

Brief Report

Paroxysmal Kinesigenic Dystonia Associated with a Medullary Lesion

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Summary: A 67-year-old man experienced the abrupt onset of intermittent spasms of tightening of his throat muscles and elevation of his tongue to the roof of his mouth. These were precipitated by initiating movements, either of his mouth (eating, drinking, speaking, yawning) or of his whole body (arising from bed or a chair, lifting heavy weights). Episodes occurred six to 20 times per day, lasted 10–30 s, then resolved spontaneously. Two years later, results of his general neurological examination, including speech, were normal. Several spasms were provoked by arising from a seated or supine position or by drinking. Objectively, there was a strained dysphonia accompanied by palpable hardening of the supralaryngeal muscles. Each episode resolved within 15 s. Magnetic resonance imaging (MRI) showed evidence

of a remote hemorrhage in the medulla. No abnormal blood vessels were seen. Phenytoin 300 mg/day abolished the spasms within days. Decreasing the dose to 200 mg/day months later led to a partial return of symptoms. Relief has persisted for 3 years. This patient has paroxysmal kinesigenic dystonia (PKD) of structures (pharynx, larynx, tongue) innervated by lower cranial motor nerves and a medullary lesion on MRI. PKD has been associated with focal lesions at all levels of the central nervous system (CNS), although never before in the medulla. PKD seems to be a nonspecific phenomenon of the CNS in reaction to injury.
Key Words: Paroxysmal kinesigenic dystonia—Medulla oblongata—Pharynx—Larynx—Tongue.

Paroxysmal kinesigenic dystonia (PKD), or choreoathetosis (PKC), is a transient movement disorder characterized by its precipitation by initiation or acceleration of movement, a duration of usually <1 min and always <5 min, a high frequency of ≤ 100 or more episodes per day, and a striking benefit from anticonvulsants (1–3). The most common precipitating movement is rising quickly from a seated position (1). Most paroxysms are preceded by sensory prodromata and followed by a refractory period (3). In the majority of cases, the movements affect the limbs in a unilateral fashion (1,3). Following is a description of a case conforming to the clinical criteria of PKD outlined above, but with a unique distribution limited to the territory of lower cranial nerves and associated with a lesion in the medulla.

CASE REPORT

A 69-year-old man complained of spasms of his throat, which had begun abruptly 2 years earlier. These episodes consisted of tightening of the throat muscles which felt “like golf balls” on either side of his throat. This was accompanied by elevation of the tongue to the palate. The net results were dysphonia and dysarthria. During ~25% of episodes he simultaneously felt a burning sensation around the eye, radiating down over the cheek and the side of the neck; he estimated that the right side was involved four times as often as the left, but never both sides at once. The episodes occurred six to 20 times per day and lasted 10–30 s each, resolving rapidly.

Spasms were provoked by a number of activities, most notably speech, especially at initiation. At times, anticipation of speech by picking up the receiver or just hearing the telephone ring could provoke a spasm. Other mouth actions could cause an episode to occur, including chewing food (particularly the first bite), swallowing, brushing teeth, and yawning. Occasionally, whole body motions such as arising from a chair or lifting a heavy weight would also result in a spasm. Immediate repetition of the

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provocative action after a spasm would never result in a second episode, and he could resume speaking, chewing, or lifting without further incident. He had been treated with clonazepam up to 2.5 mg/day, producing only disequilibrium.

His medical history was notable for hypercholesterolemia, ischemic heart disease, and a coronary artery bypass graft 10 years previously. He had also undergone a prostatectomy for benign prostatic hypertrophy and a lumbar diskectomy. His daily medications were bethanechol, simvastatin, and aspirin. The family history disclosed no neurologic illness.

Results of the general neurologic examination were unremarkable. His speech at rest was normal. Two episodes of throat spasms were produced by sitting up from a supine position, and one by drinking (videotape). Objectively, these consisted of a strained dysphonia and palpable hardening of supralaryngeal muscles bilaterally, lasting ~15 s. Speaking, eating, or answering the telephone produced no reaction.

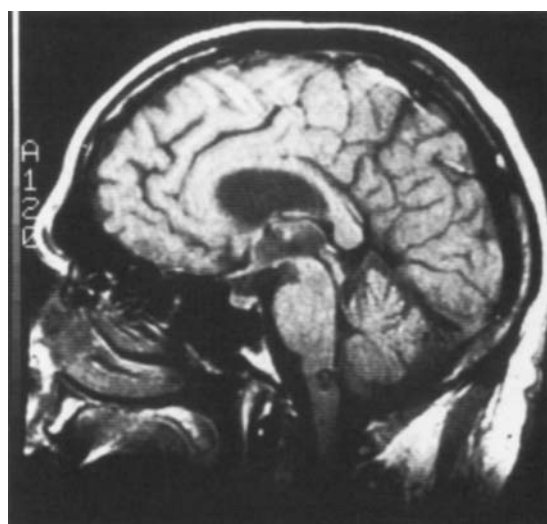
A magnetic resonance imaging (MRI) scan of the brain demonstrated a medullary lesion consistent with a remote hemorrhage (Fig. 1). No abnormal blood vessels were seen.

