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Research Article

CLINICAL FEATURES, ELECTROENCEPHALOGRAM FINDINGS AND TREATMENT OF SELF-LIMITED FOCAL EPILEPSIES OF CHILDHOOD

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ABSTRACT

Objectives: The objective is to study the clinical features of self-limited focal epilepsies of childhood and to describe their electroencephalogram (EEG) findings and treatment.

Methods: Twenty cases of self-limited focal epilepsies of childhood attending the Department of Neurology Government Medical College, Kozhikode, between August 2019 and July 2020 were included in the study. Details of history, examination, EEG, and treatment were collected.

Results: The age at first seizure was 1.5–12 years. Most children were diagnosed with self-limited focal epilepsy of childhood with centrotemporal spikes. One child had Panayiotopoulos syndrome. Most of the seizures occurred during sleep. The majority were generalized tonic–clonic seizures. The majority had bilateral centrotemporal spikes in EEG. The child with Panayiotopoulos had bilateral spikes from the posterior head region. All were treated with antiepileptics.

Conclusion: Self-limited focal epilepsy of childhood with centrotemporal spikes was the most common self-limited focal epilepsy of childhood. They had typical centrotemporal epileptiform discharges, seen bilaterally in most. The most commonly used drug was valproate. None had any major adverse effects on therapy.

Keywords: Childhood epilepsy, Rolandic epilepsy, Panayiotopoulos.

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INTRODUCTION

Self-limited focal epilepsies of childhood, formerly called benign childhood focal epilepsies are electroclinical syndromes of unknown or genetic cause that occur in developmentally and neurologically normal children. They mainly comprise three electroclinical syndromes recognized by the International League against Epilepsy [1]: The most common is self-limited epilepsy with centrotemporal spikes (SLECTS), formerly called "benign rolandic epilepsy." Others are the self-limited occipital epilepsies of childhood, with the early-onset form described by Panayiotopoulos and the late-onset form by Gastaut. A normal child with infrequent seizures and an electroencephalogram (EEG) with disproportionately severe focal epileptogenic activity is highly suggestive of self-limited focal epilepsies of childhood. There are very few studies from India looking at the clinical manifestations, EEG patterns, and treatment of self-limited focal Epilepsies of childhood. It is important to have original studies from this region as there may be variations in the clinical presentation and proportion of various entities included under this syndrome encountered here. In addition, treatment practices may also vary. The objective of our study was to describe the clinical features of self-limited focal epilepsies of childhood, their EEG findings, and treatment.

METHODS

This was a descriptive study done in the Department of Neurology, Government Medical College, Kozhikode, over 1 year from August 2019 to July 2020. Permission was obtained from the Institutional Ethics Committee.

Twenty children attending the department, who were diagnosed with self-limited focal epilepsy of childhood were included in this study after obtaining consent from their guardians. Self-limited focal epilepsies of childhood mainly include [1]:

1. SLECTS
2. Early onset self-limited occipital epilepsy of childhood (Panayiotopoulos)
3. Late-onset self-limited occipital epilepsy of childhood (Gastaut).

Data were collected on a pro forma by interviewing the guardians and the patients and from the treatment records available. Detailed clinical profiles of these children were collected including demographic details such as age, gender, and locality. Other information on the perinatal period, development, family history, and associated systemic diseases was also noted. The semiology of seizures was determined by interviewing parents/guardians/other witnesses and in some cases also with the help of home videos from parents. Other aspects of seizures such as frequency Duration, and relationship to sleep were also noted. Detailed examination was done in all children including a thorough neurological examination. EEG was taken in all patients, awake, and sleep records. Neuroimaging was available in some cases and all were magnetic resonance imaging (MRI). Treatment details including drug use, dosage, response to treatment, and any side effects of treatment were also collected.

RESULTS

Twenty children with self-limited focal epilepsies of childhood were included in this study. Nineteen (95%) of twenty children were diagnosed as SLECTS. Only one child was diagnosed with Panayiotopoulos syndrome (PS). Age at first seizure ranged from a minimum of 1.5 years to a maximum of 12 years with a median of 7.5 years for SLECTS. The child with PS had a seizure onset at 3 years. 11 (58%) were boys and 8 (42%) were girls in SLECTS. The male-to-female ratio was 1.4. PS was diagnosed in one girl child. The perinatal period was uneventful in all children. All had normal development, hearing, and vision. None had any significant systemic illness.

