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Case Report

A Recurrent Self Resolving Aphasia: Case Report Of Improved Landau-Kleffner Syndrome

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ABSTRACT

Landau-Kleffner syndrome (LKS) should be considered in the differential diagnosis in a child with acquired aphasia. Early recognition and intervention might help in prevention of further complications, such as speech impairment, cognitive impairment and seizure.
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Keywords: Landau-Kleffner syndrome; Aphasia; speech impairment; •EEG

CASE PRESENTATION

A seven-year-old boy, who was previously healthy, was brought to the emergency department with two days history of inability to speak. During the event, the patient was awake, could follow simple commands, knew family faces, and was able to play with a phone and videogames. For over one year, he had recurrent, self-resolving aphasia occurrences that last for eight to ten days, with one-month intervals, and no clinical evidence of seizures. His events usually present as expressive aphasia with intact comprehension. He had never experienced seizures in his life or any other neurological/behavioral symptoms. There were no family history of similar illness, seizures or developmental regression.

The patient was born at term, by normal vaginal delivery, with a normal developmental history. On admission, the vital signs and growth parameters were normal. The patient was alert and oriented with no facial dysmorphism. His neurological assessment was unremarkable, except for his speech abnormality in the form of expressive aphasia. His comprehension, reading, and writing abilities were normal. His vision and hearing assessments were normal.

The patient was admitted to our hospital and basic tests, including complete blood count, serum electrolytes, bone profile, blood glucose, cerebrospinal fluid study, and metabolic work up (ammonia, lactate, plasma amino acids, urine organic acids, very long-chain fatty acids, and tandem MS) were performed and all results were normal. A chromosomal study revealed normal male karyotype, and the next generation sequencing result was negative. Magnetic resonance imaging of the brain revealed no abnormalities. The admission Electroencephalography (EEG) (Figure 1) was recorded during the event and showed evidence of sleep activated continuous spike and wave activity was seen predominantly over the bilateral centrotemporal head region. The EEG findings fall within the spectrum of LKS. During hospitalization, the patient was put on valproic acid and showed significant improvement within two to three days of initiation and EEG has been normalized after one month.

Our patient has been on follow-up for about two years and has shown complete resolution of clinical aphasia. All repeated EEG results have been normal since he started on valproic acid. During follow-up, the patient has been shifted to levetiracetam, then to clobazam due to side effects, and was on medication for a total of eighteen months. The patient has been off medication for more than six months now (stopped by family), and he continues to be clinically and electrographically stable.

DISCUSSION

Landau-Kleffner syndrome (LKS) is a rare condition in a childhood age, which usually presents between ages of three to seven years (Swaiman et al., 2017). This syndrome is characterized by loss of language abilities in children who were previously normal (Swaiman et al., 2017). Some other symptoms include acquired verbal auditory agnosia, multifocal spikes, and spike–wave discharges localized mainly over the centrotemporal regions, continuously or subcontinuously during sleep, attention and behavioral problems are also involved (Swaiman et al., 2017). Patient with LKS shows normal MRI and CT scan but abnormalities in EEG is detected. In typical LKS, the defect of the language function is usually a verbal–auditory agnosia with an inability to understand language (Gordon, 1990). Receptive dysfunction seems to dominate the early stage of this disease while expressive deficits develop later (Beaumanoir, 1992). It has been reported that less than 10 percent of LKS-associated aphasia is initially predominantly expressive (Beaumanoir, 1992). Our case was expressive aphasia as the initial manifestation.

The majority (70%) of children with LKS can also present with various types of seizure (Swaiman et al., 2017). However, in our case the patient only had aphasia. Similar to our case, a study done on 29 patients found that only six patients were presented with aphasia but without seizures (Caraballo et al., 2014). Thus, such cases should also be considered at the early stage in the differential diagnosis for LKS. The EEG in the awakened state often has epileptiform abnormalities in central, parietal, and/or temporal head regions with normal background (Swaiman et al., 2017). During sleep, there is activation and a pattern, such as electrical status epilepticus in sleep (ESES) can be seen (Swaiman et al., 2017). Our patient had EEG findings which fall within the spectrum of LKS.

