

Rasmussen Encephalitis In A Nigerian Child: A Case Report

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Citation

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Abstract

INTRODUCTION Seizure disorder may be caused by a variety of medical conditions. **CASE PRESENTATION** We report a case of Rasmussen encephalitis (RE) in a 7-year-old boy of the Yoruba ethnic group in Nigeria. This to the best of our knowledge is the first reported case of this condition in a Nigerian. He had computed tomography (CT) and magnetic resonance imaging (MRI) scan of the brain and electroencephalography (EEG). **CONCLUSION** We want to emphasize the importance of imaging in the diagnosis of seizure disorder, especially when it is intractable. To diagnose RE, the index of clinical suspicion has to be high. It is probable that some cases of this condition will be missed in our environment as a result of inadequate imaging, stemming from availability and cost considerations. RE is extremely rare and many radiologists practicing in sub-Saharan Africa may not see a single case of RE in their practice life as a result of the paucity of MRI facilities (which is required for diagnosis) in this sub-region.

INTRODUCTION

Epilepsy refers to recurrent paroxysmal episodes of central nervous system dysfunction manifested by stereotyped alterations in behavior [1]. This disorder of brain function may be secondary to a variety of pathological processes such as trauma, tumour, infections and inflammations, storage diseases, hypoglycemia, hypocalcemia, hyponatremia, renal or hepatic failure, infarction, haemorrhage, drugs and toxins [2]. The aetiology of seizure is however idiopathic in about 50% of epileptics [1]. Rasmussen was the first author to report Rasmussen encephalitis (RE) or Rasmussen disease (Rasmussen syndrome) in 1959, and the syndrome consists of severe epilepsy associated with slow progressive neurologic deterioration and progressive lateralized brain dysfunction [3]. RE, now known as chronic focal encephalitis is extremely rare, and about half of the cases are preceded by an inflammatory episode such as tonsillitis, upper respiratory tract infection, and otitis media [4]. The clinical manifestation of RE include epilepsia partialis continua (EPC), progressive hemiparesis, and neuropsychological deterioration [5]. RE usually occurs in children and is often unilateral [6]. However it may be bilateral where it has a poor prognosis and is usually fatal [6]. The oldest confirmed case of RE in the medical literature was reported by Hunter et al in a 58-year-old female in Canada [7]. The occurrence of both RE and Behcet

disease in first degree relatives has raised the possibility of an autoimmune pathogenesis in both conditions [8]. Brain MRI and CT scans, as well as EEG are helpful in making the diagnosis of RE. It has however been suggested that magnetic resonance spectroscopy (MRS) can identify and quantify neuronal damage and loss in the affected cerebral hemisphere of patients with RE, including areas that appear normal on conventional MRI [9].

CASE PRESENTATION

JF is a 7-year-old abandoned child under the care of the state social services, having been brought into care in December 2009. He presented to the Neurology Unit of the Paediatrics Department of the Lagos State University Teaching Hospital Ikeja, Lagos, Nigeria, with a one week history of recurrent, focal tonic-clonic seizures, each of which lasted about three minutes. Seizure onset was sudden and frequency was about 1-2 for the day, occurs on alternate days and with consistency of nature. There was no preceding aura, automatic behavior, or associated loss of consciousness. Post-ictal sleep does not occur, but there was weakness after seizures which last for some minutes. There was no loss of continence. Antenatal, perinatal and postnatal history were absent on account of the child's status. He however recalls having been well with his faculties and abilities intact until a few months before coming into foster care, when his father made efforts to seek medical care for the weakness of the

right side of his body which was of recent origin. Examination revealed no dysmorphic facies. His head circumference was 51.3cm, which is appropriate for his age. The head shape and size are normal. His fontanelles were closed and there were no overriding sutures. Vision and hearing were intact but speech is incoherent and his words were not clear. Other cranial nerves were normal. He had a hemiplegic gait and a hemiatrophy of the right upper and lower limbs. Power was grade 3 on the right upper limb and 4 on the right lower limb, but a full 5 on the left upper and lower limbs. The muscle tone and tendon reflexes were increased on the right side. There was a flexor posture with internal rotation of the right hand. The cardiovascular, respiratory, gastro-intestinal and genito-urinary systems were grossly intact.

A brain CT scan (Figures 1 to 3) showed asymmetry of the cerebral hemispheres with dilatation of the sulci over the left cerebral hemispheres. The anterior and posterior horns and the atrium and trigone of the left ventricle were also significantly dilated, suggesting features of hemiatrophy involving the left cerebral hemisphere. There were no focal areas of calcification. The right cerebral hemisphere was normal. These findings were also corroborated with the brain MRI scan (Figures 4 to 10) conducted the next day, which also showed atrophy of the head of the left caudate nucleus.

