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COMPARING THE ASSESSMENT AND MANAGEMENT OF RASMUSSEN ENCEPHALITIS IN PEDIATRICS: A CASE SERIES

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ABSTRACT

The current theory on Rasmussen's encephalitis is that it is a continual, progressive immune-mediated process that causes apoptotic neuronal cell death and involves the neuroglial and lymphocytic responses, causing a single hemisphere to progressively deteriorate.we present a case series of two different ages of pediatrics where they have been diagnosed with MRI and EEG as Rasmussen encephalitis.They have been treated with Antiepileptics and Immunomodulatory Therapy and improved clinically.This series is being offered to clinically enlighten people about the early diagnosis of Rasmussen encephalitis and its treatment.

Key words: Rasmussen Encephalitis, cerebral Hemisphere, Epilepsia partialis continua, Focal seizures ,focal cortical Atrophy.

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INTRODUCTION

In 1958, a Canadian neurosurgeon named Theodore Rasmussen published the first report stating that Rasmussen's encephalitis (RE), a very uncommon progressive epileptic disorder with an inflammatory aetiology, is characterised by dementia, hemiparesis, chronic seizures, and encephalitis. It predominantly affects youngsters and produces severe chronic unihemispheric inflammatory illnesses of the central nervous system. [1] Epilepsy partialis continua (EPC), which is then accompanied by increasing hemiparesis and mental impairment, is a common juvenile symptom of the illness. When Rasmussen's encephalitis is present, treatment seeks to lessen seizure intensity and frequency while also enhancing functional long-term prognosis as judged by both motor and cognitive functioning. The underlying causes have not been tackled by treatments, which have simply served to reduce the symptoms. [2] Children under the age of two being affected is unusual. Smaller numbers of adolescents and young adults are also impacted. It has been calculated that 2.4/10,000,000 people under the age of 18 have new-onset Rasmussen each year. Clinical examinations, patient histories, and comprehensive neurological examinations using cutting-edge methods like magnetic resonance imaging (MRI) and electroencephalography (EEG) can all be used to diagnose Rasmussen encephalitis. The electrical impulses of the brain are captured during an EEG. These investigations might expose brain wave patterns that are particular to some kinds of epilepsy. It is typical for the diagnosis to be made following at least two scans, which will show the gradual shrinkage of the affected side of the brain. [3] Grey or white matter high-signal alterations with basal ganglia involvement, as well as increasing unilateral focal cortical atrophy, are distinctive MRI characteristics. Microglial activation, microglial nodules, and brain inflammation driven by T lymphocytes are the hallmarks of

histopathology. Neuronal degeneration and astrogliosis are the last two pathological features to be present. [4]

PATHOPHYSIOLOGY

The pathophysiology of RE has long been thought to be the outcome of improperly functioning adaptive immunity. A cell-mediated hypothesis has more and more evidence backing it up. Cytotoxic T-cells make up the majority of the infiltrating lymphocytes in RE, and MHC class I-positive neurons and astrocytes may be detected in opposition to CD8 cells that carry granzyme B. According to immunohistochemical analysis, the lymphocytic infiltrates are made up of CD3+CD8+T cells. Granzyme B was found in these cells with a polar orientation towards perikarya based on confocal laser imaging. Apoptosis occurred in single neurons. These results suggest that in Rasmussen's encephalitis, a T-cellmediated cytotoxic response causes neuronal death. [5] We will also discuss the various treatment modalities used with these two patients, including immunomodulatory therapy and anti-epileptic medication therapy.

CASE PRESENTATION

CASE 1:

A 8-year-old male presented with complaints of repeated episodes of seizures. He is the first of two children, and he was delivered by vaccum-assisted NVD. His birth weight was 2.7 kg. He had normal growth and development. During his first episode, he had an unprovoked seizure at school and lost consciousness for 1 to 2 minutes. There was no history of fever or trauma. EEG done showed a focal non-specific abnormality over the left frontotemporal region, and syrup lacosam was started. Blood investigations were done. He was started on Inj. Solumedrol 700mg IV over 5 days in view of Rasmussen's encephalitis and continued During his second episode, the semiology of the angle of mouth deviation to one side, with a duration of 1 to 2 minutes, and the syrup lacosam dose were increased. Later, He had complaints of clusters of seizure episodes. Syrup Lacosam was changed to Levetiracetam and Clobazam. An MRI brain was done and showed a normal study. And then he was presented with complaints of multiple episodes of seizure with the semiology of right hand thumb tremor for the past two days, which progressed to right hand three fingers for the past two days. He was treated with Tab. Phenytoin 100 mg HS, Tab. Clobazam 15 mg HS, Syp. Levetiracetam 3 ml–0.5 ml, and Tab. Phenytoin. The sleep EEG was done. He was treated with Brivaracetam rather than Levetiracetam.

CASE 2:

A 5-year-old boy presented with a history of episode of seizure - on January 20, 2023, the right upper limb jerks for 1 minute with an altered level of consciousness (LOC) for2 minutes on Levetiracetam orally. The Mother had a known history of LSCS due to Polyhydramnios and the child delayed a cry at birth, with a weight of 3.8 kg. He was a known case of Developmental delay and speech delay with seizure disorders. His Motor development was normal and the verbal response was good. On Examination,the child was awake, alert, and afebrile with No focal deficit. NowAdmitted with Active Seizures, Right Hand FOCALSeizures with LOC (focal generalised). CEMRI Brain The Epilepsy Protocol was completed, and the results were positive. ● Simplification in the Morphology of the Left HIPPOCAMPUS . ● Prominent Left Temporal horn of the lateral ventricle ● CSF Intensity Extra-axial Area in Right Anterior temporal fossa—likely an Arachanoid cyst. EEG was done; Paroxysmal sharp waves were noted. In Feb 2023: Daily Episodes of Seizures with a Semiology of Right Hand jerks are present. In March 2023, the boy had one episode of Right Hand Jerks with LOC for 2 Minutes and On 17.03.2023, he had reported with the same Episode of seizures. Now Admitted for further management, VEEG showed Interlectal leftoccipital and Left frontotemporal sharp present, nine recorded events were noted consists of Habitual events with EEG showing Left sided RHYTHMIC Spikes with onset from c3 evolving to involve F3 and P3, the left temporal region, and are diagnosed as Rasmussen ENCEPHALITIS.

