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(CASE REPORT)

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Adult-onset Rasmussen encephalitis: A late presentation case report

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Abstract

Rasmussen's encephalitis is a chronic neuroinflammatory disease predominantly affecting one cerebral hemisphere, with an unknown etiology but associated with potential autoimmune and infectious mechanisms. Although its incidence is highest between 6 and 10 years of age, 10% of cases occur in adults, as demonstrated by the reported case of a 39-year-old male. This progressive inflammatory condition is characterized by cerebral volume reduction, a crucial finding in the differential diagnosis of neurological disorders. Magnetic resonance imaging plays a fundamental role in its diagnosis. This case highlights the clinical and imaging features of the disease in adult patients, an uncommon but clinically relevant presentation for medical practice.

Keywords: Rasmussen Encephalitis; Encephalitis; Seizures; Hemiparesis

1. Introduction

Rasmussen's encephalitis, or chronic focal encephalitis, is a rare neuroinflammatory disorder characterized by a progressive inflammatory process predominantly affecting one cerebral hemisphere. Its pathophysiology includes unilateral hemispheric atrophy, drug-resistant focal epilepsy often in the form of epilepsia partialis continua, cognitive decline, progressive hemiparesis, and irreversible neurological deficits [1,2,4].

Diagnosis relies on clinical features such as intractable focal seizures and progressive neurological deficits, supported by neuroimaging showing unilateral cortical atrophy. EEG typically reveals localized epileptiform discharges, and histopathology may show lymphocytic infiltration and neuronal loss [16].

While its exact etiology remains unknown, autoimmune mechanisms are considered the primary contributors [3]. These include the presence of antibodies against neuronal glutamate receptors (anti-GluR3) and glutamic acid decarboxylase (anti-GAD), as well as cytotoxic T lymphocytes targeting neurons and astrocytes [5-6]. Hypotheses involving neurotropic viral infections have also been explored but lack definitive confirmation [6].

Clinically, Rasmussen's encephalitis follows a progressive course with three main stages [5]:

- **Prodromal stage** Lasting an average of 7 months, it is marked by sporadic focal seizures and mild symptoms such as intermittent hemiparesis [1,5].
- **Acute stage** Occurring over 8 months, it presents with frequent seizures, progressive hemiparesis, hemianopia, cognitive decline, and, in cases involving the dominant hemisphere, severe aphasia [1,5].
- **Residual stage** Characterized by hemispheric atrophy and irreversible neurological damage, including significant functional impairment and severe cerebral atrophy [1,5].

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Demographically, the disease primarily affects children, peaking between 6 and 10 years of age. However, up to 10% of cases occur in adults, where diagnosis is more challenging due to its low prevalence and heterogeneous clinical presentation [5,7]. Adult-onset cases tend to have a slower clinical course and less severe cognitive impairment compared to pediatric cases. Magnetic resonance imaging (MRI) is the primary diagnostic tool, enabling visualization of progressive hemispheric atrophy, white matter abnormalities, and reduced cerebral blood flow. Clinical correlation is essential for diagnosis.

2. Materials and Methods

This article presents the case of an adult patient with Rasmussen's encephalitis, emphasizing clinical features, imaging findings, and challenges in managing this condition in adults.

3. Case Report

We report the case of a 39-year-old male construction worker from the north of Ecuador (Tulcán). Ecuador, with no significant medical history. The patient presented with a sudden onset of speech difficulties and right hemiparesis 6 months ago, followed by two seizure episodes. Upon hospitalization, he reported fever and headache lasting one week. Blood tests, including CBC, biochemistry, liver function, coagulation times, HIV, VDRL, electrolytes, TSH, and serology for hepatitis B, hepatitis C, and Epstein-Barr virus, were normal. Cerebrospinal fluid (CSF) analysis revealed elevated protein levels without glucose level change; cytological and bacteriological studies were normal. An initial cranial CT showed ventricular dilation without clear evidence of atrophy [Figure1-2].

Subsequent MRI, performed using a 1.5 Tesla superconducting system, revealed diffuse hyperintensity in the white matter (T2 and FLAIR), particularly in the left temporal, insular, and parietal lobes, as well as the corpus callosum [Figure 3]. There was generalized gray matter atrophy of the left hemisphere with associated ventricular retraction. These findings were consistent with autoimmune encephalitis, particularly Rasmussen's encephalitis.

