International Journal of Medicine Research
www.medicinesjournal.com
ISSN: 2455-7404
Received: 19-06-2026, Accepted: 20-07-2026, Published: 04-08-2026
Volume 11, Issue 3, 2026, Page No. 11-14

Acquired epileptic aphasia with behavioral disturbance in an eight-year-old boy: A case of landau-kleffner syndrome managed with intravenous immunoglobulin

Vidya Nittur[1]*, Raghav Singh[1], Navitha N[1], Chandan Kumar N R[2], Anish Chakravarty[3], Prarthi Yadav[4], Harshitha Nagraj[5], Sunidhi Nayak[5], Ruthvik S N[5], Sourabha Hegde[6]

[1] Department of Psychiatry, KVG Medical College & Hospital, Sullia, Karnataka, India
[2] Department of Emergency Medicine, KVG Medical College & Hospital, Sullia, Karnataka, India
[3] Department of Radiology, Ramaiah Medical College, Bengaluru, Karnataka, India
[4] Department of Pathology, Medical College, Kolkata, West Bengal, India
[5] Department of Pediatrics, KVG Medical College & Hospital, Sullia, Karnataka, India
[6] Department of Otorhinolaryngology (ENT), SDM College of Medical Sciences and Hospital, Dharwad, Karnataka, India

Corresponding Author: Dr. Vidya Nittur

DOI: https://doi.org/10.66856/ijmr.2026.11.3.11029

Abstract

An 8-year-old boy presented with progressive language impairment beginning at two years of age and behavioral abnormalities for one year. He exhibited repetitive speech, poor response to verbal commands, episodes of abusive language, and frequent anger outbursts. Following initial assessment and stabilization in the Emergency Department, the child underwent multidisciplinary evaluation involving Neurology, Psychiatry, Pediatrics, Radiology, Pathology, and Otorhinolaryngology (ENT). Pediatric assessment confirmed isolated language delay with preserved gross and fine motor milestones. Radiological review excluded structural intracranial abnormalities, while pathological evaluation of hematological and biochemical investigations ruled out infectious, metabolic, and inflammatory etiologies. ENT evaluation excluded hearing impairment and other otorhinolaryngological causes of language regression. Based on previous neurological evaluation and current clinical findings, a diagnosis of Landau-Kleffner syndrome was confirmed. The child received intravenous immunoglobulin therapy over five days along with clobazam, risperidone, clonidine, and methylphenidate. He remained clinically stable throughout hospitalization and was discharged with advice for repeat immunotherapy after three weeks, continued speech-language therapy, and multidisciplinary follow-up.

Keywords: Landau-Kleffner syndrome, acquired epileptic aphasia, behavioral abnormalities, intravenous immunoglobulin, childhood epileptic encephalopathy

Introduction

Landau-Kleffner syndrome is a rare childhood epileptic encephalopathy first described by Landau and Kleffner in 1957 [1]. The syndrome is characterized by acquired aphasia associated with epileptiform abnormalities on electroencephalography (EEG), predominantly involving the temporal regions during sleep. The estimated prevalence is approximately one per million children, making it one of the rarest childhood neurological disorders.

The disorder typically presents between three and eight years of age in previously normal children who progressively lose receptive language, followed by expressive language abilities. In many patients, parents initially notice that the child appears not to hear spoken language despite normal hearing, resulting in inappropriate referrals for hearing impairment evaluation. Behavioral manifestations such as hyperactivity, irritability, aggression, autistic features, attention deficits, anxiety, emotional dysregulation, and sleep disturbances frequently accompany language deterioration. Approximately 70–80% of affected children develop seizures, although epilepsy may be absent in a significant proportion.

The underlying pathophysiology remains incompletely understood but is believed to involve epileptic dysfunction of the dominant temporal cortex responsible for language processing, possibly mediated by immune-inflammatory mechanisms. Consequently, immunomodulatory therapies, particularly corticosteroids and intravenous immunoglobulin (IVIG), have increasingly been incorporated into treatment protocols alongside antiseizure medications and speech rehabilitation.

We report the case of an eight-year-old boy with Landau-Kleffner syndrome who presented predominantly with acquired language impairment and behavioral disturbances and was managed with intravenous immunoglobulin therapy.

