British Journal of Research

www.britishjr.org

Case Report

Atypical Presentation of Benign Rolandic Epilepsy: A Case Report

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A B S T R A C T

Epilepsy is a common neurological disorder in childhood. Five percent of children of estimated one or more seizures in childhood, less than 1% have epilepsy. The incidence is high in preschool years. Benign rolandic epilepsy (BRE) or Benign Epilepsy with Centro Temporal Spikes (BECTS) is defined as truly benign partial idiopathic epilepsy characterised by partial motor seizures usually brief, infrequent with or without generalisation and onset during childhood. They occur mostly during sleep and these children recover spontaneously before adolescence without any neurological or cognitive impairment. We report a four year female child with Rolandic seizures who presented with abnormal behaviour and atypical EEG recordings.

Keywords: Benign rolandic epilepsy, Centro temporal spikes, Abnormal behaviour.

INTRODUCTION

Epilepsy is common neurological disorder of the childhood. It is characterised by recurrent, episodic, paroxysmal, involuntary clinical events associated with abnormal electrical activity form neurons. Benign Rolandic Epilepsy is also called as Benign Epilepsy with Centro Temporal Spikes or Sylvian Epilepsy. It is the most common epilepsy of childhood affecting previously healthy children aged 4-13 years with an incidence of 10-20 per 1,00,000 children under age of 15.[1]

It is characterised by twitching, numbness or tingling of face or tongue and may interfere with speech and cause drooling of saliva. Seizures are focal and involve unilateral sensory motor function of face, speech arrest and hypersalivation is seen. Consciousness is generally maintained although seizure may progress to become hemiconvulsive or generalised tonic clonic.

Atypical forms may present with atypical manifestation such as onset at early age, developmental delay, learning

difficulties, other seizure types, atypical EEG abnormalities.[2-4]

The same EEG pattern was later correlated with a common form of focal childhood epilepsy, then called mid temporal epilepsy, characterized by hemifacial and oro pharyngeal ictal symptoms and a favorable prognosis. Because of the localization of the ictal events to centro temporal area, Lombroso proposed the term sylvian seizures in 1967. In the same year, Loiseau and colleagues presented a series of 122 children with what they called a particular form of epilepsy in childhood, stressing its benign character and highlighting clinical and EEG features. Atypical and not so benign evolutions have been reported in some patients with this form of epilepsy.[2] This form of epilepsy is now called benign childhood epilepsy with centrotemporal spikes and is placed in the group of idiopathic localization-related epilepsies in the International Classification of Epilepsies and Epileptic Syndromes.[1] About 79 children with benign rolandic epilepsy were reported who together had more than 900 seizures over an 8.5 year period. There were no significant injuries.

A particular EEG pattern with migratory spikes originating over Rolandic (Centro temporal or Mid temporal) region was first reported in 1952 by Gastaut and in 1954 by Gibbs et al. In 1958 the first description of clinical features in association with peculiarities of EEG in those children was published by Nayrac.

CASE REPORT

A four year female child presented to casuality with history of abnormal behaviour since afternoon. The child was born to non-consanguinous parents, first in order of birth, with other sibling normal. No family history of similar complaints and any neurological diseases. She was born at full term through normal vaginal delivery without any perinatal problems. Child was immunised till date.

The Neuro Psychomotor development of the patient was globally normal. Child had presented with aggressive behaviour two months back and a presumptive diagnosis of ADHD was made by a Neuro Psychiatrist and child was on regular treatment with Sodium Valproate 2.5 ml twice daily and then Haloperidol 0.25 mg twice daily was started two days before she presented to our hospital. The child suddenly developed abnormal behaviour characterised by starring look, not able to swallow food or drink water, drooling of saliva, not able to walk and not able to speak to any one for about five minutes. When the child presented to the casuality in the evening she was active and interactive but suddenly developed ophisthotonus posture with head turning to one side for about two minutes. There was no loss of consciousness during the episode and no post ictal amnesia. There was no history of fever. The child had a head injury 6 months back. There was no history of vomiting, headache, convulsions or loss of consciousness after the head injury.

On Neurological examination the power was Grade 5 and tone was normal in all the limbs. The reflexes were normal and signs of meningeal irritation were absent. EEG showed Fronto Centro Parietal epileptiform discharges. CT Scan was normal. With the history and EEG findings, diagnosis of Benign Rolandic Epilepsy was considered and treatment with Sodium Valproate continued with appropriate dose and there were no fresh episodes.

