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## Case Report

# Role of diagnostic imaging in Rasmussen's encephalitis – A case report from Nepal <sup>☆</sup>

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## ABSTRACT

Rasmussen's encephalitis (RE) is a relatively rare chronic inflammatory neurological disease that usually only affects one hemisphere of the brain. It primarily affects children under the age of 10, although it can also affect teens and adults, causing drug-resistant seizures, progressive hemiparesis, and dementia. RE presents as a challenging diagnosis with MRI as the cornerstone of the evaluation and nuclear imaging as a complementary tool. We'd like to present a case of a 12-year-old girl who was diagnosed with RE after an MRI. In this study, we examine the diagnostic criteria, differential diagnoses, and issues that underpin the diagnostic challenge in great detail.

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## Introduction

Rasmussen's encephalitis (RE) is a rare nervous system disease that primarily affects children. It is distinguished by unilateral hemispheric atrophy, focal intractable seizures, and deterioration of neurological deficits. Rasmussen et al. [1] published a clinicopathological report of three children with a longstanding illness causing focal seizures and worsening damage to one cerebral hemisphere, who was the first to describe the condition. Though the cause of this sporadic disease is unknown, cytotoxic T cell responses against neurons have

been linked to its pathogenesis. Imaging aids in diagnosis by ruling out other potential causes and aids in disease progression and monitoring [2]. Despite having a childhood ailment, adult and teenage patients account for 10% of all cases, with the oldest patient aged 58 years as reported in the literature. Clinical manifestations occur at an average age of 6 years. The following are the 3 clinical stages that have been proposed:

1. The prodromal stage lasts an average of 7.1 months (range: 0 months to 8.1 years) and is characterized by low seizure frequency and mild hemiparesis.

**Abbreviations:** MRI, Magnetic Resonance Imaging; RE, Rasmussen's encephalitis; CSF, Cerebrospinal fluid; EEG, Electroencephalogram; PLEDs, Periodic lateralized epileptiform discharges; FLAIR, Fluid-attenuated inversion recovery; MRS, Magnetic Resonance spectroscopy; NAA, N acetyl aspartate.

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2. The acute stage lasts for eight months on average and is marked by frequent seizures. If the dominant hemisphere is affected, the neurological symptoms manifest as progressive hemiparesis, hemianopia, cognitive decline, and aphasia.
3. The residual stage is the final stage, characterized by irreversible damage and fewer seizures than the acute stage.

After 8 to 12 months, children with RE frequently enter a phase of permanent but stable neurological deficits; while the disease may progress slowly in adults and adolescents [3].

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## Case report

A 12-year-old female patient presented to the Neuromedicine outpatient department of Bir Hospital, Kathmandu with complaints of multiple seizure episodes. Her mother reported that such episodes started when she was 1-year-old. According to her mother the episode starts as focal jerking of left upper limb which spread to her lower limbs progressing to loss of consciousness and jerking of all four limbs. A thorough physical examination was performed which revealed power of 4/5 in left upper and lower limb. However, she was conscious, oriented to time, place and person with intact higher mental function. She did not show any extrapyramidal or cerebellar signs on presentation. Furthermore, her vitals and systemic examination were within normal limits. A series of blood tests were done which showed leukocytosis with counts of 11,500/cumm, however, she had normal liver and kidney function tests. Her blood glucose was within the normal range and she tested negative for vasculitic markers. She underwent a lumbar puncture to evaluate CSF which showed albuminocytologic dissociation and normal sugar level with negative culture reports. In addition blood and urine cultures also reported no growth of organism. Subsequently, she underwent an EEG which showed generalized slowing of waves in theta range. To further evaluate the condition MRI was performed whose imaging findings are reported below with relevant images. During her admission, her seizure episodes were controlled by levetiracetam and carbamazepine. Her family reported history of using a similar drug name unknown during her previous seizure episodes at home.

Our patient is a 12-year-old female who presented with a history of intractable seizures from her early life. With the clinical findings of seizures, neurological deficits and the aforementioned findings on the MRI of the brain and EEG, diagnosis of Rasmussen's encephalitis was made.

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## Discussion

RE is a chronic brain disorder of unknown etiology that causes refractory seizures with progressive neurological deficits (4). Mean age at presentation is 6-8 years of age with no gender preference. Exact incidence of this disease is not exactly known. It was first described by Theodore Rasmussen et al. [4] in the late 1950s after study among three patients who pre-

sented with intractable seizures caused by chronic progressive encephalitis.

