VOLUME - 15, ISSUE - 02, FEBRUARY - 2026 • PRINT ISSN No. 2277 - 8160 • DOI : 10.36106/gjra

Original Research Paper

Paediatrics

LANDAU–KLEFFNER SYNDROME PRESENTING WITH ACQUIRED APHASIA AND BEHAVIOURAL ABNORMALITIES IN A CHILD: A CASE REPORT

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ABSTRACT

Background: Landau–Kleffner syndrome (LKS) is a rare childhood epileptic encephalopathy characterized by acquired aphasia, behavioural disturbances, and epileptiform electroencephalographic abnormalities, often in the absence of overt seizures. Early diagnosis remains challenging due to significant clinical overlap with autism spectrum disorder and developmental language delay. **Case Presentation:** We report the case of an 8-year-old boy who presented with progressive speech regression and behavioural abnormalities over a period of two years. Neurological and systemic examinations were unremarkable. Developmental assessment revealed significantly delayed language milestones with preserved gross and fine motor development. Based on characteristic clinical features and exclusion of alternative etiologies, a diagnosis of Landau–Kleffner syndrome was established. The child was treated with intravenous immunoglobulin (IVIG), followed by antiepileptic and behavioural medications, resulting in clinical stabilization. **Conclusion:** This case highlights the importance of considering Landau–Kleffner syndrome in children presenting with acquired language regression and behavioural disturbances, even in the absence of clinically apparent seizures. Early recognition and immunomodulatory therapy may help stabilize disease progression and improve outcomes.

KEYWORDS : Landau–Kleffner syndrome; acquired aphasia; epileptic encephalopathy; childhood regression; IVIG

INTRODUCTION

Landau–Kleffner syndrome (LKS) is a rare age-related epileptic encephalopathy first described in 1957.[1] It is characterized by acquired aphasia, behavioural disturbances, and epileptiform electroencephalographic abnormalities in children with previously normal or near-normal development.[2] The typical age of onset ranges from 3 to 8 years.[3]

Due to its rarity and heterogeneous clinical presentation, LKS is frequently misdiagnosed as autism spectrum disorder or developmental language disorder, leading to delays in appropriate diagnosis and management.[4] Early recognition is essential, as prolonged disease activity may result in irreversible language impairment.[5]

We present a case of Landau–Kleffner syndrome in an 8-year-old child with progressive speech regression and behavioural abnormalities, emphasizing the clinical features, diagnostic challenges, and therapeutic approach.

Case Presentation

An 8-year-old male child was admitted to the Department of Neurology with complaints of progressive difficulty in speech since the age of two years and behavioural abnormalities for the past one year. The child initially exhibited delayed speech initiation, followed by gradual regression characterized by poor language comprehension, repetition of words, and failure to respond appropriately to questions. Behavioural changes included irritability, frequent anger spells, and use of abusive language.[6]

The child was born at term by lower-segment caesarean section with a birth weight of 3.6 kg. There was a history of neonatal intensive care unit admission for delayed cry. Gross and fine motor developmental milestones were appropriate for age; however, language milestones were significantly delayed, followed by regression. Immunization history was adequate. There was no history of seizures, head trauma, central nervous system infection, or similar illness in the family.

On examination, the child was conscious and oriented, with stable vital parameters. Neurological examination revealed a Glasgow Coma Scale score of E4V5M6. Cranial nerve examination was normal. Motor examination showed normal bulk, tone, and power (5/5) in all limbs, with symmetrical deep tendon reflexes (2+ bilaterally). Plantar responses were flexor bilaterally. No cerebellar or meningeal signs were present. Systemic examination was unremarkable.

Routine laboratory investigations, including complete blood count, liver and renal function tests, serum electrolytes, calcium levels, and random blood glucose, were within normal limits.

Based on the clinical findings and the characteristic pattern of acquired language regression associated with behavioural disturbances, a diagnosis of Landau–Kleffner syndrome was made. Electroencephalography could not be performed initially due to significant agitation.

The child was treated with intravenous immunoglobulin (IVIG) at a dose of 50 g administered over five days, along with antiepileptic and behavioural medications. During the hospital stay, the child remained hemodynamically stable with no deterioration in neurological status. He was discharged on oral medications and advised close follow-up in the neurology outpatient department, with repeat investigations and planned IVIG therapy after three weeks.[7]

DISCUSSION

Landau–Kleffner syndrome is an uncommon disorder, with an estimated prevalence of less than one per million children.[8] The hallmark clinical feature is acquired aphasia, often accompanied by behavioural abnormalities such as hyperactivity, irritability, and aggression. Notably, seizures may be absent in up to 30% of cases, making diagnosis particularly challenging.[9]

The differential diagnosis includes autism spectrum disorder, attention deficit hyperactivity disorder, and other developmental language disorders. The key distinguishing feature of LKS is the regression of previously acquired language skills, rather than a primary failure of language

development.[10]

Immunomodulatory therapies, including corticosteroids and intravenous immunoglobulin, have demonstrated benefit in reducing epileptiform activity and stabilizing language function.[11] Early intervention is critical, as prolonged untreated disease is associated with poor language recovery and long-term neurodevelopmental impairment.[12]

This case underscores the importance of maintaining a high index of suspicion for Landau–Kleffner syndrome in children presenting with language regression and behavioural disturbances, even in the absence of overt seizures.

CONCLUSION

Landau–Kleffner syndrome should be considered in children presenting with acquired aphasia and behavioural abnormalities. Early diagnosis and prompt initiation of immunomodulatory therapy may help prevent further neurological deterioration and improve long-term outcomes.[13]

Patient Consent

Written informed consent was obtained from the patient's parents for publication of this case report and accompanying clinical details.

Conflict Of Interest

The authors declare no conflict of interest.

Funding

No funding was received for this study.

Author Contributions

All authors were involved in patient management, data collection, manuscript drafting, and final approval of the manuscript.

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