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A UNIQUE CASE OF LKS WITH REVIEW OF LITERATURE

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ABSTRACT

Landau-Kleffner syndrome (LKS) is a childhood disorder. A major feature of LKS is gradual or sudden loss of ability to understand and use spoken language. All children with LKS have abnormal electrical brain waves that can be documented by an electroencephalogram (EEG), a recording of the electric activity of the brain. Approximately 80 percent of the children with LKS have one or more epileptic seizures that usually occur at night. Here we present an unusual case which had all features suggestive of LKS but she had no abnormality regarding EEG or clinically documented seizures.

KEY WORDS. Landau-Kleffner syndrome (LKS), aphasia, infantile acquired aphasia , speech therapy

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INTRODUCTION

Landau-Kleffner syndrome (LKS) is a form of rare, childhood neurological disorder characterized by gradual or sudden inability to express language or understand it, also referred to as, 'aphasia,' as well as an abnormal electroencephalogram (EEG). LKS affects the portions of the child's brain that controls their speech and comprehension. The disorder commonly occurs in children who are between the ages of five and seven. Here we present a case of LKS who had all features suggestive of the above syndrome but had no abnormal EEG findings.

Case report-A 4 year old female presented with complaint of inability to speak clearly for last one year. She had normal milestones and satisfactory schooling for last three and a half years. She gradually developed loss of speech and showed indifference to surrounding and people around her. There was no significant birth history. Earlier she could speak sentences and interacted normally with her siblings but now she used gestures for her daily needs. There was no history of trauma, seizures, altered sensorium, absence spells, hearing loss or psychological stress. She was evaluated by pediatrician, ENT specialist, speech therapist and psychologist. It was concluded that her cognitive functions had declined, she could not speak although her hearing was normal which was confirmed by BERA and LLR. (Fig. No.1,2) Motor development was normal. Comprehension was slightly impaired. There were no features suggestive of autism. MRI brain(Fig. No.3) and sleep EEG(Fig. No.4)did not show any abnormality. Patient was assessed by speech therapist and speech therapy was started which included visual clues, tactile stimulation for speech and behavioral therapy. Child is slowly responding to speech therapy. No other medication was used. She has now improved comprehension and is able to speak clearly few words and is in regular follow up. This seemed to

be a unique case of LKS syndrome as all other features of LKS syndrome were present in this child except the EEG changes.

