International Journal of Scientific Reports
Nwala CG et al. Int J Sci Rep. 2024 Apr;10(4):135-138
http://www.sci-rep.com
pISSN 2454-2156 | eISSN 2454-2164
DOI: https://dx.doi.org/10.18203/issn.2454-2156.IntJSciRep20240717

Case Report

Atypical electroclinical presentation of Landau Kleffner syndrome: a case report of an 8 year old Nigerian child

Chuks G. Nwala[1]*, Omokhuale M. Ejemierele[2]

[1]Department of Paediatric Neurology, Limi Children's Hospital Wuse 2, Abuja, Nigeria
[2]Department of Paediatrics, Limi Children's Hospital Wuse 2, Abuja, Nigeria

Received: 26 January 2024
Revised: 14 February 2024
Accepted: 05 March 2024

***Correspondence:**
Dr. Chuks G. Nwala,
E-mail: g.nwala@limihospital.org

ABSTRACT

Epileptic seizures account for common neurologic presentations in paediatric neurology units in developing countries. Atypical presentation of syndromic epilepsy may be missed especially sensory, psychic or autonomic manifestations. An 8 year old male Nigerian child presented with recurrent vomiting and delayed motor milestones, regression of speech (acquired aphasia) with unremarkable birth history or any underlying medical or surgical condition. He had delayed gross motor development and continuous bilateral polyspike waves of slow sleep on EEG. Self-limited autonomic epileptic seizure syndrome which was a strong differential of autonomic seizures in early childhood was not suggestive as his EEG findings failed to demonstrate occipital spike wave pattern nor did photic stimulation evoke any epileptiform waves. Child made significant clinical improvement with anticonvulsants, neuro vitamins, and behavioural therapy, evidenced by regain of non-verbal communication, resolution of autonomic symptoms and temper tantrums. A high index of suspicion should be applied in patients with acquired aphasia with subclinical or recurrent autonomic symptoms.

Keywords: Atypical, Landau Kleffner, Syndrome, Child, Autonomic, Nigerian

INTRODUCTION

Epileptic seizures account for most neurologic presentations to the emergency paediatric units of developing countries like Nigeria.[1] Whereas convulsive seizures may be identified without much difficulty, sensory, psychic and autonomic variants may elude clinicians with low index of suspicion as the semiology appears like other common clinical conditions.[2] Landau Kleffner syndrome (LKS) was first described in the year 1957 as a condition characterized by acquired epileptic aphasia with nocturnal EEG abnormalities. Reduction in language function and attention deficit are common accompaniment. Recovery is often delayed despite anticonvulsant therapy and communication problems are persistent. Patients present with recurrent convulsive episodes of seizures with regression of speech milestone which are not regained even following treatment.[3] Several researchers have reported cases of recurrent seizures associated with acquired auditory verbal agnosia.[4-8]

However, there is a dearth of documentations on acquired aphasia associated with autonomic seizures among the paediatric population. Autonomic seizures with occipital spikes, panayiotopoulos syndrome, now known as self-limited epilepsy with autonomic seizures is a rare epileptic syndrome associated with occipital spikes seen in young children but rarely associated with loss of acquired communication skills (acquired aphasia).[9] Children with this type of seizures will stop having symptoms within 1

to 2 years of age. Autonomic seizures with associated acquired aphasia are a rarely documented epileptic encephalopathy whose occurrence needs to be reported.

Therefore, this aims to report a case of a child with recurrent autonomic symptoms, delayed development and regression of belatedly acquired expressive verbal communication milestone, whose electroencephalogram revealed continuous polyspikes of sleep in keeping with syndromic epilepsy earlier described by Landau Kleffner.

CASE REPORT

LAA is an 8 year old male child with complaints of recurrent episodic vomiting, loss of sphincter tones and regression of speech milestones, noticed 4 months prior to presentation. Vomiting was non projectile, non-bilious, consisting of recently ingested meals. Each episode was associated with urinary and faecal incontinence and postictal sleep lasting for 2 hours. Vomiting occurred initially at monthly intervals for 2 months, then fortnightly for 6 weeks and later once weekly prior to presentation. Each bout of vomiting lasted for a few minutes and resolved spontaneously.

Child was also noticed to be laughing incongruously most times when engaged in any communication (both verbal and otherwise) and could neither comprehend nor follow parental instructions.

He had visited several primary and secondary care facilities with no medical or surgical etiopathogenic correlation with his presenting symptoms. The absence of any clear underlying causation necessitated a referral to our facility for further specialist expert care.

His birth history was unremarkable; however, gross motor development was delayed and earlier belatedly acquired speech milestone had regressed with recurrent episodic vomiting. Other aspects of his history were not contributory.

