Case Report

Landau Kleffner Syndrome: a case report

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Introduction

Landau–Kleffner syndrome (LKS), also known as infantile-acquired aphasia or acquired epileptic aphasia. It is a comparatively rare syndrome that occurs exclusively in children. The prevalence of Landau-Kleffner syndrome ranges from 44.2 to 59.6 among a population of 18 million children[(1)]. This syndrome is named after Landau and Kleffner, who described it in 1957. It occurs in previously healthy children with normal language skills and development[(2)]. Most cases present between 3-7 years of age, with male preponderance (male to female ratio being 2:1). This condition initially presents with language problems presenting as word deafness or auditory verbal agnosia (AVA), in which the affected individual is unable to comprehend speech[(3)]. The pathognomonic feature of Landau- Kleffner syndrome is acquired aphasia, whereas epileptic seizures are not a prerequisite for LKS. Other clinical manifestations include cognitive impairments and behavioral problems. These impairments may be global or focal, based on the location of the epileptic discharges, and progressive, presumably related to increasing epileptiform activity[(4)].

Here we report a case of a 5-year-old child who presented with sudden speech regression and attention deficits over the past month.

Case history

A 5-year-old male child presented to the outpatient department with gradual speech regression for the last one month. Initially, the parents noted that the boy had started responding only in a few words. Over one week, he had stopped responding to them altogether and was not following simple commands. However, he did respond to sign languages. The patient had usual language and developmental milestones as per his chronological age before the chief complaints.

On examination, the response to the name was inconsistent. The child had no dysmorphic features on general physical examination, and we did not notice any neurocutaneous markers. We observed that his anthropometric measurements were within normal limits; cranial nerves were intact, and we did not see drooling on inspection. Response to verbal commands was absent. His neurological examination and systemic examination findings were unremarkable. The patient has not been noted any behavioral problems, and a formal developmental assessment was not done. On his first examination by a neurologist, an EEG was done. His earlier electroencephalogram (EEG) in the awake state reported intermittent sharp spikes and waves from the left hemisphere with secondary generalization. No additional abnormality was brought about by hyperventilation and photic stimulation. Due to the sudden onset of auditory agnosia, we suspected epileptic encephalopathy as an underlying cause and advised a repeat electroencephalogram (EEG) under sedation. The EEG monitoring showed bilateral parietal- temporal (left>right), left central, and generalized epileptiform abnormalities with marked activation in sleep. We even noted continuous spikes and waves while the child was sleeping (CSWS). We then advised an MRI to rule out any structural abnormalities. MRI findings were reported normal. The patient had both auditory verbal agnosia and expressive

speech problems. The hearing assessment was done using Brainstem evoked response audiometry (BERA).

Based on the EEG findings, the patient was diagnosed with Landau Kleffner Syndrome.

The patient was commenced on oral valproic acid. The signs and symptoms continued Oral prednisolone 2 mg/kg was prescribed for two weeks, followed by 1.5mg/kg for the following weeks, and then 0.5 mg/kg was continued for six months. Parents were informed about the prognosis of the treatment. We tried tapering the dose gradually, but to clinical deterioration, the dose was increased intermittently. The patient was on oral steroid treatment for 18 months. Frequent follow-ups have been advised to look for other neurological signs and to monitor the adverse reactions of the drugs. Parental consent was taken before the case reporting.

Informed written consent was obtained from the patient.

Discussion

LKS is an acquired epileptic aphasia disorder, with the first common symptom among patients being the loss of speech[5]. The aphasia sets in slowly and progressively, with spontaneous improvements and aggravations persist in between. Verbal auditory agnosia is a common feature, as seen in our case. Hence the initial diagnosis is hearing loss in most of the patients[1]. Though the exact etiology isn't clear, there is a possibility that genetic factors may be involved. Mutations in the GRIN2A gene(16p13.2) have been reported. This gene encodes for a protein called GluN2A (also known as NR2A), a subunit of the N-Methyl-D-aspartate (NMDA) glutamate-gated ion channel receptor. N2RA is seen in high brain areas crucial for speech and language, and NMDA receptors are related to memory and learning. Fewer studies have shown the possible involvement of autoantibodies against brain-derived neurotrophic factors (BDNF)[6].

Despite the severe electroencephalographic abnormalities, seizures never occur in 20– 30% of LKS patients, as seen in our case. The types of seizures reported in the LKS can differ, including partial complex, partial clonic, generalized tonic-clonic, and atonic seizures[3]. Electroencephalography (EEG) usually consists of various patterns, including continuous slow spike-wave discharges during sleep (85% or more of the deep sleep recording), sharp focal waves with spikes, and centrotemporal spike[7].

The LKS is often misdiagnosed as autism, pervasive developmental disorder, hearing impairment, dyslexia, auditory/verbal processing disorder, attention deficit disorder, intellectual disability, childhood schizophrenia, or emotional/behavioral problems in the initial presentation stages[2]. Treatment of LKS can be challenging. Standard antiepileptic drugs such as valproate and lamotrigine can sometimes reduce seizure frequency and improve language and cognitive function. Persistence of the seizure and aphasia necessitates the trial of steroids. Early initiation of adrenocorticotropic hormone (ACTH) and corticosteroids show encouraging results. Still, seizure and aphasia may relapse when they are tapered, as seen in our case. Multiple subpial resections may be the treatment of choice in those with resistance to medical therapy[8].

Starting speech therapy and continuation of treatment for many years is obligatory because improvement in language function requires long period.

Thus early diagnosis and treatment of patients are essential, especially those under three years of age who may present with speech and behavioral problems and may be misdiagnosed with Pervasive Developmental disorders or Disruptive behavioral disorders. This syndrome need not present with clinically apparent seizures, as seen in our case. Since these patients may consult an otorhinolaryngologist for auditory and speech assessment, increased awareness becomes essential.

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Conflict of interest: Nil

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