Case Presentation

An eight-year-old boy was admitted to the Department of Neurology at the Institute of Medical Sciences, Banaras Hindu University, for planned administration of intravenous immunoglobulin (IVIG) following a previous diagnosis of Landau-Kleffner syndrome (LKS). Prior to admission, he underwent emergency evaluation to assess his clinical status and exclude acute neurological, metabolic, infectious, or toxic causes contributing to his behavioral and language disturbances.

According to his parents, the child had experienced progressive deterioration of language beginning at approximately two years of age. Speech development was delayed from early childhood despite preservation of gross and fine motor developmental milestones. During the preceding year, behavioral abnormalities became increasingly prominent, including repetitive verbalizations, poor response to spoken language, inability to answer simple questions, inappropriate abusive language,

irritability, and frequent anger outbursts, resulting in marked impairment of communication and social functioning.
A multidisciplinary evaluation was undertaken involving Neurology, Psychiatry, Pediatrics, Emergency Medicine, Radiology, Pathology, and Otorhinolaryngology (ENT). Pediatric assessment confirmed isolated language delay with otherwise age-appropriate growth and neurodevelopment. Emergency physicians ensured initial stabilization and coordinated baseline investigations. Radiological review of available neuroimaging excluded significant structural brain abnormalities that could explain the clinical presentation. Pathological review of routine hematological and biochemical investigations excluded infectious, inflammatory, metabolic, and hematological disorders. ENT evaluation demonstrated normal hearing and excluded otorhinolaryngological disorders contributing to language regression. Based on the characteristic clinical features, previous neurological evaluation, and exclusion of alternative diagnoses, Landau-Kleffner syndrome was confirmed, and IVIG therapy was initiated.

Birth and Developmental History
The child was delivered by lower segment cesarean section at term with a birth weight of 3.6 kg. Neonatal history was significant for delayed cry requiring admission to the neonatal intensive care unit.

Developmental assessment revealed:
- Gross motor milestones appropriate for age
- Fine motor milestones appropriate for age
- Significant delay in language milestones

Vaccinations were complete according to the national immunization schedule.

Past History
There was no history suggestive of chronic systemic illness, meningitis, encephalitis, traumatic brain injury, metabolic disease, or previous neurological disorders apart from the current illness.
No history of substance exposure or toxin ingestion was reported.

Family History
No family history of epilepsy, developmental disorders, autism spectrum disorder, language disorders, or psychiatric illness was documented.

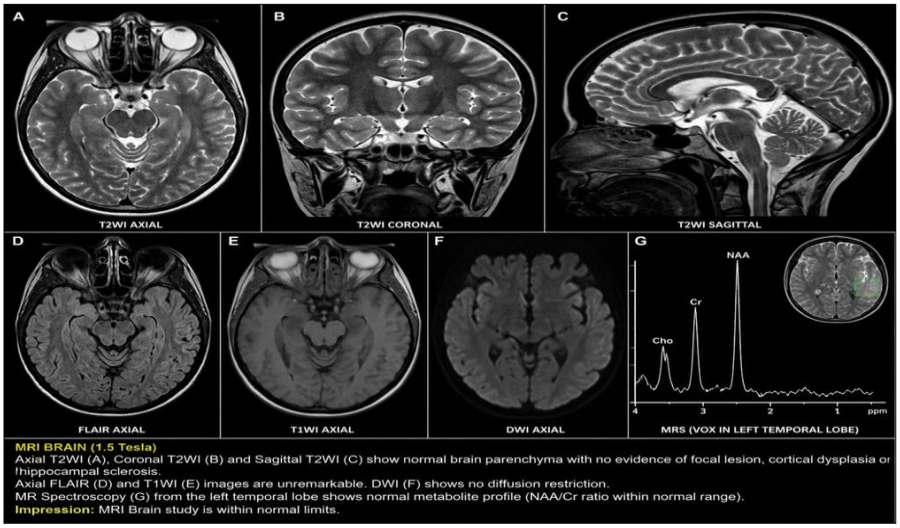
Clinical Examination
On admission, the child was conscious, alert, and oriented. General physical examination was unremarkable.

Vital signs included:
- **Blood pressure:** 102/68 mmHg
- **Pulse:** 88/min
- **Respiratory rate:** 14/min
- **Temperature:** Afebrile
- **Oxygen saturation:** 96% on room air Systemic examination was within normal limits.