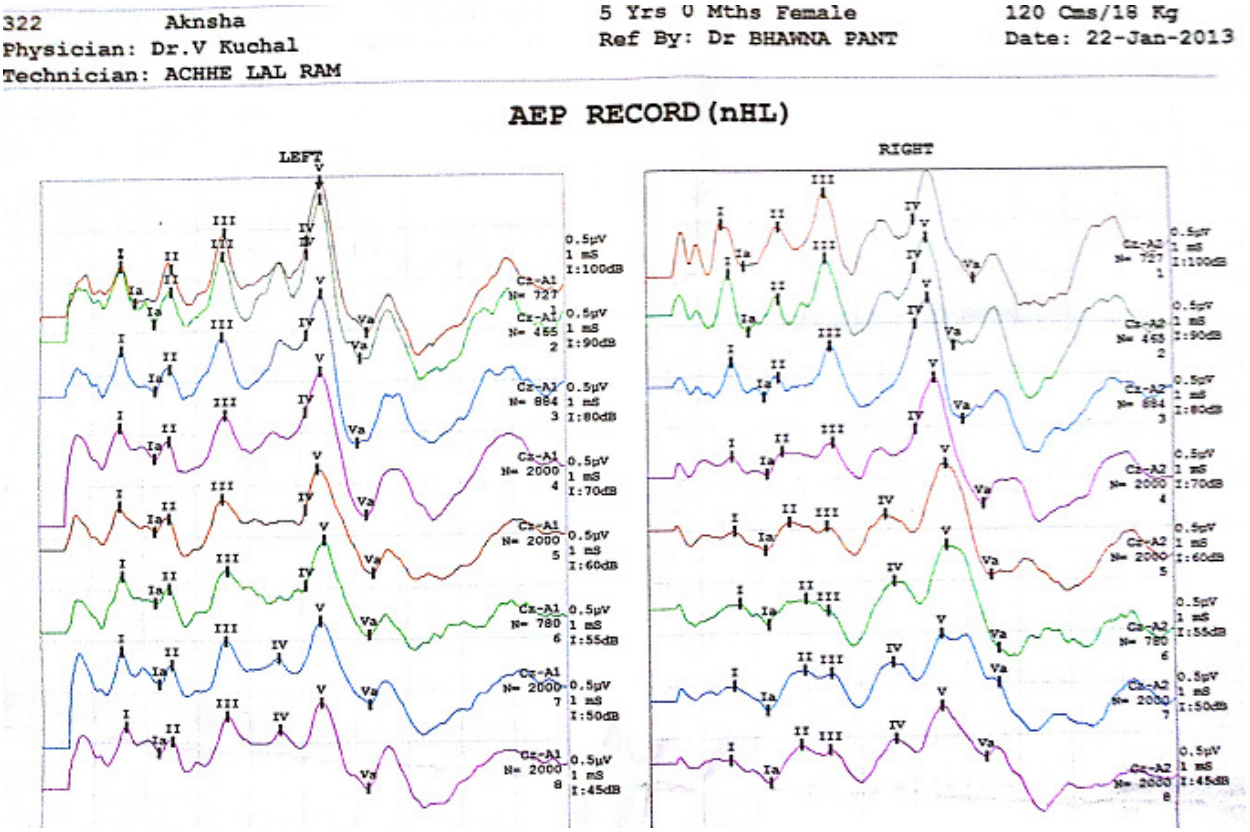

Fig. No.1-showing normal brain evoked response audiometry.

DISCUSSION

LKS may also be called infantile acquired aphasia, acquired epileptic aphasia or aphasia with convulsive disorder. This syndrome was first described in 1957 by Dr. William M. Landau and Dr. Frank R. Kleffner, who identified six children with the disorder. Landau-Kleffner syndrome (LKS) is a rare childhood neurological syndrome. There is gradual or sudden loss of the ability to understand and use spoken language. Almost all children with LKS have abnormal electrical brain waves that can be documented by an EEG.[1] Audiograms and BERA are normal. These cases are sometimes misdiagnosed as autism, pervasive developmental disorder, hearing impairment, learning disability, auditory/verbal processing disorder, attention deficit disorder , mental retardation, childhood schizophrenia emotional or behavioral problems. The early stages of LKS, with hyperkinesis and mild verbal auditory agnosia, may be confused with attention deficit hyperactivity disorder (ADHD)[2]. Personality disturbances, aggressiveness, and depression are noted[3]. Nonverbal developmental disorders may occur, but operational and intellectual capacities are usually preserved in LKS.. The autistic child's language is often stereotyped, repetitive, and idiosyncratic, with echolalia and neologisms[4] Confusing the picture is the fact that seizures may occur in autism, and EEG abnormalities are common[5]. Furthermore, at least a third of autistic toddlers demonstrate neuro developmental regression, involving language, sociability, play, and cognition[6]. LKS represents

