Available online at www.sciencedirect.com
SCIENCE DIRECT®
Epilepsy & Behavior 6 (2005) 435–439
Epilepsy & Behavior
www.elsevier.com/locate/yebeh

Brief Communication

Childhood paroxysmal kinesigenic dyskinesia: Report of seven cases with onset at an early age

Zhongzeng Li [a,*], Robert P. Turner [a,b,c], Gigi Smith [a]

[a] Department of Neurosciences, Medical University of South Carolina, Charleston, SC 29425, USA
[b] Department of Pediatrics, Bioinformatics and Epidemiology, Medical University of South Carolina, Charleston, SC 29425, USA
[c] Department of Biostatistics, Bioinformatics and Epidemiology, Medical University of South Carolina, Charleston, SC 29425, USA

Received 6 December 2004; revised 24 January 2005; accepted 26 January 2005

Abstract

We report on seven children who developed abnormal involuntary movements as early as $1\frac{1}{2}$ years after unremarkable term births. The paroxysmal episodes of abnormal movements were typically precipitated by sudden, voluntary movements, or a startle. The clinical features in each case were consistent with the diagnosis of paroxysmal kinesigenic dyskinesia (PKD). The episodes of abnormal movements are described. EEG was obtained in all cases, and video/electroencephalography (VEEG) monitoring was performed to exclude the possibility of epilepsy in six patients. VEEG studies revealed multiple events consistent with PKD; no ictal epileptiform discharges were recorded. The apparent benign nature of the disorder, as well as treatment options with antiepileptic drugs, was discussed with the parents, and most chose no pharmacologic treatment. We discuss clinical characteristics of PKD, treatment with anticonvulsant therapy, and recent insights into its possible pathophysiology.

Keywords: Antiepileptic drug; Movement disorder; Paroxysmal kinesigenic dyskinesia; Paroxysmal kinesigenic choreoathetosis; Regional cerebral blood flow; Video/electroencephalography

1. Introduction

Paroxysmal kinesigenic dyskinesia (PKD), also called paroxysmal kinesigenic choreoathetosis (PKC), is an uncommon movement disorder characterized by brief, frequent episodes of abnormal involuntary movements that are precipitated by sudden voluntary movement of the body, primarily face, arms, and legs, especially after a period of rest [1–5]. Though rare cases of paroxysmal episodes of abnormal movements with preserved consciousness may be secondary to a lesion in the supplementary motor area [6,7], this is not present in children diagnosed with PKD. Electroencephalography (EEG) or video/EEG (VEEG) monitoring is often required to exclude epilepsy, though the clinical manifestations of PKD are stereotypic and uniquely different from seizures. Etiology in a majority of children with PKD is apparently idiopathic, and age of onset is usually in later childhood or adolescence [1,3–5,8]. We report seven cases of early-onset (<5 years) childhood PKD seen in our pediatric epilepsy program. Review of home videotape recordings (two cases) and VEEG monitoring (six cases) suggested the clinical diagnosis from characteristics of abnormal movements and the absence of scalp EEG changes. The clinical data on each case are summarized in Table 1.

2. Case reports

2.1. Case 1 (K.G.)

A $3\frac{1}{2}$-year-old female was referred for evaluation of abnormal involuntary movements, present over the

* Corresponding author. Fax: +1 843 792 0029.
E-mail address: LiZhong@musc.edu (Z. Li).

1525-5050/$ - see front matter © 2005 Elsevier Inc. All rights reserved.
doi:10.1016/j.yebeh.2005.01.011

Table 1
Clinical data of PKD patients

Case No./gender	Age (years)	Age at onset (years)	Duration of episode (s)	Frequency of events (No./ day)	Movement-induced	Family history of movement disorder	VEEG
1/F	3.5	1.5	3–5	Multiple	Yes	No	Multiple events, no IED[a]
2/F	3	2.8	5–30	Multiple	Yes	No	Multiple events, no IED
3/F	5.5	3	5	Few	Yes	No	Multiple events, no IED
4/F	4.5	3.5	3–5	Multiple	Yes	No	Multiple events, no IED
5/F	4	1.5	5–10	Multiple	Yes	No	Multiple events, no IED
6/F	3	1.5	3–30	Multiple	Yes	Yes	Multiple events, no IED
7/M	3	2	5–30	Multiple	Yes	No	n/a

n/a, not available; multiple, >10 attacks/day.
[a] IED, ictal epileptiform discharge.

