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Caffeine, Stress and Sleep Deprivation Trigger Disabling Secondary Paroxysmal Dyskinesia

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Abstract

Background: Secondary paroxysmal dyskinesia may appear as episodic disabling dystonia in individuals with previous brain injury.

Methods: We describe two young individuals with cerebral palsy and attacks of prolonged, rocking movements triggered by caffeine, stress and sleep deprivation. Both episodes were severely disabling.

Results: Triggering factors were identified as excessive caffeine drinking, insomnia, stress and possibly hypoglycemia. Paroxysmal dyskinesia resolved with lifestyle modification and pharmacological treatment (clonazepam, carbamazepine, levetriacetam, trihexiphenidyl, clobazam).

Discussion: Individuals with dystonia may develop secondary paroxysmal dyskinesia and integrated management can alleviate such attacks, including those resembling primary paroxysmal dyskinesia.

Keywords: Cerebral palsy; Clonazepam; Clobazam; Carbamazepine; Levetriacetam; Trihexiphenidyl

Introduction

Paroxysmal dyskinesia is sudden, episodic, involuntary movement disorders that may include any combination of dystonia, chorea, athetosis, or ballism. The majority of reported cases are familial or idiopathic; however, there have been several reports of secondary paroxysmal dyskinesia [1]. Lifestyle modifications are part of the therapeutic regimen of individuals with primary dyskinesia to prevent events of paroxysmal dyskinesia. The triggers of secondary paroxysmal dyskinesia in individuals with previous brain injury are unknown [1,2]. We present two cases of secondary paroxysmal dyskinesia triggered by factors documented in the primary form of this disorder.

Case 1

A 34 year old woman with quadriplegic dystonic cerebral palsy following peripartum asphyxia was referred for new onset paroxysmal head rocking. Nine months earlier an intrathecal baclofen pump had been implanted for increasing spasticity and painful dystonia. Because the improvement in motor symptoms was accompanied by lowered blood pressure, the patient began ingesting more than a liter of Coca-Cola with caffeine to increase blood pressure. After a couple of weeks on a regular diet of Coca-Cola, she developed paroxysmal head rocking (no-no), tongue protrusion, tonic gaze and head tilt accompanied by jaw tremor without loss of consciousness (Video 1); video-EEG performed during an attack was free of paroxysmal activity. A 72-hour-EEG failed to demonstrate electrographic changes during episodes of head nodding. Brain MRI was normal but there was significant cervical spine atrophy at C3-C5 level. Extensive paraneoplastic work up and DYT1 were negative. Fasting glucose level was within normal limits. Fatigue and hunger exacerbated the attacks, which appeared many times daily, each lasting a few minutes. Treatment with clonazepam (0.5mg/daily) resulted in a reduction in frequency and duration of the attacks. She was instructed to reduce caffeine (one can of cola instead of 4 cans/day), maintain a rich carbohydrate diet and sleep at least 8 hours a night. Within a month, there was a dramatic reduction in head rocking attacks to just one daily. Her overall neurological examination was unchanged and complete resolution of attacks was achieved with carbamazepine.

One year later, the patient was readmitted for emergency management of insomnia, dystonia and pain. She had uncontrolled dyskinetic (no-no) movements in her neck and face, and severe pain leading to depression and a suicide attempt. Treatment was commenced with clobazam (15mg nocte) for its sedative and anti-

Video 1: Paroxysmal head rocking in a 34 year-old woman with quadriplegic dystonic cerebral palsy.
These uncontrolled, rocking "no-no" movements associated with tongue protrusion and tonic gaze developed following high caffeine ingestion. The uncontrolled nature of the movements prevented sleep and eventually led to depression and suicidality. The dyskinesia initially responded to lifestyle modification, however anti-convulsants and benzodiazepines were later required.

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dystonic effects, carbamazepin was increased to 400mg bid, oxycodone hydrochloride (5mg bid) for pain and mirtazipine (7.5mg) for depression. There was overall improvement in sleep, pain and mental state as well as decreased motor symptoms. Since she continued to have daily episodes of dyskinesia, levetriacetam (1500mg bid) was introduced (carbamazepin withdrawn) with resolution of symptoms.

Case 2

An 18 year old girl with dystonic cerebral palsy due to birth asphyxia was referred for assessment of paroxysmal arm twisting. She did not have spasticity. Trihexiphenidyl (2.5mg mane) had previously been used for hyperkinetic cerebral palsy with generalized dystonia, with improvement of proved salivation and speech. Over the previous summer, when the patient had vacation-induced sleep deprivation, she had episodes of rhythmic back and forth movements of her wrists, which ceased with positioning of her hands over her knees. We worked with the patient's family to improve her sleep hygiene and the attacks disappeared within a day following a night sleep of at least 7 hours.

A similar episode occurred 4 months later, triggered by stress related insomnia. The dyskinesia was treated successfully by increasing trihexiphenidyl to 2.5mg bid. A third attack of paroxysmal dyskinesia appeared during school examinations; the sleep pattern was not altered. Trihexiphenidyl was reintroduced and symptoms resolved within 30 minutes of the first dose. Treatment was continued for 10 days until completion of her examinations.