DISCUSSION

Benign Rolandic epilepsy represented 9.6–10.3 percent of all childhood epilepsies, determined at presentation and 2 years later. In its pure form, it is not associated with

structural lesions or severe neurocognitive deficits.[5]

Familial EEG studies suggest that the benign epilepsy with centro temporal spikes follows an autosomal dominant mode of inheritance with high but incomplete penetrance and age dependant expression.

Loiseau and Douche provided 5 criteria for diagnosis of benign childhood epilepsy with centro temporal spikes:[6]

1. Onset between 2-13 yrs age
2. Absence of neurological or intellectual deficit before onset
3. Partial seizures with motor signs, frequent association with somato sensory symptoms or precipitated by sleep.
4. A spike focus located in centro temporal (Rolandic) area with normal background activity on the interictal EEG.
5. Spontaneous remission during adolescence.

When awake, children experience brief focal twitching of one side of face, anarthria, drooling of saliva and parasthesia of face, gums, tongue or inner cheeks. These manifestations may be followed by hemiclonic movements or hemitonic posturing. These diurnal seizures are simple and consciousness is preserved. Post ictal weakness of the involved face and limbs may occur.

Most of the children have purely nocturnal seizures that usually become secondarily generalised. In such cases, the focal onset of seizures usually is not observed, but parents are alerted by sounds of secondarily generalised convulsion.

The typical rolandic seizure is hemifacial, characterised by a clonic manifestations involving hemi face, sometimes preceeded by unilateral parasthesia involving tongue, lips, gums, cheek; the jerks can be associated with a lateral tonic deviation of mouth involving lips and tongue as well as pharyngeal and laryngeal muscles and result in anarthria or speech arrest and drooling of saliva due to sialorrhoea and saliva pooling.[7] The seizure lasts from less than a minute to 2 minutes can spread to homolateral arm and rarely leg.[8] Sometimes the seizures consists of a brief somatosensory phenomena and in 4.3% cases seizures are partial complex.[9] Seizure frequency is usually low.

In children aged 2-5 years Hemi clonic seizures are more frequent, sometimes lasting from more than 30 to 60 minutes followed by transient homolateral deficit generally not including face.[9] In all case intra venous Benzodiazepines immediately stop status even if it is of long duration. No permanent deficits are seen.

EEG findings are distinctive and diagnostic in benign focal epilepsy with central-mid temporal spikes. Focal di or triphasic sharp waves of almost invariant morphology occur in the central and mid temporal regions. Epileptiform discharges usually are of high voltage (greater than 100 mV), tend to occur in clusters, and activate dramatically during sleep, when they may seem almost continuous. In a single EEG, discharges may be unilateral, but with prolonged or repeated recordings, they are almost always bilateral. Lateralization may switch in serial tracings. Generalized spikes and spike-and-wave activity occasionally occur, although more often the spikes are maximal in either central region and are simply bisynchronous. No correlation has been found between EEG findings and seizure occurrence or frequency. As a rule, EEG abnormalities are much more impressive than clinical seizure activity. Indeed, when central-midtemporal spikes are recorded in children without seizures, in whom EEGs are performed for other reasons, typical seizures eventually develop in only approximately half of the children. In symptomatic children, EEG abnormalities persist long after seizures cease. Thus, EEG does not provide assistance in making decisions about when or how long to

treat.[6] Epileptiform activity may be augmented in sleep and rarely other focal discharges or generalised spike waves have been reported. Three quarters of Rolandic seizures occur during Non-Rapid eye movement sleep, often during transition.

Wirrell and colleagues published a retrospective case series design of 42 children diagnosed with benign rolandic epilepsy in whom they looked for atypical features, which they found in 50% of the patients.[4] These features do not coincide with those that we now consider atypical. Since then, more comprehensive and prospective studies have focused on the correlation between EEG abnormalities and neuropsychological impairments in children with benign rolandic epilepsy. Atypical features in benign childhood epilepsy with centrotemporal spikes can be seen on clinical grounds like daytime-only seizures, postictal Todd paresis, prolonged seizures, or even status epilepticus, or in EEG features like atypical spike morphology, unusual location, or abnormal background.[10] Early age of onset of seizures seems to be one of the most important items among atypical features.[2]

Treatment is often not necessary due to low seizure frequency. The first line drug used is Carbamezepine while Sodium Valproate, Phenytoin, Gabapentin, Leviteracitam are also effective.

