleep [4].

2. CLINICAL CASE

We report the case of a 13-year-old child, with good psychomotor and cognitive development, of 1st degree inbred parents, there was no family history of childhood epileptic disorder, his sister and brother are in good health, no notion of taking tonixes or trauma (physical or psychological), he presented six months ago 15 minutes tonic-clonic straight hemicrises and a dysgraphia settled gradually, against a background of apyrexia. These symptoms progressed six months later, towards a very significant regression of the oral language with difficulties of articulation and a total written regression. On the general physical examination, the GCS score is 15/15. The child measured 154 cm (M) in height and 37 Kg (M) in weight and has a head circumference of 51 cm, without signs of dysmorphia. The sensory and motor examination was normal. The cranial nerves were intact. Cerebellar signs and nystagmus were absent. The walk was normal. The systemic examination was without abnormality. Her lab tests, including full blood count, kidney function tests, and liver function tests were normal. Studies on cerebrospinal fluid (CSF) were normal (cytological, biochemical tests, immune-electrophoresis of proteins in CSF, anti NMDAr antibodies, anti VGKC and lactate). Amino acid chromatography, acylcarnitine profile, lactaemia and ammonia were up to standard. MRI of the brain with contrast is normal. The GET objectified short short bursts, mainly centro-temporal biphasic, and during sleep, the presence of a central theta focuses. The patient was put on sodium valproate, levetiracetam, clobazam, intravenous immunoglobulins and corticosteroids and speech therapy with good clinical and EEG improvement.

3. DISCUSSION

LKS, acquired epileptic aphasia, childhood aphasia or aphasia with convulsive disorder [5,6], is characterized by the triad: epileptic seizures in a healthy child (generally male), aphasia receptive and expressive, EEG abnormalities [7]. It generally appears between 2 and 13 years old [5]. A predominance in male children is noted, with a male / female ratio of 2:1 [2].

The neurophysiological mechanism of stagnation and involution of acquired speech in affected patients is not yet very clear. Landau and Kleffner [7] had suggested that it could be secondary to a functional ablation of the primary cortical language by persistent rejections in these regions, while Gascon et al. [8] mentioned the possibility that EEG discharges are cortical manifestations of a low-level subcortical

differentiation process. Sometimes it can be secondary to low-grade brain tumors, head trauma, demyelinating disease or neuro-cysticercosis [8].

The overall aphasia of this child may be linked to a constant peak EEG activity in the left temporo-occipital region and no other secondary cause was found in our case.

Bilateral clonic tonic can be observed in 70% to 85% of patients, behavioral disorders in 78% of cases, and hyperactivity and a decrease in the duration of concentration are observed in 80% of patients. [8].

EEG abnormalities are constant (clinical or subclinical). EEG abnormalities consist of a discharge of spikes, patterns of spikes or acute waves of high synchronous amplitude either in foci of variable location, usually temporal or temporo-occipital, or in both hemispheres. Sleep activates anomalies, which may be absent in wakefulness recordings [9]. Brain imaging (CT or MRI) is generally without abnormality, and positron emission tomography can show a united or bilateral hypo- or hyper-metabolism [10].

Treatment aims to reduce the frequency of seizures and improve language and cognitive function. It is based on the use of standard antiepileptics such as valproate and lamotrigine [11]. If the crisis and aphasia persist, the use of steroids may be considered: oral prednisolone 2 mg/kg/day for 2 months, followed by a gradual reduction to 0.5 mg/kg both days. If corticosteroids are beneficial, this dosage can be maintained for 6 to 8 months [12]. Intravenous immunoglobulins can be used [13]. Speech therapy will be prescribed for several years because the improvement only occurs over a prolonged period. Sub-cortical transection may be indicated in clinically refractory cases [14]. The prognosis depends mainly on the age of onset of aphasia. Early onset indicates that the chances of complete recovery are low. The objective of this article is to educate pediatricians about this syndrome because early management can present a relatively better prognosis.

4. CONCLUSION

LKS is rare and you have to think about it in front of the triad: seizures, aphasia and EEG abnormalities without forgetting the other differential diagnoses. Multidisciplinary treatment will be considered early for a better prognosis.

CONSENT AND ETHICAL APPROVAL

As per university standard guideline, participant consent and ethical approval have been collected and preserved by the authors.

COMPETING INTERESTS

Authors have declared that no competing interests exist.

REFERENCES

1. Deonna TW. Acquired epileptiform aphasia in children (Landau-Kleffner syndrome). J Clin Neurophysiol. 1991;8:288-298.
2. Pearl PL, Carrazana EJ, Holmes GL. The landau-kleffner syndrome. Epilepsy Curr. 2001;1:39- 45.
3. Ramanathan RS, Ahluwalia T, Sharma A. Landau-Kleffner syndrome-a rare experience. East J Med. 2012;17:36-39.
4. Nemati R, Nabipour I, Javadi H, Chabi N, Assadi M. Regional cerebral blood-flow with 99mTc- ECD brain perfusion SPECT in Landau-Kleffner syndrome: Report of two cases. Case Rep Radiol. 2014;1-5.
5. Raybarman C. Landau-Kleffner syndrome: A case report. Neurol India. 2002;50: 212-3.
6. Bishop DV. Age of onset and outcome in 'acquired aphasia with convulsive disorder' (Landau-Kleffner syndrome). Dev Med Child Neurol. 1985;27:705-12.
7. Landau WM, Kleffner FR. Syndrome of acquired aphasia with convulsive disorder in children. Neurology. 1957;7:523-30.
8. Gascon G, Victor D, Lombroso CT. Language disorders, convulsive disorder and electroencephalographic abnormalities. Acquired syndrome in children. Arch Neurol. 1973;28:156-625.
9. Tuchman RF. Acquired epileptiform aphasia. Semin Pediatr Neurol. 1997;4:93-101.
10. Maquet P, Hirsch E, Dive D, Salmon E, Marescaux C, Franck G, et al. Cerebral glucose utilization during sleep in Landau-Kleffner syndrome: A PET study. Epilepsia 1990;31:778-83.
11. Duran MH, Guimarães CA, Medeiros LL, Guerreiro MM. Landau-Kleffner syndrome: Long-term follow-up. Brain Dev. 2009;31: 58-63.
12. Beumonoir A. The Landau Kleffner syndrome. In: Roger J, Beureau M, Drovet CH, Dreifuss FE, Perret A, Wolf P, editors.

Epileptic syndrome in infancy, childhood and adolescence. 2nd ed. London, England: John Libbey Eurotext. 1992;231-43.

13. Arts WF, Aarsen FK, Scheltens-de Boer M, Catsman-Berrevoets CE. Landau-Kleffner syndrome and CSWS syndrome: Treatment with Intravenous Immunoglobulins. Epilepsia. 2009;50 (Suppl 7):55-8.
14. Morrell F, Whisler WW, Smith MC, Hoeppner TJ, de Toledo-Morrell L, Pierre-Louis SJ, et al. Landau-Kleffner syndrome. Treatment with Subpial Intracortical Transection. Brain. 1995;118(Pt 6):1529-46.

Peer-review history:
The peer review history for this paper can be accessed here:
http://www.sdiarticle4.com/review-history/55936