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 6years old male, presented to emergency department with complaint of intractable severe seizures, progressive hemiparesis and deteriorated cognition followed by an episode of encephalitis. His course of illness was focal seizures and right-sided weakness (hemiparesis) for 1 year. In addition to classical clinical presentation of Rasmussen's Encephalitis, MRI Brain showed gliotic area involving the left superior and middle frontal gyrus that supports diagnosis of Rasmussen's Encephalitis.

Introduction : Rasmussen's encephalitis (RE) is a rare neurological disorder of inflammatory aetiology characterised by encephalitis, intractable seizures, hemiparesis, variable motor deficits, and dementia. [1] Historically, the condition was first described by Rasmussen et al. who published a clinico-pathological report of three children with alongstanding illness causing focal seizures and worsening damage to one cerebral hemisphere.

The average age of clinical presentation is six years[2,3,4] 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), has a low seizure occurrence, and mild hemiparesis. [2,3,4]
2. The acute stage has a median duration of eight months 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.[2,3,4]

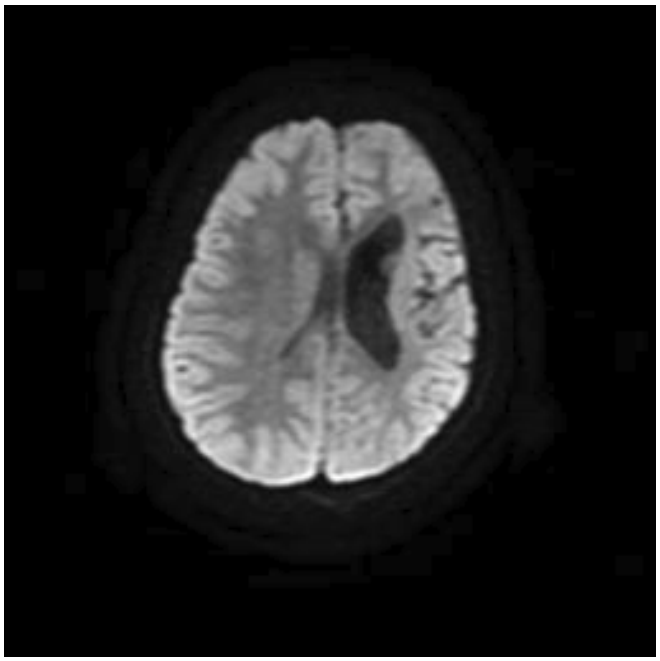
The residual stage is the last stage with permanent damage and seizures being less frequent than in the acute stage.[2,3,4]

Here we report a case of an child presenting with a common symptom of seizure caused by Rasmussen's encephalitis.

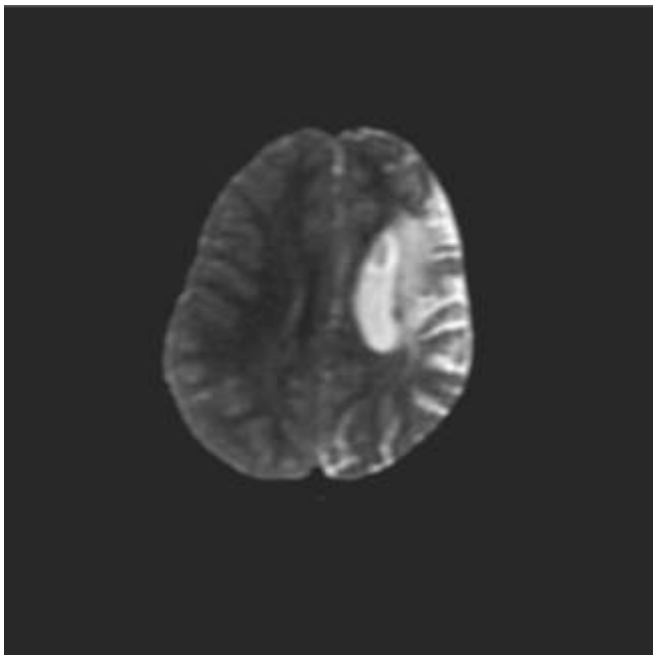
Case report : A 6years male child presented with history of three episodes of Convulsion since one year and was associated with right sided hemiparesis.Initialy there was clonic movements of the right upper and lower limb for 5 months. Later on, they progressed to focal movement of the right leg associated with difficulty in walking and weakness in movement of right hand

