



Paroxysmal Kinesigenic Dystonic Choreoathetosis Associated with a Thalamic Infarct

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Summary: We describe a patient with the development of paroxysmal kinesigenic dystonic choreoathetosis (PKDC) after a thalamic infarct. PKDC consists of brief episodes of dystonia or choreoathetosis triggered by movement. PKDC improves with anticonvulsants, and in some cases, with L-Dopa or anticholinergics. We review PKDC, and relate its salient features to idiopathic and secondary torsion dystonia. We postulate a similar underlying pathophysiology. **Key Words:** Thalamic infarct—Paroxysmal kinesigenic dystonic choreoathetosis—Idiopathic torsion dystonia.

Paroxysmal kinesigenic dystonic choreoathetosis (PKDC) is a disease consisting of episodes of dystonia or choreoathetosis lasting less than 5 min (usually less than 1 min) and triggered by quick movements or startle. Episodes can occur many times per day and may be preceded by paresthesias or vague sensory phenomena. Some patients report the ability to abort an episode, that they have episodes of varying severity, and that they have pain with episodes. The level of consciousness is unaltered during an episode. Patients are usually asymptomatic between episodes. PKDC improves with anticonvulsants.

We report a case of PKDC after a thalamic infarct. Thalamic infarction has not previously been described in association with PKDC.

CASE REPORT

A 53-year-old woman presented to the emergency room with a 2-week history of dystonic episodes involving her left side, triggered by movement, and lasting less than 1 min. Episodes initially involved only her left hand, but progressed to involve her left arm, face, and then her leg. The frequency of epi-

sodes increased to a maximum of two dozen attacks daily. She remained awake and alert during the episodes. The right side was never involved and she denied symptoms between episodes. Episodes were preceded by a sensation of tightness in the left wrist and dorsum of the left hand. She could sometimes abort the episode by flattening the fingers of her left hand or running hot water over her hand.

She had a history of hypertension and type II diabetes. She had no history of seizures. There was no family history of movement disorders.

The patient was mildly obese with a diastolic blood pressure of 90-100. Her pulse was regular. She was without heart murmur or carotid bruits. Mental status, cranial nerve, and sensory testing were normal. A left pronator drift was present on exam. Rapid alternating movements were clumsy in the left leg. Motor testing was otherwise normal.

Movements such as bending over to untie a shoelace or walking from chair to examining table would trigger an attack. Her left fingers flexed and contracted at the metacarpophalangeal and proximal interphalangeal joints. Next, there was flexion of her left wrist and elbow and internal rotation of her left arm. Her head turned to the left, with mouth and lower face also drawing up to the left (Fig. 1). Her left leg extended at the hip and knee. She was unable to speak during the attack, although she was

A videotape segment accompanies this article.
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able to comprehend speech. She could walk during an episode by circumducting her left leg.

Her blood chemistries, blood counts, and thyroid functions were normal. Doppler studies of the carotid and vertebral arteries were normal. An EEG was normal between and during attacks. A T2-weighted magnetic resonance imaging (MRI) with 5-mm slices revealed a large area of increased signal involving the right thalamus in the area of the ventral posterolateral, lateral posterior, and ventral lateral nuclei. In addition, the medial globus pallidus and posterior limb of the internal capsule may be involved. The lesion was only seen on one slice. Small areas of increased signal in the centrum semiovale bilaterally and left greater than right paratrial periventricular white matter, compatible with multiple infarcts, were present. There was mild to moderate atrophy (Figs. 2, 3, and 4).

Therapy with carbamazepine to 200 mg three times per day reduced the frequency of attacks. Because of increased weakness and fatigue, her medication was changed to phenytoin 300 mg once a day which resulted in complete resolution of attacks. Approximately once a week she still senses "tightness" in her left hand for less than a second.

The presence of a thalamic infarct and contralateral dystonic choreoathetosis implicate the thalamic infarct as the source of her PKDC.