Neurological examination demonstrated
- Glasgow Coma Scale score of E4V5M6
- Cranial nerves intact
- Normal muscle bulk and tone
- Muscle strength 5/5 in all four limbs
- Deep tendon reflexes 2+ and symmetrical
- Bilateral flexor plantar responses
- Normal sensory examination
- No cerebellar signs
- No meningeal signs

The principal neurological abnormality consisted of severe impairment in language function with marked deficits in communication.

Investigations
Routine hematological investigations revealed hemoglobin of 12.5 g/dL, total leukocyte count of 7,370/mm³, and platelet count of 362 × 10³/mm³. Biochemical investigations demonstrated serum creatinine of 0.57 mg/ dL, blood urea of 12.6 mg/dL, serum sodium of 133 mmol/L, serum calcium of 8.86 mg/dL, and random blood glucose of 90 mg/dL. All laboratory parameters were reviewed by the Department of Pathology and were within acceptable limits, excluding significant infectious, inflammatory, metabolic, or hematological abnormalities.
Available neuroimaging was reviewed by the Department of Radiology and did not reveal any structural abnormalities to account for the patient's acquired aphasia or behavioral disturbances. ENT assessment demonstrated normal hearing and excluded peripheral auditory or upper airway pathology. Sleep EEG could not be performed during the present admission because of marked behavioral agitation.



Diagnosis
A diagnosis of Landau-Kleffner syndrome (acquired epileptic aphasia) with associated behavioral disturbances was established based on previous neurological evaluation and current clinical findings.

Differential Diagnosis
Differential diagnoses included autism spectrum disorder, childhood disintegrative disorder, developmental language disorder, hearing impairment, electrical status epilepticus during sleep (ESES/CSWS), and other developmental and epileptic encephalopathies. Multidisciplinary evaluation involving Pediatrics, Radiology, Pathology, and ENT helped systematically exclude developmental, structural, metabolic, infectious, and hearing-related causes, supporting the diagnosis of Landau-Kleffner syndrome.

Therapeutic Intervention
The child received intravenous immunoglobulin therapy at a cumulative dose of 2 g/kg administered over five consecutive days.

Concomitant medications at discharge included
- Clonidine (Arkamin)
- Risperidone (Sizodon)
- Clobazam (Cloba)
- Methylphenidate (Inspiral)

Behavioral stabilization was achieved during hospitalization.
Repeat IVIG therapy was planned after three weeks, with continued neurological follow-up. Management was coordinated through a multidisciplinary team comprising neurologists, psychiatrists,
pediatricians, emergency physicians, radiologists, pathologists, speech-language therapists, and rehabilitation specialists. Following exclusion of alternative etiologies, the child received IVIG at a cumulative dose of 2 g/ kg administered over five consecutive days along with adjunctive pharmacotherapy.

Outcome and Follow-up
The child remained hemodynamically stable throughout hospitalization. No immediate adverse effects related to intravenous immunoglobulin administration were observed. At discharge, behavioral symptoms had stabilized sufficiently for outpatient management. Parents were advised regarding continuation of prescribed medications, regular speech-language therapy, neurological follow-up, laboratory monitoring, and repeat IVIG administration.
Long-term assessment of language recovery and neuropsychological functioning was planned during subsequent follow-up visits.

Discussion
Landau-Kleffner syndrome is a rare but potentially reversible cause of acquired aphasia in childhood. Unlike developmental language disorders, affected children typically demonstrate regression of previously acquired receptive language abilities, often followed by expressive aphasia. This distinction is crucial because early diagnosis allows initiation of therapies that may alter disease progression.
Behavioral disturbances frequently complicate the clinical picture and often lead to initial psychiatric referral. Hyperactivity, aggression, irritability, autistic behaviors, emotional dysregulation, attention deficits, and social withdrawal are common. Our patient exhibited repetitive speech, poor responsiveness to verbal communication, abusive language, and anger outbursts, features that may mimic autism spectrum disorder or disruptive behavior disorders.
The present case highlights the value of a multidisciplinary approach in the evaluation of children with acquired language regression. Pediatric assessment established preserved overall neurodevelopment apart from language impairment, while Emergency Medicine ensured stabilization and timely referral.
Radiological evaluation excluded structural intracranial pathology, pathological assessment ruled out metabolic, infectious, and inflammatory causes, and ENT consultation excluded hearing impairment as an explanation for language regression. Such systematic exclusion of differential diagnoses is essential because the clinical manifestations of LKS frequently overlap with autism spectrum disorder, developmental language disorders, and other epileptic encephalopathies.
The exact pathogenesis remains uncertain. Functional disruption of language networks by continuous epileptiform activity, particularly during non-rapid eye movement sleep, is considered central. Emerging evidence also supports immune-mediated mechanisms, explaining the favorable response observed with corticosteroids and intravenous immunoglobulin in selected patients.
Although corticosteroids remain first-line immunotherapy in many centers, IVIG has demonstrated benefit in steroid-resistant or recurrent cases and in patients with suspected immune-mediated disease. Proposed mechanisms include modulation of autoantibodies, suppression of pro-inflammatory cytokines, complement inhibition, and restoration of immune homeostasis.
Successful management requires a multidisciplinary approach involving pediatric neurologists, psychiatrists, speech-language pathologists, psychologists, occupational therapists, and special educators. Long-term prognosis depends largely on age at onset, duration of untreated disease, severity of EEG abnormalities, and prompt initiation of therapy.
The present case emphasizes the importance of recognizing language regression associated with behavioral changes as a neurological emergency rather than attributing symptoms solely to psychiatric or developmental disorders.

