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Rasmussen Encephalitis: Case Report and Review of the Literature

Yao Christian Hugues Dokponou[1,2*], Fernand Nathan Imoumby[1,2], Napoléon Imbunhe[1,2], Sofia El Akroud[1,2], Miloud Gazzaz[1,2]

[1]Mohammed V Military Teaching Hospital, Rabat, Morocco
[2]Faculty of Medicine and Pharmacy, Mohammed V University, Rabat, Morocco
Email: *dokponou2407@gmail.com, ifernandnathan@gmail.com, imbunhe@gmail.com, sofiaelakroud16@gmail.com, gazzazmiloudi@hotmail.com

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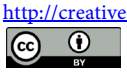

Abstract

Rasmussen encephalitis (RE), also called Rasmussen syndrome, is a rare, progressive, chronic encephalitis affecting one hemisphere of the brain. It occurs mainly in children. However, around 10% of all cases are adolescents and adults. RE occurs usually in healthy individuals. It is estimated that no more than two new cases per year are identified in large epilepsy centers. Rasmussen's encephalitis is characterized by intractable focal seizures, often in the form of epilepsia partialis continua (EPC) with motor and cognitive deterioration. Neuroimaging shows the progressive damage of the affected hemisphere, and histopathology is consistent with a T-cell dominated encephalitis with activated microglial cells and reactive astrogliosis. Cerebral hemispherectomy remains the only cure for seizures, but there are inevitable functional compromises. We report the rare case of an 8-year-old girl without any clinical history presented with recent left hemiparesis and seizures, and whose investigations based on the brain MRI and Electroencephalogram data, conducted to the diagnosis of Rasmussen's encephalitis. Our management was exclusively medical, with a good outcome.

Subject Areas

Neurology

Keywords

Rasmussen Encephalitis, Seizures, Management, Positive Outcome, Case Report

1. Introduction

Theodore Rasmussen, a Canadian neurosurgeon, was the first to report in 1958,

a rare, chronic inflammatory brain disease of unknown etiology, affecting mostly children and will be called Rasmussen encephalitis (RE). Rasmussen encephalitis is a devastating neurological disorder characterized by seizures, brain inflammation, and progressive hemispheric atrophy. Focal drug-resistant epilepsy, hemispheric atrophy, progressive intellectual decline, and neurological deficits are common clinical and radiological presentations of the disease [1] [2] [3]. Although high-dose steroid and intravenous immunoglobulin therapies lead to temporary alleviation of seizures, the only known therapy for interruption of this progressive disorder is surgical resection of the affected hemisphere, *i.e.*, hemispherectomy [4]. For those patients that are treated surgically, Histopathological findings demonstrate the presence of inflammation dominated by CD8+ T-cells, widespread microglial activation, perivascular lymphoid cells, and glial scarring sometimes with cystic cavitation to confirm the diagnosis in case the clinical and radiological finding is not specific.

The outcome depends on the choice of the management; either medical, surgical, or both. The aim of this study is to report a rare type of encephalitis and its positive outcome of conservative medical treatment.

2. Case Presentation

An 8-year-old girl a second child of a family of two children, born to nonconsanguineous parents with unremarkable past medical history was admitted, to the pediatric department of Military Teaching Hospital of Rabat on November 11th, 2020, with severe progressive headache, and vomiting, complicated by left hemiparesis. Neurological examination, imaging (**Figure 1**), and laboratory tests concluded on a diagnosis of auto-immune encephalitis and she was treated as such. The patient did well on corticotherapy and was discharged a week later.

