



Rasmussen's Encephalitis: A Case Report

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ABSTRACT	Rasmussen's Encephalitis is a rare, chronic inflammatory neurological disease of unknown origin that usually affects only one hemisphere of the brain. It is usually seen in children less than 10 years of age, in 10% cases it has been reported in the adult population too. It is characterized by intractable seizures with progressive neurological deterioration. Our case is of a 15 years old female, who presented to our hospital with complaint of generalised tonic clonic seizures and hemiparesis. On the basis of clinical presentation, MRI findings and EEG she was diagnosed as a case of Rasmussen's Encephalitis.
KEYWORDS	

Introduction

Rasmussen's encephalitis (RE) is an uncommon disease of the nervous system that is usually seen in children. It is characterised by unilateral hemispheric atrophy, focal intractable seizures, and worsening neurological deficits. Historically, the condition was first described by Rasmussen et al. who published a clinico-pathological report of three children with a longstanding illness causing focal seizures and worsening damage to one cerebral hemisphere.¹ Though considered as an illness in children, adult and adolescent patients account for 10 per cent of all cases² with 58 years being the oldest reported age.³ Onset of symptoms in the adult age group is associated with a more protracted and milder clinical course with less residual functional deficit, lower degree of hemiatrophy, and more frequent occipital lobe seizure onset.^{2,4}

The average age of clinical presentation is six years.^{2,5} Three clinical stages have been proposed:

1. The prodromal stage, which has a mean duration of 7.1 months (range: 0 months to 8.1 years),^{2,5-7} has a low seizure occurrence, and mild hemiparesis.
2. The acute stage has a median duration of eight months^{2,5} and is characterised by frequent seizures. The neurological symptoms become apparent in the form of worsening hemiparesis, hemianopia, and cognitive deterioration and aphasia if the dominant hemisphere is involved.^{2,5}
3. The residual stage is the last stage with permanent damage and seizures being less frequent than in the acute stage.^{2,5}

This complex case describes an adult patient who presented with what was eventually diagnosed as Rasmussen's encephalitis.

Case Report:

A 16-year-old female presented to the emergency department with sudden onset of generalised tonic clonic movements of both upper and lower limbs associated with loss of consciousness, and frothing at the mouth. On physical examination she was in status epilepticus with generalised tonic clonic seizures. Her pulse rate was 110 beats/minute, blood pressure was 130/90mmHg, and interictal respiratory rate was 20 cycles per minute. On CNS examination patient had left upper and lower limb hypertonia, with power of 3/5, hyperreflexia, and an extensor plantar response. The rest of the central nervous system and systemic

examination were within normal limits. She had normal body temperature, oxygen saturation of 95 per cent on room air and Glasgow coma scale of 3/15. She was intubated and started on infusion of midazolam (as seizures could not be controlled with lorazepam, phosphenytoin, and levetiracetam) with which her seizures stopped.

She had history of similar episode seizure before 12 months following which she had weakness of left upper and lower limb with slurring of speech which recovered gradually and completely over period of 4 months. Following recovery patient stopped antiepileptic therapy which she was put on following this 4 month interval. She remained seizure free until she presented to the emergency department with episode of seizure.

Blood investigations of patient revealed leucocytosis with neutrophilic predominance (12,000/cu.mm and 61 per cent neutrophils). CSF analysis showed cytoalbuminodissociation [total wbc count-2 (all lymphocytes) and protein of 172mg%], and normal sugar level. The rest of the blood counts, renal function tests, liver function tests, vasculitic markers, electrolytes, blood glucose level, chest radiograph, and electrocardiogram were all within normal limits. The urine, blood, and cerebrospinal fluid cultures were all sterile. An MRI of the brain with gadolinium enhancement was performed. This showed atrophy of the right cerebral and left cerebellar hemispheres with dilatation of the right lateral and third ventricles, and hyperintensity of the grey and white matter of the right cerebral hemisphere on FLAIR. EEG was performed which showed presence of bilateral hemispheric dysfunction (more on the right hemisphere) with active epileptiform activity.

With the clinical findings of seizures, cortical deficit, and the aforementioned finding on the contrast-enhanced MRI of the brain and EEG, a diagnosis of Rasmussen's encephalitis was made.

Discussion:

This patient probably presented in the acute stage of Rasmussen's encephalitis as she exhibited hemiparesis and other neurological symptoms, despite the large time span between the first and second seizures. Most of the tissue loss occurs within the first year of onset of the acute stage.^{2,5} The aetiology and pathophysiology of RE remain elusive. Four groups have been described to assist in classifying the pathophysiology. Group 1 has inflammation with microglial nodules; group 2 reveals at least one gyrus segment of

complete necrosis; group 3 shows neuronal loss and gliosis; and group 4 displays gliosis and glial scarring.^{2,5,6,7}

There is no laboratory test that positively supports the diagnosis of RE. In a large series of CSF tests, in about one-half of the examinations cell counts and protein levels were within the normal range, and in the remainder, elevated cell counts (16–70 cells/ml, predominantly lymphocytes), and increased protein level (50–100mg/dl) were observed.² Standard CSF tests are not useful in confirming or excluding the diagnosis. An MRI of the brain in most RE patients shows unilateral enlargement of the CSF compartment with maximum accentuation in the insular and peri-insular region, and increased signal intensity in the cortical, subcortical, or both regions. EEG shows unihemispherical impairment of background activity and sleep spindles, focal slow activity, multifocal ictal discharges, and subclinical ictal discharges. Our patient had MRI and EEG findings that suggested bilateral involvement, a relatively rare entity in patients with RE. However, the caudate nucleus, insular, and peri-insular regions were normal. Other investigations that can be performed to help confirm the unilateral nature of suspected RE include positron emission tomography (PET), single photon emission computed tomography (SPECT), and magnetic resonance spectroscopy (MRS).² Brain biopsy is not needed for all patients as diagnosis can be made without it as well. The goals of therapy are to decrease inflammation, restore functional capacity, and control seizures. The various modalities of treatment available to achieve these goals include pharmacological, immunotherapeutic, surgical, and rehabilitative techniques.

The female presented here had a reasonable quality of life before this episode of status epilepticus, hence although surgical intervention would result in amelioration of seizure symptoms, it would also cause a disabling post-operative functional deterioration. The early institution of long-term immunotherapy to prevent functional decline was the recommended line of therapy.^{2,8} However, due to financial constraints (cost of immunotherapy), and after a detailed discussion with the family about the potential benefits and side effects, it was decided to manage him with antiepileptic drugs and physiotherapy. Monitoring of clinical improvement in these patients can be undertaken by follow-up of the hemiparesis,^{2,5} neuropsychological performance in patients without hemiparesis,² and hemispheric ratio on neuroimaging. The hemispheric ratio is the ratio of affected/unaffected hemisphere on planimetry of axial and coronal slices, including the sylvian fissure.

Conclusion:

RE is an uncommon cause of seizure and bilateral cortical involvement, such as that seen in this case, is a rare entity. Although rare in adults, RE should be considered as a possible diagnosis in patients who present with intractable seizures. As the aetiology is largely uncertain, the treatment is mainly symptomatic with other modalities being far from definite, and in India, being available only to those who can afford it.

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