Landau-Kleffner Syndrome : A Case Report

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Summary

A healthy 5 year old boy developed aphasia, attention disorder and hyperkinesia preceded by transient formed visual hallucinations and emotional outburst, immediately after a stressful event of forced separation from his father. EEG showed generalized epileptiform activity. He was diagnosed as Landau-Kleffner syndrome (LKS). CT and MRI of the brain were normal. SPECT showed left mesial temporal hypoperfusion. He improved on antiepileptics and ACTH.

Key words : Aphasia, Epileptiform activity, Stressful event.

Introduction

Landau-Kleffner syndrome (LKS) is a rare epileptic syndrome characterized by acquired aphasia with epilepsy.

Case Report

A 5 year old boy developed episodes of sadness and crying for about one hour, immediately after the departure of his father for higher study. He also had transient formed visual hallucinations for about three to five minutes in the form of ‘lion killing his father’. This was followed by inattentiveness, marked receptive and expressive aphasia and hyperkinesia. The child became totally non interactive with his environment within a week’s time. He was seen by the author a month later. He could talk well and write simple sentences in his vernacular language before his illness. There was no family history of epilepsy, mental retardation or developmental speech disturbances. On examination, the child was hyperactive but communication ability was interrupted. Cranial nerves were normal and there were no long tract signs.

EEG, done in sleep state, showed generalized epileptiform activity (Fig 1). CT and MRI of the brain were normal. 99m-Technetium brain single photon emission computed tomography (SPECT) study showed left mesial temporal hypoperfusion. He was put on antiepileptic drug (AED) supplemented with a course of injectable adrenocorticotrophic hormone (ACTH). Follow-up revealed significant improvement in ten months, in verbal output, comprehension, attention span and hyperkinesia. Repeat EEG after 8 months showed persistent epileptiform activity.

Discussion

The diagnosis of LKS, in this case, was made on the basis of rapid loss of language in a previously normal child, associated with abnormal EEG compatible with the diagnosis of epilepsy. While this disorder appears to be relatively uncommon, its frequency is questionable due to its unfamiliarity among the professionals and the likelihood of misdiagnosis. It is imperative that communication specialists become alert to the characteristic symptoms of LKS.1,2 All children diagnosed as LKS have abnormal EEG compatible with the diagnosis of epilepsy, however, only 70% have clinical seizures.2 The subjective emotional outburst of sadness and crying accompanied by formed visual hallucination in this patient may constitute a seizure. This type of seizure is rare in LKS. The epileptiform activity in LKS is thought to result in a functional ablation of eloquent speech areas. In one study, 13 of the 19 patients had perisylvian magneto-encephalography (MEG) spikes.3

The cause of the syndrome remains unknown. Relationship between the structural brain lesions and the clinical manifestations of LKS has been tried but not proved.4-6 Cysticercosis, tumors, arteritis and
acute inflammatory conditions have been reported to produce LKS. CT and MRI of the brain did not reveal structural brain lesion in the present case. However, 99m Tc brain SPECT study showed left mesial temporal hypoperfusion. A stressful event, like forced separation from his father, preceding the onset of interruption in communication ability, as observed in this case, has not been reported. LKS might have been provoked by a stressful event, however, this is only conjectural. LKS is difficult to treat. Treatment modalities used include antiepileptic drugs (AED), corticosteroids, intravenous immunoglobulin, ketogenic diet and multiple subpial transections. Accepting the possible role of the epileptic discharge in producing the symptomatology of LKS and the possible autoimmune reactions in the etiopathogenesis of LKS, the present case was put on AED supplemented with ACTH. He had significant improvement in verbal output, comprehension, attention span and hyperkinesis, in ten months. He had no recurrence of overt seizures, although repeat EEG, eight month after the initiation of the treatment, showed persistent epileptiform activity.

References


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