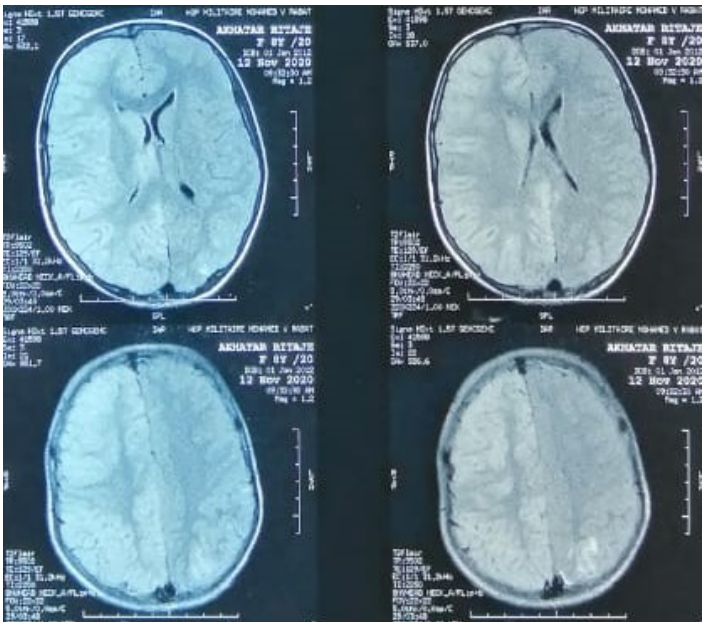

Figure 1. Axial FLAIR-weighted hyperintense signal of the right hemisphere.

On the 15th of December 2020, the patient was readmitted with status epilepticus and left hemiplegia. At the admission, she was unconscious with a GCS of 9, pupillary size is normal, no stiffness of the neck, fever, the Babinsky sign was positive on the left side, and left hemiplegia of 2/5 was confirmed at the neurological examination. The patient was in good general status and normal psychomotor development.

The complete blood count shows only severe thrombocytopenia (60.000/µL with normal range 239.000/µL - 357.000/µL). All other items were normal. The cerebrospinal fluid (CSF) was collected by lumbar puncture and the laboratory (bacteriological and biochemical) analysis was almost normal: an isolated high glucose level of the CSF (0.72 g/l with normal range 0.40 - 0.70) was found. The CSF is also used for searching for cytomegalovirus, enterovirus, herpes simplex, herpes human, parechovirus, varicella-zoster, Escherichia coli, Hemophilus influenzae, Listeria monocytogenes, Neisseria meningitides, streptococcus agalactiae, streptococcus pneumoniae, and cryptococcus were all negative. The head CT-Scan revealed a right-hemispheric tumor-like lesion with a mass effect on the homolateral ventricle with a shift of the median line and right temporal lobe herniation. There was right cerebral hemisphere atrophy on the magnetic resonance imaging (MRI) (**Figure 2**). The electroencephalogram (EEG) was abnormal with sharp waves and spike-and-wave complexes in the right hemisphere being the site of the seizure's activity. With the results of these investigations, we concluded a Rasmussen encephalitis.

Surgical treatment of hemispherectomy was raised as a possible therapy to

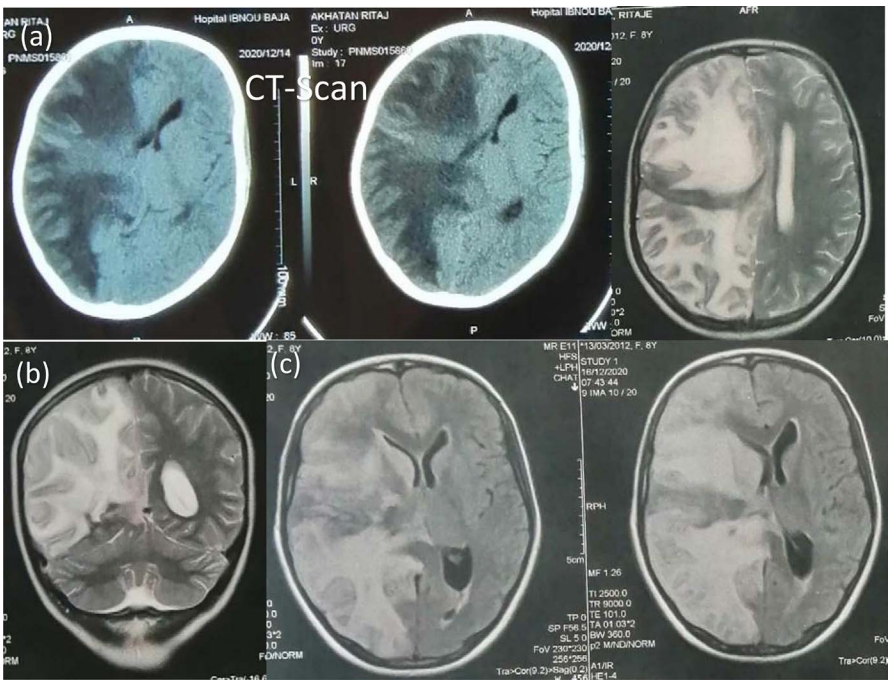

Figure 2. Bain CT-Scan axial section showing right hemisphere lesion with a total hemispheric oedema. (a)-(c): T2 & FLAIR-weighted MRI showing affected right hemisphere that is hyperintense with cortical atrophy.