selective loss of language in association with an abnormally paroxysmal EEG, eventually characterized by electrographic status epilepticus of slow-wave sleep (ESES). Only 10% of children with LKS regress before three years [7]. As regression in autism occurs early, it usually entails the loss of single words, versus more drastic changes in LKS children who are typically older and have more developed vocabulary and language. The presence of additional forms of aphasia complicates achievement of a diagnosis of LKS[8]. Acquired means the aphasia has happened after the child has already begun to develop language skills. Aphasia may also occur in children who have experienced an infection such as encephalitis, a brain tumor, head trauma, cerebrovascular accidents, or additional brain disorders. Researchers have not discovered a connection between aphasia and a child's gender, age, or race.When a child experiences a brain injury or a stroke, they are assessed for any affects to their language skills. Computed tomography scans (CT) or magnetic resonance imaging (MRI) may be used to map the area of the child's brain that has been affected, or to detect a SOL . PET scanning is useful. LKS affects the parts of the brain that control comprehension and speech (Broca's area and Wernicke's area).This disorder affects children between age of 3-7 years. Typically these children develop normally and then lose their language skills. First indicator of language problem is auditory verbal agnosia. LKS is a disorder of higher-level auditory processing. In a younger child without advanced language development, the effect is devastating, as the normal auditory route leading to acquisition of language is blocked. Word deafness can deteriorate into total unresponsiveness and impaired expressive communication. Expression is` marked by a gradual increase in misarticulations and telegraphic speech; a fluent jargon, or total mutism can occur.

In the present case child slowly lost interest in surroundings, stopped conversation with siblings and at the time of presentation she was totally mute although her hearing seemed normal.LKS is confirmed by obtaining overnight sleep EEGs, including EEGs in all stages of sleep. Most cases of LKS do not have a known cause. Occasionally, the condition may be induced secondary to other diagnoses such as low-grade brain tumors, closed-head injury, CNS vasculitis, neurocysticercosis, and demyelinating disease. An interdependent relationship between language and epileptic manifestations has been described [910],although not all studies have suggested this [11]. EEG changes may not be accompanied by a change in aphasia . Aphasia may not respond to conventional anticonvulsants despite seizure control [12,13] and persists into adulthood despite normalization of EEG. [14,15] The pharmacologic treatment of LKS is problematic due to several confounding observations. The devastating language impairment, fluctuating course of aphasia, lag of improvement in relation to the EEG and possibility of spontaneous remission render multiple barriers to controlled clinical trials in contrast to the benign course of epilepsy . The determination of treatment efficacy is difficult. There is relatively scarce mention in the literature regarding antiepileptics of choice. Marescaux et al. [18] observed that phenobarbital, carbamazepine, and phenytoin were ineffective or even aggravating. Phenobarbital, having no effect on language, intensified behavioral problems, particularily hyperkinesis. Corticosteroids have been an efficacious treatment for both clinical and EEG abnormalities. This was reported by McKinney and McGreal [24], leading to the speculation of chronic encephalitis as the etiology of LKS. Effectiveness may be increased by early introduction [25]. A recurrence of epileptiform EEG followed by an aphasic relapse has been described after tapering steroids . Prolonged, chronic, or intermittent therapy may be warranted if significant improvement of neuropsychological function is attained .Another recent addition is IVIG [26,27,28]. The rationale for its use in LKS lies in the refractory nature of the epileptiform abnormalities, and the reports of beneficial effects in other intractable childhood epilepsies. Speech/language therapy is indispensable with periodic language and neuropsychological evaluationsA speech therapist works with a child affected by aphasia in order to strengthen their remaining language

skills and to discover ways to compensate for the skills they have lost. Various techniques used by the therapist are repetition of words, learning sign language, exercising the child's facial muscles, completing reading and writing , exercises in workbooks, flash cards to improve the child's memory of object names, use of picture of objects and activities to communicate with others, using computer programs to aid in speech, hearing, and reading comprehension, as well as recall. Surgery is used only in relation to the cause of aphasia ie. to reduce pressure from a brain tumor or to reduce swelling from head trauma. One interesting surgical procedure which has been tried with variable outcomes is Subpial transactions severing the pathways in the brain where abnormal activity occurs without damaging the cortical function [31,32]. Prognosis of LKS is variable. Some affected children may have a permanent severe language disorder, while others may regain much of their language abilities (although it may take months or years). Exact relationship between structural brain lesions and clinical manifestation of LKS has not been established [33,34]. In some cases, remission and relapse may occur. The prognosis is improved when the onset of the disorder is after age 6 and early institution of speech therapy. Adverse prognostic factors include onset before 4 years, duration of aphasia longer than one year, duration and continuity of ESES.