The third episode was triggered by stress accompanied by insomnia. This event was effectively treated with Trihexiphenidyl (5mg bid). Clobazm (5-10 mg) was also effective in stopping the dyskinesia and is her preferred treatment before stressful events such as exams.

Discussion

These 2 young individuals presented with attacks best classified as secondary paroxysmal dyskinesia: episodic movements characterized by sudden onset of involuntary posture or movements not explained by cortical epileptic discharge [3]. Most cases (75-78%) of paroxysmal dyskinesia are reported as primary, namely appearing in people with normal posture and movement and further classified as: Kinesigenic (PKD), Nonkinesigenic (PNKD), Exertion Induced (PED), and Hypnogenic (PHD) depending on triggers and duration of attacks [3,4]. While primary Paroxysmal dyskinesia precipitants and treatment have been described, Paroxysmal dyskinesia secondary to an identifiable cause has not been delineated and is probably under-diagnosed and undertreated. These cases demonstrate several characteristics of secondary Paroxysmal dyskinesia**.**

Rocking movements: The attacks in our patients are best described as PNKD. However, they had unique rocking movements (case1- neck, case 2- hands) that could not be classified as chorea, dystonia or tremor.

Precipitants are similar to primary NKPD: Episodes of primary and secondary dyskinesia are aggravated by psychological stress, fatigue and sleep deprivation [5-9], however to our knowledge this is the first report of excessive caffeine as a secondary paroxysmal dyskinesia trigger.

Attacks can become prolonged and disabling: Typical attacks of NKPD are usually short-lived[6] requiring various drug treatments. In our cases, the secondary dyskinesias were long lasting and disabling. Case 1 evolved to a prolonged continuous disabling condition as previously described [9]; however since the response to multiple high dose medications was adequate she did not require Deep Brain Stimulation [9].

Anti dystonic treatments are effective: Episodes failing to respond to lifestyle modification were effectively treated with medication. Case1 improved with anti-convulsant medication and benzodiazepines as recommended for primary cases. In case 2 anti-dystonic medication was effective when administered at the beginning of the attack.

Lifestyle changes trigger dyskinesia in individuals with dystonia due to brain injury: We argue that individuals with dystonia secondary to brain injury are sensitive to known triggers of genetic paroxysmal dyskinesia. While it is well known that lifestyle modification such as stress reduction improves overall tonic dyskinesias, other factors such as caffeine may also trigger attacks. Thus identification of possible triggers and appropriate treatment, discomforting symptoms can be alleviated.

Conclusion

Secondary Paroxysmal dyskinesia can present as rocking movements that may last for days. Identification of triggers is essential for treatment and prevention of future disabling events. Prolonged disabling attacks should be managed with life style changes and pharmacologic treatment, including anti-dystonic medication.

Acknowledgment

We thank the patients and parents for agreeing to publish their story and video.

Ethics Statement

Between writing the case report and the submission for publication, the young woman (case 1) passed away following a severe bout of pneumonia. Before the incident she had provided consent to publish her video. Her parents (legal guardians) gave their consent to publish the case and video**.**

References

1. Blakeley J, Jankovic J. Secondary causes of paroxysmal Dyskinesia. Adv Neurol. 2002; 89: 401-420.

2. Mehta SH, Morgan JC, Sethi KD. Paroxysmal dyskinesias. Curr Treat Options Neurol. 2009; 11: 170-178.

3. Demirkiran M, Jankovic J. Paroxysmal dyskinesias: clinical features and classification. Ann Neurol. 1995; 38: 571-579.

4. Netravathi M, Pal PK, Indira Devi B. A clinical profile of 103 patients with secondary movement disorders: correlation of etiology with phenomenology. Eur J Neurol. 2012; 19: 226-233.

5. Fink JK, Hedera P, Mathay JG, Albin RL. Paroxysmal dystonic choreoathetosis linked to chromosome 2q: clinical analysis and proposed pathophysiology. Neurology. 1997; 49: 177-183.

6. Bruno MK, Lee HY, Auburger GW, et al. Genotype-phenotype correlation of paroxysmal nonkinesigenic dyskinesia. Neurology. 2007; 68: 1782-1789.

7. Stefanova E, Djarmati A, Momcilović D, et al. Clinical characteristics of paroxysmal nonkinesigenic dyskinesia in Serbian family with

Myofibrillogenesis regulator 1 gene mutation. Mov Disord. 2006; 21: 2010-2015.

8. Sorgun MH, Akbostancı MC, Yücesan C, Erdoğan S, Mutluer N. Striatal infarct with paroxysmal nonkinesigenic dyskinesia. Acta Neurol Belg. 2013; 113: 197-198.

9. Kaufman CB, Mink JW, Schwalb JM. Bilateral deep brain stimulation for treatment of medically refractory paroxysmal nonkinesigenic dyskinesia. J Neurosurg. 2010; 112: 847-850.

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