About 5 (25%) of these children had a history of febrile seizures in the SLECTS group. Out of this, one child had complex febrile seizures, rest had simple febrile seizures. Family history of seizures was present in 6 (30%) children, five in the SLECTS group, and one child with PS. Out of this 3 (16%) children had a family history of epilepsy and 1 (5%) had a family history of febrile seizures, one child had a family history of both epilepsy and febrile seizures (5%). PS child had a family history of febrile seizures.

Seizures were focal in 5 (25%), focal to bilateral tonic–clonic in 3 (20%), and generalized tonic–clonic in 11 (55%) in SLECTS. The child with PS had focal to bilateral tonic–clonic seizures. Seizures of SLECTS occurred only during sleep, especially in the early morning hours in 15 (79%) children and during sleep and while awake in 4 (21%). In PS seizures occurred in sleep alone mostly early in the morning before waking up and were focal to the bilateral tonic–clonic. At enrolment, in SLECTS 6 (32%) had only one seizure, 8 (42 %) had two seizures, 2 (11%) had three seizures and 3 (16%) had four or more seizures. The child with PS had three seizures. Examination findings were normal in all children enrolled.

All children had EEG tests including sleep records. Background rhythms were normal in nineteen children (95%). Only one child had mild asymmetric hemispheric slowing (5%) and belonged to the SLECTS group. In this group, in seven children (37%) spikes were unilateral, whereas in 12 (63%) were bilateral spike foci. All showed activation of spikes by sleep and had typical morphology of high amplitude and diphasic waveforms in centrotemporal locations. The child with PS had bilateral epileptiform discharges from the posterior head region.

MRI brain was taken in 9 (45%). All were normal. Other routine blood investigations were normal in all children.

Antiepileptic was started in all children. Of 15 (79%) were started on valproate, 2 (10.5%) on levetiracetam, and 2 (10.5%) on oxcarbazepine in the SLECTS group (Table 1).

The child with PS was started on oxcarbazepine. None required polytherapy. The median dose of valproate was 20 mg/kg/day, levetiracetam 20 mg/kg/day, and oxcarbazepine 15 mg/kg/day. Only one child had seizure recurrence while on therapy (valproate group in rolandic epilepsy on dose 10 mg/kg/day). None had any significant side effects to treatment.

DISCUSSION

All children enrolled in the study were diagnosed with SLECTS except for one child diagnosed with PS. We did not have any case of late-onset self-limited occipital epilepsy of childhood (idiopathic childhood occipital epilepsy-Gastaut [ICOE-G]).

SLECTS is the best-known and most common self-limited focal epilepsy of childhood. The prevalence of PS may be high but is practically absent in designed controlled epidemiological studies [2-5] as this syndrome was only recently formally recognized, its features imitate many other conditions and often manifest with a single seizure only. The ICOE-G is a relatively rare form of pure occipital epilepsy accounting for about 2–7% of benign childhood focal seizures [6-15].

The age of onset for SLECTS ranged from 1.5 to 12 years, with 63% starting between 4 and 10 years. The child with PS had onset at 6 years. These were similar to prior published studies. The male-to-female ratio was 1.4 (11 males and 8 females) for SLECTS consistent with many previous reports of male predominance. The child with PS was a girl child. Both genders are probably equally affected by PS, though a female preponderance was found in some studies [16].

All children had normal perinatal history and development. About 5 of 19 children with SLECTS (26%) had a history of febrile seizure. Reported occurrence of febrile seizures ranges from 10% to 20% of patients [17]. Here febrile seizures were slightly higher probably related to the higher incidence of febrile seizures in Indian children compared to the Western studies.

About 4 of 19 (21%) of children with SLECTS gave a family history of epilepsy which is in the reported range from previous studies. Rolandic epilepsy is genetically determined although conventional genetic influences may be less important than other mechanisms [16,18]. The child with PS had a family history of febrile seizures. PS, like SLECTS, is probably genetically determined. Usually, there is no family history of similar seizures, although siblings with PS or PS and SLECTS have been reported [11,16].

Seizures were focal in 5 (26%), focal to bilateral tonic–clonic in 3 (16%), and generalized tonic–clonic in 11 (58%) of 19 cases of SLECTS. Based on prior studies 70–80% of the seizures are focal [19]. They may be the only type of attack or may alternate with generalized seizures, occurring in 24–80% of patients. Most partial seizures are motor. Generalized seizures may be the only ictal manifestation of SLECTS. However, because they are mainly nocturnal, their focal onset may go unrecognized. Progression to hemiconvulsions or generalized tonic–clonic seizures occurs in around half of the children. The child with PS had focal to bilateral tonic–clonic seizures. The parents did not notice any evident autonomic manifestations. The hallmark of PS is ictal autonomic aberrations that may involve any function of the autonomic system mainly emesis. Autonomic manifestations may not be apparent at seizure onset even in witnessed diurnal seizures. They may be absent, mild, or missed in clinical observation.

