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CASE REPORT

Rasmussen syndrome in a pediatric patient: case report

Síndrome de Rasmussen em paciente pediátrico: relato de caso

André Vinícius Soares Barbosa[1], Arthur Andrade Resende[2], Clarissa Milbratz de Castro[3], Luiza Sousa Vilano[3], Patrícia Laura de Lacerda Viggiano[3], Vivian Proença Lara[3]

ABSTRACT

Introduction: Rasmussen's Syndrome (RS) is a rare, chronic, and progressive encephalitis, presumably of autoimmune origin, that predominantly affects children. It is characterized by drug-resistant focal seizures, progressive neurological deterioration, and cerebral hemispheric atrophy. **Objective:** To describe the clinical course, therapeutic challenges, and outcome of a pediatric patient with Rasmussen's syndrome. **Case Report:** This case describes an 8-year-old female patient with a history of delayed neuropsychomotor development who developed drug-resistant focal epilepsy. The diagnosis of RS was confirmed following clinical and imaging investigations. Functional hemispherectomy was indicated due to refractoriness to medical treatment. The procedure was temporarily postponed due to SARS-CoV-2 infection and was later performed successfully. The patient developed complications postoperatively, including hemodynamic instability, electrolyte disturbances, diabetes insipidus, respiratory infections, and acute kidney injury. Complete seizure control was achieved after approximately 65 days of hospitalization. **Results:** This case highlights the complexity of managing RS in pediatric patients and emphasizes the importance of an individualized approach. **Conclusion:** Hemispherectomy proved effective in controlling seizures and stabilizing neurological progression.

Keywords: Rasmussen's syndrome; Drug-resistant epilepsy; Hemispherotomy; Autoimmune encephalitis; Pediatric neurology.

[1] Department of Neuropediatrics, Santa Casa de Misericórdia de Belo Horizonte, Minas Gerais, Brazil.

[2] Centro Universitário de Belo Horizonte (UniBH), Belo Horizonte, Minas Gerais, Brazil.

[3] Santa Casa de Misericórdia de Belo Horizonte, Minas Gerais, Brazil.

Associate Editor Responsible:
Cássio da Cunha Ibiapina
School of Medicine of the Universidade Federal de Minas Gerais.
Belo Horizonte/MG, Brazil.

Corresponding author:
André Vinícius Soares Barbosa
Santa Casa de Misericórdia de Belo Horizonte, Minas Gerais, Brazil.
Email: andrevbarbosa@email.com

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RESUMO

Introdução: A Síndrome de Rasmussen (SR) é uma encefalite rara, crônica e progressiva, presumivelmente de origem autoimune, que afeta predominantemente crianças. Caracteriza-se por crises epilépticas focais farmacorresistentes, deterioração neurológica progressiva e atrofia hemisférica cerebral. **Objetivo:** Descrever a evolução clínica, os desafios terapêuticos e o desfecho de uma paciente pediátrica com síndrome de Rasmussen. **Relato de Caso:** Este caso descreve uma paciente do sexo feminino, de 8 anos de idade, com história de atraso no desenvolvimento neuropsicomotor, que evoluiu com epilepsia focal farmacorresistente. Após investigação clínica e por imagem, o diagnóstico de SR foi confirmado. Devido à refratariedade ao tratamento medicamentoso, foi indicada hemisferectomia funcional. O procedimento foi temporariamente adiado devido à infecção por SARS-CoV-2 e posteriormente realizado com sucesso. No pós-operatório, a paciente apresentou complicações, incluindo instabilidade hemodinâmica, distúrbios hidroeletrolíticos, diabetes insipidus, infecções respiratórias e lesão renal aguda. Após aproximadamente 65 dias de internação, foi obtido controle completo das crises. **Resultados:** Este caso evidencia a complexidade do manejo da SR em pacientes pediátricos e reforça a importância de uma abordagem individualizada. **Conclusão:** A hemisferectomia mostrou-se eficaz no controle das crises e na estabilização da progressão neurológica.