Prognosis is excellent. Remission occurs within 2 to 4 yrs of onset nd prior to the age of 16 yrs and rarely child may develop language and cognitive dysfunction.

In the present case the child presented at the age of 4 years which fulfils the age criteria of Rolandic epilepsy. The symptomatology like drooling of saliva, unable to talk, abnormal posturing also suggests that child may have Rolandic epilepsy but the episodes were only during day time and the EEG showed Fronto Centro Parietal epileptiform discharges which are atypical features of Benign Rolandic Epilepsy. Figure 1 shows spikes in the right and left central and parietal areas, left frontal area, and left frontal polar site. Figure 2 shows spikes in right and left frontal and central areas. Figure 3 shows spikes in left frontal, central and parietal areas and left frontal polar site. Figure 4 shows spikes in right and left central and parietal areas and left frontal area. So it can be considered as Atypical form of Benign rolandic epilepsy. The dose of Sodium Valproate was increased and the child did not have any fresh episodes.

ACKNOWLEDGEMENT

We would like to thank Mr. C Lakshminarasimha Rao and Dr. V Suryanarayan Reddy, CAIMS, Karimnagar for giving for permission to publish the case report and also Dr. Sanjay, Asst. Professor, Neurology, Ramesh, Stenographer, Pediatric department for their valuable help.

Conflict of interest

Nil.

Source of funding

Nil.

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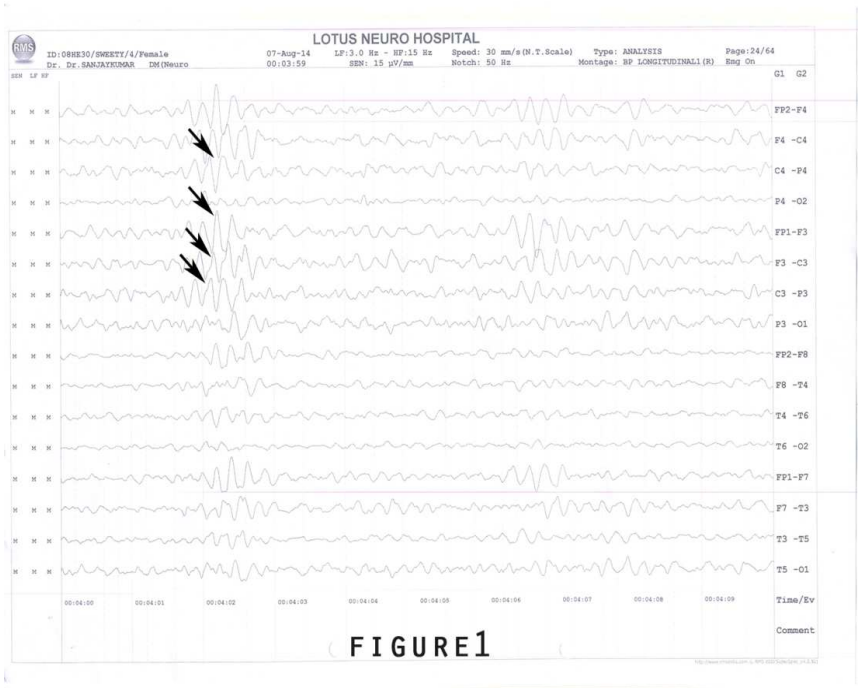