Table 1: Laboratory Investigations

LABORATORY PARAMETERS	Case 1	Case 2	REFERENCES
Hemoglobin	12.7	11.4	11.2-14.5 g/dl
Platelet	282000	264400	150–450 × 109/L
CRP	1.0	1.0	0-5.0 mg/dl
Sodium	140	140	136-145 mmol/L
Potassium	4.53	4.10	3.4-4.7 mEq/L
Calcium	10.4	9.7	8.010.8mg/dl-
Magnesium	2.0	1.98	1.5-2.3mg/dl
Blood urea	24	22	5-18 mg/dl
Creatinine	0.42	0.43	0.8-1.3 mg/dl
Bilirubin Total	0.2	0.2	0.2-1.2 mg/dl

Bilirubin Direct	0.06	0.10.	0.0- 0.30 mg/dl
SGOT	26	23	0-35 U/L
SGPT	13.0	0.9	0-34 U/L
Total Protein	7.4	7.1	6.0-8.0 g/dL
Albumin	4.5	4.4	2.8-5.4g/dl
Globulin	2.9	2.6	2.0-3.5g/dl
Alk phosphatase	235	181	0-120U/L
GGT	13	11	0-55 U/L

Table 2: Diagnostic Criteria

Part A	
1.Clinical	Focal seizures (with or without Epilepsia partialis continua) and unilateral cortical deficit
2.EEG	Unihemispheric slowing with or without epileptiform activity and unilateral seizure onset.
3.MRI	Unihemispheric focal cortical atrophy and atleast one of the following
	Grey or white matter T2/FLAIR hyperintense signal
	Hyperintense signal or atrophy of the ipsilateral caudate head

Part B	
1.Clinical	Epilepsia partialis continua or progressive unilateral cortical deficit
2.MRI	Progressive unihemispheric focal cortical atrophy.
3.Histopathology	T cell dominated encephalitis with activated microglial cells (typically ,but not necessarily forming nodules) and reactive astrogliosis
	Numerous parenchymal macrophages,B cells or plasma cells or viral inclusion bodies exclude the diagnosis of RE.

Table 3: TREATMENT CHART:

CASE 2 DRUG NAME	DOSE	FREQUENCY	ROA	DURATION
INJ .BRIVARACETAM	50 MG /5ML	BD	IV	2 DAYS
SYP.BRIVRACETAM	(10MG/ML)5 ML	BD	CONVERTED FRM IV TO PO	3 DAYS
INJMETHYLPREDNISOLONE SODIUM SUCCINATE	700MG	OD (OVER 3 HOURS)	IV	5 days
INJ PANTOPRAZOLE	20MG	OD	IV	4 days
INJ FOSPHENYTOIN	150MG/2ML	BD	IV	3 days
T.CLOBAZAM	5MG	HS	PO	4 days
SYP.PHENYTOIN	(30MG/5ML) 10ml	BD	PO	2 days

DISCUSSION:

Here we report cases of two boys. The first boy presented with a history of a first episode of seizure, right upper limb jerks for one minute, and loss of consciousness for two minutes after 72 months of normal motor development, and the Second boy presented with the chief complaints of clusters of seizure episodes. The first episode was unprovoked seizures with LOC for 1 to 2 minutes. The second episode dealt with the semiology of angle of mouth deviation to one side, with a duration of 1 to 2 minutes. The diagnosis of RE is made using clinical and neuroradiological criteria that are based on the 2005-proposed European consensus criteria. The chronic inflammatory nature of the disease is indicated by the progressive MRI degradation.

CONCLUSION

A rare condition of the central nervous system known as Rasmussen encephalitis, sometimes known as Rasmussen syndrome, is characterised by chronic,

increasing inflammation (encephalitis) of one cerebral hemisphere. This case series emphasises the difficulties in diagnosing and treating paediatric-onset RE. Because early and quick intervention with surgery can enhance patient prognosis, its detection is crucial.

REFERENCES

1. Vimal J, Nagarajan R, Sriram D. (2015). Rasmussen's encephalitis: A case report. *Australas Med J* 8, 80–4.
2. Hammed A, Badour M, Baqla S, Amer F. (2021). Diagnosis and treatment of Rasmussen's encephalitis pose a big challenge: Two case reports and literature review. *Ann Med Surg (Lond)* 68, 102606.
3. Rasmussen Encephalitis. National Organization for Rare Disorders 2015. https://rarediseases.org/rare-diseases/rasmussen-encephalitis/ (accessed July 21, 2023).
4. Wycliffe ND, Holshouser BA, Bartnik-Olson B, Ashwal S. (2017), Pediatric Neuroimaging. *Swaiman's Pediatric Neurology, Elsevier*; 78–86.
5. Bien CG, Bauer J, Deckwerth TL, Wiendl H, Deckert M, Wiestler OD, (2002). Destruction of neurons by cytotoxic T cells: A new pathogenic mechanism in rasmussen's encephalitis. *Ann Neurol* 51, 311–8.

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