ESES is likely that continuous epileptiform discharges during sleep, for the most part in the peri-sylvian region, disrupt networks involved in normal language processing (Swaiman et al., 2017). It is likely that LKS is on the severe end of the spectrum, which also includes benign childhood epilepsy with centrotemporal spikes (BECTS) and continuous spike waves during sleep (CSWS) (Swaiman et al., 2017). These EEG abnormalities must be differentiated from epilepsy with BECT and from CSWS. In CSWS, abnormal awake records are more common, and seizures are more severe than in LKS (Bureau, 1995). In BECT, the EEG pattern is spikes in the centrotemporal head regions, which are high voltage, but blunted that can be unilateral or bilateral and become more frequent during drowsiness and non-rapid eye movement (REM) sleep (Swaiman et al., 2017) and usually no EEG activation during sleep such as ESES.

Treatment of this condition is challenging because of its fluctuating course, lag in improvement relative to the EEG, and response to seizures, but not in aphasia (Appleton et al., 1993; Glauser et al., 1995). Seizures usually respond to treatment very well, and the treatment of choice is valproic acid with or without adjunctive

benzodiazepine. Corticosteroids or adrenocorticotropic hormones (ACTH) have been used in an attempt to normalize the EEG and prevent long-standing language deficits (Swaiman et al., 2017). In some patients, intravenous immunoglobulin has been found to be useful (Mikati & Saab, 2000). Multiple subpial resections is the surgery of choice for those who fail to respond to medical treatment (Mikati & Saab, 2000).

The long-term prognosis is uncertain, although improvements in language ability seem to be related to the age of onset (pre-language or post-language development), with early onset resulting in a poorer prognosis (Swaiman et al., 2017). The older the child is at LKS onset, the better the prognosis for salvaging language skills (Bishop, 1985). Our patient was seven-year-old when he had the first event. He was treated with Valproic acid has shown complete resolution of aphasia with no recurrence of events or cognitive decline, and with normalized repeated EEG results over the next two years of follow-up. A similar case was reported by the author Motwani et al., where a 11-year-old patient was presented with convulsions and developed aphasia subsequently. He also showed similar diagnosis profile with abnormal EEG and thus LKS was diagnosed. He also showed improvement after treatment with drugs (sodium valproate, levetiracetam and steroids) (Motwani, 2015). In another case, a 3.5-year-old patient with no verbal agnosia and EEG with multiple spike and wave discharges and was diagnosed with LKS. She also showed improvement in her speech after treatment with valproic acid, ACTH, intravenous immunogobulin therapy followed by clobazam therapy (Tütüncüoğlu et al, 2002).

Furthermore, in a previous review of 45 reported cases followed to at least age 12 years, the author reported that to a certain degree prognosis correlated with age of onset. The patients affected before the age of five had a poorer language recovery outcome than those who affected later (Bishop, 1985). However, other studies did not confirm a relationship between the long-term prognosis and the age of onset (Bishop, 1985). Most of the cases of LKS do have residual speech abnormalities, although in some conditions the aphasia has responded fairly well to drug treatment (Raybarman, 2002). Consistent with this observation language skills of the patient were recovered, and EEG abnormalities normalized in our case over a period of two years. In conclusion, our study and previous studies showed the importance of early diagnosis of LKS and significance of its proper management for better outcomes. Furthermore, our case also presents a unique presentation of self-resolving acquired aphasia and thus adds to the literature about the recurring and resolving type of LKS cases".

POINTS TO LEARN

- LKS should be considered in the differential diagnosis in a child with acquired aphasia.
- Early recognition and intervention might help in prevention of further complications, such as speech impairment, cognitive impairment and seizure.
- EEG is diagnostic in cases of LKS.
- LKS is one of the few conditions that abnormal EEG needs to be treated.
- Follow-up is essential to assess the response to treatment.

