In most cases, regardless of the etiology, cerebral lesions (sometimes associated with additional spinal cord lesions) are causative. There are just a few reports of PKD associated with pure lesions of the spinal cord (9). The case reported here corresponds to this form.

Although the spinal cord lesion seems to correspond to an area of demyelination, we prefer to use the term "central demyelinating disease" as Sethi et al. (4) because not all the diagnostic criteria forms were met. The global involvement of the posterior columns may be responsible for the bilateral and alternating dystonic attacks as well as the lack of impairment of the face.

There are some reports of PKD precipitated by a mild tactile stimulus (10), and we have found only one case of PKD precipitated by a deep nociceptive stimulus like the Lhermitte's sign (11). In our patient, this was a remarkable feature.

Although the anatomical basis of secondary PKD is variable, we believe that deep sensory systems play an important role in the pathogenesis of the attacks, not only because the thalamus is one of the most frequently affected sites, but because in our patient there was a marked involvement of the posterior columns without cerebral lesions.

Antiepileptic drugs are often very effective in treating PKD. The excellent response to sodium valproate in our patient might indicate that γ-aminobutyrate activity was diminished, which correlates with some animal models of paroxysmal dystonia (12). Although our patient did not have hyperventilation-induced attacks, acetazolamide resulted in an excellent response.

Because there are only a few reports in the literature, we describe another case in which PKD was associated with an isolated spinal cord lesion and the attacks were precipitated by both movement and deep nociceptive stimulus.

Legend to the Videotape

This patient shows dystonic posturing of either the left or the right side of the body after voluntary movement of upper limbs, after flexion of the neck, and while writing. There is no alteration of the mental state. Facial grimacing is sometimes observed.

C. Cosentino
L. Torres
M. Flores
J. M. Cuba
Movement Disorder Unit
Instituto de Ciencias Neurológicas
Lima, Peru

References

1. Berger JR, Sheremata WA, Melamed E. Paroxysmal dystonia as the initial manifestation of multiple sclerosis. *Arch Neurol* 1984;41:747–750.
2. Camec A, Greene P, Khandji A. Paroxysmal kinesigenic dystonic choreoathetosis associated with a thalamic infarct. *Mov Disord* 1990;5:235–238.
3. Aladro-Benito Y, Villagra Cocco P, Carrillo Padilla F. Paroxysmal dystonia as the first manifestation of multiple sclerosis. *Rev Clin Esp* 1993;193:64–66.
4. Sethi KD, Hess DC, Huffnagle VH, Adams RJ. Acetazolamide treatment of paroxysmal dystonia in central demyelinating disease. *Neurology* 1992;42:919–921.
5. Drake ME Jr, Jackson RD, Miller CA. Paroxysmal choreoathetosis after head injury. *J Neurol Neurosurg Psychiatry* 1986;49:837–843.
6. Fishbeck K, Layzer R. Paroxysmal choreoathetosis associated with thyrotoxicosis. *Ann Neurol* 1986;6:453–454.
7. Newman RP, Kinkle WR. Paroxysmal choreoathetosis due to hypoglycemia. *Arch Neurol* 1984;41:341–342.
8. Clark JD, Pahwa R, Koller WC. Diabetes mellitus presenting as paroxysmal kinesigenic dystonic choreoathetosis. *Mov Disord* 1995;10:353–355.
9. Previdi P, Buzzi P. Paroxysmal dystonia due to a lesion of the cervical cord: case report. *Ital J Neurol Sci* 1992;13:521–523.
10. Nijssen PCG, Tijssen CC. Stimulus-sensitive paroxysmal dyskinesias associated with a thalamic infarct. *Mov Disord* 1992;7:364–366.
11. Kuroiwa Y, Araki S. Lhermitte's sign and reflex tonic spasm in demyelinating diseases with special references to their localizing value. *Kyushu J Med Sci* 1963;14:29–38.
12. Fredow G, Loscher W. Effects of pharmacological manipulation of GABAergic neurotransmission in a new mutant hamster model of paroxysmal dystonia. *Eur J Pharmacol* 1991;192:207–219.

