

Case Report

A Case of Rasmussen's Encephalitis in a 9-year-old girl from Tanzania

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Abstract

Introduction: Rasmussen's encephalitis (RE) is a rare but chronic inflammatory neurodegenerative disorder with a poorly understood etiopathogenesis. It is characterized by progressive unilateral inflammation of the cerebral cortex leading to intractable seizures, gradual neurocognitive decline and unilateral hemispheric atrophy. Despite its relatively pathognomonic clinico-radiological picture, RE confers a diagnostic dilemma to practitioners all over the globe. Over 80% of cases demonstrate pharmacoresistance to anti-seizure drugs while similar rates are reported as far as post hemispherectomy seizure freedom is concerned.

Case presentation: A 9-years-old female of African descent presented with a history of recurrent seizures, right-sided weakness, speech difficulties, memory loss and poor school performance. Her early childhood was uneventful with a good weight increment and normal psychomotor development until from the age of 4½ years when she developed afebrile seizures. Central nervous system examination revealed right-sided hemiparesis with disuse atrophy. There were no signs of meningeal irritation but she had a moderate mental retardation. Cerebral spinal fluid (CSF) analysis was normal but electroencephalogram (EEG) revealed focal epileptiform discharges and subclinical ictal discharges originating from the left fronto-temporal region. Computed tomography (CT) scan of the brain showed diffuse volume loss in the left cerebral hemisphere with dilatation of the ipsilateral lateral ventricle. Based on the clinical picture, physical findings and radiographic features, a diagnosis of Rasmussen's encephalitis was reached.

Conclusions: Despite its infrequency, early recognition and timely hemispherectomy is potentially curative for RE. In this regard, clinicians should raise an index of suspicion for RE when faced with a previously healthy child with a history of refractory seizures and unilateral motor deficits accompanied by a unihemispheric cortical atrophy.

Key Words: Rasmussen's encephalitis; chronic focal encephalitis; Rasmussen's syndrome; Epilepsia partialis continua; focal seizures; encephalitis; case report

Introduction

Worldwide, neurological disorders are associated with developmental disability, functional impairment, stigma, increased complexity and cost of care, and reduced life expectancy. Rasmussen's encephalitis (RE) is a rare but chronic inflammatory neurodegenerative disorder that is characterized by progressive unilateral inflammation of the cerebral cortex leading to intractable seizures, gradual neurocognitive decline and unilateral hemispheric atrophy. Despite several hypothesis postulations (i.e.

humoral auto-antibodies and viral infection) regarding its etiology, the pathogenesis of RE remains poorly understood. Recent studies however have demonstrated a cytotoxic T-cell mediated immune reaction with astrocytosis as the most likely cause.¹

Majority (85%) of RE cases occur in children aged 10 years and below.² On the other hand, the incidence in adults is rising with the routine CT/MRI screening among those with difficult to control seizures.³ Although at times an infectious or inflammatory entity is notable couple of months prior the onset of RE, majority of cases have an unremarkable past medical history. Rasmussen's encephalitis has an insidious onset which makes its diagnosis delayed or missed, nevertheless its characteristic clinico-radiological features are usually enough to reach a diagnosis. With regards to management, over 80% of cases show signs of pharmacoresistance

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to antiepileptic drugs and thus far only cerebral hemispherectomy has proven to be highly effective in achieving seizure freedom and halting disease progression.⁴ We report a case of RE in a 9 years old female of African descent who presented with a history of recurrent seizures, right-sided weakness, speech difficulties, memory loss and poor school performance.

Case Presentation

A 9-year-old female child of African descent presented with a history of recurrent seizures, right-sided weakness, speech difficulties and progressive cognitive decline. She was born to a 14-year old woman (now 23 years) by spontaneous vertex delivery at term. The child weighed 2,000 grams and had APGAR scores of 7 and 9 at 1st and 5th minute respectively. She had received all vaccinations as per the Tanzanian immunization and vaccine development program. Her early childhood was uneventful with a good weight increment and normal psychomotor development until from the age of 4½ years when she developed afebrile seizures. The seizures were tonic-clonic in nature, lasted an average of 5 to 10 minutes with a prolonged postictal state and they would recur several times within an hour regardless of the child's wakefulness or sleep status. Consequently, she succumbed to several hospital admissions and outpatient visits and was treated for malaria and meningitis despite negative evidence for the conceivable diseases. Since the onset of seizures, the child developed right-sided hemiparesis with a slurred speech. Thus far, the child has never been seizure free despite long term use of phenobarbital 60mg once daily and carbamazepine 200mg once daily. The mother however reported that the frequency of seizures is now lesser than before and that the last episode was 2 weeks prior this index visit. Additionally, although the right-sided weakness and slurred speech have remained fairly steady, the child is progressively forgetful and performs poorly in class.

On examination, the child was alert and oriented with stable vital signs. Central nervous system examination revealed right-sided hemiparesis with disuse atrophy. She had a slurred speech but otherwise unremarkable cranial nerves examination. Her sensation was normal with no deficits on the cerebellar or extrapyramidal systems. Furthermore, hypertonia, hyperreflexia, reduced power (distal [1/5] > proximal [3/5]), and a positive Babinski sign were also seen on the right-sided limbs. There were no signs of meningeal irritation but she had a moderate mental retardation (IQ 35-40). Other systemic examinations were unremarkable.