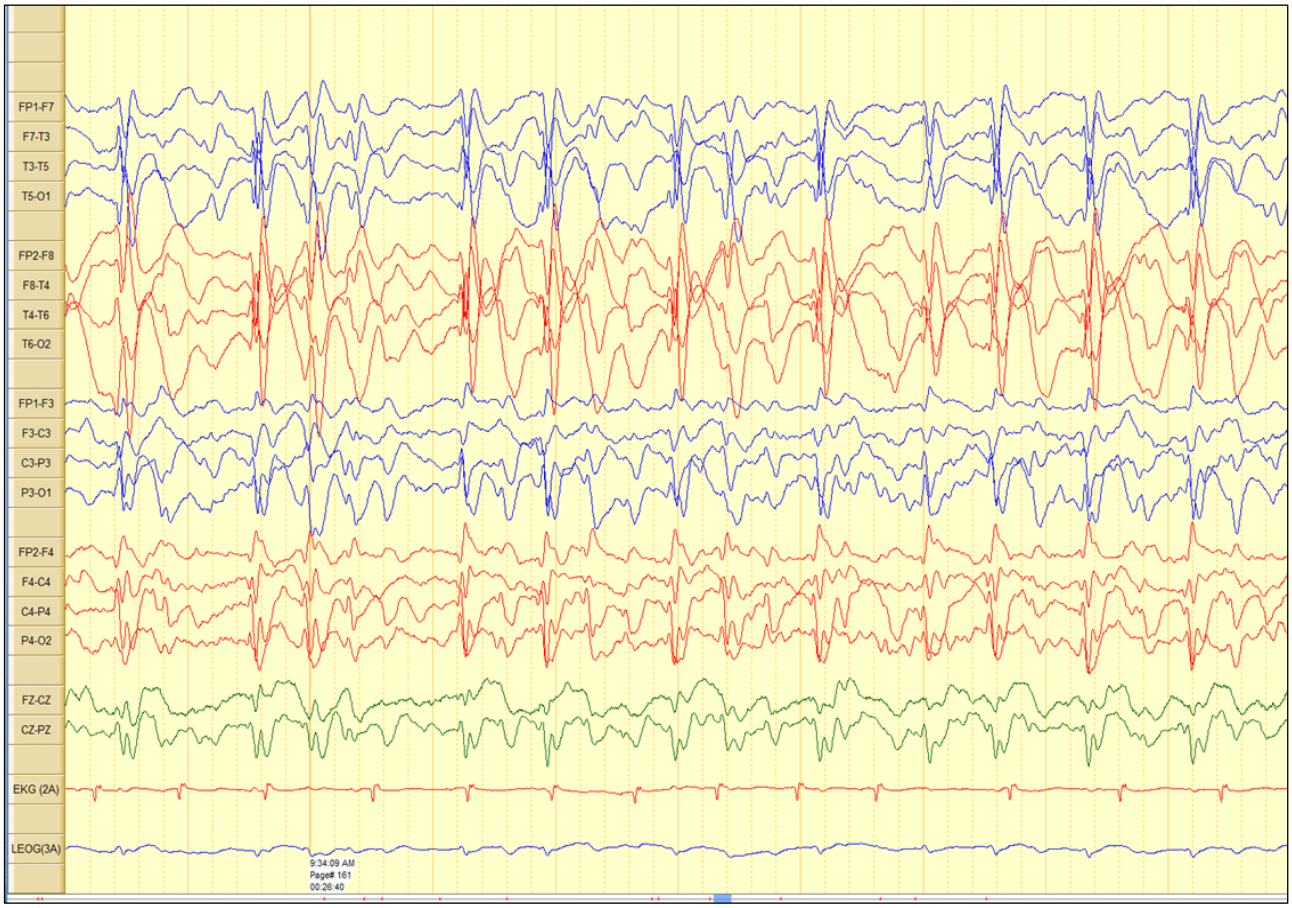

Figure 1: The EEG of patient showing evidence of sleep activated continuous spike and wave activity confirming the diagnosis of LKS.

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ETHICAL CONDIDERATIONS

Ethical approval was waived since publishing patients' information for research purposes should be approved by the patient upon admission as a part of the King Abdullah Specialist Children at King Abdulaziz Medical City policy. All information was kept confidential and was not accessed except for scientific research purposes.

CONFLICTS OF INTEREST

There is no funding agency / research support / conflict of interest for this study.

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