previous 2 years, that appeared to be brought on by sudden movements. Her past medical history included twin gestation birth at 37 weeks with a weight of 6 lb, 4 oz. Delayed fine and gross motors skills, without regression, were noted during the first year after birth, leading to a diagnosis of choreoathetoid cerebral palsy. She had one possible seizure at 1 year, with nondiagnostic EEG and normal brain MRI. Abnormal movements initially involved arms only, but progressed to involve the legs and facial muscles as well. The episodes consisted of hand twisting, grinding of teeth, and closure of her eyes, lasting 3–5 seconds. These occurred up to 50 times per day, and she remained alert during the episodes, and was at baseline between episodes. Many episodes occurred in car seats while riding or in a high chair at home. Her parents never noted abnormal movements while she was asleep. VEEG demonstrated multiple stereotypic events, with asynchronous arm movements, followed by facial grimacing and twisting of the torso, lasting less than 20 seconds. However no ictal epileptiform discharges (IEDs) were recorded during the events. Treatment sequentially with carbamazepine, levetiracetam, and clonazepam failed to affect the episodes.

2.2. Case 2 (S.H.)

A 3-year-old female was evaluated for a 2-month history of abnormal movements in her arms and hands, most noted in her high chair and while sitting in her car seat. Her past medical history revealed blow-by oxygen at term birth for mild meconium and APGARs of 7/8. She was hospitalized 5 days after birth for a reflux episode and treated with antireflux medication for 8 weeks. She had bronchiolitis and pneumonia at the age of 1 year. Family history included a maternal cousin with seizures, which were outgrown at 8 years of age; the patient's father had a history of attention-deficit hyperactivity disorder (ADHD) as a child. Normal waking and sleep EEG was obtained. Normal laboratory values included complete blood count, glucose, creatinine, ceruloplasmin, liver enzymes, lead level, thyroid panel, DNase-B antibody, ANA screen, ASO, and ESR. Her abnormal movements were reported to consist of dystonic posturing of both arms and hands. Her head turned to the left or right side while she grimaced, and she thrusted her tongue repeatedly. These abnormal movements lasted from 5 to 30 seconds, and would occur many scores of times during the day. They were brought on by sudden movements of her hands. VEEG monitoring documented multiple clinical episodes where the patient would be sitting quietly in her high chair and suddenly exhibit tonic extension of her arms associated with distal choreoathetotic movements of the hands and extension of the legs. Head deviation most commonly to the left side associated with tongue thrusting was also noted. All events lasted 3 to 8 seconds, and alertness and responsiveness to parents were documented. Several episodes occurred during meals but did not alter chewing or swallowing. EEG demonstrated normal waking background during all events. Treatment was started with gabapentin. Initially, a decrease in the frequency of events was noted, but this seemed to wane over several months. A gradual increase in dosing resulted in mood swings, hyperactivity, and anger outbursts, and her parents tapered the medication. Over the ensuing year, events have decreased in frequency to none to five per day, and seemingly increase with fatigue and overstimulation.

2.3. Case 3 (E.J.)

A $5\frac{1}{2}$-year-old female was referred for evaluation of abnormal involuntary movements that appeared to be brought on by sudden movements for the past $2\frac{1}{2}$ years. Her past history included trisomy 21, mild gastroesophageal reflux, and development of infantile spasms by 7 months after birth that resolved before her first birthday. MRI demonstrated mild prominence of subarachnoid spaces and ventricles, without hydrocephalus, and small symmetric hippocampal formations. Complex partial epilepsy developed at the age of 3 years and was treated sequentially with valproate, topiramate, and zonisamide. She improved in seizure frequency with zonisamide monotherapy. She also developed episodes of abnormal movements consistent with PKD by the age of 3 years. These occurred one or two times a day