

Rasmussen's Encephalitis: A Case Report

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Abstract

We report a case of Rasmussen's Encephalitis. It is a rare, chronic inflammatory neurological disease of unknown origin that usually affects only one hemisphere of the brain. It is common in children under the age of 10 with average age at disease onset around 6 years but uncommon in adults, adult variant that accounts for about 10% of the cases only. Rasmussen's Encephalitis is characterized by intractable severe seizures, loss of motor skills and speech, paralysis on one side of the body (dysfunctions associated with the affected hemisphere). Our case is a 21 years old female, presented to emergency department JMPC Karachi with complaint of intractable severe seizures, progressive hemiparesis and deteriorated cognition followed by an episode of encephalitis. Her course of illness was focal seizures and right-sided weakness (hemiparesis) for 5 months. In addition to classical clinical presentation of Rasmussen's Encephalitis, MRI Brain showed hemispheric atrophy of one cerebral hemisphere and old gliotic changes that further supports diagnosis of Rasmussen's Encephalitis.

Abbreviations: CT: Computed Tomography; MRI: Magnetic Resonance Imaging; RE: Rasmussen's Encephalitis; HT: Hemispherectomy

Introduction

Rasmussen's encephalitis is a chronic inflammatory disease of unknown origin, brain inflammation resulting in unilateral brain atrophy that was first described by Rasmussen et al. in 1958 [1]. Clinically this disease is characterized by drug-resistant focal seizures, progressive weakness, worsening of motor and cognitive functions [1,2]. It is common in children under the age of 10 with average age at disease onset around 6 years [3] but uncommon in adults, adult variants account for around 10% of the total cases [4].

Various etiological hypotheses have been postulated about Rasmussen's encephalitis, some researchers revealed cell-mediated immunity relates the pathogenesis of RE, more precisely T-cell immunoreaction against neurons and astrocytes [5,6]. Cell mediated hypothesis is further supported by a recent study by Granata et al. that suggests that cytotoxic T cells may be directed against a viral protein presents in both neurons and astrocytes [7]. But there is another school of thought which supports autoimmune pathology [8]. In 1994, Rogers et al. found role of antibodies against glutamate receptor (GluR3), other triggers have also shown to be associated with etiology, like herpes simplex virus and cytomegalovirus found [8].

Diagnosis is based on clinical, imaging, and histopathological findings, focal seizures and unilateral cortical deficit on examination are important clues to help in diagnosis. However, imaging studies could be challenging for diagnosis in initial stages of Rasmussen's encephalitis [9].

There are no biomarkers for RE to diagnose RE, the presence of anti-GluR3 is not considered specific for the diagnosis, since it does not discriminate between RE and other non-inflammatory epilepsies [10]. However histopathology of brain by open biopsy provides an important finding to diagnose RE. Biopsy for the diagnosis of RE shows multifocal changes of the T cell dominated encephalitis with activated microglia and reactive gliosis [11], but some believe that CSF standard tests are not suitable to confirm or to exclude the diagnosis of RE [2].

Treatment strategies vary among different patients at different stages, treatment options are selected according to need of patient. There is insufficient evidence and data for most effective treatment of RE [12], In medical treatment corticosteroids are often used and shown robust and effective response [13], similarly intravenous immunoglobulin (IVIg) and Plasmapheresis (PEX) have been used and proven to be effective for short term response but inefficient to achieve long terms results [14]. Whilst comparing to medical treatment options, surgically hemispherectomy is generally considered more useful and widely accepted treatment option because it is highly effective to eliminate seizures and one of modern treatment option for RE [12]. Surgical approach is tailored to remove affected hemisphere by hemispherotomy, and the only potential treatment that able to control seizures and prevent from further mental deterioration in these patients [15].

Case Presentation

21 years unmarried female known case of focal epilepsy came to emergency department of JMPC Karachi, patient presented with uncontrolled intractable seizures and hemiparesis. On arrival she was give diazepam and phenytoin infusion, same time investigated with CT scan of brain (stat), after acute management, patient was transferred on floor in neurology ward. After moving to ward she was started on phenytoin and clonazepam orally but fits did not cease and became worsened and uncontrollable. Soon after this condition levetiracetam infusion was started promptly, After 72 hours of treatment, patient continued to develop seizures, besides seizures her right-sided weakness persisted, progressed and became worsen. During these episodes of fits,

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blood sample was collected for baseline investigations (complete blood picture, ESR, blood culture & sensitivity, comprehensive metabolic profile, thyroid function test, blood Sugar, urine toxicology, blood toxicology, ANA profile). Laboratory tests were reported soon after ordering and results shown within normal limits. Lumbar puncture and MRI Brain with contrast were also performed. The CSF report showed normal glucose, increased protein, increased lymphocytes, and also (PCR) for Herpes Simplex Virus appeared to be negative. The CT scan brain shown atrophy of left hemisphere. The MRI showed high signal intensity area is seen in bilateral temporal lobes and right partial cortex with adjacent effacement of cortical sulci and also asymmetry between two cerebral hemisphere with atrophy of one cerebral hemisphere represent old gliotic changes. In the suspicion of Herpes Simplex Virus Encephalitis empirical antiviral treatment was given but patient continued to have seizures without any clinical stability, and focal seizures labelled as "epilepsia partialis continua".

Discussion

Typically management of Rasmussen's encephalitis is tailored to control intractable seizures and alter disease progression with the aid of various treatment strategies, for example, antiepileptic drugs, immune suppressants, immune modulators and Immunoglobulin, plasmapheresis and surgical treatment like Hemispherotomy. Our patient presented with severe seizures and one-sided weakness, she was managed with conventional antiepileptic drugs, diazepam and phenytoin. levetiracetam, clonazepam, These antiepileptic drugs were used at different times while on admission and transferring to floor, AED were switched to one on other for next 72 hours, but regimen remained unsuccessful, IV Immunoglobulin or Plasmapheresis were not applied due to unavailability of resources, similarly Hemispherectomy was not offered due to unavailability of specialized neurosurgeon for required procedure, evidence and data (No Clinical Trial) have shown fair results to abort uncontrollable seizures with Hemispherectomy [2,12,15]. Our patient was qualified for surgical procedure due to weakness in arm but due to limited sources, generally accepted Hemispherectomy was not performed. So the patient was conservatively managed during the hospital stay and referred to another tertiary care hospital for surgery. We tracked our patient to the referred hospital; patient had refused to sign high-risk consent for surgery and discharged on antiepileptic medicine.

Conclusion

Our patient presented with the clinical history of a Rasmussen's encephalitis. Though we diagnosed our patient on basis of clinical judgment and imaging studies, we tried available conventional therapies to treat our patient at our best.

Authors' Contributions

SK developed the idea for the case report and headed the study, and then he revised the draft of the case report, and provided a critical review of the study. SA and FM helped to write case report. SA, RA, FM helped to write discussion and Case presentation section. SA and SK worked on endnote to fix references. All authors read and approved the final manuscript.

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