The cause of RE is uncertain, with some research pointing to viral infections and others defining it as an autoimmune disease involving antibodies against a glutamate receptor protein. RE manifests clinically as epilepsy partialis continua, which is followed by hemiparesis and cognitive impairment as the disease advances [1]. There are no known specific EEG findings in epilepsy partialis continua patients, but they can include spikes, sharp waves, and periodic lateralized epileptiform discharges (PLEDs) with slow-wave activity.

RE is diagnosed using a combination of clinical, radiographic, and pathological symptoms, with a focus on clinico-radiological features because brain biopsy is not always performed due to its invasive nature.

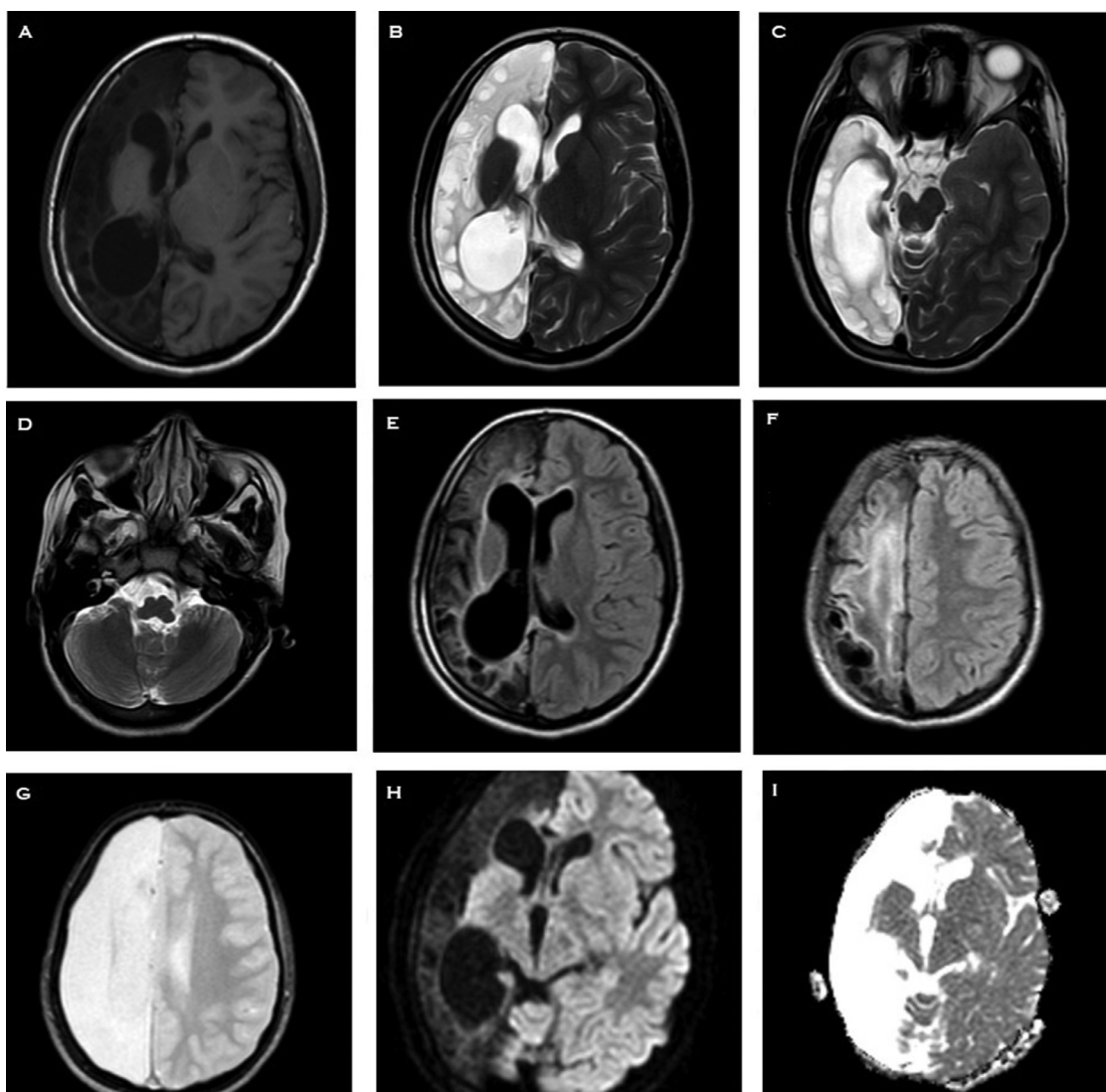
There is a three stage natural history of RE on the basis of long term observation of 13 patients as performed by Bien et al. [5]. The first stage being nonspecific and presenting with relatively low seizure frequency and on rare occasion mild hemiparesis. The second stage characterized by increased frequency of seizures and increase in degree of hemiparesis. The final stage is characterized by permanent and stable neurological deficits and severe hemiparesis but decrease in seizure frequency [3].