His physical findings revealed a well-nourished child, unwilling to sit still in the consulting room and not following instructions. He had mild palpebral conjunctival pallor, global hypotonia and aphasia. Other findings were essentially normal.

Investigations

Complete blood count, serum electrolytes, calcium and magnesium showed normal parameters. However, EEG revealed bilateral continuous polyspike waves of slow sleep (CSWSS).

Treatment

He received haematinics, calcium supplements, risperidone and AED (Valproic acid). He has been on follow up with the child neurology unit with good seizure control, better attention span, now sits calmly and follows instructions (regained non-verbal speech milestone). However, he is yet to regain lost expressive speech milestone.

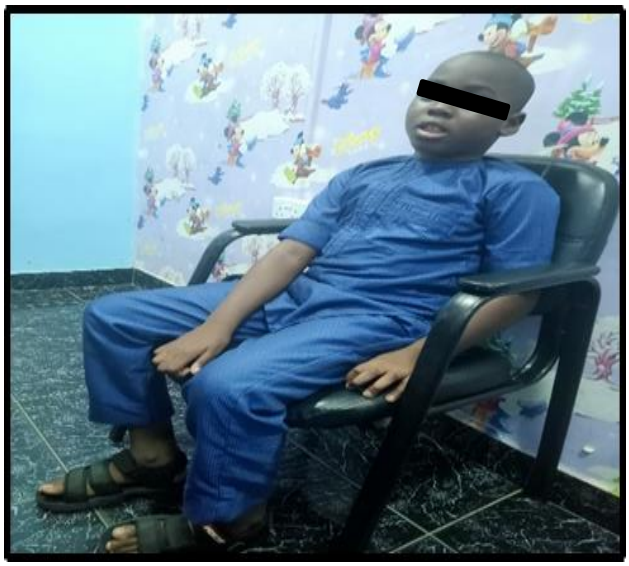

Figure 1: Clinical photograph of patient at follow up.

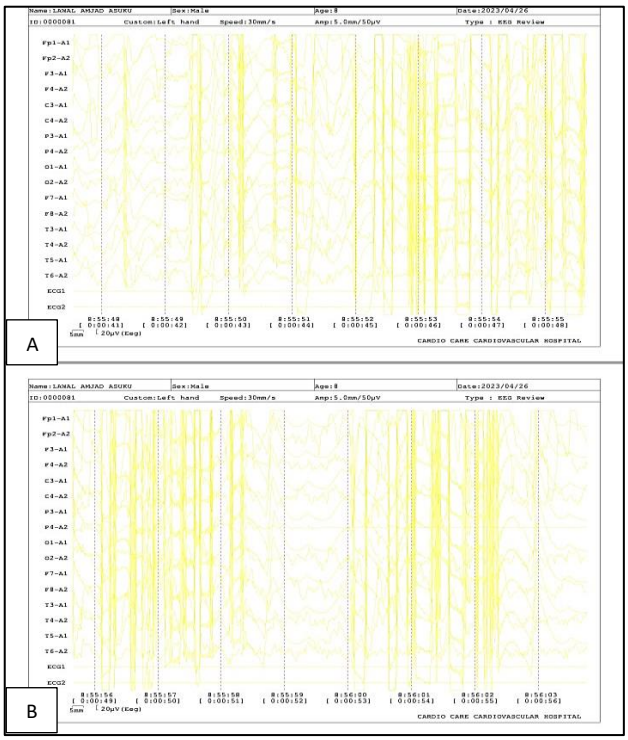

Figure 2 (A and B): EEG of child showing continuous bilateral polyspike waves of sleep.

DISCUSSION

According to developmental epilepsy clinic and family information group report of 2016, LKS occurs in 1 in a million children, mostly male children between 3 and 9 years of age.[10]

Most documented seizures among these individuals are convulsive and mostly occur during sleep. However, before the convulsive seizures, subclinical seizures may go unnoticed during the active phase of the disorder leading to delayed or regression of already acquired developmental milestones.[11-13]

Acquired aphasia in childhood present with recurrent seizures and is a known cause of epileptic seizures globally. Although there is little coordinated data and isolated reports of syndromic epileptic seizures documented in different resource constrained settings.[14-16] Aina et al documented overall poor quality of life ascribed to language difficulties.[17] Other authors on the other hand, have reported varying degrees of limitation of daily activities due to recurrent seizures and the negative impact of the condition to overall quality of life of patients.[18-20]

The common clinical features in children with this condition include recurrent seizures and regression of verbal communication skills. The index patient may have had silent/subclinical seizures which may have contributed to the gross motor delays noticed during his early development and subsequently, loss of communicative skills noticed 4 months prior to presentation. In addition, the autonomic features he had like vomiting, inattention and irrational behaviour, are rare accompaniment of LKS by most documented series to the best of authors' knowledge.[21] The uncommon autonomic symptomatology eluded the earlier facilities caregivers sought care, until the clinical suspicion of LKS was entertained.