FIGURE1

 ISSN: 2394-3718

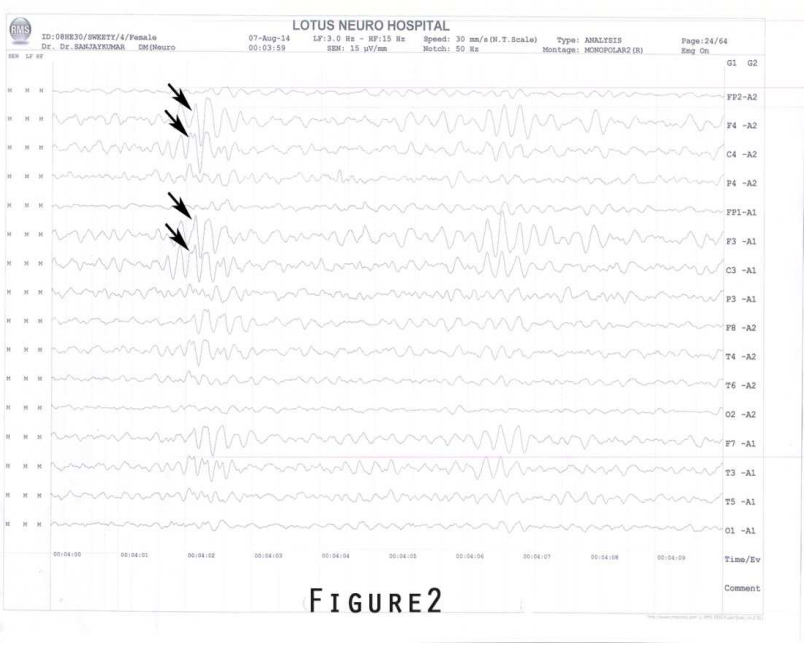

FIGURE2

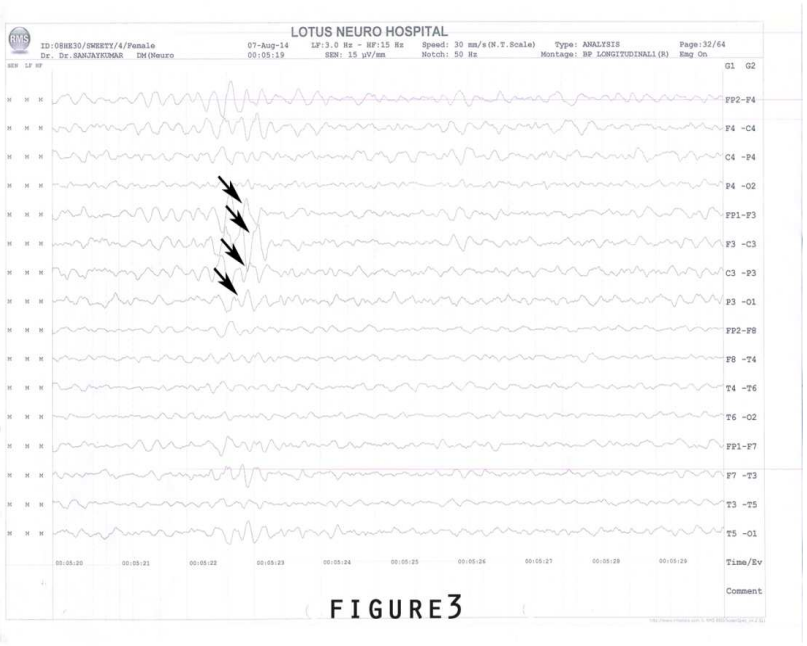

FIGURE3

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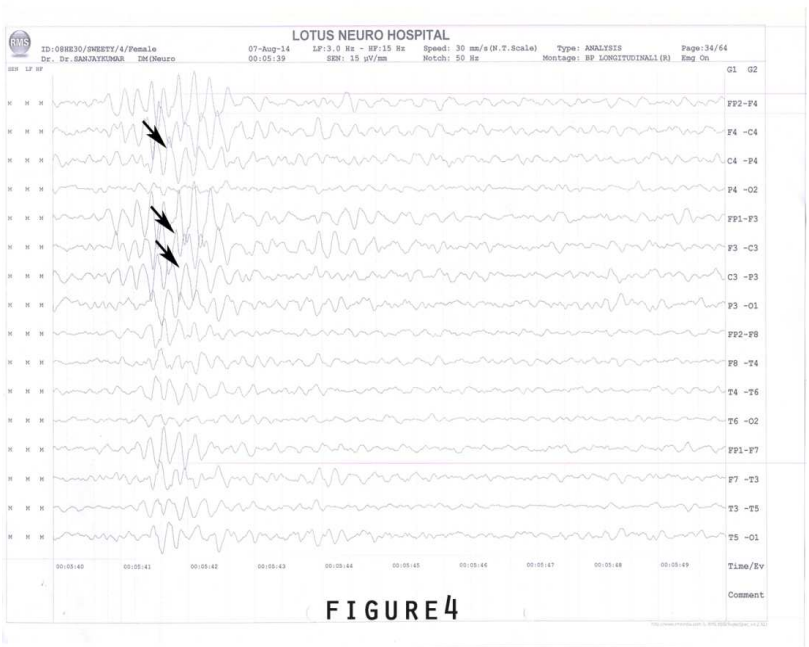

FIGURE 4