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Rasmussen's Encepalitis: A Rare Case Report With Atypical Features

KEYWORDS: Rasmussen's encephalitis, intractable epilepsy, MRI, neurological deficit, atypical features

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ABSTRACT *Rasmussen's encephalitis (RE) is a devastating syndrome of uncertain origin which is associated with chronic encephalitis and focal refractory seizures. The disease usually tends to localize in one hemisphere, most commonly involving fronto-insular regions. The slow insidious progression of disease results in varying degree of neurological deficit and mental deterioration. The seizures may eventually evolve into epilepsia partialis continua (EPC). We present a case of Rasmussen's encephalitis in a 6 year old child occurring at the unusual site and associated with gelastic seizures without any neurodeficit.*

INTRODUCTION

Rasmussen encephalitis is a chronic inflammatory disease which is of uncertain origin and usually affects one hemisphere. [1-3] Rasmussen and his colleagues in 1958 were first to report a syndrome of chronic encephalitis and refractory focal seizures now is termed as Rasmussen's encephalitis (RE). [1-7] The disease typically begins in childhood with refractory focal motor seizures which gradually evolve into epilepsia partialis continua (EPC) which is followed by progressive hemiparesis and mental deteriotation. [2,4, 6-8] Here we report a case of RE in a child presenting with atypical clinical features (gelastic seizures with absent hemiparesis) and predominantly involving insular cortex and temporo-parieto-occipital lobes with sparing of frontal lobes.

CASE REPORT

A 6 year old male child presented to us with the history of seizures since 3 and a half years. The seizures consists of involuntary shaking of left upper limb and twitching of left eye followed by generalized tonic clonic activity lasting approximately for 5 to 10 minutes. The seizures were progressively increasing in frequency and severity despite of antiepileptic treatment. Sometimes the seizures were followed by continuous involuntary blinking of left eye which persisted during activity and sleep. Since last 15 days child had developed gelastic seizures in the form of abnormal episodic outbursts of laughing lasting for 4 to 5 minutes. On clinical examination, the child was temperamentally stubborn. The cognition was impaired. There was no any evidence of apparent hemiparesis. EEG showed slow activity with multifocal polyphasic bursts and spikes on the right side. MRI brain revealed right hemispheric atrophy and increased signal intensity in right insular cortex and temporo-parieto-occipital lobes on T2W and FLAIR sequences with dilatation of ipsilateral lateral ventricle and adjacent sulcal spaces. The T2W and FLAIR MR findings are shown in Figure 1 and 2 respectively. MR spectroscopy (MRS) revealed decreased N-acetylaspartate (NAA) levels in the affected hemisphere which is depicted in Fig. 3.

DISCUSSION

RE is a disease of uncertain origin which primarily affects the children below 10 years of age with mean age of presentation between 6 to 8 years. [4, 6, 7] The disease usually tends to localize in one hemisphere. [2,6] The patient initially presents with intractable focal seizure with a predominant motor component. [4, 6, 8-10] The slow and insidious progression of the disease may result in varying degree of hemiparesis, aphasia and cognitive impairment .[2,4, 6, 8]

Our patient presented with progressive refractory simple motor partial seizures with secondary generalization which is second most common type of seizure reported in RE patients.[7] In addition, our patient also developed gelastic seizures lately. Gelastic seizures are an extremely rare form of epilepsy defined as automatic bouts of laughter without mirth commonly associated with a hypothalamic hamartoma. [10,11] None of the previous studies have reported any association between RE and gelastic seizures.

Although cognitive impairment was present in our patient there was no apparent hemiparesis. Few studies have reported similar findings where no apparent hemiparesis was present in the RE patients despite of diffuse hemispheric atrophy. [1,5]

The diagnosis of RE rests on clinical features, EEG and MRI findings. [12]EEG findings in our patient are in agreement with the previous studies and are consistent with diagnosis of RE. [4, 7]

The typical MRI findings of RE are described in literature and include predominant unilateral distribution, focal cortical atrophy usually involving fronto-insular region, white matter or cortical high signal intensities on T2W and FLAIR sequences, atrophy of head of caudate nucleus and enlargement of lateral ventricles. [3- 6, , 8-10, 12]

In our patient, the disease was limited to right insular cortex and temporo-parieto-occipital lobes while frontal lobes were not involved. Very few studies in literature have reported involvement of temporo-parietal and occipital lobes. [1, 8] The high signal intensity lesions in the gray and white matter on T2W and FLAIR are attributed to reactive gliosis caused by chronic brain damage. [5-8]We also noticed cortical and white matter hyperintensities in affected hemisphere on T2W and FLAIR in our patient. Although head of caudate nucleus was not atrophic in our case but ipsilateral lateral ventricle and adjacent sulcal spaces were dilated.