Palavras-chave: Síndrome de Rasmussen; Epilepsia farmacorresistente; Hemisferotomia; Encefalite autoimune; Neurologia pediátrica.

Introduction

The diagnosis is based on three main pillars: (1) unilateral cortical inflammation and hemispheric atrophy, (2) drug-resistant epilepsy, and (3) progressive neurological deterioration. The etiology remains unclear, although autoimmune mechanisms involving T-cell-mediated cytotoxicity and microglial activation are implicated. Treatment remains challenging. Immunotherapy may delay progression, but surgery is often required. Hemispherectomy remains the most effective treatment for seizure control, despite functional deficits.

Methods

This study is a retrospective clinical case report based on the analysis of a pediatric patient diagnosed with Rasmussen's Syndrome. Data were collected from medical records, imaging studies, and multidisciplinary evaluations. The study was approved by the Research Ethics Committee (CEP) of Pesquisa Clínica Santa Casa BH (approval no. 7.720.307/2025).

Case Report

An 8-year-old female patient, with no neonatal complications, had a history of delayed neuropsychomotor development (sitting at 12 months, walking at 24 months, and beginning speech around 2 years of age). No specific etiology was identified for the developmental delay. On neurological examination prior to surgery, the patient presented with right-sided hemiparesis, reduced functional autonomy, limited verbal communication, and incomplete sphincter control. At 4 years of age, she began experiencing focal epileptic seizures. Despite treatment with levetiracetam (30 mg/kg/day) and topiramate (5 mg/kg/day), the epileptic seizures remained uncontrolled. The patient also received clobazam, valproic acid, and carbamazepine, without achieving satisfactory seizure control, thus fulfilling the criteria for drug-resistant epilepsy. Multiple focal seizures were observed during hospitalization when the patient was 8 years old, prompting further diagnostic evaluation. Neurologically, she presented with persistent left hemiparesis, compatible with the expected postoperative outcome following right hemispherectomy. She maintained partial functional dependence but was

clinically stable. A cranial computed tomography (CT) scan revealed signs of cerebral hemispheric atrophy, supporting the suspected diagnosis of Rasmussen Syndrome. Brain magnetic resonance demonstrated right hemispheric atrophy, more pronounced in the mesial temporal region, with signal alteration in the right hippocampus, suggestive of mesial temporal sclerosis (Figure 1).

During hospitalization, the patient was transferred to the pediatric intensive care unit due to refractory seizures, where she developed status epilepticus, requiring invasive ventilatory support and continuous sedation with midazolam. After clinical stabilization, she was referred to a tertiary center. Following the recurrence of seizures, video-electroencephalography (VEEG) demonstrated epileptiform discharges predominantly in the right hemisphere, associated with unilateral slowing of background activity, consistent with the epileptogenic focus. These findings were consistent with the MRI abnormalities, reinforcing the anatomo-electroclinical correlation.

Due to refractoriness to medical treatment, the case was discussed at a multidisciplinary meeting, and a functional hemispherectomy was indicated. The procedure was temporarily postponed due to SARS-CoV-2 infection and was later performed successfully. The postoperative period was marked by complications, including hemodynamic instability, electrolyte disturbances, diabetes insipidus, respiratory infection, and acute kidney injury, which were managed with intensive multidisciplinary care. Despite these complications, the patient showed progressive clinical improvement, with stabilization of metabolic parameters and resolution of infectious conditions. Postoperative imaging demonstrated the expected findings after right functional hemispherectomy, with no evidence of acute complications (Figure 2).

At hospital discharge, the patient remained free of epileptic seizures. She was discharged on anti-seizure medications, including levetiracetam and topiramate, with outpatient follow-up scheduled.

Discussion

Rasmussen's Syndrome (RS) is a rare, chronic, and progressive encephalitis, presumably of autoimmune origin, that primarily affects children and adolescents. It is characterized by focal epileptic seizures, progressive neurological deterioration, and cerebral hemispheric atrophy evident on imaging studies[1-8].