Figure 1

Figure 1: Rasmussen Encephalitis. Axial brain CT scan showing dilated cerebral sulci on the left.

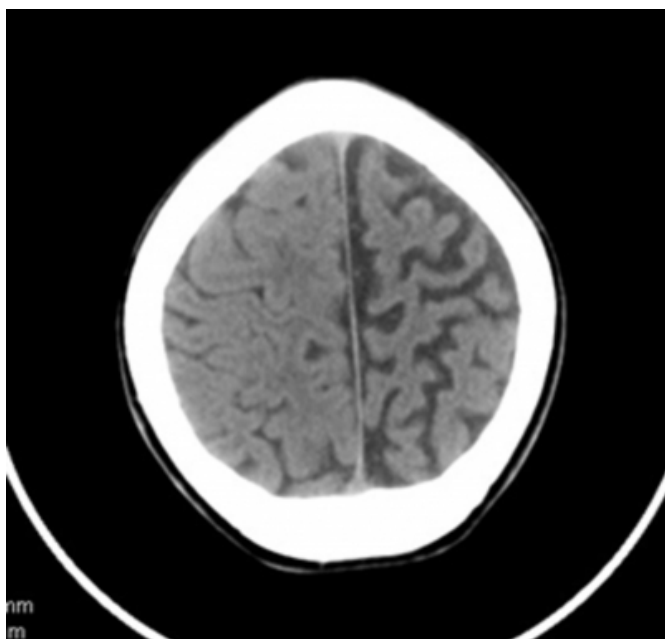


Figure 2

Figure 2: Rasmussen Encephalitis. Axial brain CT scan showing dilated left lateral ventricle and prominent left sulci, in keeping with left hemiatrophy.



Figure 3

Figure 3: Rasmussen Encephalitis. Axial brain CT scan, showing hemiatrophy of the left temporal lobe.



Figure 4

Figure 4: Rasmussen Encephalitis. Axial FSE T2 MRI scan of the brain showing dilated left cerebral sulci and dilated left lateral ventricle.

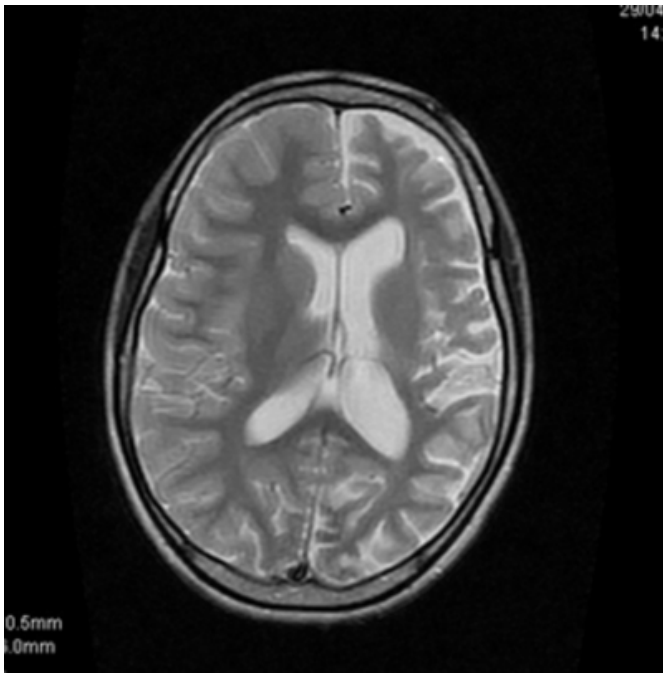


Figure 6

Figure 6: Rasmussen Encephalitis. Axial T1 FLAIR MRI scan of the brain showing prominent sulci, dilated lateral ventricle, and hemiatrophy on the left.

