

## Bilateral Rasmussen's Encephalitis in an African Child: A Case Report

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### Abstract

Rasmussen's encephalitis is a rare neurological disease characterized by unilateral hemispheric atrophy, focal intractable seizures, and progressive neurological deficits. The bilateral subtype is extremely rare with few reported cases in the literature. We present a seven year old boy with bilateral Rasmussen's encephalitis which could have been missed if there is no imaging facility. We also highlighted the possible associated etiological factors and sociocultural challenges in the management.

**Keywords:** Bilateral Rasmussen's Encephalitis; Intractable seizures; Hemispheric atrophy; Sociocultural challenges

### Introduction

Rasmussen's Encephalitis has been described as an uncommon cause of childhood intractable seizures which usually presents as intractable focal seizures associated with progressive neurological decline and imaging findings of unilateral cerebral atrophy<sup>[1-3]</sup>. Bilateral Rasmussen's encephalitis is reserved for cases in which both clinical and radiological findings are in keeping with the bilateral hemispheric deficit. The affected brain tissue shows chronic inflammatory histopathology and an autoimmune reaction has been suspected as an etiological factor<sup>[3]</sup>. The average age at presentation is 6 years of age<sup>[4]</sup>. In the acute stage, the neurological deficits that appear include cognitive deterioration and aphasia if the dominant hemisphere is affected<sup>[5]</sup>.

Management of Rasmussen's encephalitis is tailored to control epilepsy and modify disease progression with antiepileptic drugs, immune suppressants, immune modulators, plasmapheresis and intravenous immunoglobulin<sup>[6]</sup>. Surgery is the only treatment modality of choice to provide complete seizure control, but it is usually offered to patients with established neurological deficits, such as hemiparesis<sup>[4,7-10]</sup>. The choice of therapy needs to be reviewed for each patient with the treating physician, neurologist or pediatric neurologist, in order to provide an individually tailored therapy<sup>[10]</sup>.

### Case Report

OQ was a 7-year old boy who was initially managed for probable cerebral malaria at a Private Hospital and he was referred to Federal Teaching Hospital Ido-Ekiti, Nigeria on account of refractory generalized tonic-clonic seizures of 6 weeks duration. There was a history of fever and diarrhoea at the onset of illness. There was no preceding