The lost milestones may be regained, depending on how urgent interventional measures are applied.[22]

Children with LKS may recover other lost milestones but not expressive speech deficit. This could be as a result of delayed identification of recurrent seizures and consequently, the delay in getting the required therapy for proper control.[23]

Patients that receive treatment early during the active phase of illness, may regain all lost milestones while others that had delayed presentation and interventions like the index case, may not regain all the lost milestones, especially verbal communication skills as reported.[24]

The index patient presented with extreme temper tantrums that led to some level of frustration in his caregivers as the child could hardly communicate his concerns to parents who got overwhelmed by such an experience. Head banging and inability to follow parents' instructions noticed in index child have been reported by the national organization of rare diseases in 2018.[25]

Recovery of auditory verbal agnosia (language comprehension) and aphasia (verbal expression) varies from person to person.[26] In this case, the child was able to comprehend parental communication after four months of treatment, had regained nonverbal communication skills like following parental instructions. He was then able to follow conversations and sang with some clear words but was still unable to use these words to effectively communicate with caregivers. Although parents were still expecting full regain of his verbal expression, the degree of recovery was significant.

In 2022, the international league against epilepsy (ILAE) recommended that Landau-Kleffner syndrome be considered a subtype of developmental/epileptic encephalopathy with spike wave activation on sleep (DEE-SWAS). These conditions are a spectrum of epilepsy syndromes in children characterized by a specific abnormal EEG pattern and variable degrees of cognitive regression. A child with a diagnosis of LKS may transition to another DEE-SWAS syndrome over time. The EEG recording in index demonstrated polyspike waves during sleep with a constellation of clinical symptoms in keeping with an acquired aphasia on background recurrent autonomic seizures. Although most seizures in LKS are convulsive, reports had suggested that seizures may even be subclinical prior to the overt motor manifestations identified at presentation.[27] This rarely reported but evident autonomic symptoms in a child with continuous polyspike waves of slow sleep obtained was strongly suggestive of LKS.

In summary, clinicians' index of suspicion for LKS should be heightened in children with gross developmental delays and speech regression associated with recurrent seizures. Although most children with this neurologic disorder do not usually regain lost milestones, timely and focused intervention hold some promise in ensuring good quality of life and allaying parental anxiety.

Limitation

A single case report may not be truly reflective of all neurologic presentations of LKSs in childhood.

CONCLUSION

A child with recurrent seizures with acquired loss of verbal communication associated with demonstrable electroencephalogram of continuous spikes of sleep should be considered for syndromic epilepsy like LKS even if the seizures are non-convulsive.

Funding: No funding sources
Conflict of interest: None declared
Ethical approval: Not required

REFERENCES

1. Iloeje SO, Ojinnaka NC. Epilepsy and Epileptic Seizures: Paediatrics and Child Health in a Tropical Region. 3rd Ed, Educational Printing and Publishing. 2016.