Belly Dystonia Induced by Levodopa and Biperiden in a Case of Suspected Multiple-System Atrophy

A variety of involuntary abdominal movements have been reported recently, including belly dancer's dyskinesia, spinal myoclonus, recumbent tic, paroxysmal axial spasms of spinal origin, and focal dyskinesia of the abdominal wall (1–5). We report a parkinsonian patient who developed focal involuntary movements of the abdominal wall that were closely related to treatment with levodopa or biperiden.

A 73-year-old Chinese man first noticed rest tremor of his right limbs for 5 years and progressive difficulty in walking for 2 years. He received treatment with levodopa/benserazide and biperiden for 4 years and bromocriptine for 5 months. Two months earlier, shortly after the addition of selegiline, involuntary movements of the abdominal wall appeared, consisting of slow, undulating and stereotypic protrusion and retraction, with occasionally slow writhing movements to both sides. The movements were semirhythmic and repetitive at a frequency ~0.6 Hz, sometimes associated with repetitive flexion–extension of the right hip and knee at a frequency ~1.4 Hz. They appeared ~1 h after he took his first morning dose, gradually built up in intensity, persisted in the day, and gradually subsided after 4:00 p.m. He might have a feeling of shortness of breath, and occasionally the involuntary movements made him fall. He did not describe any pain or urge to make the movements, nor could he suppress them on will. There were no superimposed jerks, no gurgling noises, and no sniffing sounds.

After titrating the dosages, it became evident that levodopa/benserazide 100/25 mg alone was able to induce the dyskinetic movements, lasting up to 3 h (video segment 1). One tablet of biperiden (2 mg) alone was also capable of inducing the dyskinetic movements, although of a smaller amplitude, lasting ~4 h. A half tablet of levodopa/carbidopa (100/10 mg) induced dyskinetic movements of much smaller amplitude. Bromocriptine or selegiline alone was unable to induce the movements (video segment 2). All the laboratory findings, including thyroid function and cranial computerized tomography, showed normal results.

He underwent suprapubic cystostomy in the following year. Cystometry and a urethral sphincter electromyogram disclosed detrusor hyperreflexia but a coordinated sphincter. Three months after the operation, he developed visual hallucinations, delusions of persecution, and hypersexuality, which gradually subsided after all medications were discontinued. Motor examinations disclosed a man with marked rest tremor, moderate rigidity and marked bradykinetic movements bilaterally, and marked postural instability. He obtained a score of 57 on the Columbia University Rating Scale. In addition, a supine blood pressure of ~130/80 mm Hg and a blood pressure while standing of ~70/50 mm Hg were found. There was no supranuclear downgaze palsy, no history of early falls, and no pyramidal, cerebellar, or peripheral nerve signs. One capsule of Madopar-HBS (100/25 mg) was given without eliciting the dyskinetic movements and was given three times a day thereafter. Together with the use of fludrocortisone, he obtained an 18% improvement in scores when evaluated 1 month later. He died in mainland China due to undetermined cause the following year.

It is quite obvious that the involuntary movement in this patient cannot be categorized as myoclonus or tics. It is more akin to the unusual focal dyskinesia of the abdominal wall reported previously (5). It has been assumed in the latter that the movement fits into the spectrum of dystonia and that peripheral factors may play a role in its pathogenesis.

Although lacking electromyographic support in this patient, the slow stereotypic movements and the lack of spreading to other parts of the body make the categorization of dystonia very likely. If this assumption is true, this patient presents with an unusual form of on state, rather than peak-dose, focal dystonia that involves mainly the abdomen, unlike the usual pattern of levodopa-induced choreiform movements that involve mainly the limbs (6,7). In addition, unlike most forms of focal dystonia that may respond to anticholinergics, biperiden elicited focal dyskinesia in this patient.

It is not likely that the involuntary movements in this patient resulted from dyskinetic movements of the diaphragm. Levodopa-induced respiratory dyskinesia manifests mainly with tachypnea with a respiratory rate up to 80/min (8). Orobuccal dyskinesia elicited by trihexyphenidyl therapy has also been reported in a patient with Parkinson's disease (9). It appears that there are no reports of involuntary movements related to antiparkinson medication similar to those seen in this patient.

