



Journal Homepage: - www.journalijar.com
**INTERNATIONAL JOURNAL OF
 ADVANCED RESEARCH (IJAR)**

Article DOI: 10.21474/IJAR01/5621
 DOI URL: <http://dx.doi.org/10.21474/IJAR01/5621>



RESEARCH ARTICLE

LANDAU- KLEFFNER SYNDROME: A RARE CASE REPORT.

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Manuscript Info

Manuscript History

Received: 14 August 2017
 Final Accepted: 16 September 2017
 Published: October 2017

Key words:-

Electroencephalography, Steroids,
 Acquired aphasia, Seizures.

Abstract

A 9 year old male presented with history of loss of speech and seizure disorder for two years duration. The boy was developmentally normal before the onset of seizures. Examination of higher mental function revealed excessive hyperactivity, self destructive behaviour and decreased attention span. Audiological evaluation showed bilaterally normal hearing thresholds and he started communicating with signs. EEG showed repetitive spike and epileptiform activity from bilateral parieto-occipital regions. Clinical features and EEG led to a diagnosis of Landau Kleffner Syndrome (LKS). Child was started on steroid, anti-epileptic drug, speech and behavioural therapy immediately and improvement in the symptoms and reduced seizure activity was observed.

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Introduction:-

Landau- Kleffner Syndrome (LKS) was described by Dr. William.M. Landau and Dr. Frank. R. Kleffner in 1957 as Acquired Epileptic Aphasia (AEA). This syndrome is characterized by a triad of receptive and expressive aphasia, epileptic seizures in an apparently normal child and Electroencephalography (EEG) changes. It is a rare childhood epileptic aphasia (AEA) of unknown etiology. Landau Kleffner syndrome may also be called by the following names - Aphasia with convulsive disorder, Infantile acquired aphasia, Worster Drought Syndrome [1]. It is commonly seen in male child [2]. The aim of this case report is to create awareness among otorhinolaryngologists and other health care professionals in making an appropriate diagnosis and to plan the treatment. Because, the children with LKS Syndrome are referred to otorhinolaryngologists initially for the reason of acquired auditory loss, speech impairment and auditory agnosia.

Case Report:-

A 9 year old male was referred to our institute with the complaint of progressive deterioration of speech and seizure disorder for the past two years. A detailed history and general physical examination and a systemic examination was done. The child was developmentally normal before the onset of aphasia and speech deteriorated following seizures. The seizure was of generalized tonic clonic type together with an episode of partial seizure that occurred during sleep, for which child was not under any medications.

Objective testing of all aspects of speech such as naming, reading and writing, repetition of words, comprehension, fluency was done by a speech pathologist. Both 'receptive' and 'expressive' type of speech was affected and 'auditory agnosia' was determined. Speech became garbled and he started communicating mainly through non-

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verbal communication. A complete audiological evaluation was done to be within normal limits and bilateral hearing sensitivity was found to be within normal limits.

A neurologist opinion was taken and diagnosis of Landau Kleffner syndrome was confirmed after doing set of investigations such as Hematology, Liver function test, Renal function test, CSF examination, Ophthalmoscopy, EEG and MRI Brain. The blood investigations were within normal limits. The EEG revealed repetitive spike and epileptiform activity from bilateral parieto-occipital regions.

The MRI Brain was also normal (No focal lesion of the brain parenchyma, no evidence of intracranial space occupying lesion). The child was started on, Lamotrigine and steroid (Prednisolone) along with speech therapy.

The patient showed abnormal behaviour along with attention deficit and did not respond verbally to any command. Excessive hyperactivity, self destructive behaviour, decreased attention span were noticed during clinical examination and the counselling was done by the clinical psychologist. The child had weak gait. So, the child was referred to physiotherapist for physiotherapy.

The patient showed gradual improvement in clinical symptoms, the number of seizure episodes reduced and he started understanding the short sentences. The child is under regular follow-up.

Discussion:-

The diagnosis of LKS, in this case, was made on the basis of progressive deterioration of speech in a previously normal child, associated with abnormal EEG findings compatible with the diagnosis of seizures. The prevalence of clinical seizures in Landau Kleffner Syndrome is 70% and this syndrome is commonly seen in male child, with a male to female ratio of 1.7:1[3,4].

LKS usually appears at 4-7 years of age. However, the onset of symptoms has been described in patients as young as 18 months and in those as old as 13years[5]. Due to relatively rare and uncommon disorder, there is a high chances of misdiagnosis or delayed treatment by the health care professionals. So, it is imperative to become alert to the characteristic symptoms of LKS.

The first symptom is language problem seen in 50% of the cases. Speech defects of those patients could be classified as: Paraphasia, mutism, perseveration and pronunciation deficit. The complete loss of both receptive and expressive speech, confuses the doctors, makes the diagnosis challenging and referring the child to the otorhinolaryngologists, thinking that deterioration of speech may be due to loss of audition. However, hearing tests shows normal hearing [6]. In our patient auditory evaluation was normal.

The aetiology of the syndrome remains unknown. Some author believe a functional disconnection of brain cortex areas by reception language(temporal medial cortex) and cortex areas responsible by spoken language, in the junction of frontal temporal and parietal in the left brain hemisphere[7]. The epileptic form activity in LKS is thought to result in functional ablation of eloquent speech areas. Neurocysticercosis, arteritis, brain tumours, head injury and acute inflammatory conditions have been reported to produce LKS [8].

Around 80% of the patients shows behavioural disturbances such as hyperactivity and decreased attention span. Aggressive and oppositional behaviour, intellectual deficits and personality disturbances are common in Landau Kleffner Syndrome (LKS) patients and wrongly looked as manifestation of psychosis. This may be due to primary functional disinhibition at limbic or diencephalic level or as a secondary effect due to loss of comprehension.

EEG is the accepted diagnostic test in LKS, but EEG changes may vary. Bilateral centrotemporal, posterior temporal and parieto-occipital spikes and waves were seen in awake patients. Sleep EEG may reveal continuous and diffuse slow spikes and waves, mainly at 1.5-2.5Hz persisting through all the slow-sleep stages. This pattern of continuous spikes and waves during slow sleep (CSWS) or electrical status epilepticus in sleep (ESES) in patients with LKS tends to be unilateral or clearly lateralized [9]. Our patient EEG pattern was similar to those features.

Early diagnosis and treatment is essential in case of LKS. Antiepileptic drug such as valproate an Lamotrigine, along with oral steroid (Prednisolone 2mg/Kg/ day for 2 months followed by 0.5mg/Kg/every other day) is required to reduce the seizure frequency and to improve the language and cognitive function. Intravenous immunoglobulin is

also used as the drug of choice by many researchers [10]. Apart from the medical line of treatment, the child needs psychiatrist opinion and counselling followed by physiotherapy by the occupational therapy

Conclusion:-

The otorhinolaryngologist and other health care professionals should be alert in their clinical practice in making the diagnosis of a child presented with a rapid loss of speech and language along with seizure activity as Landau Kleffner syndrome even though it is a very rare entity. A short course of steroid with antiepileptic drugs should be started along with speech and educational rehabilitation as early as possible for the better outcome.

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