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Case Report

Paediatrics Section

Rasmussen Encephalitis Closely Mimicking Infantile Spasms: A Case Report

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ABSTRACT

Rasmussen Encephalitis (RE) is a rare, progressive neurological disorder primarily affecting children, with early-onset cases posing significant diagnostic challenges. In infants, RE may closely resemble more common conditions such as infantile spasms, often leading to delayed diagnosis and suboptimal treatment. The present case reported a six-month-old male who presented to the paediatric neurology department with the chief complaint of repeated jerky movements of the left hand for the past three weeks. His seizure semiology mimicked infantile spasms. Initial Electroencephalography (EEG) and Magnetic Resonance Imaging (MRI) findings were unremarkable, contributing to diagnostic uncertainty. However, over subsequent weeks, the child developed progressive left hemispheric atrophy on neuroimaging, along with persistent, treatment-resistant seizures. These evolving features guided the diagnosis towards RE. Management included oxcarbazepine, levetiracetam, and corticosteroids, which resulted in only partial seizure control. This case underscores the importance of maintaining a high index of suspicion for RE in infants with refractory spasm-like seizures and emphasises the diagnostic value of serial imaging. Early recognition and initiation of immunotherapy may potentially alter disease progression and improve outcomes. Multidisciplinary collaboration remains essential for optimising long-term neurological function and quality of life in affected patients.

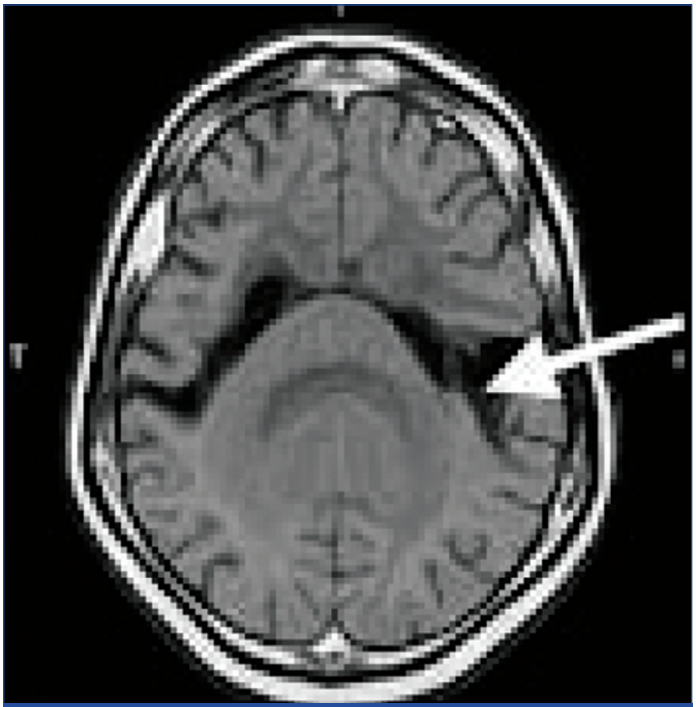
Keywords: Corticosteroids, Hemispheric atrophy, Immunotherapy, Intractable seizures

CASE REPORT

A six-month-old male infant was brought to the paediatric neurology department with the chief complaint, described by his parents, of repeated jerky movements of the left hand for the past three weeks. These episodes occurred multiple times daily, each lasting 15 to 30 seconds, without any associated fever, loss of consciousness, or postictal drowsiness. The child had been developmentally normal before the onset of seizures and had achieved appropriate milestones for age. The parents noted that the seizure frequency had gradually increased to six to eight episodes per day, prompting them to seek further medical care. He was born at term via normal vaginal delivery, with an uneventful antenatal and perinatal history. The mother had no known comorbidities during pregnancy. There was no family history of epilepsy or autoimmune disease.

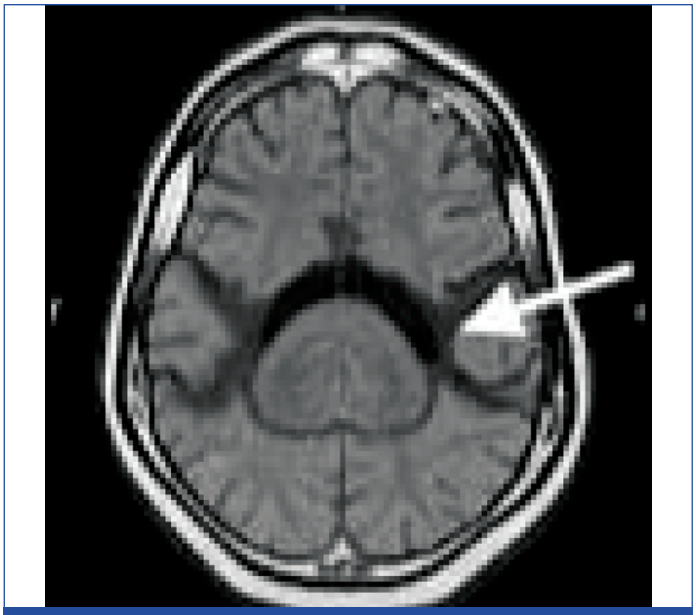
Initially evaluated at a local facility, the child was suspected of having infantile spasms and was started on empirical antiepileptic therapy. However, despite two weeks of treatment, the seizures persisted. At the time of presentation to our centre, he was alert, active, feeding well, and not in respiratory distress. No seizure activity was witnessed during examination. His vital signs were within normal limits, with a temperature of 36.8°C, heart rate of 128/min, respiratory rate of 36/min, oxygen saturation of 98% on room air, and blood pressure of 82/58 mmHg. Birth and antenatal history were unremarkable, with no perinatal complications.