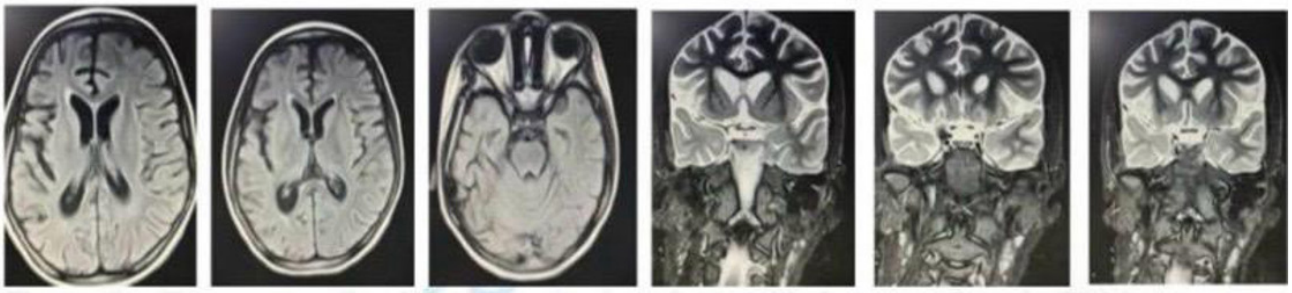


Figure 1. Preoperative brain magnetic resonance imaging (MRI), T2-weighted/FLAIR sequences, demonstrating right hemispheric atrophy with volume reduction and signal alteration in the right hippocampus, suggestive of mesial temporal sclerosis.

Caption: Preoperative imaging study performed on November 23, 2024.
Source: Authors; Research data (2025).

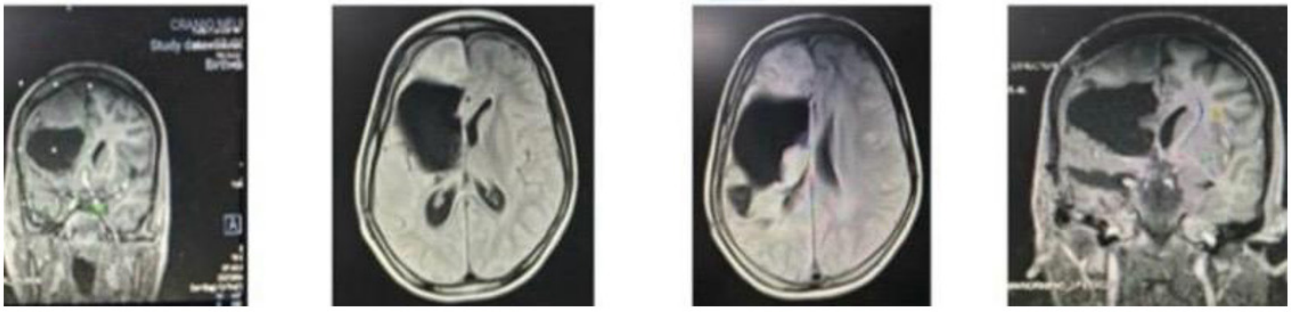


Figure 2. Postoperative brain magnetic resonance imaging (MRI), including T2-weighted and FLAIR sequences, demonstrating findings consistent with right functional hemispherectomy, characterized by an extensive surgical cavity in the right cerebral hemisphere, ipsilateral cerebral volume loss, and compensatory dilatation of the right lateral ventricle, with no evidence of acute postoperative complications.

Caption: Postoperative imaging studies performed on March 27, 2025.
Source: Authors; Research data (2025).

The clinical case presented describes a child with a history consistent with the development of RS, including delay in neuropsychomotor development and early onset of focal epilepsy, progressing to seizures that were drug-resistant to conventional medical treatment. The diagnosis was confirmed based on the combination of clinical presentation, imaging findings, and epileptogenic activity identified on video-electroencephalography.

