



## Brief Report

# Tourettism and Dystonia After Subcortical Stroke

Carolyn H. Kwak, MS, PA-C, and Joseph Jankovic, MD\*

*Baylor College of Medicine, Parkinson's Disease Center and Movement Disorders Clinic, Houston, Texas, USA*

**Abstract:** The term "tourettism" has been used to describe Tourette syndrome (TS)-like symptoms secondary to some specific cause. Tics associated with attention deficit hyperactivity disorder (ADHD), obsessive-compulsive disorder (OCD), or both, are commonly present in TS, but this constellation of symptoms has been rarely attributed to stroke. We describe two boys who suffered a subcortical stroke and subsequently developed hemidystonia, tics, and behavioral comorbidities. Both had right hemispheric stroke involving the basal ganglia at 8 years of age, and in both the latency from the stroke to the onset

of left hemidystonia was 2 weeks. In addition to ADHD and OCD, both exhibited cranial-cervical motor tics but no phonic tics. The temporal relationship between the stroke and subsequent TS-like symptoms, as well as the absence of phonic tics and family history of TS symptoms in our patients, argues in favor of a cause and effect relationship, and the observed association provides evidence for an anatomic substrate for TS and related symptoms. © 2002 Movement Disorder Society

**Key words:** tourettism; stroke; dystonia; obsessive compulsive disorder; attention deficit hyperactivity disorder

Tourette syndrome (TS) is a neurological disorder of childhood onset characterized by tics that vary in distribution, severity, and frequency which are commonly associated with neurobehavioral conditions such as attention deficit hyperactivity disorder (ADHD) and obsessive-compulsive disorder (OCD).<sup>1</sup> The term tourettism has been used to describe TS-like symptoms secondary to some specific cause.<sup>2</sup> Besides TS, tics have been reported secondary to numerous causes such as infection, head trauma, medication use, and in association with other neurodegenerative disorders.<sup>3–13</sup> Tourettism secondary to cerebral infarction have been described previously in only three patients.<sup>14–16</sup> We describe here two boys who suffered a subcortical stroke, both at 8 years of age, with subsequent onset of hemidystonia, tics and behavioral comorbidities.

## PATIENT 1

This 12-year-old, right-handed boy, a product of a full term, uncomplicated pregnancy and delivery with normal developmental milestones, suffered a right subcortical hemorrhagic stroke secondary to the formation of a blood clot of unknown etiology in the region of the right caudate and putamen, 4 years earlier while assisting his father unload wood boards. At age 8, he had a sudden onset of severe headache with subsequent weakness and numbness of the left side of the body. Within 2 weeks after the stroke, he regained his motor strength and the left hemi-paresthesia completely resolved. He, however, developed involuntary flexion of his left forearm muscles and left-hand deformity. On examination he had left hemidystonia manifested by flexion of the left metacarpophalangeal (MCP), proximal interphalangeal (PIP), and distal interphalangeal (DIP) joints of the second and third digits with hyperextension of the left MCP, PIP, and DIP joints of the fourth and fifth digits, involuntary left foot inversion with toe flexion of digits 2 to 5 and extension of the left big toe (see Video, Segment 1). He also held his arm behind his back, with supination and abduction of the left arm and elevation of left shoulder while walking. His symptoms have stabilized within the past 2 years. Levodopa, trihexyphenidyl, and baclofen

A videotape accompanies this article.

\*Correspondence to: Joseph Jankovic, MD, Department of Neurology, 6550 Fannin, Suite 1801, Houston, TX 77030. E-mail: josephj@bcm.tmc.edu

Received 15 December 2000; Revised 4 May 2001, 4 September 2001; Accepted 2 October 2001

Published online 5 April 2002 in Wiley InterScience (www.interscience.wiley.com). DOI 10.1002/mds.10207

failed to provide any improvement of his dystonia. A trial of clonazepam partly improved his symptoms, but the drug was discontinued due to side effects of severe personality changes and mood fluctuations. Botulinum toxin A (BTX) injections in the left forearm and left foot, administered every 3 months for the past 3 years, markedly improved his left foot dystonia.

Six months after the stroke, the patient developed compulsive tics of repetitive blowing of air onto his palm, compulsive scratching and a persistent urge to sniff (particularly food and people). He was started on fluoxetine with notable improvement of his compulsive symptoms. When the medication was discontinued he had exacerbation of all symptoms and developed depression. In addition, the patient's parents noted the onset of behavioral changes including restlessness, hyperactivity, difficulty focusing, and low distraction tolerance. He was diagnosed with ADHD based on the Diagnostic and Statistical Manual of Mental Disorders (DSM-IV),<sup>17</sup> and his concentration and academic performance markedly improved with dextroamphetamine-racemic amphetamine 20 mg without exacerbation of compulsive tics. There is no family history of dystonia, tics, ADHD, or OCD and his parents claim that none of these symptoms were present before the stroke. At age 11 years, he exhibited additional tics including bursts of rapid, clonic, head nodding followed by slower, dystonic, cervical extension and flexion, and more sustained stretching and thrusting movement of the neck forward, which his parents noted