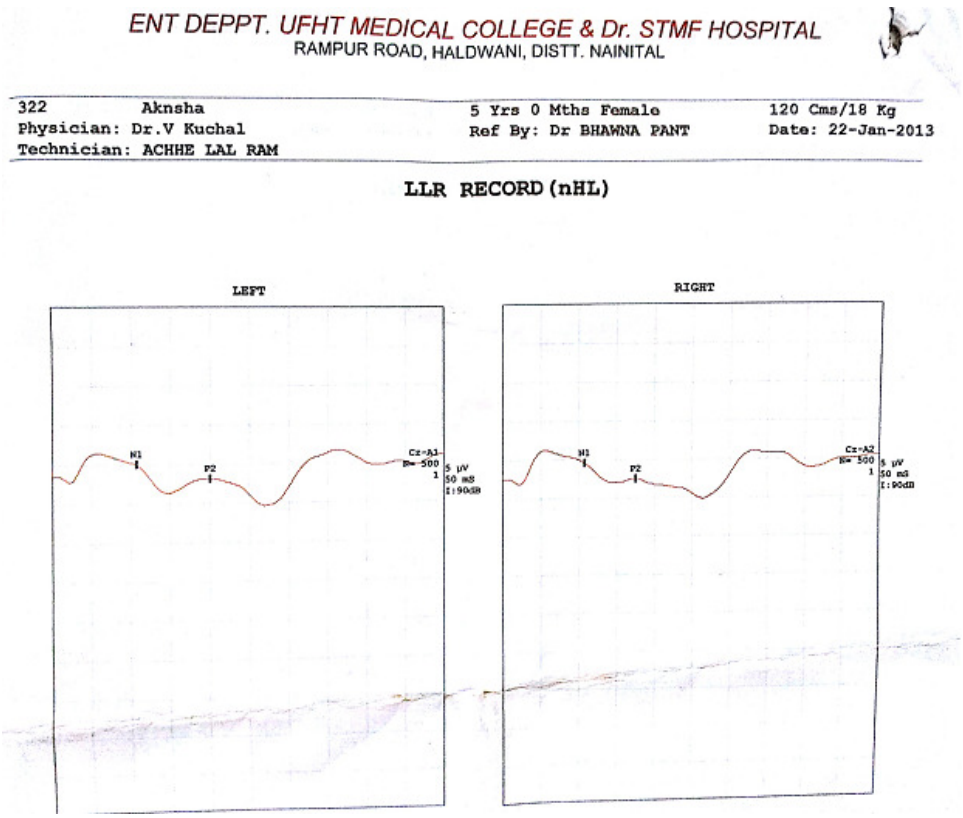

Fig. No.2-showing late .latency response

CONCLUSION

Landau-Kleffner syndrome (LKS) is a form of rare, childhood neurological disorder characterized by gradual or sudden inability to express language or understand it, also referred to as, 'aphasia,' as well as an abnormal electroencephalogram (EEG). The disorder commonly occurs in children who are between the ages of five and seven. One of the unusual presentation of LKS was discussed here where the child had all features suggestive of LKS but she had no abnormality regarding EEG or clinically documented seizures . Patient was assessed by speech therapist

and speech therapy was started which included visual clues, tactile stimulation for speech and behavioral therapy. The patient is responding to it and now showing improved comprehension and speech. A high degree of suspicion is required to make an early diagnosis which is imperative as the clinical outcome is dependent on early institution of speech and behavioral therapy.

