ADULT-ONSET RASMUSSEN ENCEPHALITIS MIMICKING CHRONIC CEREBRAL INFARCT – A CASE REPORT

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ABSTRACT

Background: **Rasmussen Encephalitis (RE) is a rare, chronic, immune-mediated inflammatory disease of the brain that results in progressive unilateral cerebral atrophy, severe neurological deterioration, and drug-resistant focal seizures. Although classically described in children, particularly those under the age of ten, a small but growing number of adult-onset cases have been documented in recent years, with a notable predominance among females.**

Neuroimaging plays a pivotal role in the diagnosis and differentiation of RE from other unilateral cerebral pathologies. Given the scarcity of imaging mimics and the significant therapeutic implications, it is essential that radiologists and clinicians recognize its characteristic imaging patterns to facilitate early and accurate diagnosis, thereby preventing delays in the initiation of appropriate treatment.

Case Summary: **We present a case of Rasmussen Encephalitis in an adult female, which was initially misdiagnosed as a chronic cerebral infarct on magnetic resonance imaging (MRI). A detailed review of subsequent imaging revealed typical features of RE, including progressive unilateral cortical atrophy and signal changes confined to one cerebral hemisphere.**

Conclusion: This case highlights the diagnostic challenge of adult-onset RE, which may mimic chronic vascular insults on imaging. Awareness of its distinctive radiological features and characteristic electroencephalogram (EEG) abnormalities are crucial for early and accurate diagnosis. Early recognition also allows for timely institution of appropriate therapy, which may slow disease progression, improve seizure management, and preserve neurological dysfunction.

Keywords: Rasmussen, MRI, EEG, Seizure, Epilepsia partialis continua

INTRODUCTION

Rasmussen Encephalitis (RE) is a rare chronic immune-mediated neurological disease characterized by progressive unilateral hemispheric atrophy, intractable focal seizures, and gradual neurological deterioration[1]. First described by the neurosurgeon, Theodore Rasmussen, and his colleagues in 1958[1–3] , since then, several cases of RE have been reported.

Classically described as childhood encephalopathy with average age of disease manifestation been at 6 years[4], multiple case reports of adolescent and adult-onset RE have been published in literature casting doubts on the exclusivity of RE as a childhood disease[5].

The European association in 2005 gave a consensus on the pathogenesis, diagnostic criteria, including characteristic imaging findings, and treatment options of RE[2]. These criteria, grouped into two parts (Table 1), remain central to the diagnosis and management of RE and continue to be widely accepted in clinical practice[3].

Despite advances in understanding the clinical evolution and pathobiology of RE, its precise aetiology remains unknown[6]. Current evidence supports an immune-mediated mechanism, although no specific serological or intrathecal biomarkers have been identified[4]. The immune-mediated mechanism of RE perhaps explains the variable clinical response to immunomodulatory therapy[3,5].

We report a case of adult-onset Rasmussen encephalitis in a 68-year-old woman initially managed as having a chronic cerebral infarction. Subsequent clinical reassessment and further imaging evaluation enabled a confident diagnosis of RE. This case underscores the potential for RE to mimic other unilateral cortical disorders—including chronic infarction, Dyke–

Table 1: Diagnostic criteria for RE

Category	Criteria
Part A	**All three required**
Clinical	Focal seizures (with or without epilepsia partialis continua) and unilateral cortical neurological deficits
EEG	Unihemispheric slowing with or without epileptiform activity and unilateral seizure onset
MRI	Unihemispheric focal cortical atrophy, with at least one of the following: • Grey or white matter T2/FLAIR hyperintensity • Hyperintensity or atrophy of the ipsilateral caudate head
Part B	**Two of three required**
Clinical	Epilepsia partialis continua or progressive* unilateral cortical neurological deficits
MRI	Progressive* unihemispheric focal cortical atrophy
Histopathology	T cell dominated encephalitis with activated microglial cells (typically, but not necessarily forming nodules) and reactive astrogliosis. Numerous parenchymal macrophages, B cells or plasma cells or viral inclusion bodies exclude the diagnosis of RE.