Initial investigations revealed normal routine blood tests and metabolic screening. Cerebrospinal fluid analysis, including Polymerase Chain Reaction (PCR) for Herpes Simplex Virus (HSV), measles, Japanese encephalitis, and enterovirus, was unremarkable. A sleep-deprived EEG showed normal background activity without hypsarrhythmia or epileptiform discharges. Initial MRI revealed mild diffuse atrophy of the right cerebral hemisphere, with sulcal prominence and reduced cortical and white matter volume, suggestive of early RE [Table/Fig-1-4]. Based on clinical presentation, imaging findings, and lack of response to antiepileptics, a provisional diagnosis of RE was made. Differential diagnoses considered were infantile spasms, which were ruled out due to a normal EEG and absence of hypsarrhythmia, as well as focal cortical dysplasia and post-encephalitic epilepsy, both of which were unlikely due to the absence of structural lesions or preceding febrile illness. Metabolic causes were excluded based on a normal metabolic profile. Hence, a final diagnosis of RE was made.

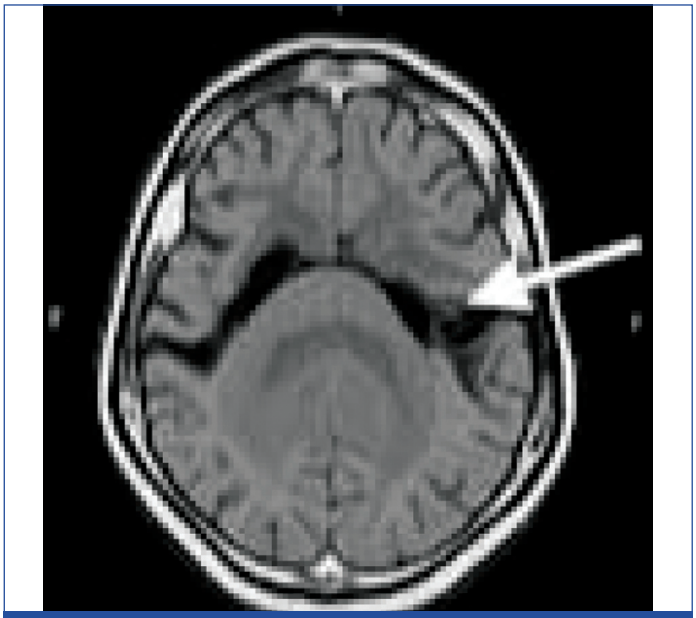


[Table/Fig-1]: MRI brain showing mild diffuse atrophy of the right cerebral hemisphere.

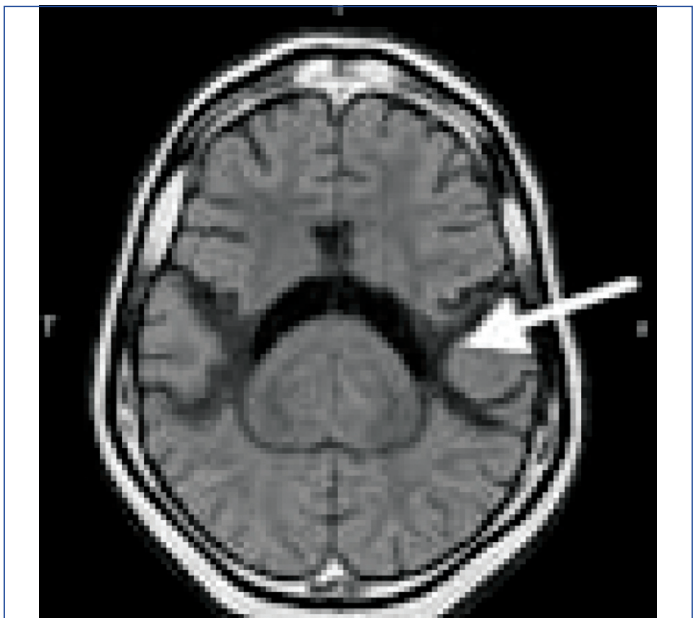
Treatment was initiated with oxcarbazepine at 10 mg/kg/day in two divided doses, gradually increased to 20 mg/kg/day, and levetiracetam at 20 mg/kg/day, titrated to 40 mg/kg/day. Due to ongoing seizures and emerging neurological concerns, oral corticosteroids (prednisolone) were added at 2 mg/kg/day. Following



[Table/Fig-2]: MRI brain showing sulcal prominence (widened grooves) in the right cerebral hemisphere.