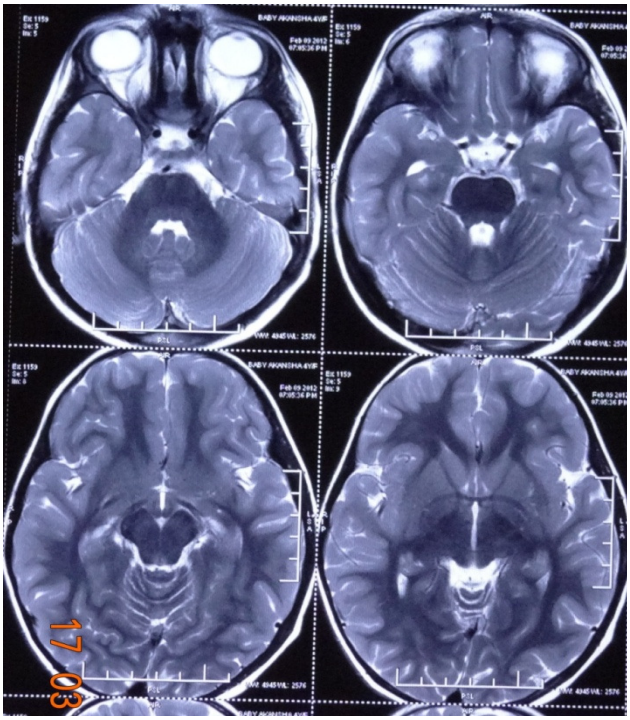

Fig. No.3-Showing normal MRI Brain.

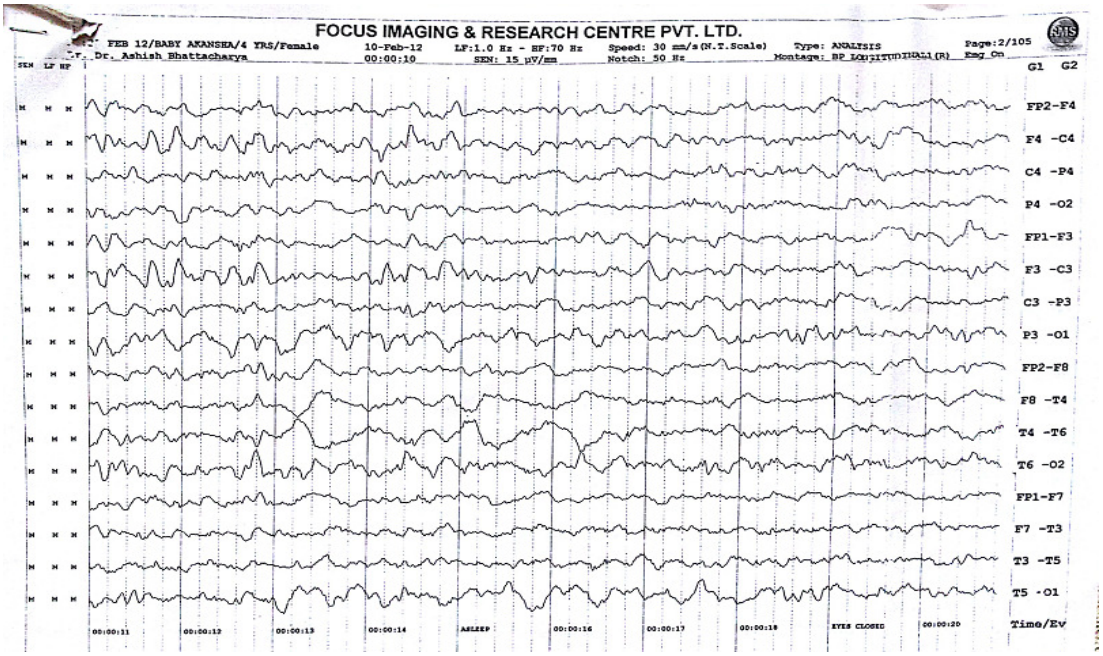

Fig. No.4-Showing normal sleep EEG Recording

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