6 months before his initial visit. The patient did not have a history of phonic tics nor did he exhibit vocalizations during the initial visit. Clonidine 0.25 mg per day provided no meaningful improvement of his tics or ADHD. His parents note the motor tics waxed and waned in severity and frequency, but the compulsive type behaviors such as the urge to smell and pick on scabs have persisted over the past 4 years. On our initial physical examination he had severe excoriations and scabs on both legs (see Video, Segment 1). The most recent MRI showed infarct in the right putamen and caudate (Fig. 1).

He continues to be an active child who enjoys riding his bike and playing basketball, although he is unable to dribble with his left hand. He is unable to play baseball because he has difficulty releasing the bat due to his left hand dystonia.

## PATIENT 2

This is a 22-year-old, right-handed man who was first seen in our clinic at 8 years of age, shortly after he suffered a right ischemic stroke in the distribution of the right middle cerebral artery secondary to migraine headache, with basal ganglia involvement. This was manifested by a migrainous headache, unsteady, propulsive gait and left side weakness. At the time of hospital discharge, he regained his motor strength and ability to walk, but had occasional clonic seizures. Approximately 2 weeks after the stroke, he developed left-sided upper extremity dystonia. During his initial physical examina-

**TABLE 1.** *Acquired tics/obsessive-compulsive disorder secondary to brain lesion*

Study	Lesion site	Etiology	Symptom
Rodrigo et al., 1997 <sup>23</sup>	Bilateral globus pallidus	Carbon monoxide poisoning, infarction	OCD
Simpson et al., 1995 <sup>24</sup>	Right inferior parietal	Infarction	OCD
Swoboda et al., 1995 <sup>22</sup>	Right posterior frontal, right internal carotid occlusion	Infarction	OCD
Ward et al., 1988 <sup>16</sup>	Fronto-arietal lobe, left frontal, lacunar infarcts in right superior cerebellar peduncle and left basal ganglia	Infarction, tumor	Premonitory urge
Demirkol et al., 1999 <sup>28</sup>	Bilateral, symmetrical globus pallidus "tiger's eye" lesion	Unknown	Tics, ADHD, OCD
Hugo et al., 1999 <sup>25</sup>	Temporal and frontal lobes	Hypoperfusion and compensatory hyperperfusion	OCD
Berthier et al., 1996 <sup>26</sup>	Frontal and temporal lobes, cingulate, caudate	Tumor (lipoma, hygroma, angioma, hamartoma, tuberous sclerosis), arachnoid cyst, post-encephalitic hydrocephalus, head trauma, infarction	OCD
Peterson et al., 1966 <sup>29</sup>	Ventral striatum, corpus collosum, thalamus, midbrain	Malignancy	Tics, OCD, with temporal progression of tics and OCD with tumor progression
Max et al., 1995 <sup>27</sup>	Frontal and temporal lobe lesions	Head trauma	OCD
Ward et al., 1988 <sup>16</sup>	Fronto-parietal lobe, left frontal, lacunar infarcts in right superior cerebellar peduncle and left basal ganglia	Infarction	Transient feelings of compulsion

OCD, obsessive-compulsive disorder.



**FIG. 1.** Flair/90 MRI of the brain of Patient 1 showing infarction of right putamen and caudate.

tion in our clinic, he exhibited involuntary flexion of the left hand, abduction of the left arm, drawing of the left arm behind his back with a clinched left fist. He was able to passively open the fingers of the left hand, but when released the fingers remained flexed (see Video, Segment 2).

Four weeks after the stroke, he developed excessive bilateral eye blinking and transient blepharospasm with occasional upward oculogyric deviation. He exhibited increased frequency of blinking, transient upward oculogyric deviation, facial grimacing with contraction of the paranasal muscles and jaw opening, but no phonic or other motor tics were present at the time of initial examination in our clinic. Although he was a "straight-A" student before the stroke, his academic performance steadily declined (grades D and F), mainly attributed to difficulty sustaining attention. The MRI showed infarction in the distribution of the right middle cerebral artery with involvement of the basal ganglia, specifically the head of the caudate nucleus. Migraine prophylaxis with verapamil was started at the time of his stroke.

The patient was delivered after a full term, uncomplicated pregnancy to a healthy mother and had appropriate developmental milestones. There is a multigenerational family history of seizures but no tics, ADHD, OCD, or dystonia. His father died of a ruptured aneurysm at age 24. No prior history of dystonia, tics, or ADHD was noted in the patient.