According to the criteria proposed (10), this patient may be classified as having probable multiple system atrophy because of the presence of prominent orthostatic hypotension, urinary problems, and poorly levodopa-responsive parkinsonism. Unusual patterns of levodopa-induced dyskinetic movements, involving mainly the axial muscles, have been reported in patients with multiple system atrophy (10).

In this patient, no history of peripheral trauma could be traced. The close time links between the administration of levodopa or biperiden and the appearance of focal dystonia indicate that the dopaminergic or anticholinergic system may be responsible for the involuntary movement, and that a central mechanism may also be important in generating focal dyskinesia of the abdominal wall.

Legends to Videotape

Segment 1. Semirhythmic and repetitive movements of the abdominal wall were elicited by giving levodopa/benserazide (100/25 mg) alone.

Segment 2. Disappearance of the involuntary movements after levodopa/benserazide and biperiden were discontinued.

Din-E Shan
Shang-Yeong Kwan
Hsu-Hua Ho
Ming-Shung Su
Neurological Institute
Veterans General Hospital
Department of Neurology
School of Medicine
National Yang-Ming University
Taipei, Taiwan, Republic of China

References

1. Iliceto G, Thompson PD, Day BL, Rothwell JC, Lees AJ, Marsden CD. Diaphragmatic flutter, the moving umbilicus syndrome, and "belly dancer's" dyskinesia. *Mov Disord* 1990;5:15–22.
2. Kono I, Ueda Y, Araki K, Nakajima K, Shibasaki H. Spinal myoclonus resembling belly dance. *Mov Disord* 1994;9:325–329.
3. Davies L, King PJ, Leicester J, Morris JG. Recumbent tic. *Mov Disord* 1992;7:359–363.
4. Brown P, Thompson PD, Rothwell JC, Day BL, Marsden CD. Paroxysmal axial spasms of spinal origin. *Mov Disord* 1991;6:43–48.
5. Caviness JN, Gabellini A, Kneebone CS, Thompson PD, Lees AJ, Marsden CD. Unusual focal dyskinesia: the ears, the shoulders, the back, and the abdomen. *Mov Disord* 1994; 9:531–538.
6. Marconi R, Lefebvre-Caparros D, Bonnet AM, Vidailhet M, Dubois B, Agid Y. Levodopa-induced dyskinesias in Parkinson's disease phenomenology and pathophysiology. *Mov Disord* 1994;9:2–12.
7. Luquin MR, Scipioni O, Vaamonde J, Gershanik O, Obeso JA. Levodopa-induced dyskinesias in Parkinson's disease: clinical and pharmacological classification. *Mov Disord* 1992;7:117–124.
8. Jankovic J, Nour F. Respiratory dyskinesia in Parkinson's disease. *Neurology* 1986;36:303–304.

9. Hauser RA, Olanow CW. Orobuccal dyskinesia associated with trihexyphenidyl therapy in a patient with Parkinson's disease. *Mov Disord* 1993;8:512–514.
10. Quinn N. Multiple system atrophy. In: Marsden CD, Fahn S, eds. *Movement disorders 3*. Oxford: Butterworth-Heinemann, 1994:262–281.

Cervical Dystonia as the Initial Presentation of Huntington's Disease

We report a 36-year-old man with cervical dystonia for 10 years before the onset of conspicuous manifestations of Huntington's disease (HD). The diagnosis of HD was confirmed by the presence of the (CAG)*n* repeat expansion within the Huntington gene. Although dystonia is an unusual or late feature in HD, our case suggests that it may occur as the presenting manifestation of HD.

