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Rasmussen Encephalitis: A Pediatric Case Report on Diagnosis and Management

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ABSTRACT

We report the case of a 16-year-old female, presenting with seizure activity consistent with Rasmussen encephalitis, a rare, progressive neurological disorder characterized by drug-resistant focal seizures and progressive neurological decline. The patient had a prior history of epilepsy, right hemiparesis secondary to a cerebrovascular accident (CVA) at age six, and was on multiple antiepileptic drugs. Diagnostic imaging revealed left cerebral hemisphere atrophy with gliotic changes, confirming Rasmussen encephalitis. Management included an intensive antiepileptic regimen, supportive care, and immunotherapy with IV immunoglobulins. This case highlights the diagnostic and therapeutic challenges associated with Rasmussen encephalitis, emphasizing the importance of early recognition and multidisciplinary care.

KEYWORDS: Rasmussen encephalitis, neurological disorder, focal seizures, hemiparesis.

I. INTRODUCTION

Rasmussen's encephalitis (RE) is a chronic inflammatory brain disorder accompanied by progressive impairment of neurological functions associated with the affected hemisphere and intractable seizures. It is characterised by drug-resistant focal epilepsy, progressive hemiplegia, and cognitive decline, with unilateral hemispheric brain atrophy. The disorder is rare and affects mostly children or young adults [1] [2] [3].

Usually, a progressive course of focal onset seizures, characteristically epilepsia partialis continua (EPC) as well as neurological deterioration (more often progressive motor defect) occurs. The diagnosis of RE is also based on neuroradiological findings, with characteristic MRI features of areas of cortical hyperintense T2/fluid attenuated inversion recovery (FLAIR) signal and atrophy of the affected hemisphere. The pathogenesis of RE has been linked to autoimmune process including antiantibodies and cytotoxic T cells [4].

Although the etiology of RE is not completely clear, the main treatment is aimed at symptoms and inflammation, to control epilepsy, and to prevent further deterioration of neurological function, which means treatment with antiepileptic drugs (AEDs), immune treatments and surgery. Of course, symptomatic treatments such as vagus nerve stimulation (VNS) and transcranial stimulation also are means of sporadic palliative therapy according to the literature. AEDs treatment has only a limited effect on seizures. Immunotherapy, which is used mainly in the early stage of the disease, can relieve seizures or prevent immune-mediated hemispheric damage; however, these treatments also have only slight effects. Surgical treatment is a more satisfactory therapeutic strategy for RE. There are three main types of surgical operation: anatomical hemispherectomy, functional hemispherectomy, and disconnection hemisphere [5] [6].

II. CASE REPORT

A 16-year-old female patient, presented to the emergency department with a complaint of seizure activity lasting approximately one hour, which began one day prior to admission. The patient had a known history of epilepsy diagnosed in 2014, managed with multiple antiepileptic medications, including levetiracetam 1 g BD, carbamazepine 100 mg BD, brivaracetam 75 mg BD, and clobazam 10 mg OD. She also had a history of cerebrovascular accident (CVA) at the age of six, resulting in right-sided hemiparesis, and was brought to the emergency department with complaints of seizures. On the day of admission, she developed focal right-sided seizures without impaired awareness and head deviation towards the right side. These episodes were not associated with uprolling of the eyes, frothing, tongue biting, or postictal confusion. On physical examination, her blood pressure was 100/60 mmHg, and her pulse rate was 90 beats/minute. The respiratory system examination revealed bilateral air entry positive (BAE+) with no added sounds whereas the cardiovascular system (CVS) examination revealed normal

heart sounds, S_1 and S_2, with no murmurs or added sounds. Central nervous system examination shows bilateral pupils normal in size and reactive to light (NSRL), plantar reflexes were bilaterally downgoing, power in the upper limbs was reduced (right 2/5, left 4/5), while power in the lower limbs was normal (5/5 bilaterally) and increased tone in some areas with normal tone in others. Per abdomen examination revealed a soft, non-tender abdomen with no organomegaly or masses palpable. There was no history of fever, vomiting, or loose stools. No complaints of headache, blurring of vision, slurring of speech, or other systemic complaints.

On neurological examination, the patient had persistent right-sided hemiparesis, consistent with her history of CVA. Laboratory investigations revealed hyponatremia with a serum sodium level of 125 mmol/L, along with hypoalbuminemia, with levels decreasing from 5.8 g/dL to 4.0 g/dL. Renal and liver function tests were within normal limits. MRI of the brain with contrast revealed the reduced volume of the left cerebral hemisphere, midbrain, and pons, indicating left-sided hemi atrophy. Additionally, T1 hypointense and T2/FLAIR hyperintense areas were noted in the left cerebral lobe, with ex vacuo dilatation of the left lateral ventricle and cystic encephalomalacia with gliotic changes, confirming the diagnosis of Rasmussen encephalitis. The EEG recording, conducted while the patient was awake, revealed abnormal findings characterized by posterior dominant alpha activity that was asymmetrical and synchronous, with noted hemispheric asymmetry. No epileptiform transients or generalized epileptiform activity were observed; however, non-epileptiform variants were present. These irregularities in occipital dominant alpha activity and hemispheric asymmetry led to the classification of the EEG as an Abnormal Awake EEG.