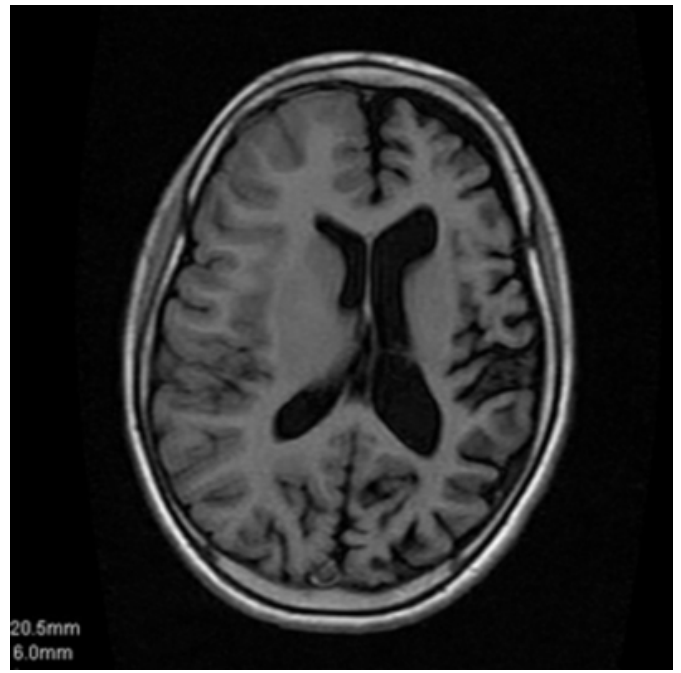


Figure 5

Figure 5: Rasmussen Encephalitis. Axial FSE T2 MRI scan of the brain showing left hemiatrophy.

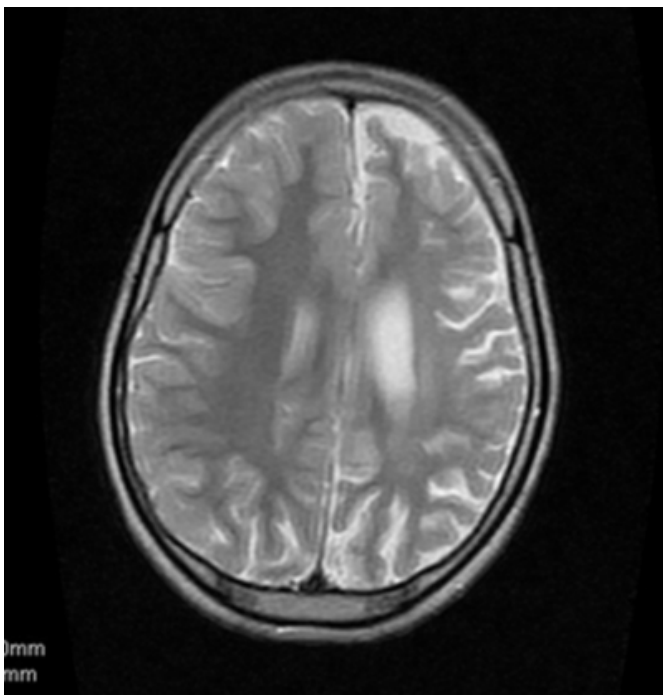


Figure 7

Figure 7: Rasmussen Encephalitis. Axial T1 FLAIR MRI scan of the brain showing dilated sulci and cerebral hemiatrophy on the left.

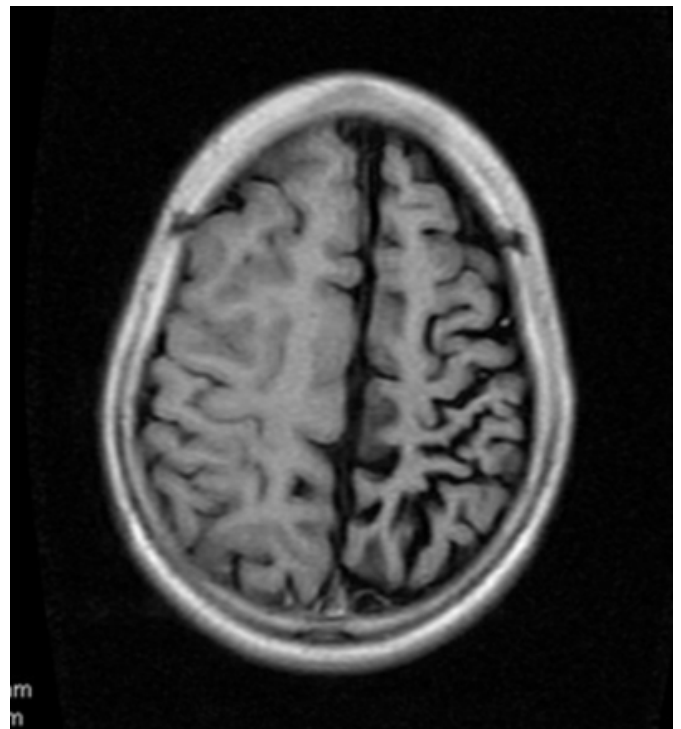


Figure 8

Figure 8: Rasmussen Encephalitis. Sagittal T1 FLAIR MRI scan of the brain showing dilated cerebral sulci on the left.