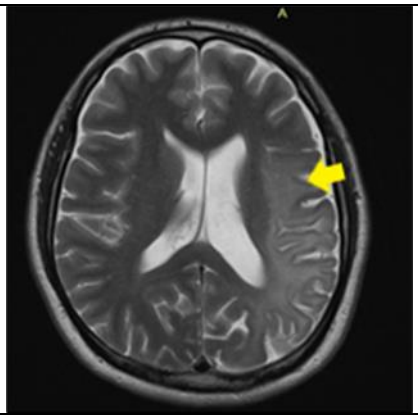

Figure 1 Axial T2: A reduction in volume and hyperintensity is noted in the left parietal subcortical gray and white matter

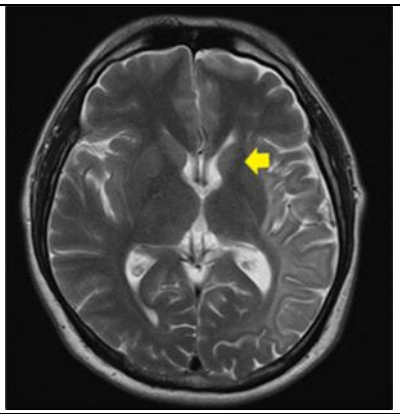

Figure 2 Axial T2: Atrophy of the ipsilateral caudate nucleus is observed at the basal ganglia section

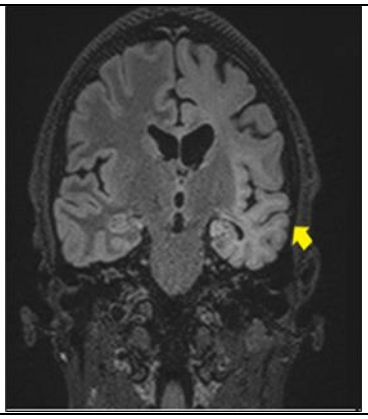

Figure 3 Axial FLAIR T2: Diffuse involvement is observed at the basal left temporal level, extending beyond the arterial territory

The patient exhibited persistent aphasia and partial right hemiparesis with recurrent seizures responsive to valproic acid. Due to suspected autoimmune etiology, the patient received five pulses of systemic corticosteroids, followed by oral therapy.

4. Discussion

Rasmussen's encephalitis (RE) is a rare disorder primarily affecting children, characterized by chronic, progressive inflammation of one brain hemisphere. Adult-onset RE (LORE) is even rarer and presents unique challenges in diagnosis and management due to its distinct clinical and pathological features. Unlike pediatric RE, LORE typically has a slower progression and a better overall prognosis. While focal epilepsy is the most common manifestation, atypical symptoms can complicate early recognition [8].

In the presented case, the adult patient exhibited an unusually aggressive course, with MRI revealing rapid hemispheric atrophy and ipsilateral ventricular enlargement. This imaging pattern is critical for distinguishing LORE from conditions like multiple sclerosis, Alzheimer's disease, Parkinson's disease, Huntington's disease, chronic alcoholism, or normal aging, all of which typically show bilateral cerebral volume loss and more generalized symptoms affecting cognition, motor function, and emotion [9,10,11].

Diagnostic precision is essential in LORE. While MRI remains a cornerstone, recent evidence supports the utility of PET/CT with 18F-FDG in detecting early disease changes. This technique may help identify metabolic abnormalities before structural damage becomes evident, underscoring the need for further research into its role in diagnosis [8].

Treatment strategies for LORE differ from pediatric cases and must account for the disease's slower progression. Immunotherapy is the mainstay, with corticosteroids being the most robust first-line treatment. Intravenous immunoglobulin (IVIG) may serve as an adjunct in certain cases. [12] For refractory disease, second-line agents like azathioprine, adalimumab, and rituximab have demonstrated efficacy in controlling seizures and slowing progression. In this case, valproic acid was successfully used for initial seizure management, while definitive therapy continues to be tailored to the patient's evolving needs [13,14,16].

Clinically, the patient presented with refractory focal seizures with a predominant motor component and progressive unilateral neurological deficits, including hemiparesis. These findings fulfill two of the three Group A diagnostic criteria for RE. Although epilepsia partialis continua (EPC)—a hallmark feature of RE—was absent, its absence does not rule out the diagnosis [3].

Histopathological confirmation, a key diagnostic criterion, was not available due to the invasive nature of brain biopsy. Ideally, a biopsy would reveal chronic encephalitis with CD8+ T-cell infiltration and microglial activation. Nevertheless, the combination of clinical findings, imaging results, and therapeutic response supported a working diagnosis of RE [15].

Adult-onset RE often progresses more subtly than pediatric cases, complicating early diagnosis. This case underscores the importance of integrating neuroimaging findings and therapeutic responses into the diagnostic framework, particularly when classical features like EPC or characteristic EEG findings are absent. The patient's favorable response to corticosteroid therapy highlights the immunopathological basis of RE and the potential for effective intervention when the disease is recognized promptly.

5. Conclusion

In conclusion, this case illustrates the diagnostic and therapeutic challenges associated with adult-onset Rasmussen's encephalitis. While not all diagnostic criteria were met, the constellation of clinical, imaging, and therapeutic findings strongly supports the diagnosis. This report highlights the need for heightened clinical suspicion, a multidisciplinary approach, and potentially revised diagnostic criteria to account for atypical presentations in adults. Further research into the pathophysiology and management of LORE is essential to improve outcomes for this rare condition.

Compliance with ethical standards

Disclosure of conflict of interest

No conflict of interest to be disclosed.

Statement of ethical approval

Ethical approval was obtained

Statement of informed consent

Informed consent was obtained from all individual participants included in the study.

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