** 'Progressive' means that at least two sequential clinical examinations or MRI studies are required to meet the respective criteria. To indicate clinical progression, each of these examinations must document a neurological deficit, and this must increase over time. To indicate progressive hemiatrophy, each of these MRIs must show hemiatrophy, and this must increase over time.Adapted from Bien et al[2].*

Davidoff-Masson syndrome, Sturge–Weber syndrome, hemimegalencephaly, and unihemispheric cerebral vasculitis—and highlights the importance of heightened awareness of this diagnostic challenge among neurologists and neuroradiologists.

Although few cases of childhood RE have been reported from Nigeria[7,8], no adult-onset cases have been documented to the best of our knowledge. This may reflect either extreme rarity adult-onset RE in Nigeria or clinical misdiagnosis as other unilateral cortical disorders, further emphasizing the need for increased recognition of this rare condition.

CASE DESCRIPTION

We present a 68-year-old woman with background systemic hypertension and diabetes mellitus. She reportedly suffered a cerebrovascular event of undocumented subtype approximately five years prior to presentation, resulting in residual right-sided

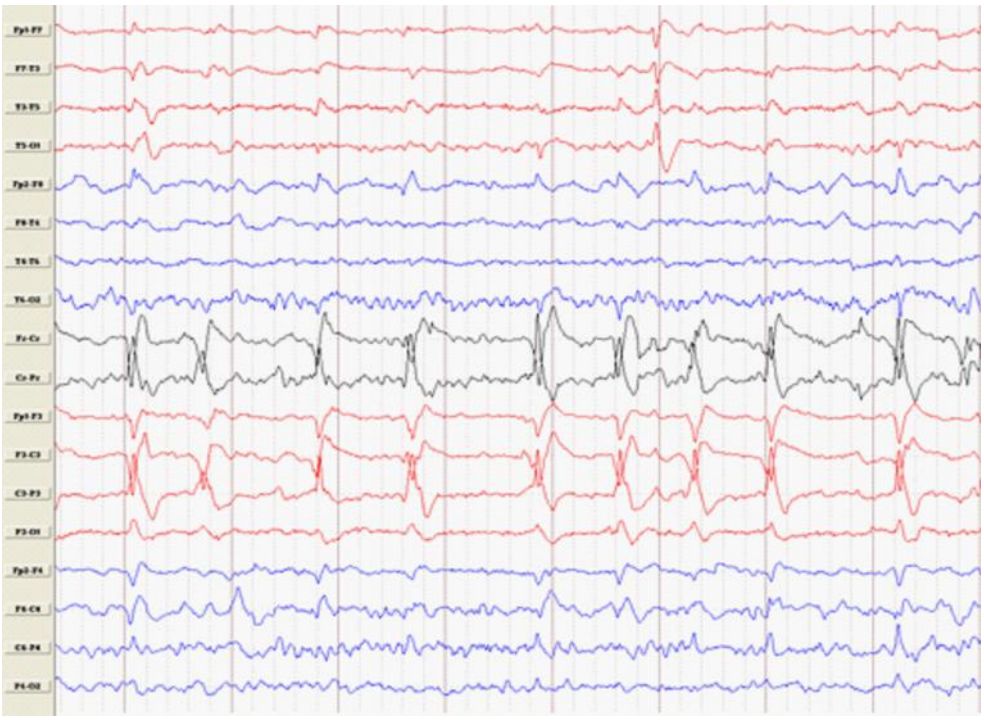

Fig. 1: Electroencephalogram showing left hemispheric slowing, left lateralized periodic discharges, and left mid-temporal focal epileptiform discharges.