manage this case, but we decided to start with conservative medical management with corticoid (solumedrol) 1400 mg per day, 40 mg of Cortancyl each two days, 400 mg three times a day of Aciclovir (zovirax), phenobarbital and Third generation cephalosporin perfusion, and Valproate acid (dépakine). From the 4th day of treatment, there was no more seizure, and the motor palsy was progressively subsiding. This medical therapy continues for three weeks with a good outcome, the child was doing good and was discharged from the hospital without surgery.

3. Discussion

RE is a rare disease that should be envisaged as sporadic since there is no evidence for a genetic component. There is, at present, no conclusive evidence of why and how RE starts. A viral etiology was already suggested by Rasmussen based on the constituents of the immune reaction in the brains such as lymphocyte infiltration and microglial nodules. Three disease stages have recently been proposed. Initially, there may be a rather non-specific "prodromal stage" with a relatively low seizure frequency and rarely mild hemiparesis with a median duration of 7.1 months (range: 0 months to 8.1 years). Following this, all patients enter an "acute stage" of the disease, although, for a third of cases, this appears to be the initial clinical disease manifestation. It is characterized by frequent seizures, mostly simple partial motor seizures often in the form of epilepsia partialis continua (EPC). The neurological deterioration becomes manifest by progressive hemiparesis, hemianopia, cognitive deterioration and if the language dominant hemisphere is affected, aphasia. The median duration of this stage is 8 months (range 4 - 8 months). After that, the patients pass into the "residual stage" with permanent and stable neurological deficits and still many seizures, although less frequent than in the acute stage. At this stage, not all patients are hemiplegic. hemiparesis is the most useful marker as this feature is most consistently found, and it allows quantitative evaluation, even in children. The ages of these patients ranged between 7 and 16 years, and the mean age at onset of seizures was 7.1 ± 2.2 years. [5] [6]. These findings correspond to what we got in our case, except for the serology test that was negative for all possible viral infections. In our study, the patient also had a prodromal stage with a relatively mild hemiparesis with a median duration of 1 month before being admitted the second time in the clinical disease manifestation stage with a status epilepticus or epilepsia partialis continua.

Radiologically, characteristic MRI features are areas of cortical hyperintense T2/fluid-attenuated inversion recovery signal and progressive atrophy in the affected cerebral hemisphere [7]. We have the same imaging finding with our patient's right hemisphere being totally affected. Rasmussen encephalitis is a pathophysiologically fascinating condition because both seizures and inflammation are seen only in 1 cerebral hemisphere, making it a disease that can be diagnosed with a simple combination of signs and symptoms couple with those specific radiological changes.

Robust data on incidence and most effective treatment of RE are missing (Bi-

en & Schramm, 2009). There is broad consensus (but no trial evidence) that hemispherectomy in one of its modern variants is highly effective in eliminating epileptic seizures. HE is usually reserved for patients in the residual disease stage with dense hemiparesis. The surgical methods included anatomical hemispherectomy, functional hemispherectomy, hemispherectomy, lesion resection, multilobar resection, selective resection, and bipolar electrocoagulation of functional cortexes [3] [8] [9] [10]. The outcome is diverse and multiform depending on the stage of the disease before operation and which type of surgical procedure has been carried out. On contrary, our patient did well with remission of seizures under medical treatment. Furthermore, in 2009 Terra Bustamante [11] has reported that mental and language impairment was observed in 15 and 12 patients, after surgery, respectively. Eight patients presented post-operative cognitive decline, while only two patients had cognitive improvement. This means, even though surgery can cease seizures in RE patients, the post-operative consequents can be a lifetime handicap for patients.