On two subsequent office visits, he developed throat spasms upon arising from his chair in the waiting room. The only other episode was provoked by drinking water. He began treatment with phenytoin 300 mg every morning. He stopped having attacks the following day. When he reduced his daily dosage to 200 mg, he felt a rare sensation of tightness in his jaw "almost like it was trying to do it again." However, he has had no throat spasms since starting phenytoin 3 years ago.

DISCUSSION

This patient's story is typical of PKD in every respect save for the distribution of the muscles affected. PKD almost always involves the limbs, usually in a unilateral fashion; no prior case of PKD of the pharynx, larynx, and tongue has been reported. The anatomic location of the lesion shown in Fig. 1 suggests strongly that it is responsible for the unique clinical presentation because the cranial nerve nuclei (ambiguus, hypoglossal) predominantly responsible for motor innervation of the pharynx, larynx, and tongue are located at the medullary level.

The mechanism for production of PKD is unknown. Most investigators speculating on this topic have chosen to regard PKD as either a disorder originating in the basal ganglia (1,4) or a form of reflex epilepsy (5,6). The evidence in favor of a basal ganglia lesion includes the dystonic (or choreic or athetotic) character of the involuntary movement, the coexistence in a minority of patients of other dyskinesias suggesting basal ganglia disease, and a lone report of a patient responding to levodopa (7). Kertesz reasoned that PKD, at least in idiopathic or familial cases, could represent "a paroxysmal release phenomenon from striatal control" caused by "insufficient maturation of some of these control pathways," leading to "pathological oscillation in the circuits of the basal ganglia" due to a disordered feedback mechanism (1).



1A



1B

FIG. 1. MRI shows a large, hypointense area in the dorsal medulla oblongata on T1-weighted sagittal (A) and T2-weighted axial (B) views. The appearance is consistent with hemosiderin deposition, most likely due to a remote hemorrhage. No abnormal vasculature is seen.

Support for the notion of PKD as an epileptic phenomenon comes from its episodic, stereotyped occurrence, analogous to that of seizures, and its response to anticonvulsants. Features inconsistent with epilepsy are the preservation of consciousness, even during bilateral episodes, and the conspicuous absence of paroxysmal epileptiform activity from electroencephalograph recordings during attacks. Franssen et al. found an enhanced cortical potential (the "slow negative wave") in an electrophysiologic study of the contingent negative variation after a signal warning of anticipated tasks in a patient with PKC (8). However, they were careful to conclude that the elec-

TABLE 1. Focal lesions causing paroxysmal kinesigenic dystonia

Site of lesion	Nature of lesion	Reference
Frontal lobe	Cortical scar	10
Frontal lobe	Trauma	11
Putamen	Infarct	12
Thalamus	MS plaque	13,14
Thalamus	Infarct	14,15
Medulla oblongata	Hemorrhage	This case
Cervical spinal cord	SLE vasculitis	16
Cervical spine cord	Herpes simplex I myelitis	17

Only those cases with typical manifestations of PKD convincingly related to a single lesion were listed.

MS, multiple sclerosis; SLE, systemic lupus erythematosus.

trical abnormality could have arisen from a disturbance in an unknown part of the "medial thalamic-prefrontocortical-nucleus reticularis thalami system," and its relationship to PKC was not established.

Most cases of secondary or symptomatic PKD are due to multiple sclerosis or brain trauma (9) and cannot be correlated with a specific focal lesion of the central nervous system. The small number that can be assigned a localization show a diversity of clinical-pathological correlation. PKD is associated with focal lesions at all levels of the neuraxis, not just within the cerebral cortex or basal ganglia but also in the thalamus, brain stem, and spinal cord (Table 1). This suggests that PKD is not a manifestation of dysfunction within a specific region or pathway of the nervous system, but rather a shared expression of disorders of varying etiology and location, whose distribution depends on the site of the lesion. Details of the pathophysiology remain obscure, but fundamentally there appears to be a disordered response to movement or anticipation of movement. The occurrence of dyskinesias with anticipation, and preceding actual movement, indicates that the aberrance occurs in the planning stage of actions rather than as a result of the influx of sensory information from the movement itself, such as proprioceptive changes. At the very least, patients such as the one described here compel us to broaden our consideration of the pathophysiology of PKD beyond the scope of epilepsy and basal ganglia disease.

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LEGENDS TO VIDEOTAPE

Segment 1. Speech at baseline is normal.

Segment 2. After sipping water, he feels hardening of his throat muscles and has dysphonia for ~10 s.

Segment 3. On another occasion, he develops a spasm after sitting up from a supine position.

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