This case fulfills the three main diagnostic pillars of Rasmussen's encephalitis[5]: (1) unilateral cortical inflammation and hemispheric atrophy demonstrated on neuroimaging, (2) drug-resistant epilepsy, evidenced by failure of multiple anti-seizure medications, and (3) progressive neurological deterioration, observed through the patient's clinical evolution.

Additionally, there was clear concordance between electrophysiological and neuroimaging findings, with VEEG demonstrating epileptiform activity and unilateral slowing in the right hemisphere, corresponding to MRI findings of right-hemispheric atrophy and mesial temporal involvement. This anatomo-electroclinical correlation further supports the diagnosis and reinforces the lateralization of the epileptogenic zone.

Functional hemispherectomy is considered the treatment of choice in cases of drug-resistant epilepsy associated with RS, aiming to eliminate epileptic seizures and halt the progression of neurological dysfunction[9]. In the present case, the patient underwent the procedure after careful multidisciplinary evaluation, an approach considered essential in patients with progressive neurological impairment.

Although technically successful, the postoperative course was marked by clinical complications, including electrolyte disturbances, respiratory infections, hemodynamic instability, diabetes insipidus, and acute kidney injury. Although significant, these complications are recognized in major neurofunctional procedures and should be interpreted in the context of the severity of the underlying disease.

Despite these postoperative complications, the patient showed a favorable clinical course, with a marked reduction in seizure frequency and progressive clinical stabilization, indicating a satisfactory response to surgical treatment. Although complete seizure freedom may be achieved in some patients, this outcome is not universal, and a substantial reduction in seizure frequency and severity is already considered a highly relevant therapeutic result. In the present case, the absence of new seizures during the follow-up period reinforces the clinical benefit of the surgical intervention.

From a clinical perspective, this case highlights the importance of early recognition of the diagnostic triad and timely surgical intervention, which may significantly impact long-term neurological outcomes and quality of life.

Finally, this report contributes to the understanding of a rare neurological condition, emphasizing the diagnostic challenges and the need for an individualized, multidisciplinary therapeutic approach.

Conclusion

This case report highlights the complexity of managing Rasmussen's Syndrome (RS) in pediatric patients, reinforcing the importance of a multidisciplinary approach combined with careful surgical planning, especially in clinical scenarios marked by significant comorbidities, such as active infections.

The decision to postpone hemispherectomy due to COVID-19 infection proved to be appropriate, ensuring greater safety during the perioperative period.

The favorable clinical response following the surgical procedure confirms the effectiveness of hemispherectomy in cases of drug-resistant epilepsy, supporting its role as a key therapeutic option in selected patients.

Although it is a high-risk intervention, hemispherectomy can be a viable therapeutic alternative in selected cases to control epileptic seizures in patients with severe unilateral brain damage. Available treatment options for chronic encephalitis with epilepsy, including anticonvulsant medications, immunosuppressants, and plasmapheresis, have variable outcomes depending on the individual. Therefore, treatment should be individualized.

Finally, this report contributes to the understanding of a rare and unpredictable neurological condition, highlighting the diagnostic and therapeutic challenges involved. Further research aimed at elucidating disease mechanisms, identifying reliable biomarkers, and developing more effective therapies is essential to improve outcomes and quality of life for affected patients.

Author's Contributions

We describe contributions to the paper using the taxonomy (CRediT) provided above:

Conceptualization, Investigation, Methodology, Visualization & Writing - review & editing: André Vinícius Soares Barbosa; Luiza Sousa Vilano. *Project administration, Supervision & Writing - original draft*: André Vinícius Soares Barbosa; Luiza Sousa Vilano. *Data curation & Formal Analysis*: Arthur Andrade Resende; Clarissa Milbratz de Castro. *Visualization*: Patrícia Laura de Lacerda Viggiano; Vivian Proença Lara. *Validation*: Vivian Proença Lara. *Resources & Funding acquisition*: Not applicable.

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