Introduction

Landau–Kleffner syndrome (LKS) also called infantile acquired aphasia, acquired epileptic aphasia\(^1\) or aphasia with convulsive disorder is a rare childhood neurological syndrome. It is named after William Landau and Frank Kleffner, who characterized it in 1957.

Acquired epileptic aphasia (AEA) typically develops in healthy children who acutely or progressively lose receptive and expressive language ability coincident with the appearance of paroxysmal electroencephalographic (EEG) changes\(^2\). In most cases described in detail, a clearly normal period of motor and language development occurs before acquired epileptic aphasia symptoms appear. Because this syndrome appears during such a critical period of language acquisition in a child’s life, speech production may be affected just as severely as language comprehension. The onset of LKS is typically between 18 months and 13 years. The prevalence of clinical seizures in acquired epileptic aphasia (LKS) is 70-85\%. The syndrome can be difficult to diagnose and may be misdiagnosed as autism, pervasive developmental disorder, hearing impairment, learning disability, auditory/verbal processing disorder, attention deficit disorder, intellectual disability, childhood schizophrenia, or emotional/behavioural problems. An EEG (electroencephalogram) test is imperative to a diagnosis.

Key words: Aphasia, Epilepsy.

Case Report

A Case of Landau-Kleffner Syndrome

Manjunath S Pandit*, Ashok Gupta**, Priyanshu Mathur***, Manish Sharma****, Manisha Garg*****

*Resident, **Professor and Unit Head, ***Assistant Professor, ****Consultant Paediatrician and In Charge Accident Emergency, *****Senior Resident, Department of Paediatrics, SMS Medical College, Jaipur

ABSTRACT

The Landau-Kleffner syndrome or the syndrome of acquired epileptic aphasia was first described in 1957. The disorder is characterized by gradual or rapid loss of language in a previously normal child. Acquired epileptic aphasia (AEA) typically develops in healthy children who acutely or progressively lose receptive and expressive language ability coincident with the appearance of paroxysmal electroencephalographic (EEG) changes\(^3\). In most cases described in detail, a clearly normal period of motor and language development occurs before acquired epileptic aphasia symptoms appear. Because this syndrome appears during such a critical period of language acquisition in a child’s life, speech production may be affected just as severely as language comprehension. The onset of LKS is typically between 18 months and 13 years. The prevalence of clinical seizures in acquired epileptic aphasia (LKS) is 70-85\%. The syndrome can be difficult to diagnose and may be misdiagnosed as autism, pervasive developmental disorder, hearing impairment, learning disability, auditory/verbal processing disorder, attention deficit disorder, intellectual disability, childhood schizophrenia, or emotional/behavioural problems. An EEG (electroencephalogram) test is imperative to a diagnosis.

Key words: Aphasia, Epilepsy.

Correspondence
Dr Manjunath S Pandit, Resident
Department of Paediatrics, SMS medical college, Jaipur,
E-mail: manju_sp18@yahoo.com

DOI-10.21304/2016.0302.00123
years after onset of language loss. Forty percent of the seizures will precede the language regression, 40 percent will follow, and 20 percent will occur concurrently with onset of language regression.

Case Report
A 10 year old boy was brought in Emergency Room in status epilepticus, with a history of generalized tonic-clonic seizures on and off since the age of 21 months with poor drug compliance since the inception of treatment. Child was developmentally normal till the age of 21 months, when he could hear commands and could speak a sentence of 6-8 words which was appropriate for his age. Then child started having regression of language, deafness and seizures since the age of 21 months. Now the child is deaf with no reception of any sounds and has aphasia and speech which is reduced to only one word.

On clinical examination general and systemic examination were normal, child has normal cognition with no sensory/motor/cranial deficit. Fundus examination was normal, blood counts renal and liver functions tests, CSF were in normal limits. MRI Brain showed altered signal intensity in the right frontal lobe likely a post-ictal edema. Audiometry displayed bilateral profound hearing loss and BERA reported non recordable peaks at 100 dBnHL in both the ears with poor wave morphology and amplitude EEG (Figure 1) showed Bilateral-synchronous bursts of spikes and slow spike-wave complexes over the temporal region. On the basis of the presentation, EEG and MRI, Landau Kleffner syndrome was diagnosed and the child was started on methylprednisolone, sodium valproate and speech therapy; the child is under follow-up.

Discussion
The pathophysiology of LKS remains unknown, although several possibilities have been suggested. Some maintain that the aphasia is the result of the auditory agnosia, implicating a cortical or subcortical dysfunction of the auditory system. Unfortunately, children with LKS can easily be
misdiagnosed, since the outstanding features of this disorder may be suggestive of autism, peripheral hearing loss, emotional or behavioural disorder, or other types of acquired aphasia. Whether seizures and epileptiform discharges cause language dysfunction in acquired epileptic aphasia (AEA) is disputed. Aphasia and electroencephalographic (EEG) abnormalities might have a common cause. The pathogenesis and etiology of landau Kleffner syndrome are unknown and probably complex. The agnosia/aphasia may represent an ‘epileptic’ phenomena caused by paroxysmal spike and slow wave activity within the appropriate temporal lobe. However this may be difficult to accept in the absence of clinically occurring epileptic seizures. An alternative hypothesis is that there is underlying brain pathology in an area or areas concerned with speech, which may be responsible both for the comprehension/speech difficulties and abnormal EEG findings and subsequently, for the development of epileptic seizures. Concrete substantiation of this hypothesis is the existence of poor speech in patients who are affected early and whose condition does not respond to anticonvulsant measures. Other patients with acquired epileptic aphasia appear to have worsened language skills during periods of increased epileptiform activity. However, some reports describe no correlation between EEG abnormality and language dysfunction. The prognosis for children with LKS varies. 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). In some cases, remission and relapse may occur. The prognosis is better when the onset of the disorder is after the age of 6 years and when speech therapy is started early. Seizures generally disappear by adulthood. Short-term remissions are not uncommon in LKS but they create difficulties in evaluating a patient’s response to various therapeutic modalities.

Conclusion

In conclusion Acquired epileptic aphasia is a rare entity and easily misdiagnosed as autism, pervasive developmental disorder, hearing impairment, learning disability, auditory/verbal processing disorder, attention deficit disorder, intellectual disability, childhood schizophrenia, or emotional/behavioural problems. Early recognition and early initiation of speech therapy, anti-convulsants and steroids may benefit.

Source of Funding: None  Conflict of Interest: None

References

1. “Landau-Kleffner syndrome” at Dorland’s Medical Dictionary