2. Epileptic Encephalopathies in infancy and early childhood in which the epileptiform abnormalities may contribute to progressive dysfunction. In Panayiotopolous CP. The Epileptic seizures, syndromes and management. Oxfordshire (UK) Blade Medical Publishing. 2005.
3. Robinson F. Landau Kleffner Syndrome current issues. Neurology. 2003;19:53-56.
4. Kliegman RM, Staton BF, St Gene KW, Schor NE. Nelson Textbook of Paediatrics, 19th ed. 2011;119.
5. Aina OF, Morakinyo O. Landau-Kleffner syndrome (acquired epileptic aphasia) in a Nigerian child. Niger Postgrad Med J. 2002;9(1):29-31.
6. López-Ibor MI, López-Ibor JJ, Hernández Herreros M. Landau-Kleffner syndrome (acquired aphasia with epilepsy). Etiopathology and response to treatment with anticonvulsants. Actas Luso Esp Neurol Psiquiatr Cienc Afines. 1997;25(6):410-6.
7. Guevara-Campos J, González-de Guevara L. Landau-Kleffner syndrome: an analysis of 10 cases in Venezuela. Rev Neurol. 2007;44(11):652-6.
8. Singh MB, Kalita J, Misra UK. Landau Kleffner syndrome: electroclinical and etiopathogenic heterogeneity. Neurol India. 2002;50(4):417-23.
9. Covanis A. Panayiotopolous Syndrome: a benign childhood autonomic epilepsy frequently imitating encephalitis, syncope, migraine, sleep disorder, or gastroenteritis. Pediatrics. 2006;118(4):e1237-43.
10. Landau-Kleffner Syndrome. Available at: https://www.ninds.nih.gov/health-information/disorders/landau-kleffner-syndrome#:~:text=What%20is%20Landau%2DKleffner%20syndrome,express%20language)%20and%20repeated%20seizures. Accessed on 05 January, 2024.
11. Landau Kleffner syndrome. Available at: https://www.gosh.nhs.uk/conditions-and-treatments/conditions-we-treat/landau-kleffner-syndrome/#:~:text=In%20subclinical%20seizure%20activity%2C%20there,subclinical%20and%20occur%20during%20sleep. Accessed on 05 January, 2024.
12. Trevathan E. Seizures and epilepsy among children with language regression and autistic spectrum disorders. J Child Neurol. 2004;19(1):S49-57.
13. Overvliet GM, Besseling RM, Vles JS, Hofman PA, Backes WH, Van Hall MH, et al. Nocturnal epileptiform EEG discharges, nocturnal epileptic seizures, and language impairments in children: review of the literature. Epilepsy Behav. 2010;19(4):550-8.
14. Ahmad MM, Ahmed H. Childhood Epileptic Encephalopathy in Sokoto, Northwestern Nigeria. J Pediatr Neurol Neurosci. 2021;5(2):164-8.
15. Landau-Kleffner Syndrome. Available at: https://www.hopkinsmedicine.org/health/conditions-and-diseases/landaukleffner-syndrome. Accessed on 05 January, 2024.
16. Tuft M, Årva M, Marit B, John AW, Karl ON. Landau Kleffner Syndrome: Clinical Review. Tidsskr Nor Legeforen. 2015;135(22):2061-4.
17. Deonna T, Roulet-Perez E. Early-onset acquired epileptic aphasia (Landau-Kleffner syndrome, LKS) and regressive autistic disorders with epileptic EEG abnormalities: the continuing debate. Brain Dev 2010;32:746-52.
18. Landau-Kleffner Syndrome. Available at: https://my.clevelandclinic.org/health/diseases/22699-landau-kleffner-syndrome-lks. Accessed on 05 January, 2024.
19. Motwani N, Afsar S, Dixit SD, Sharma N. Landau-Kleffner syndrome: an uncommon dealt with case in Southeast Asia. BMJ Case Rep. 2015;2015:bcr2015212333.
20. Aina OF, Morakinyo O. Landau-Kleffner syndrome (acquired epileptic aphasia) in a Nigerian child. Niger Postgrad Med J. 2002;9(1):29-31.
21. Smith MC, Hoeppner TJ. Epileptic encephalopathy of late childhood: Landau-Kleffner syndrome and the syndrome of continuous spikes and waves during slow-wave sleep. J Clin Neurophysiol. 2003;20:462-72.
22. Cockerell I, Bølling G, Nakken KO. Landau-Kleffner syndrome in Norway: long-term prognosis and experiences with the health services and educational systems. Epilepsy Behav. 2011;21:153-9.
23. Landau WM, Kleffner FR. Syndrome of acquired aphasia with convulsive disorder in children. Neurology. 1957;7(8):523-30.
24. Caraballo RH, Cejas N, Chamorro N, Kaltenmeier MC, Fortini S, Soprano AM. Landau-Kleffner syndrome: a study of 29 patients. Seizure. 2014;23(2):98-104.
25. Wang S, Zhang YH, Bao XH, Wu Y, Jiang YW, Liu XY, et al. Clinical characteristics and long-term prognosis of Landau-Kleffner syndrome. Zhonghua Er Ke Za Zhi. 2006;44(2):105-9.
26. Hughes JR. A review of the relationships between Landau-Kleffner syndrome, electrical status epilepticus during sleep, and continuous spike-waves during sleep. Epilepsy Behav. 2011;20(2):247-53.
27. Deonna T, Prelaz-Girod AC, Mayor-Dubois C. Sign language in Landau-Kleffner syndrome. Epilepsia. 2009;50(7):77-82.
28. Baird G, Robinson RO, Boyd S, Charman T. Sleep electroencephalograms in young children with autism with and without regression. Dev Med Child Neurol. 2006;48(7):604-8.
29. Hrdlicka M. EEG abnormalities, epilepsy and regression in autism: a review. Neuro Endocrinol Lett. 2008;29(4):405-9.

Cite this article as: Nwala CG, Ejemierele OM. Atypical electroclinical presentation of Landau Kleffner syndrome: a case report of an 8 year old Nigerian child. Int J Sci Rep 2024;10(4):135-8.