Fig1. Right sided hemiparesis

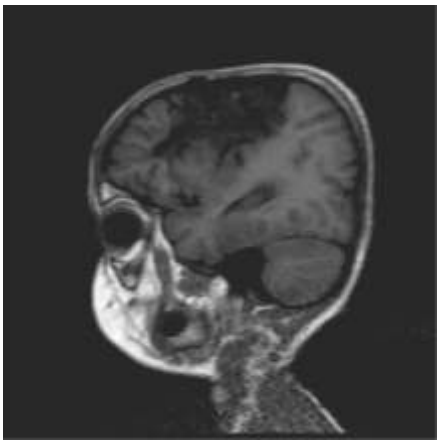
For the past one year, he is having mild orbito-frontal headache and decreased vision of both eyes . There is no significant birth history and developmental milestones had been normal. On examination, visual acuity in both eyes was 6/24 and the right upper & lower limb showed decreased tone and power (3/5).Routine blood and cerebrospinal fluid investigations and metabolic tests were within normal limits. His electroencephalogram shows focal left fronto-temporal epleptiform theta activity left>right and poly spikes left>>right, diagnostic of complex partial seizures. Neuroimaging studies revealed gliotic area involving the left superior and middle frontal gyrus.The patient was initially treated with antiepileptic medication. Treatment with prednisolone was started later based on the diagnosis of RE. Motor function of the right leg improved mildly. Partial control of the seizures was attained. The clinical condition remained almost static with medication on follow-up for seven months.



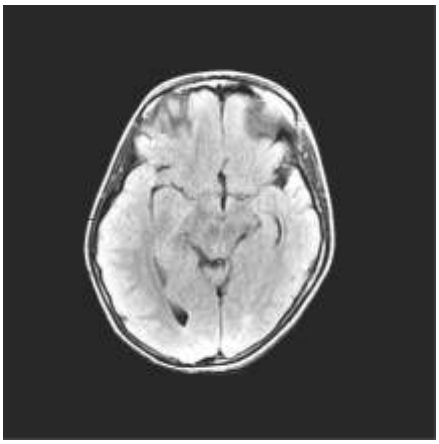
Cross sectional view (Plain)



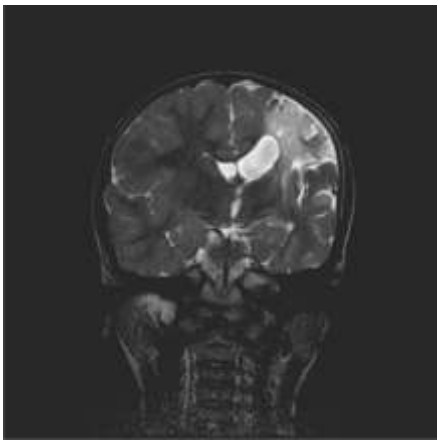
Cross sectional view(Contrast)



Sagittal view



Cross sectional view



Coronal view

The patient was initially treated with antiepileptic medication. Treatment with prednisolone was started later based on the diagnosis of RE. Motor function of the right leg improved mildly. Partial control of the seizures was attained. The clinical condition remained almost static with medication on follow-up for seven months.

Discussion : RE is a sporadic chronic inflammatory disease of the central nervous system occurring mostly in the pediatric population, first reported by Theodore Rasmussen in 1958. The mean age of presentation is between 6 to 8 years. Both sexes are equally affected.1The etiology of RE is unknown, with some earlier studies suggesting the role of viral infections, while others describing it as an autoimmune phenomenon involving antibodies against a protein of glutamate receptor.[3,4]The differential diagnoses for RE include Dyke-Davidoff-Masson syndrome, Sturge-Weber syndrome, hemimegalencephaly, and unihemispheric cerebral vasculitis. The early institution of long-term immunotherapy to prevent functional decline is the recommended line of therapy.

Conclusion : RE is an uncommon cause of seizure and cortical involvement, such as that seen in this case, is a rare entity. 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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