FIG. 1. Abnormal posture of patient during an episode of PKDC. Still picture from videotape.

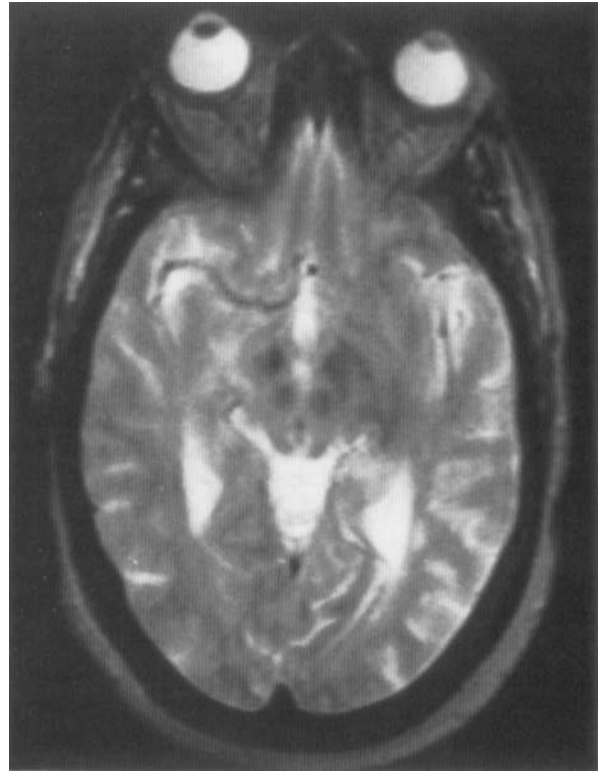


FIG. 2. T2-weighted MRI 5 mm caudal to thalamic lesion.

DISCUSSION

Attacks with the phenomenology of PKDC have been associated with: cortical seizures (1), multiple sclerosis (2), thyrotoxicosis (3), head trauma (4), birth hypoxia (5), basal ganglia calcifications (6), hypoparathyroidism (7), progressive supranuclear palsy (8), infarction (9), and possibly Wilson's disease (10). It is unlikely that all these cases share similar pathophysiology.

Few reports have associated specific lesions with PKDC, but these may be critical in separating similar clinical syndromes with different causes. Reports have associated PKDC with lesions of the putamen (9), brainstem (11), and right frontotemporal region (12). Falconer et al. (1) reported a 35-year-old man with right paroxysmal kinesigenic attacks associated with a seizure focus on the medial surface of the left hemisphere near the central sulcus. This patient, however, had occasional clouding of consciousness with his attacks.

Shibasaki et al. (2) report a 44-year-old man with multiple sclerosis and brief, painful tonic spasms triggered by voluntary or passive movement. Autopsy revealed plaques in the spinal cord, midbrain, optic chiasm, and subcortical white matter. This pa-

tient had not responded to anticonvulsant therapy, although some multiple sclerosis patients with painful spasms do respond to anticonvulsants. Most reports of paroxysmal dystonia in patients with multiple sclerosis describe a prominent sensory component and pain, but this is not always the case (13). It has been observed that many patients with multiple sclerosis and tonic spasms have symptoms or signs referable to the brainstem or spinal cord (2).

A comparison of PKDC with nonparoxysmal idiopathic torsion dystonia (ITD) may improve our understanding of dystonia in general. Although the pathophysiology of ITD is unknown, lesions associated with secondary hemidystonia are usually located in the putamen, globus pallidus, caudate nucleus, or thalamus (14). Cranial dystonia has been associated with brainstem lesions, including multiple sclerosis plaques (15). Recently, ITD has been associated with abnormalities of norepinephrine, serotonin, and dopamine in the brainstem (16).