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On MRS, the affected areas in RE reveal decreased NAA which correlates well with brain atrophy and neuronal loss. [3,5, 6, 8]In our case also, NAA and choline levels were remarkably decreased in the atrophic areas consistent with neurodegenerative changes and implying neuronal death.

Treatment of children with RE has so far been disappointing. [8] Medication with antiepileptic agents is ineffective or are usually only partially effective. [6, 7] Surgical management with hemispherectomy is the only successful alternative. [2,4, 10]

Thus, despite of atypical clinical presentation and unusual site, the characteristic EEG and MRI findings may help in diagnosis of RE.

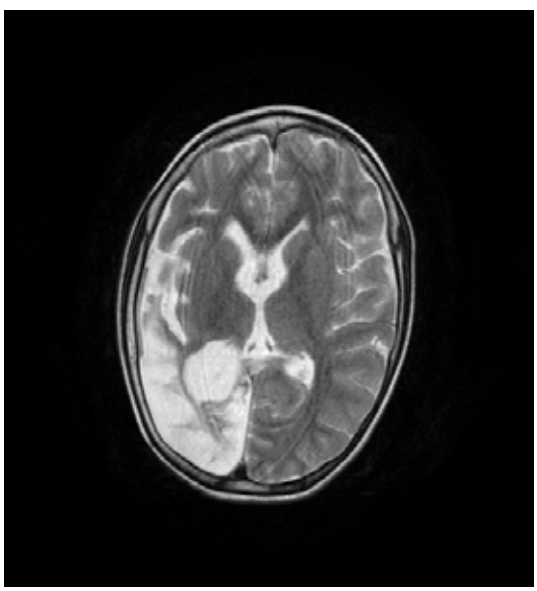

Figure 1: Axial T2W MRI brain reveals increased signal intensity in right insular cortex and temporo-parieto-occipital regions with dilated ipsilateral lateral ventricle with prominent adjacent sulcal spaces.

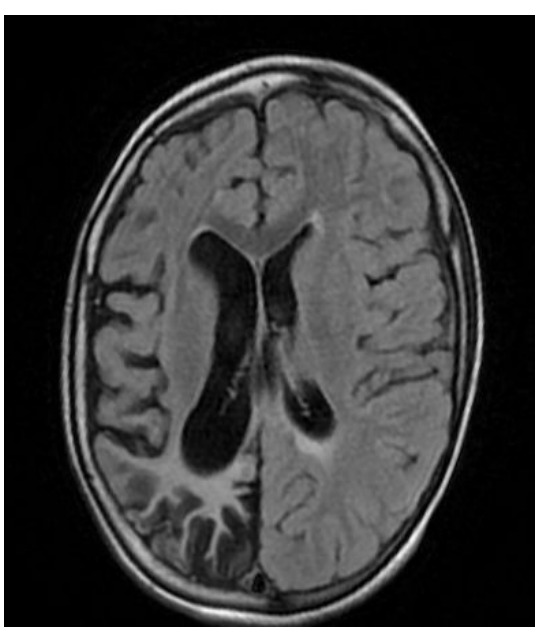

Figure 2: Axial FLAIR MRI brain reveals atrophy of right hemisphere with hyperintensities in periventricular deep white matter and dilated ipsilateral lateral ventricle.

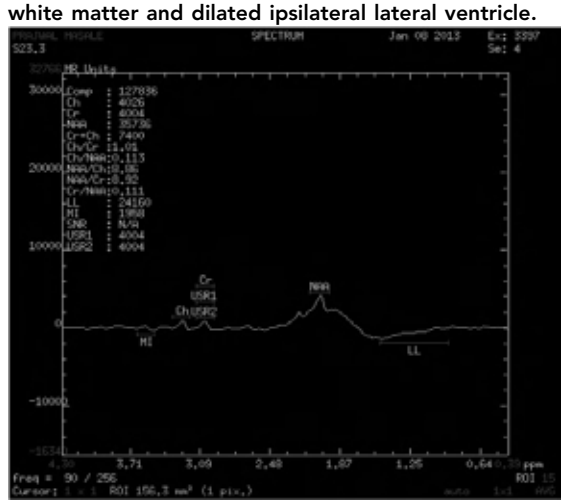

Figure 3: MR spectroscopy reveals decreased NAA and choline levels in atrophic areas in right hemisphere

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