[Table/Fig-3]: MRI brain showing reduced cortical and white volume of the right cerebral hemisphere.



[Table/Fig-4]: MRI brain showing white arrows indicate areas of interest in the right hemisphere of the right cerebral hemisphere.

initiation of corticosteroids, the patient showed a partial response, with approximately 50% reduction in seizure frequency, now experiencing three to four seizures per day with reduced severity. At follow-up at nine months of age, the child remained seizure-prone but without new neurological deficits and continued to achieve developmental milestones appropriately. He is under regular follow-up for further management, including consideration of long-term immunotherapy or surgical intervention, if required. The parents were thoroughly counselled regarding the chronic nature of the condition, treatment options, and potential outcomes. The patient was last followed up on one month ago, with ongoing monitoring of seizure frequency and developmental progress.

DISCUSSION

The RE is a rare, progressive, chronic inflammatory neurological disorder that primarily affects children. Although it typically presents between 6 and 10 years of age, cases in early infancy are increasingly recognised, often posing significant diagnostic challenges. The estimated incidence is approximately 2.4 cases per 10 million children annually, making it an uncommon but serious cause of intractable epilepsy. In infants, RE may initially mimic more common epileptic syndromes, particularly infantile spasms, with normal early EEG and MRI findings complicating timely diagnosis [1].

The exact aetiology of RE is not fully understood, but current evidence supports an autoimmune mechanism, with cytotoxic T-cell–mediated neuronal injury playing a central role. Antibodies against glutamate receptor subunits, such as GluR3, and other neuronal antigens have been reported in some cases, although their diagnostic utility remains limited. Histopathological findings typically include T-cell infiltration, microglial nodules, neuronal loss, and gliosis, supporting the theory of immune-mediated neurodegeneration [2].

Differential diagnoses in such presentations are broad and include hemimegaloencephaly, focal cortical dysplasia, stroke, and unilateral infections [3]. Hemimegaloencephaly is usually evident on early neuroimaging as hemispheric enlargement and cortical malformations. Focal cortical dysplasia may present similarly, but often shows cortical thickening or blurring of the grey-white junction on MRI. Perinatal stroke, another consideration, typically follows a known vascular territory and is often associated with a perinatal insult. Unilateral infectious aetiologies such as herpes simplex encephalitis or other viral encephalitis may be suggested by fever, Cerebrospinal Fluid (CSF) abnormalities, or diffusion restriction on MRI. However, in RE, these differentials are often ruled out through serial imaging, clinical evolution, and negative infectious or metabolic evaluations [4].

RE should be suspected when seizures become progressively unilateral and refractory despite initially normal investigations. In the present case, the child presented with seizures closely resembling infantile spasms but showed no abnormalities on early EEG or imaging. As the clinical course progressed, follow-up neuroimaging revealed right hemispheric atrophy with sulcal prominence and cortical and white matter volume loss, which are hallmark features of RE [5]. This parallels other reported cases in which initial imaging was unremarkable, and diagnostic clarity emerged only through serial radiological evaluation [6]. For instance, a recent case described a five-year-old girl with initially normal MRI and focal seizures, later developing right hemispheric atrophy and characteristic EEG findings that confirmed the diagnosis of RE following serial imaging. Similarly, another case involved a young child with early, subtle imaging changes that were initially overlooked; the diagnosis of RE became evident only as the clinical symptoms progressed and imaging evolved [7].

These cases highlight the importance of longitudinal follow-up and a high index of suspicion in children presenting with atypical seizure patterns and inconclusive early investigations [8].

EEG findings may also be nonspecific in early stages, sometimes showing normal or mildly abnormal patterns, but frequently evolve to demonstrate asymmetric background slowing or focal epileptiform discharges over time [9]. Therapeutic approaches to RE remain challenging. Early initiation of immunotherapy-such as corticosteroids, Intravenous Immunoglobulin (IVIG), cyclophosphamide, or Mycophenolate Mofetil (MMF)- may offer temporary seizure control and slow disease progression in selected patients [10]. However, these interventions often have limited long-term efficacy. Hemispherectomy remains the only definitive treatment for refractory cases, though it carries significant risk, particularly in very young children, and is associated with permanent neurological deficits, including hemiplegia and visual field loss [11].

In our case, early immunotherapy with corticosteroids in conjunction with antiepileptic drugs achieved partial seizure control and may have delayed further neurological deterioration. At the time of follow-up, seizure frequency had decreased by approximately 50%, and no new neurological deficits had emerged. The child remains under close follow-up with plans for serial EEG, MRI, and neurodevelopmental assessments to monitor progression and guide future treatment decisions.

Previous studies also reported the chronic nature of the disease, in which subjects were followed up without complete cure of the disease [12-14].

CONCLUSION(S)

Early-onset RE may closely mimic infantile spasms, leading to misdiagnosis. Clinicians should maintain suspicion when focal seizures persist despite normal initial evaluations. Serial imaging and early immunotherapy may improve outcomes. Further research and reporting of such cases are needed to refine diagnostic and therapeutic approaches.

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