Table 2: Differential diagnoses to RE

Condition	Clinical Features	Laboratory/CSF Findings	MRI Findings	EEG Findings
Chronic Infarction	History of stroke; fixed neurological deficit; late-onset focal seizures; no disease progression.	Normal	Encephalomalacia; gliosis; volume loss in vascular territory; ex-vacuo ventricular dilatation.	Focal slowing +/- epileptiform discharges over infarcted cortex.
Dyke-Davidoff-Masson Syndrome	Childhood onset hemiparesis; seizures; intellectual disability; facial asymmetry; symptoms non-progressive after childhood.	Normal	Unilateral cerebral atrophy with ipsilateral ventricular dilatation; calvarial thickening; hyperpneumatized frontal sinus	Focal slowing and epileptiform discharges over atrophic hemisphere.
Sturge-Weber Syndrome	Early-onset seizures; facial port-wine stain; progressive hemiparesis.	Normal	Leptomeningeal enhancement; cortical atrophy; tram-track calcifications.	Focal slowing and epileptiform activity over affected hemisphere.
Unihemispheric Cerebral Vasculitis	Progressive focal seizures; headaches; stepwise or progressive neurological deficits; stroke-like episodes.	Elevated ESR/CRP; CSF pleocytosis and elevated protein; autoimmune markers may be present	Patchy cortical-subcortical infarcts; gyriform or leptomeningeal enhancement; progressive unilateral atrophy in chronic cases	Focal or hemispheric slowing; epileptiform discharges corresponding to ischemic regions.
Unihemispheric Cerebral Vasculitis	Progressive focal seizures; headaches; stepwise or progressive neurological deficits; stroke-like episodes.	Elevated ESR/CRP; CSF pleocytosis and elevated protein; autoimmune markers may be present	Patchy cortical-subcortical infarcts; gyriform or leptomeningeal enhancement; progressive unilateral atrophy in chronic cases	Focal or hemispheric slowing; epileptiform discharges corresponding to ischemic regions.
Hemimegalencephaly Cerebral Vasculitis	Infancy onset; severe epilepsy; developmental delay; congenital hemiparesis	Normal	Enlarged hemisphere; abnormal gyration; thickened cortex; ventricular enlargement; static malformation	Markedly asymmetric background; multifocal discharges from malformed hemisphere
Infectious Chronic Encephalitis (e.g HSV, SSPE)	Seizures with cognitive decline; systemic prodrome; myoclonus (SSPE).	CSF pleocytosis; elevated protein; viral PCR or antibody positivity	Cortical/subcortical signal change; often bilateral; variabe atrophy	Diffuse slowing; periodic complexes (SSPE)

ESR – Erythrocyte Sedimentation Rate; CRP – C-Reactive Protein; CSF – Cerebrospinal Fluid; HSV –Herpes Simplex Virus; SSPE - Subacute Sclerosing Panencephalitis; PCR-Polymerase Chain Reaction.

hemiparesis. Details of the acute management and neuroimaging from that period were unavailable for review.

Approximately two years after the presumed stroke, she developed recurrent seizures. Her typical seizures started as focal motor seizures with impaired awareness which evolved to bilateral tonic-clonic seizures. The semiology was as follows: no aura; leftward neck version; tonic posturing of the upper limbs; and then generalized convulsions. Each seizure typically lasted a few minutes and was associated with impaired consciousness. She had amnesia for each event and had a post-ictal somnolent state lasting hours.

Initially, her seizure frequency was 1-2 per year. However, about a year before the index presentation, seizure frequency increased to 3-4 per year despite adherence to antiseizure medications (ASMs); she also had a flurry of seizures in the weeks preceding the evaluation. Further, her seizure semiology also became more varied, with development of several episodes of right-sided epilepsia partialis continua and new focal motor seizures with preserved awareness. Indeed, the reason for a neurological consultation was the recent increase in seizure frequency, as well as change in the seizure semiology. She had suffered no falls or head injury, and had no headaches, fever, or features of meningism. The clinical diagnosis after initial review was post-stroke epilepsy with a suspicion of a (new) structural/metabolic insult causing the new seizure types. To this end, a metabolic panel, complete blood count, electroencephalogram (EEG), and brain magnetic resonance imaging (MRI) scan were requested.

Her metabolic panel which included blood glucose, glycated hemoglobin, renal function, and liver function tests were all essentially normal, as was her complete blood count. The EEG showed left hemispheric slowing, left lateralized periodic discharges (LPDs), and left mid-temporal focal epileptiform discharges distinct from the LPDs (Figure 1).

Brain MRI (using SIGNA™ Creator GE 1.5T MRI scanner) revealed left unihemispheric cerebral atrophy involving the ipsilateral head of caudate nucleus, thalamus, and cerebral peduncle, with associated ex-