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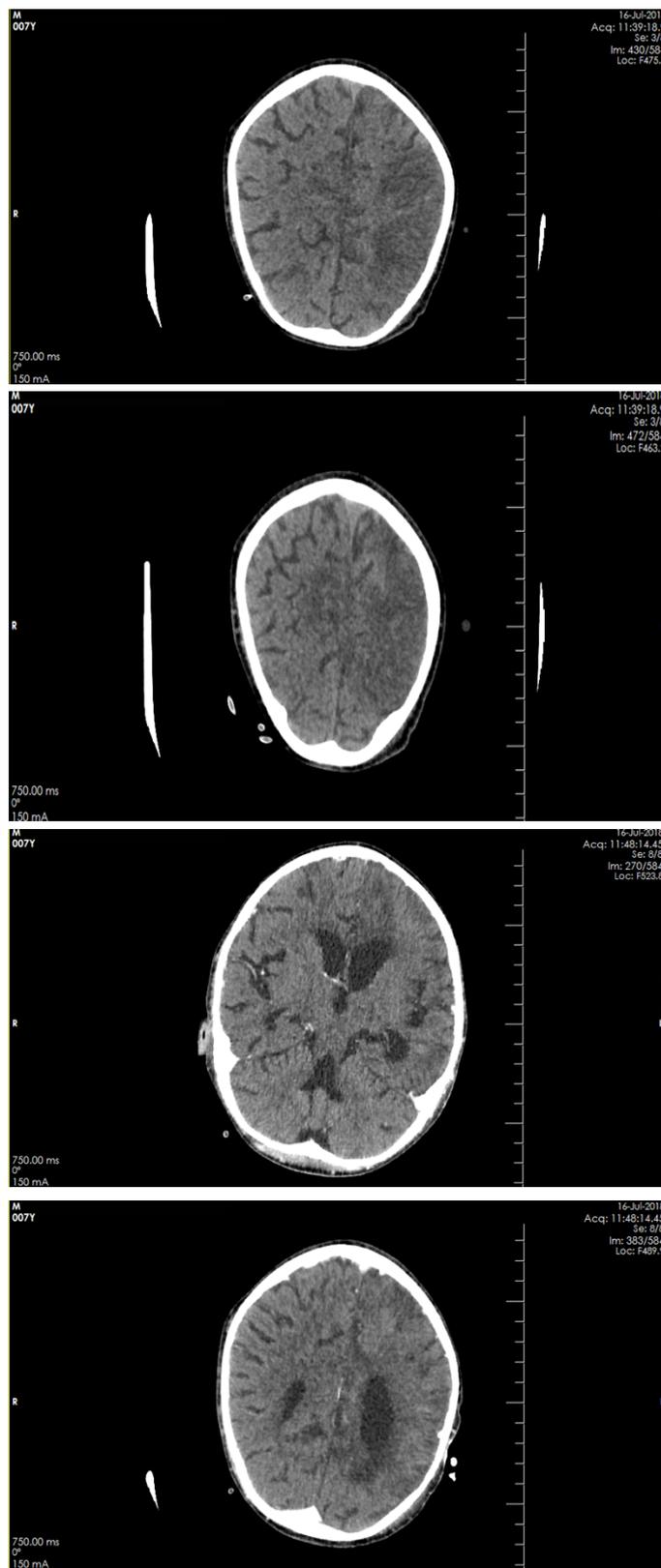
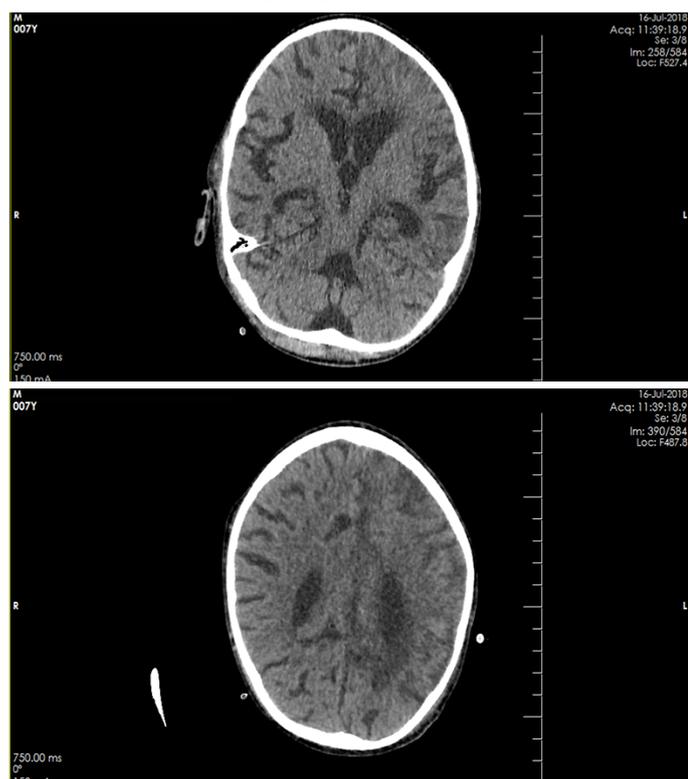
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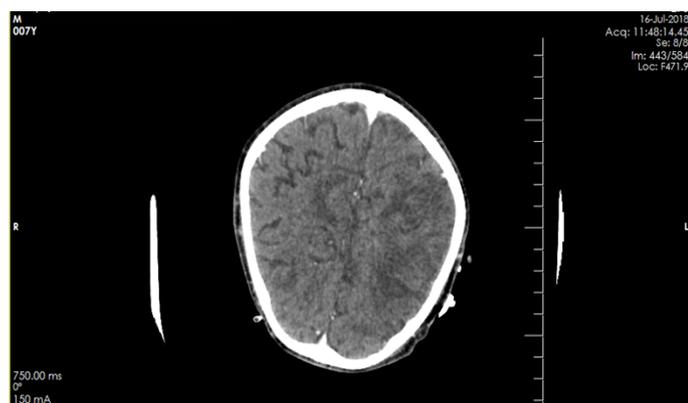
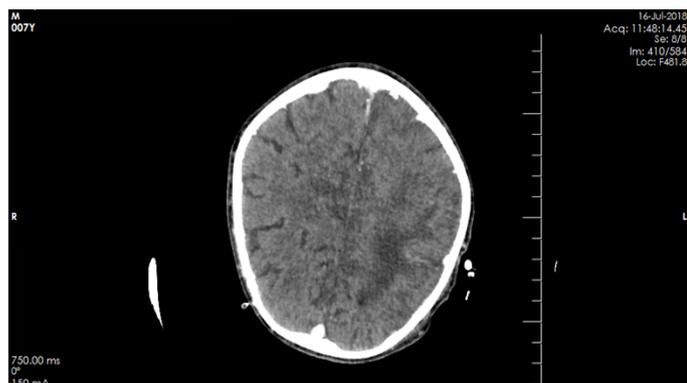
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illness. He was the third of 4 children, only male child of his parents. The parents were separated and he was residing with his maternal aunty. Examination revealed a young cachectic boy, with normal vital signs. He was awake but aphasic. His pupils were 3mm in size and pupillary response to light was brisk. The Brudzinski's and Kernig's signs were negative. He was spastic globally with flexion of the limbs and fisting of the hands worse on the right. The plantar response was extensor bilaterally. He had no dysmorphic feature, both the shape and size of his head were normal. His fontanelles were closed. The examination of other systems was normal. Cerebrospinal fluid (CSF) analysis was not suggestive of meningitis. An initial clinical diagnosis of encephalitis with neurological sequelae was made. Cranial CT scan (8 weeks post-ictus) showed extensive left hemispheric and right parietal cortical infarction with differential ventriculomegaly in favour of the left lateral ventricle and gross global brain atrophy (figure 1). The diagnosis of probable bilateral Rasmussen's encephalitis was made in view of refractory seizures and cranial CT scan findings. Brain MRI and EEG could not be obtained due to financial constraint. We planned to optimize the anticonvulsants and nutritional status but the parents declined further care and discharged against medical advice.