All routine laboratory investigations were normal except for the iron deficiency anemia (Hb 9.7g/dL, MCH 21 pg/cell, MCV 69 fL, RDW 14.9%) in the full blood count. Cerebral spinal fluid (CSF) analysis was normal but electroencephalogram (EEG) revealed focal epileptiform discharges and subclinical ictal discharges originating from the left fronto-temporal region. Computed tomography (CT) scan of the brain showed diffuse volume loss in the left cerebral hemisphere with dilatation of the ipsilateral lateral ventricle, Figure 1.

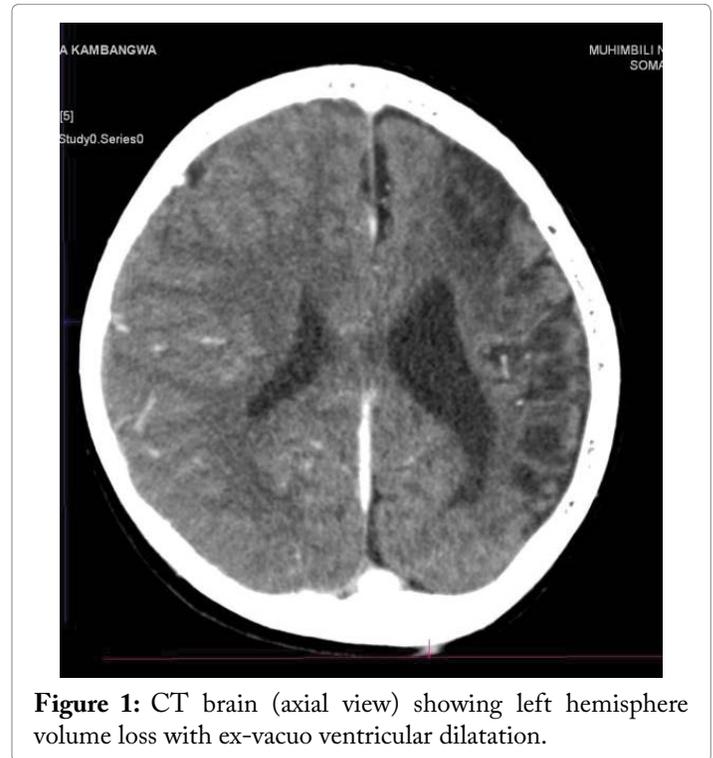


Figure 1: CT brain (axial view) showing left hemisphere volume loss with ex-vacuo ventricular dilatation.

Based on the clinical picture, physical findings and radiographic features, a diagnosis of Rasmussen's encephalitis was reached. Pediatric neurosurgeon together with child psychologist, physiotherapist, occupational and speech therapists were consulted. The mother was counseled regarding the prognosis and hemispherectomy.

Discussion

Rasmussen's encephalitis is a rare inflammatory neurological disease with a poorly understood etiopathogenesis. Polymorphous intractable seizures is the hallmark of RE while other manifestations including progressive hemiparesis, hemianopia, cognitive deterioration, and aphasia (if dominant hemisphere affected) are characteristic. It is predominantly a disease of childhood and the mean age at presentation is around 7 years.⁵ Clinically, RE can be categorized into three stages; (i) prodromal stage, where non-specific symptoms, low frequency of seizures and hemiplegia may be observed, (ii) acute stage which presents with frequent seizures, progressive hemiparesis and cognitive regression, and the (iii) residual stage characterized by continuing seizures with stable and permanent neurological deficits.⁶ Our patient was symptomatic for seizures, hemiparesis and progressive cognitive decline for 4 years before the diagnosis was reached in the residual stage of the disease.

Imaging plays a fundamental role in reaching the diagnosis of RE as well as in monitoring the disease progression. Unilateral cortical atrophy with ex-vacuo ventricular dilatation are the typical RE features on both CT scan and MRI.⁷ The prognosis of RE remains disappointing although early initiation of immunotherapy

is known to delay the disease progression and improve the overall quality of life.⁸ Unfortunately, due to the fact that the diagnosis of RE was missed until referral to our institution, the child was never put on any immunotherapy regimens. Cerebral hemispherectomy has shown excellent results with postsurgical seizure freedom rates of up to 85%, however, the decision of whether or not to perform surgery especially in the absence of significant neurological deficits remains a dilemma.⁹ Moreover, even when surgery is opted for, the most favorable timing for it to achieve the best neurocognitive outcomes remains enigmatic and consequently functional compromises remain inevitable. At the moment cerebral hemispherectomy is not performed in Tanzania and processes are on going to refer the child overseas.

Conclusions

In conclusion, despite its infrequency, early recognition and timely hemispherectomy is potentially curative for RE. In this regard, clinicians should raise an index of suspicion for RE when faced with a previously healthy child with a history of refractory seizures and unilateral motor deficits accompanied by a unihemispheric cortical atrophy.

Consent

Written informed consent was obtained from the patient's legal guardian for publication of this case report and any accompanying images. A copy of the written consent is available for review by the Editor-in-Chief of this journal.

Abbreviations

CSF: Cerebral spinal fluid
 CT: Computed tomography scan
 EEG: electroencephalogram
 HB: Hemoglobin
 MCH: Mean Cell Hemoglobin
 MCV: Mean Corpuscular Volume
 MRI: Magnetic Resonance Imaging
 RDW: Red Cell Distribution Width
 RE: Rasmussen's encephalitis

Ethical approval

Ethical clearance was sought from the Directorate of Research of the Jakaya Kikwete Cardiac Institute

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Authors' contributions

PP and PN took the history and performed physical examination. HM and FL interpreted the CT scan images. PP wrote the initial draft of the manuscript. All authors reviewed and contributed to the final version of this case report.

Competing interests

The authors declare that they have no competing interests

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