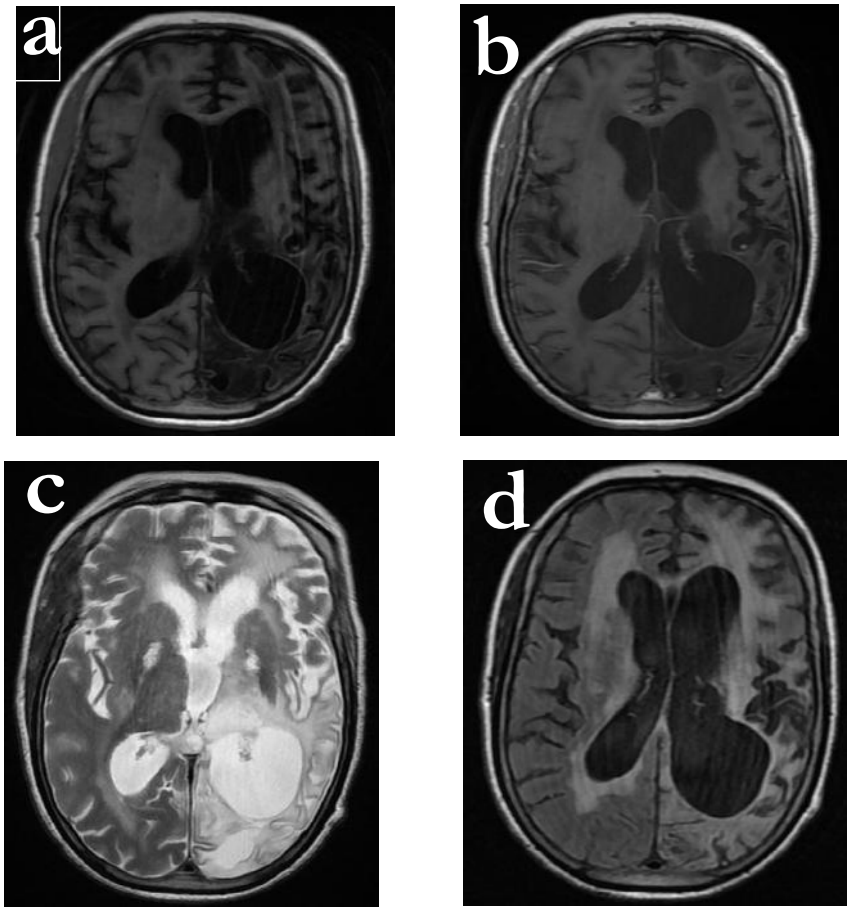

Fig.2 (a-d): Axial T1W, T1W+c, T2W and FLAIR brain MRI showing severe left unihemispheric cerebral atrophy involving the ipsilateral head of caudate nucleus, thalamus, and cerebral peduncle, with associated ex-vacuo dilatation of all components of the ipsilateral lateral ventricle and the third ventricle. No significant area of contrast enhancement.

vacuo dilatation of all components of the ipsilateral lateral ventricle and the third ventricle. No significant area of contrast enhancement was noted (Figure 2).

Given the progressive seizure burden, evolving semiology, EEG abnormalities, and characteristic imaging findings, a diagnosis of adult-onset Rasmussen encephalitis was strongly suggested. The previous attribution of her epilepsy to a cerebrovascular event was considered unlikely, and metabolic causes for the new seizure types were excluded.

The patient and her relatives were counselled regarding the revised diagnosis and the recommended diagnostic and therapeutic interventions. However, given her age, chronic comorbidities, and established neurological deficits, immunosuppressive or surgical options were not pursued. Instead, management focused on seizure control and quality of life. Her existing antiseizure regimen of levetiracetam and sodium valproate were up titrated, resulting in significant reduction in seizure frequency, and was scheduled for follow-up, and supportive rehabilitation.

DISCUSSION

Rasmussen Encephalitis (RE) was first described by Rasmussen *et al.* in 1958 as a syndrome of progressive focal epilepsy, hemiparesis, and cognitive decline with unilateral cerebral atrophy[1–3]. Initially, a viral aetiology was suspected, but mounting evidence now supports a T-cell–mediated immune mechanism causing neuronal destruction[2]. Despite this understanding, the specific triggers remain unidentified[3].

Although typically a childhood disease, adult-onset RE has been increasingly recognized. Castellano *et al.*[5] reported three adult female patients aged 44–56 years, while Dupont *et al.*[4] identified 102 cases of late-onset RE, predominantly female (M:F = 4:1). The current case—a 68-year-old woman—fits this demographic trend. Villani *et al.*[9] also observed left hemispheric predominance in adult RE, consistent with our patient's MRI findings.