The pathology in our patient links secondary PKDC and secondary torsion dystonia, since the phenomenology of both may be produced by damage to the putamen, thalamus, and possibly brainstem. Many reports attributing paroxysmal kinesigenic movements to cortical seizures lack demonstration of electroencephalographic (EEG) changes during the paroxysm. Some, like the case of Falconer et al. (1), represent a similar but not identical syndrome that can be distinguished from PKDC on

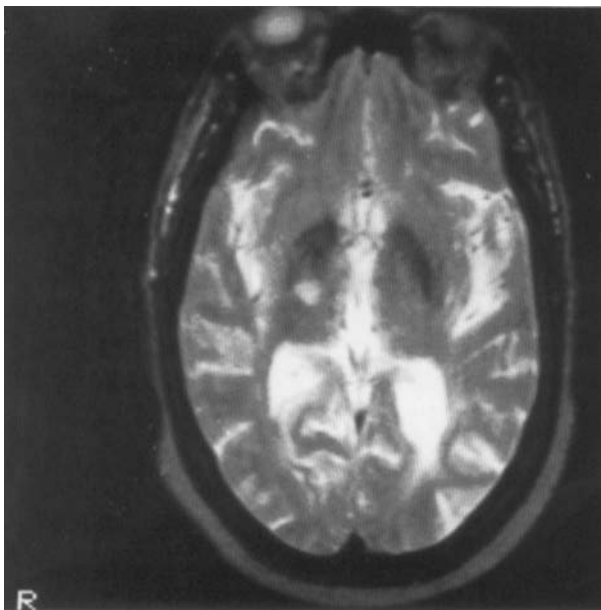


FIG. 3. T2-weighted MRI revealing increased signal in the right thalamus.

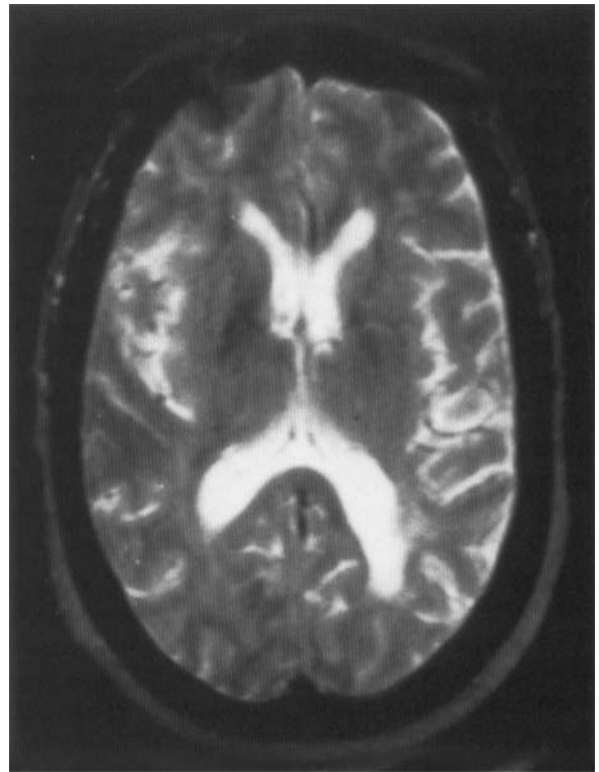


FIG. 4. T2-weighted MRI 5 mm rostral to thalamic lesion.

clinical grounds. The tonic spasms in multiple sclerosis are often accompanied by striking sensory symptoms, severe pain, and sometimes autonomic phenomena. It is not clear if these share a common physiology with PKDC. Those painless, kinesigenic attacks in multiple sclerosis as in Berger et al. (13) may be produced via a similar mechanism to that which produces paroxysmal dystonia in multiple sclerosis.

We postulate that a related alteration in neural transmission is the basis for both PKDC and torsion dystonia.

LEGENDS TO THE VIDEOTAPE

SEGMENT 1. First dystonic episode.

SEGMENT 2. Examination between episodes.

SEGMENT 3. Second dystonic episode.

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