

Case Report

Paroxysmal Kinesigenic Dyskinesia-like Symptoms in a Patient with Tourette Syndrome

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Abstract

Background: Paroxysmal kinesigenic dyskinesia (PKD) is characterized by episodic dystonia or choreiform movements provoked by sudden voluntary movement. PKD is not commonly reported in Tourette syndrome (TS). We describe a unique case of TS with PKD-like episodic dyskinesia that responded to carbamazepine.

Case Report: A 36-year-old male with long-standing TS developed paroxysmal “cramping”. Attacks were provoked by quick, sudden arm movements, which induced dystonic cramping, or by reaching overhead, which caused painful contraction of truncal muscles. The spells typically lasted 5–20 seconds and occurred multiple times daily. The patient’s mother suffered from intermittent dystonic toe curling. In view of the similarity of symptoms to PKD, carbamazepine was prescribed at 400 mg daily. The symptoms resolved completely. Inadvertent discontinuation led to relapse, and resumption led to recapture of benefit.

Discussion: This case demonstrates the possibility that PKD-like symptoms may co-occur with TS and may be responsive to carbamazepine.

Keywords: Tourette syndrome; paroxysmal kinesigenic dyskinesia

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Introduction

Paroxysmal kinesigenic dyskinesia (PKD) is characterized by episodic dystonia or choreiform movements provoked by sudden voluntary movement.^{1–3} PKD is not commonly reported in Tourette syndrome (TS). We describe a case of TS with PKD-like episodic dyskinesia, which responded to carbamazepine.

Case Report

A 36-year-old male with TS presented with new-onset “cramping” in both his arms and his trunk. His tics began at age 6 years with humming, followed by facial grimacing and shoulder shrugging. In his youth, he also had loud vocalizations, which disrupted the classroom. In addition, he was inattentive, hyperactive and had a compulsive need to make sure all the tongs of his fork lined up perfectly before eating. Haloperidol, at an unrecalled dose, yielded partial improvement and was taken for 6 years despite leaving him feeling “like a zombie”. His tics gradually remitted by age 13 years and haloperidol was discontinued. At that point, he had mild residual tics, which he found tolerable.

At age 34 years he developed paroxysmal forceful “cramps” of his arms and back. The attacks were provoked by quick, sudden movements such as sharply turning the steering wheel, which induced dystonic hand cramping, or reaching overhead while bathing, which caused painful contraction of his truncal muscles lasting 5–20 seconds. He occasionally had involuntary clawing of the hand while writing. The spells could occur multiple times daily—on average, three to four times a day. Neurological examination performed by a fellowship trained movement disorder specialist was completely normal between spells, with the exception of very rare blinking tics. Over serial examinations in the clinic, one spell was witnessed. Strength testing of his right bicep provoked a typical attack comprising forceful flexion at his right elbow and tightening of his brachioradialis lasting approximately 15 seconds. For his tics, he reported sporadic right shoulder shrugging and nose scrunching, which were never witnessed in the clinic. He also reported a rare compulsion to repeat a sentence internally. Serum electrolytes and limited EEG leads during polysomnography were normal.

Given that his symptoms were reminiscent of PKD, although not classic, carbamazepine was prescribed. At 200 mg twice a day his action-induced paroxysmal cramping and dystonic posturing resolved almost completely. When the patient ran out of medication his symptoms returned to their baseline frequency of multiple episodes a day, within 2 weeks. On resuming the medication the symptoms were suppressed. He elected to reduce his dose to 200 mg daily to minimize sedation and found that cramping or posturing spells occurred only once or twice a week at that dose. He had no premonitory urge, sense of relief, or ability to suppress the cramps, in direct contrast to his tics. His other established tics were not affected by the medication.

The patient's 55-year-old mother reported that similar cramping of the extremities, typically dystonic toe curling, had occurred throughout her life. Her cramping, however, was not always movement-induced, had a longer duration (≥ 20 minutes) and even woke her from sleep.

Discussion

PKD is characterized by involuntary hyperkinesias provoked by sudden movements. The attacks typically last from seconds to minutes, ranging in frequency from 100 a day to one a month. The pathogenesis is unknown, but channelopathy has been hypothesized due to analogous characteristics, and proximity of associated genetic loci to known ion channel genes on chromosome 16. Anticonvulsants such as carbamazepine and phenytoin have had reported efficacy.¹⁻³

This case, in addition to fulfilling DSM-IV criteria for TS, had adult onset of PKD-like cramps with kinesigenic triggers, short duration, preserved consciousness, and excellent responsiveness to carbamazepine. The spells lacked any sense of urgency, relief, or suppressibility in contrast to his tics, which were unaffected by carbamazepine. Atypical features for PKD were the presence of pain, adult onset, and lack of clear typical family history as his mother's dystonic cramping was more prolonged and not always movement-induced. Although he did not meet strict criteria for PKD, his symptoms had interesting parallels to that disorder.

This patient's movements, in the context of known "classic" TS, could certainly represent atypical tics or, less likely, might present a potential overlap of these two conditions. Alternative diagnoses could include psychogenicity, although he lacked identifiable risk factors or satisfaction of criteria for psychogenic movement disorders.⁴ Thyroid/electrolyte imbalances were ruled out, and he did not have clinical myotonia with prolonged grip or with percussion of the thenar eminence. Simple cramps remain a possibility.

TS is not known to co-occur with PKD, but in one review, seven of 121 patients with PKD reported a family history of TS.¹ Another report describes a PKD case with tics and obsessive-compulsive disorder.⁵ This case highlights the symptom overlap between the two conditions, the diagnostic difficulty in characterizing PKD, or less likely, a common genetic predisposition. Perhaps a unifying genetic mutation might have phenotypic variability in its presentation. For example, in the case of GTP cyclohydrolase 1 mutations, heterozygotes have manifested dystonia, parkinsonism, and TS,⁶ but have also manifested exercise-induced dystonia and increased frequency of obsessive-compulsive disorder.⁷

This case demonstrates an interesting presentation of PKD-like symptoms in a patient with TS. Whether his symptoms represent atypical tics or a distinct phenomenon is not clear, but it raises the possibility that a subset of patients with similar symptoms might benefit from carbamazepine therapy.

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