The 2005 European Consensus diagnostic criteria remain the cornerstone for diagnosis (Table 1). Part A includes: (1) clinical features—focal seizures or *epilepsia partialis continua* with progressive unilateral deficits; (2) EEG showing unilateral slowing or epileptiform discharges; and (3) MRI demonstrating progressive unilateral atrophy with cortical and subcortical T2/FLAIR hyperintensities[2,10]. Our patient fulfilled all three: recurrent focal seizures with *epilepsia partialis continua*, left-lateralized EEG abnormalities, and unilateral hemispheric atrophy.

Adult-onset RE may mimic other unilateral cortical disorders such as chronic infarction, Dyke–Davidoff–Masson syndrome, Sturge–Weber syndrome, hemimegalencephaly, unihemispheric cerebral vasculitis, and infectious chronic encephalitis. Differentiation is aided by the progressive nature of cortical atrophy, evolution of seizure semiology, typical radiologic findings, and characteristic EEG localization (Table 2). Post-stroke epilepsy was initially suspected in this patient due to her history, but the clinical progression and imaging findings were more consistent with RE. The chronic progressive course of RE results from sustained immune-mediated neuronal loss leading to hemispheric atrophy and neurological decline. Histopathological examination—though not performed in this case—typically shows cortical neuronal loss, microglial nodules, perivascular lymphocytic infiltration, and T-cell–mediated cytotoxicity[3].

Differentiating RE from stroke-related epilepsy is essential. While both can manifest with focal seizures and unilateral findings, RE demonstrates ongoing seizure evolution, progressive atrophy across multiple regions, and EEG features consistent with continuous hemispheric irritability. Chronic infarction, by contrast, leads to static neurological deficits without progressive imaging changes.

In the present case, the patient's EEG demonstrated characteristic left hemispheric slowing and LPDs, while MRI revealed global left-sided atrophy involving cortical and subcortical structures, extending to the thalamus and cerebral peduncle. These findings collectively fulfill the criteria for RE and exclude other aetiologies[2].

The pathophysiological mechanism of hemispheric atrophy in RE involves immune-mediated inflammation with activated cytotoxic T lymphocytes and microglia infiltrating cortical neurons. This results in progressive tissue loss and secondary ventricular dilatation[2,3]. While paediatric RE often presents with severe cognitive and motor decline, adult forms may have a more indolent course with predominant seizure manifestations and slower progression[5].

Therapeutically, treatment is directed toward halting the inflammatory process and controlling seizures[3,5]. Immunomodulatory therapies—including corticosteroids, intravenous immunoglobulin (IVIG), plasmapheresis, and immunosuppressive agents such as tacrolimus or azathioprine—have shown varying efficacy in slowing progression, particularly when instituted early[3–5]. Hemispherectomy remains the definitive treatment in paediatric RE with intractable

seizures, though outcomes in adults are less favourable due to established hemispheric dominance and comorbidities[5]. In elderly patients, therapy is largely conservative, focusing on optimizing seizure control and functional stability. The index patient achieved significant reduction in seizure frequency upon up-titration of her current antiseizure regimen, while awaiting her consent to proceed with further diagnostic and therapeutic interventions.

Adult-onset RE poses diagnostic and therapeutic challenges. Clinicians must maintain high suspicion when evaluating older patients with new or evolving focal seizures, particularly those with unilateral cortical atrophy unexplained by vascular or neoplastic causes. Early recognition is crucial, as timely initiation of immunotherapy may delay or attenuate disease progression.

CONCLUSION

Although traditionally considered a childhood disorder, RE can present in adults and the elderly, as shown by this 68-year-old patient. Diagnosis relies on recognizing progressive focal seizures, epilepsia partialis continua, unilateral cortical deficits, characteristic EEG abnormalities, and progressive unihemispheric atrophy on MRI. This case fulfills established diagnostic criteria and demonstrates left hemispheric involvement in late-onset disease. Awareness of these features is crucial to distinguish RE from stroke-related epilepsy and other unilateral cortical disorders.

ADDENDUM

Conflict of Interest:

The authors declare no conflict of interest.

Contribution of Authors:

Conceptualization: SJA and AIM; Methodology: SJA and AIM; Formal analysis and investigation: SJA and AIM; Writing - original draft: SJA, MML and OIL; Writing - review and editing: SJA and AIM; Resources: AIM and SJA; Supervision: AIM.

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