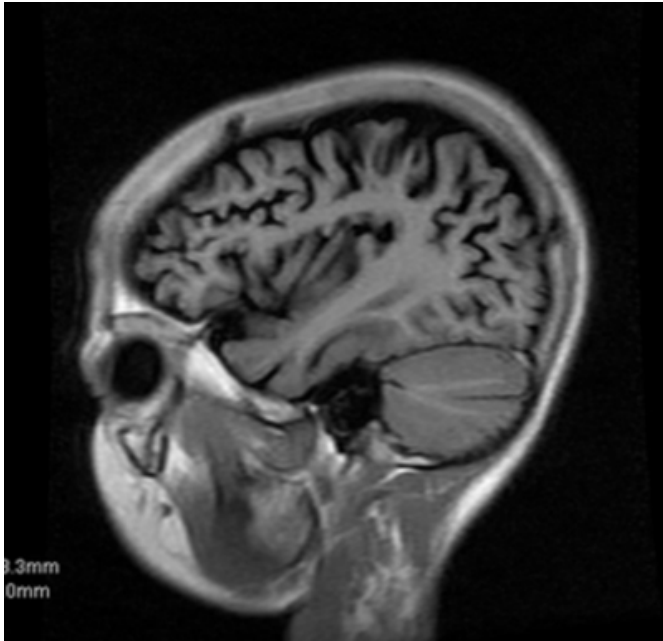


Figure 9

Figure 9: Rasmussen Encephalitis. Axial SE T1 MRI scan of the brain showing dilated left lateral ventricle, dilated left cerebral sulci, and left cerebral hemiatrophy.

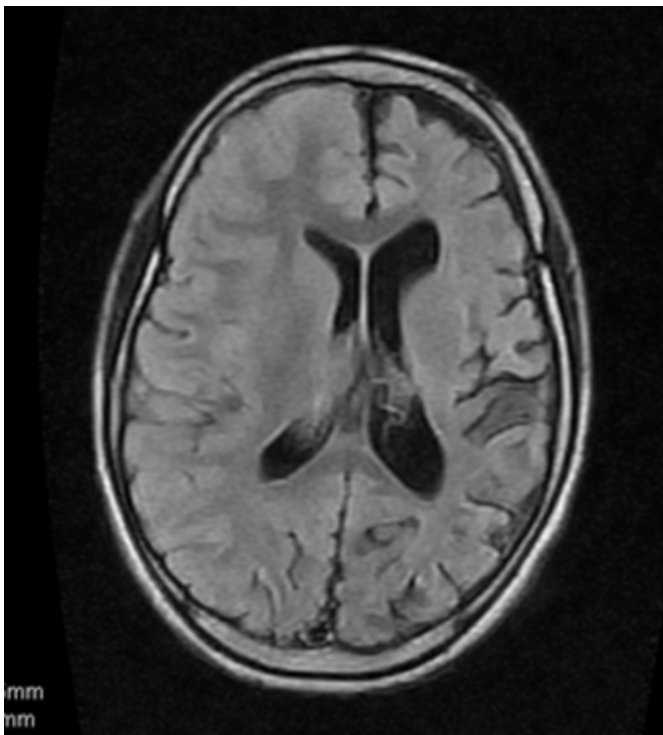
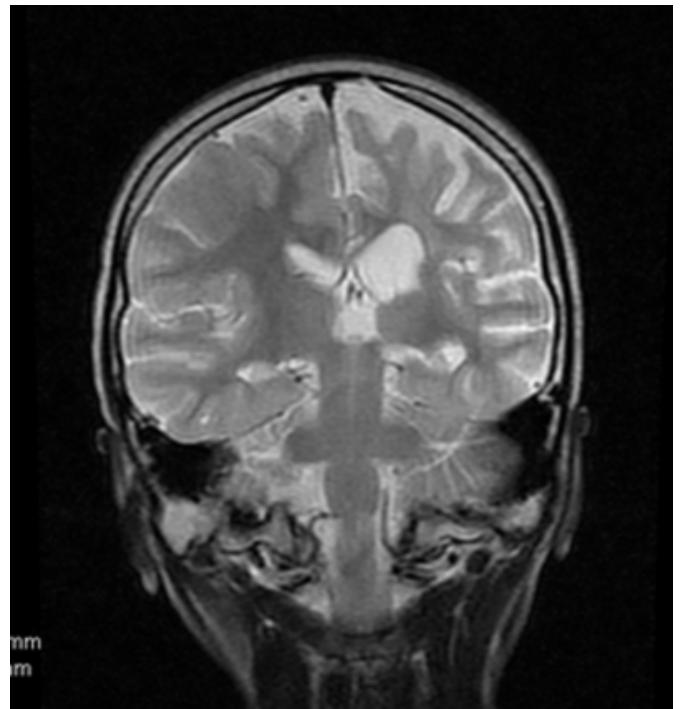


Figure 10

Figure 10: Rasmussen Encephalitis. Coronal FSE T2 MRI scan of the brain showing dilated left lateral ventricle and cerebral sulci on the left.