Dystonia is a neurologic syndrome dominated by involuntary, sustained (tonic) or spasmodic (rapid or clonic), patterned, and repetitive muscle contractions, frequently causing twisting and other abnormal movements or postures (1). The sustained nature of dystonia has been emphasized in traditional descriptions; hence, rapid movements are often unrecognized as dystonic. In some patients, dystonic movements are so rapid that they appear myoclonic (myoclonic dystonia). In contrast to chorea, which consists of brief, jerk-like movements that flow randomly from one body part to another, dystonia is characteristically repetitive and patterned, involving the same group of muscles. While chorea is typically present at the onset of adult HD, this hyperkinesia may not be evident until late in the course of the disease (2). Myoclonus can be the presenting feature in juvenile cases (2–4). Rarely, tics and other manifestations of Tourette's syndrome may precede symptoms of HD (5). The prevalence of dystonia in HD has not been documented in large series, but this hyperkinetic movement disorder is rarely seen in early stages of the disease and it has not been described as the initial manifestation of HD (6,7).

We studied and prospectively followed a patient with family history of HD who had cervical dystonia for 10 years before typical features of HD became evident. We confirmed the diagnosis of HD based on an expanded (CAG)*n* repeat within the Huntington gene (8).

Case Report

A 38-year-old chemist developed repetitive pulling of the head toward the right shoulder at age 26. These movements were initially noticeable mostly at night when he was fatigued, but became conspicuous during daytime within 6 months after onset. The involuntary neck movements and abnormal posture progressed, and they were accompanied by severe pain in the left side of the neck. He became unable to play racquetball or basketball because of these symptoms. He was diagnosed with "spasmodic torticollis" by a local neurologist and was unsuccessfully treated with haloperidol. Ethopropazine (Parsidol) 350 mg/day provided only partial relief of the pain and abnormal movement. Neither sodium valproate, biofeedback therapy, nor cervical traction was effective. Diazepam 40 mg/day improved the tilting of the head, but the patient could not tolerate the doses because of drowsiness and confusion. He had never taken dopamine receptor blocking drugs prior to the onset of the cervical dystonia.

His father had been suffering from "jumping movements," slurred speech, changing personality, violent behavior, and a history of "nervous breakdown" and a suicide attempt. He was diagnosed with HD at age 46 years. A paternal uncle, 63 years old, had also been suffering from similar symptoms since age 57. The paternal grandfather who died in his early sixties had had involuntary movements, incoordination, and dementia since age 55, and his two sisters died in their thirties in a mental institution after developing similar symptoms.

The initial examination of the patient at age 28 in our Movement Disorders Clinic showed severe cervical dystonia. The head was markedly tilted to the right, and he was unable to elevate his head >4 cm above the right shoulder. There was slight dystonic-type tremor in his neck, which appeared to be a corrective-type movement in an effort to overcome the marked pulling. The right sternocleidomastoid, splenius capitis, and trapezius muscle showed severe spasmodic contractions with hypertrophy. There was a moderate amplitude, ~8- to 10-Hz flexion–extension wrist tremor when arms were held outstretched. He seemed somewhat anxious, but there was no evidence of chorea. The remainder of the neurological examination was entirely normal, including his mental status, cerebellar function, reflexes, and gait. He was able to maintain tongue protrusion, and there was no evidence of "milkmaid's grip."

Computed tomography of the brain was unremarkable; brain magnetic resonance imaging was not obtained. Cervical spine roentgenographs were normal. Serum and urine copper levels, ceruloplasmin levels, thyroid functions, heavy metal screen, serum protein electrophoresis, electroencephalography, and a neuroophthalmological examination were normal. Neuropsychological evaluation showed poor visuospatial skills, poor planning and organization ability, slightly impaired short-term spatial and long-term verbal memory, and erratic tracking and sequencing ability. These abnormalities were attributed to high-dose anticholinergic therapy. Overall cognitive abilities were at an average level. The patient continued to work as a chemist.

Tetrabenazine, a monoamine-depleting and dopamine-blocking drug (9), initially alleviated the cervical dystonia, but the treatment was complicated by mild parkinsonism. Subsequently, he was successfully treated with repeat botulinum toxin injections 75–100 units each in the right splenius capitis, trapezius, and sternocleidomastoid muscles every 3–4 months. After 3 years, however, he became refractory to the treatment and anti-botulinum toxin A antibodies were documented by a mouse bioassay (10). He was referred to the National Institutes of Health where the diagnosis of cervical dystonia was confirmed and the patient responded well to repeat injections of botulinum toxin F. Although a repeat bioassay 16 months after the initial test no longer showed the presence of