## Discussion

Rasmussen's encephalitis is a rare neurological disease characterized by unilateral hemispheric atrophy, focal intractable seizures, and progressive neurological deficits<sup>[3,6,11]</sup>. The bilateral subtype is extremely rare with few reported cases in the literature and the diagnosis is usually challenging<sup>[3,10]</sup>. It tends to affect male more than females<sup>[12-14]</sup>. The index patient was a male child that presented with intractable generalized seizures, progressive neurological deficit including global aphasia and cranial CT scan findings of bilateral cerebral infarctions worse on the left hemisphere which suggests a case of probable bilateral Rasmussen's encephalitis.

Cases of Rasmussen's encephalitis may not be as rare as previously documented because with the increasing availability of neuroimaging facilities, more cases are being reported<sup>[15,16]</sup>. The first neuroimaging facility in our region was acquired about 5 months prior to the presentation of this case of probable bilateral Rasmussen's encephalitis which could have been missed if there was no imaging facility. There was a preceding history of fever associated with diarrhoea in this patient which may suggest a viral aetiology<sup>[17,18]</sup>. The response of Rasmussen's encephalitis to steroids may suggest an inflammatory and immunological process underlies the manifestation of this rare disease<sup>[13]</sup>. For any inflammatory process, the immune effector cells or antibodies usually originate from the bloodstream, thus the encephalitis is expected to be bilateral<sup>[3]</sup>. Poor socioeconomic status has been noted in a similar case of Rasmussen's encephalitis as in this index patient<sup>[1]</sup>. Late referral and presentation could also have worsened the prognosis for this patient. Seizure disorders still carry a lot of stigmatization in our region. There is sociocultural

and religious belief that it is caused by a spiritual attack, for this reason, many victims of this type of disease tend to be taken out of the hospital for spiritual intervention which might have played out in this index case.

## Conclusion

Bilateral Rasmussen's encephalitis is reported to be extremely rare with few reported cases in the literature but the trend may change with the increasing availability of neuroimaging facility in the rural communities. This may further strengthen the theory of immunological response to chronic inflammation.

**Conflict of interest:** Authors declare no conflict of interest.

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