Electroencephalography revealed moderate voltage record showing mixed activity comprising ill-defined 8-10 Hz alpha rhythm plus 4-7Hz waves and random background delta components at 3Hz frequency. The alpha waves were symmetrical in distribution, better formed and of higher amplitude on the left than the right hemisphere. On a couple of occasions, a 5-6Hz activity stood out of the background features in form of bilateral “bursts”.

His hematological and biochemical parameters were within normal limits. A clinical diagnosis of focal tonic-clonic epilepsy was made secondary to Rasmussen encephalitis, based on the features on CT and MRI scan findings of the brain. He was commenced on oral Carbamazepine-CR. The guardian however noticed an increase in the frequency of his seizure to three times a day everyday despite anticonvulsant therapy. He is currently on outpatient review, and a more aggressive treatment (including surgical intervention) is being entertained.

DISCUSSION

The diagnosis of RE was facilitated in this case by the finding of cerebral hemiatrophy on CT and MRI scans, in the clinical setting of hemiparesis and intractable seizure disorder. MRI has been shown to demonstrate the

progression of RE and may suggest the diagnosis in the early stages, often before the appearance of neurological deficits [10]. Apart from atrophy of the head of the caudate nucleus, MRI may also show associated secondary changes such as atrophy of the contralateral cerebellar hemisphere, the ipsilateral hippocampus, and the brainstem [10]. There are four recognized stages of RE based on T2 weighted MRI criteria [4]. These are swelling with hyperintense signal (stage 1); normal volume with hyperintense signal (stage 2); atrophy with hyperintense signal (stage 3); and progressive atrophy and normal signal (stage 4) [4]. Our patient presented in stage 4. Ictal single photon emission computed tomography (SPECT) is useful for the localization of the epileptogenic focus in the respective cerebral hemisphere in RE patients [11]. The use of SPECT in this case report would have assisted us in localizing the epileptogenic focus, but this facility is not available in our centre. The age at presentation in our case report is close to the median age at disease onset of 6 years as reported by Ramesha KN et al in 2009 [12]. As high as 84.2% of RE patients present with hemiparesis, while EPC was noted in 73.6% [12]. A patient may present with dysarthria due to tongue EPC without hemiparesis [12]. Our patient presented with hemiparesis, focal tonic-clonic seizures, incoherent speech, and hemiatrophy of the right upper and lower limb. Radiologically, RE should be differentiated from other causes of cerebral hemiatrophy like Sturge-Weber syndrome, hemispheric infarction (Dyke-Davidoff-Masson), and MELAS (mitochondrial encephalopathy, lactic acidosis, and stroke-like episodes). Our patient satisfied the part A criteria of the European consensus statement of Bien et al for the diagnosis of RE [13]. We did not administer gadolinium during the MRI study, but we performed a brain CT scan to document the absence of calcifications to exclude the differential diagnosis of a unihemispheric vasculitis.

Immunomodulatory treatment with either high-dose steroids or intravenous immunoglobulin is a treatment option for patients with RE, and this can achieve more than 50% reduction in the seizure frequency [14]. Better results are obtained in RE patients who undergo either of the two options of surgical resection, namely focal cortical resection and functional hemispherectomy [14]. Despite the controversy surrounding the introduction of functional hemispherectomy, it is still the only curative treatment for RE [5,14,15]. Though rare, a patient can develop contralateral hemispheric disease following successful hemispherectomy [12]. The use of positron emission tomography (PET) has been advocated for the presurgical

evaluation of RE patients billed for surgery to ascertain whether or not they will benefit from surgery [16].

Unfortunately, this imaging modality is rarely available in sub-Saharan African countries including Nigeria. Early diagnosis of RE is crucial for selecting patients for aggressive medical therapy or major surgical interventions [10].

CONCLUSION

RE has not been previously described in a Nigerian in the medical literature, to the best of our knowledge. The diagnosis of this condition was aided and confirmed by brain CT and MRI scans. The authors want to emphasize the importance of these imaging modalities in the diagnosis of patients presenting with hemiparesis and seizure in our environment. RE is extremely rare and many radiologists practicing in sub-Saharan Africa may not see a single case of RE in their practice life as a result of the paucity of MRI facilities (which is required for diagnosis) in this sub-region.

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