The earliest abnormal MRI finding is cortical swelling with high signal intensity in the affected hemisphere in T2/FLAIR sequences, which lasts for up to 12 months. In the second stage findings include focal or multifocal T2 and FLAIR high signal intensity areas involving the cortex or white matter of the affected hemisphere, which lasts for up to 22 months. Later on, there is atrophy of affected hemisphere characterized by widening of sulci and dilatation of lateral ventricle on the affected side (Stage 3). The final stage (stage 4) is characterized by the disappearance of the abnormal signal, leaving a substantially atrophied cerebral hemisphere [5].

CT is inferior to MRI study although there may be similar imaging findings. The superiority of MRI study over CT scan is that signal changes in the affected hemisphere appear earlier which is detected in MRI. The role of contrast use in the patients with RE is not well identified. MRS study may show increase choline and decrease in NAA owing to neuronal loss (Fig. 1) [6].

There is no laboratory test that supports the diagnosis of RE. Some studies suggest increase in lymphocyte count and protein level in CSF study which was observed in a few numbers of cases. Standard CSF testing, on the other hand were ineffective in confirming or eliminating the diagnosis [3].

Similar MRI findings can be observed in Dyke-Davidoff-Masson syndrome, Sturge-Weber syndrome, and hemimegalencephaly. Dyke-Davidoff-Masson syndrome (DDMS) is characterized by unilateral cerebral atrophy with ipsilateral calvarial thickening and hyper-pneumatization of paranasal sinuses. The cause may be congenital or acquired. In the congenital variety, cerebral insult usually has a vascular origin. In acquired type, cerebral insults occur during perinatal period or later in life which may be as a result of trauma, intracranial hemorrhage, infection, and ischemia. Since, insult to brain in DDMS occurs earlier in intrauterine or perinatal life, there is compensatory overdevelopment of paranasal sinuses,



**Fig. 1 – (A–I) Right cerebral hemiatrophy showing low signal intensity in T1WI (A), high signal intensity in T2WI (B and C), Affected hemisphere show few areas of FLAIR high signal intensity, likely gliotic changes (E and F), No blooming foci noted in the affected hemisphere on GRE sequences (G), No areas showing diffusion restriction in the affected hemisphere in DWI/ADC sequences (H and I). Ex-vacuo dilatation of lateral ventricle in the affected right cerebral hemisphere, likely due to volume loss. No calvarial changes, dilated paranasal sinuses noted. Normal MRI findings in left cerebral hemisphere.**

mastoid air cells and calvarial thickening of the affected side which helps the differentiation from RE [1].

Sturge-Weber syndrome is a neurocutaneous syndrome which manifests with capillary telangiectasia and capillary-venous malformations in the distribution of trigeminal nerve, leptomeningeal angiomas, seizures, dementia, and hemiplegia. Distinctive imaging findings of Sturge-Weber syndrome are cerebral atrophy with gyral or curvilinear calcification (tram track) which differentiates this entity from RE [5].

Hemimegalencephaly is characterized by enlargement of unilateral cerebral hemisphere with ipsilateral ventriculomegaly which occurs as a result of partial or total hamartomatous overgrowth of all or part of the cerebral hemisphere. There may be focal or diffuse neuronal migration defects, with areas of polymicrogyria, pachygyria, lissencephaly, and agyria.

Typical features of the affected hemisphere include increased lateral ventricle size, shallow sulci and enlarged gyri, enlarged/thickened calvarium and contralateral displacement of the posterior falx [7].

Hemispherectomy has been the most efficient option to eradicate seizures and prevent further deterioration in cognitive function in RE patients [1].

## Conclusion

Rasmussen's encephalitis is a rare disease and is a diagnostic dilemma for many clinicians. A history of refractory seizures starting in a young age should raise suspicion for Rasmussen's

encephalitis. Knowledge about the clinical stages and radiological diagnosis is essential to solve this diagnostic puzzle. In a country like Nepal with scarce diagnostic imaging centers in rural areas, it is even more difficult for patients to get timely diagnosis and treatment albeit treatment is only supportive.

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**Peer review**

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**Patient consent statement**

Consent for this case report was taken from the mother of the patient in written form, after explaining about the case.

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