4. Conclusions

Rasmussen encephalitis (RE) is a very rare, progressive, chronic encephalitis that usually affects only one hemisphere of the brain. This disease is characterized by focal seizures, with motor and cognitive deterioration that occurs mainly in children under the age of 10. Investigations of the brain are based on MRI and electroencephalograms leading to diagnosis. The management is medical (to reduce seizure severity and frequency or surgery) treatment against the primary process. The prognosis of the patient with Rasmussen encephalitis varies.

To better understand the Rasmussen encephalitis, we propose these four stages of the disease evolution that we found in this reported case as well as described in the literature (Figure 3).

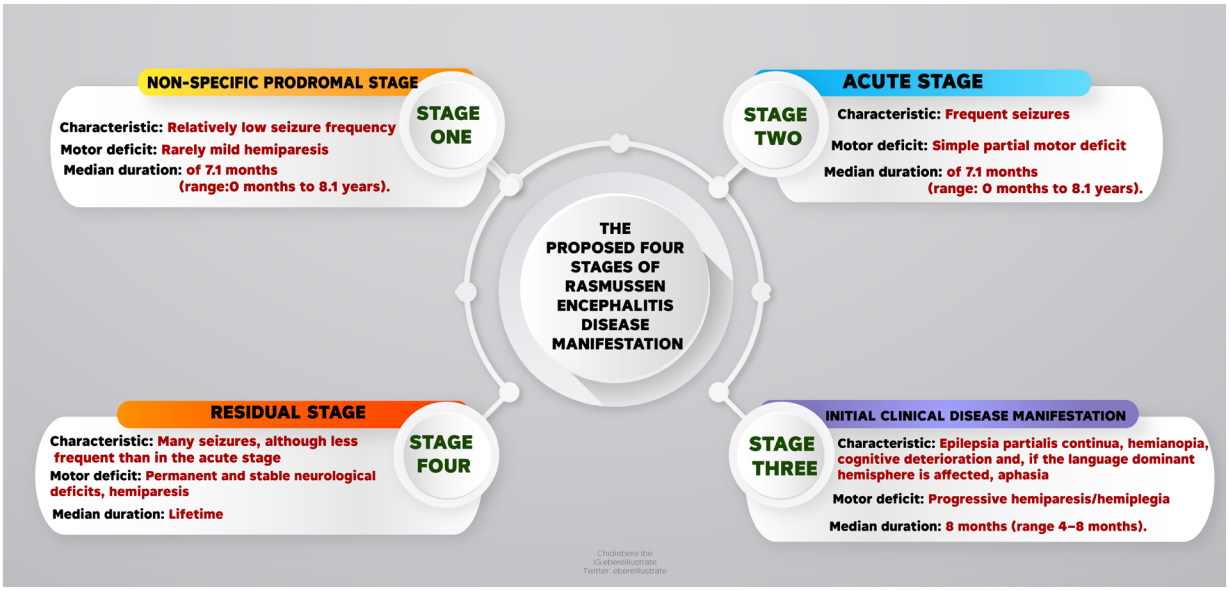

Figure 3. Four stages of rasmussen encephalitis.

Disclosure

The authors did not receive any funding for the preparation of this case report.

This article is an original work that is not being considered or reviewed by any other publication, and has not been published elsewhere in the same or a similar form.

All authors of the manuscript have read and agreed to its content and are accountable for all aspects of the accuracy and integrity of the manuscript.

Informed Consent

The patient gave his informed consent to publish his case.

Authors' Contributions

Yao Christian Hugues Dokponou: Conceptualization, Writing an original draft, & editing. Fernand Nathan IMOUMBY: Writing—review & editing. IMBUNHE Napoléon: Writing & editing. EL AKROUD Sofia: Writing & editing. Miloud GAZZAZ: Supervision, review, & Validation.

Ethics and Reporting Guidelines

Informed consent and verbal permission were obtained from the patient's family prior to the submission of this article. Also, this article respects both the Consensus-based Clinical Case Reporting Guideline and the Recommendations for the Conducting, Reporting, Editing, and Publication of Scholarly Work in Medical Journals. [12] [13]

Conflicts of Interest

The authors declare no conflicts of interest regarding the publication of this paper.

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