The patient was managed with an extensive antiepileptic regimen, including intravenous (IV) Inj. levetiracetam 1.5 g BD, Inj. valproate 1 g stat followed by 500 mg BD, phenytoin 20 mg/kg via infusion, and oral carbamazepine 200mg/OD, Tab. Brivaracetam 75mg/BD, Tab. clobazam 10mg/OD, Tab. perampenal 2mg/OD. She also received supportive therapy, including IV immunoglobulins (0.4 mg/kg/day) for 5 days, Inj. pantoprazole 40 mg, Inj. ondansetron 4mg, and Inj. midazolam as needed for acute seizure control.

Despite aggressive management, the patient's condition highlights the refractory nature of seizures in Rasmussen encephalitis and underscores the need for early recognition and multidisciplinary treatment.

III. DISCUSSION

Rasmussen's encephalitis is a rare, chronic, progressive neurological disorder that presents a formidable clinical challenge due to its intractable seizures, progressive neurological decline, and resistance to conventional treatments. The clinical manifestations of RE typically involve focal seizures, often progressing to epilepsia partialis continua, as well as hemiparesis and cognitive deterioration [1]. The pathogenesis of RE is believed to be autoimmune-mediated, involving cytotoxic T cells and autoantibodies that target neuronal structures, leading to chronic neuroinflammation and cortical destruction [3].

In the presented case, the patient's history of epilepsy, persistent hemiparesis following a CVA, and focal seizures aligned with the classical presentation of RE [1]. However, the prolonged duration of seizure activity and the refractory nature of the condition highlight the aggressive course of the disease, particularly in cases with a delayed or complex diagnosis.

Hyponatremia and hypoalbuminemia noted during the hospital stay underscore the importance of monitoring systemic complications and comorbidities, which can further exacerbate the neurological condition. Such findings necessitate a multidisciplinary approach to manage not only the neurological symptoms but also the systemic derangements [7].

The diagnosis of RE relies on a combination of clinical, radiological, and laboratory findings. The MRI findings in this case – including unilateral cerebral atrophy, T1 hypointensity, T2/FLAIR hyperintensity, and evidence of cystic encephalomalacia – are characteristic of RE. These imaging features corroborate the neurodegenerative process driven by an autoimmune and inflammatory response localized to one hemisphere [5].

The pathogenesis of RE, though not completely understood, is thought to involve autoimmune mechanisms, including cytotoxic T-cell-mediated neuronal damage and autoantibody formation [3]. This highlights the importance of early and accurate diagnostic workup to differentiate RE from other progressive epileptic syndromes or structural brain pathologies, enabling timely therapeutic intervention.

The refractory nature of RE underscores the limitations of conventional AED's in controlling seizures in this population. Despite aggressive antiepileptic therapy, including the use of multiple AED's and intravenous immunoglobulin (IVIG), the patient's seizures remained resistant to treatment [6]. This aligns with the broader literature indicating that AEDs alone are insufficient for long-term seizure control in RE, necessitating adjunctive immunotherapy or surgical interventions [2].

Immunotherapy, including IVIG and corticosteroids, is most effective during the early stages of the disease. While such therapies aim to mitigate the autoimmune-mediated inflammatory process, their efficacy in halting disease progression is often limited. In advanced cases, surgical interventions such as functional or anatomical hemispherectomy offer the most definitive treatment for seizure control, albeit at the cost of significant neurological deficits [4].

The current case emphasizes the importance of early intervention with immunomodulatory therapies and, where appropriate, consideration of surgical options before the disease advances to a stage where neurological deficits become irreversible.

This case illustrates the complex interplay between systemic, neurological, and psychological aspects of managing RE. The progressive nature of the disease underscores the need for heightened awareness among clinicians, especially in pediatric populations with refractory epilepsy and hemispheric neurological deficits. Furthermore, the lack of definitive curative treatments for RE calls for ongoing research into its pathophysiology and the development of targeted therapies [3].

IV. CONCLUSION

This case illustrates the diagnostic challenges and therapeutic strategies in Rasmussen encephalitis, particularly in patients with neurological comorbidities. Early recognition and a multidisciplinary approach, including immunotherapy and antiepileptic medications, are essential to optimize outcomes.

CONFLICT OF INTEREST

The authors declare that they have no conflict of interests.

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LIST OF ABBREVIATIONS

CVA	Cerebrovascular accident
RE	Rasmussen's encephalitis
EPC	Epilepsia partialis continua
AEDs	Antiepileptic drugs
VNS	Vagus nerve stimulation

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