Seizures of SLECTS occurred only during sleep in 15 (79%) children mostly early morning time and during sleep and while awake in 4 (21%). Generally, in more than half of patients, seizures occur only during sleep; in the remainder, they occur while asleep and awake. Only 5–25% of attacks occur exclusively while awake. Approximately 25% of nocturnal seizures occur during the middle part of the night; 20% appear on falling asleep and 35% either on awakening or the 2 h before awakening [20]. Seizures in our child with PS occurred exclusively in sleep early in the morning. Two-thirds of seizures in PS start in sleep according to most studies.

At enrolment, in SLECTS 6 (32%) had only one seizure, 8 (42%) had two seizures, 2 (11%) had three seizures, and 3 (16%) had four or more seizures. SLECTS usually has a low seizure frequency. Approximately one-fourth of the patients of SLECTS have only one attack. The child with PS had three seizures. Seizures are relatively rare in PS, and up to 30% of children have only a single episode [11].

All children with SLECTS had seizures lasting <10 min. SLECTS seizures are usually brief lasting for 1–3 min. The child with PS had seizures lasting for 30 min. In PS the seizures are usually lengthy over 6 min and almost half of them last for >30 min to many hours.

All children had EEG tests including sleep records. Background rhythms were normal in 19 children (95%). Only one child had mild asymmetric hemispheric slowing (5%) and belonged to SLECTS group. All in this group had the typical location of epileptiform discharges at the centrotemporal region. In 7 (37%) spikes were unilateral, whereas in 13 (63%) there were bilateral spike foci. All showed activation of spikes by sleep and had typical morphology of high amplitude and diphasic waveforms. The child with Panayiotopoulos had bilateral epileptiform discharges from the temporoparietooccipital region. In about 90% of cases, the EEG reveals mainly multifocal, high amplitude, sharp slow wave complexes that may appear in any area, often shifting from one region to another in the same or the contralateral hemisphere in sequential EEGs of the same child. Occipital spikes predominate but they do not occur in a third of patients.

MRI brain was taken in 9 (45%) including the one child with PS. All were normal. In its typical form, SLECTS and PS have no underlying

Table 1: Treatment in SLECTS

Drug	Number (%)
Valproate	15 (79)
Levetiracetam	2 (10.5)
Oxcarbazepine	2 (10.5)

SLECTS: Self-limited epilepsy with centrotemporal spikes

structural brain abnormalities, and neuroimaging is normal, except for incidental findings.

Antiepileptic was started in all children. 15 (79%) were started on valproate, 2 (10%) on levetiracetam, and 2 (10%) on oxcarbazepine in SLECTS. The child with PS was started on oxcarbazepine. None required polytherapy. The median dose of valproate was 20 mg/kg/day, levetiracetam 20 mg/kg/day, and oxcarbazepine 15 mg/kg/day. Only one child had seizure recurrence while on therapy (valproate group in SLECTS on dose 10 mg/kg/day). None had any significant side effects to treatment necessitating discontinuation of therapy.

In carbamazepine is most commonly used in SLECTS [21]. However, rarely there may be a possible aggravating effect, with increased frequency and diffusion of the spikes, leading in some cases to status epilepticus or continuous spike-waves of slow sleep [21]. Some use valproate rather than carbamazepine to avoid this risk of aggravation. The treatment of self-limited occipital epilepsy is similar to that of SLECTS. Some avoid antiepileptics altogether, as patients may experience only a low number of seizures.

CONCLUSION

SLECTS was the most common self-limited focal epilepsy of childhood seen in this part of the world in the study. There was a male preponderance and the majority had seizure onset at 4–10 years of age. Most of the seizures occurred during sleep and were short-lasting. The majority were generalized tonic–clonic seizures. SLECTS had typical centrotemporal epileptiform discharges, seen bilaterally in most. PS had bilateral temporoparietooccipital epileptiform discharges. All those who underwent MRI scan had normal findings. All were treated with antiepileptics. Seizures were well controlled, none required polytherapy. The most commonly used drug was valproate. None had any major adverse effects on therapy.

AUTHORS CONTRIBUTION

Dr. Harsha T. Valoor: First author (primary researcher). Dr. James Jose: Research guide. Dr. Abdul Gafoor: Research co-guide.

CONFLICTS OF INTERESTS

There is no conflict of interest.

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