Conclusion
Landau-Kleffner syndrome is a rare but potentially reversible childhood epileptic encephalopathy characterized by acquired language regression, epileptiform EEG abnormalities, and behavioral disturbances. Owing to its varied clinical presentation, it is frequently misdiagnosed as autism spectrum disorder, hearing impairment, or primary developmental language disorders, resulting in delayed treatment. This case emphasizes the importance of maintaining a high index of suspicion in children presenting with progressive language deterioration and behavioral changes. Early diagnosis through multidisciplinary assessment and timely initiation of immunomodulatory therapy, antiepileptic medication, behavioral interventions,

and intensive speech rehabilitation may significantly improve neurological and functional outcomes. Increased awareness among pediatricians, neurologists, psychiatrists, and rehabilitation specialists is essential for prompt recognition and optimal long-term management of this uncommon disorder.

Acknowledgements
The authors thank the patient and his family for providing informed consent for publication of this case report.

Funding
The authors received no financial support for the research, authorship, or publication of this article.

Conflict of Interest
The authors declare that there are no conflicts of interest.

Ethical Approval
Institutional ethical approval was not required for this single case report as per institutional policy. Written informed consent for publication was obtained from the patient's parent/legal guardian.

Consent for Publication
Written informed consent was obtained from the patient's parent/legal guardian for publication of this case report and accompanying clinical information. Patient identity has been protected.

References

1. Landau WM, Kleffner FR. Syndrome of acquired aphasia with convulsive disorder in children. Neurology,1957:7(8):523–530.
2. Caraballo RH, Cersósimo RO, Fejerman N. Landau-Kleffner syndrome: A study of 29 patients. Seizure,2014:23(2):98–104.
3. Pearl PL, Carrazana EJ, Holmes GL. The Landau-Kleffner syndrome. Epilepsy Currents,2001:1(2):39–45.
4. Stefanatos GA. Changing perspectives on Landau-Kleffner syndrome. The Clinical Neuropsychologist,2011:25(6):963–988.
5. Smith SJ. EEG in the diagnosis, classification, and management of patients with epilepsy. Journal of Neurology, Neurosurgery, and Psychiatry,2005:76(2): ii2–ii7.
6. Specchio N, Fusco L, Vigevano F. Epileptic encephalopathy in children. Epilepsy Research and Treatment,2011:2011:1–10.
7. Tuchman R. Autism and epilepsy: What has regression got to do with it? Epilepsy Currents,2013:13(6):262–264.
8. Nickels KC, Wirrell EC. Electrical status epilepticus in sleep. Seminars in Pediatric Neurology,2008:15(2):50–60.
9. van den Munckhof B, van Dee V, Sagi L, *et al*. Treatment of Landau-Kleffner syndrome: A systematic review. Epilepsia Open,2020:5(4):498–507.
10. Sánchez Fernández I, Loddenkemper T, Peters JM, Kothare SV. Electrical status epilepticus in sleep: Clinical presentation and pathophysiology. Pediatric Neurology,2012:47(6):390–410.
11. Hirsch E, French J, Scheffer IE, *et al*. ILAE clinical practice recommendations for the treatment of epilepsies. Epileptic Disorders,2022:24(2):183–232.
12. Wirrell EC, Shellhaas RA, Joshi C, *et al*. How should children with epileptic encephalopathy be evaluated and treated? Epilepsia,2015:56(8):1185–1197.