## DISCUSSION

Our two patients exhibit the rare combination of dystonia, tics, ADHD, and OCD. Both had right hemisphere

strokes involving the basal ganglia at 8 years of age and in both the latency from the stroke to the onset of left hemidystonia was 2 weeks. In addition to ADHD and OCD, both exhibited cranial-cervical motor tics, but no phonic tics. The temporal relationship between the stroke and subsequent TS-like symptoms, as well as the absence of phonic tics and family history of TS symptoms in our patients, argues in favor of a cause and effect relationship and against a simple coincidental occurrence of a stroke and TS.

The coexistence of tics and dystonia has been well documented<sup>18,19</sup> but the association of tourettism (consisting of tics, ADHD, and OCD) with post-stroke hemidystonia has not been described previously. Three reports have described individual cases of TS-like symptoms secondary to vascular lesions.<sup>14–16</sup> In one report, a 43-year-old man developed motor tics after a four-artery angiography.<sup>14</sup> Although neuroimaging was not available when the case was first reported, a vascular lesion in the basal ganglia was suspected to be responsible for subsequent tics including tongue clicking and protrusion, eye closure, sniffing, frowning, and numerous other motor tics as well as palilalia. Neurobehavioral changes were not reported in this patient. Masso and Obeso<sup>15</sup> described a 66-year-old man with post-anoxic hemiballism and concomitant onset of coprolalia, but without motor or phonic tics. Both the hemiballism and coprolalia improved with tetrabenazine. More recently, Ward<sup>16</sup> reported a 62-year-old woman with acute onset of dysphasia and a subsequent suppressible urge to shake her right arm. A CT scan showed lacunar infarcts in the right superior cerebellar peduncle and left basal ganglia.

The occurrence of tics, ADHD and OCD, in our two patients after a subcortical stroke provides additional support for the notion that these TS symptoms have a biological and neuroanatomical substrate. Well documented cases of ADHD after stroke are lacking, but hemi-inattention, or neglect, is a common phenomenon after a stroke.<sup>20</sup> Although idiopathic OCD is usually not associated with any identifiable, anatomic lesion<sup>21</sup> several cases of secondary OCD have been documented.<sup>16,20,22–29</sup> Acquired obsessive-compulsive symptoms have been reported secondary to infarction of inferior parietal lobe,<sup>24</sup> posterior frontal lobe,<sup>22</sup> both globus pallidi,<sup>23</sup> both caudate nuclei,<sup>20</sup> superior cerebellar peduncle, and left basal ganglia.<sup>16</sup> Several documented cases of acquired OCD associated with cerebral malignancy, trauma or perfusion abnormalities have been attributed to lesions involving the frontal, parietal, temporal lobes and cingulate areas.<sup>16,25–27</sup> There is now a substantial body of evidence that implicates basal ganglia dysfunction in the pathogenesis of OCD.<sup>30–33</sup> In a study

of patients with bilateral basal ganglia lesions secondary to trauma, anoxic or toxic encephalopathy stereotyped and obsessive-compulsive behaviors were seen in many of these patients after the reported injuries.<sup>30</sup> We and others have observed patients with Parkinson's disease and atypical parkinsonism who have developed features of OCD, such as obsessions with bowels, compulsive gambling, and other obsessive-compulsive behaviors as their disease progresses.<sup>32</sup>

Acquired TS symptoms of tics and neurobehavioral comorbidities have been reported secondary to a variety of other lesions, usually involving the basal ganglia.<sup>30,34-39</sup> Furthermore, various imaging and biochemical studies provide support for frontal-subcortical involvement in mediating human behavior.<sup>40</sup> In TS, the cortico-striatal-thalamic-cortical circuit plays an important role in the pathogenesis of TS and related disorders.<sup>39</sup> A dysfunction in the dorsolateral prefrontal circuit, which links Brodmann's area 9 and 10 with the dorsolateral head of the caudate has been implicated in an impairment of executive functions and possibly in ADHD. The lateral orbitofrontal circuit originates in the inferior lateral prefrontal cortex (area 10) and projects to the ventral medial caudate. An abnormality to this circuit is associated with personality changes, mania, disinhibition, and irritability. Lastly, the anterior cingulate circuit, which arises in the cingulate gyrus (area 24), receives input from the amygdala, hippocampus, medial orbitofrontal cortex, entorhinal and perirhinal cortex, and projects to the ventral striatum has been linked to a variety of behavioral problems including OCD.<sup>41</sup>

In conclusion, this unique presentation of left hemidystonia and tourettism in our two young patients after a right subcortical hemorrhagic stroke involving the basal ganglia support the neuroanatomical substrate for tics, associated neurobehavioral comorbidities and dystonia.

## LEGENDS TO THE VIDEOTAPE

**Segment 1.** Patient 1 demonstrates left hemidystonia and mild blinking, head jerking and shoulder shrugging, but most of his motor tics are suppressed. He also describes his urges and other features of OCD and shows severe markings on his legs from self scratching.

**Segment 2.** Patient 2 shows blinking, oculogyric deviations, facial grimacing, jaw opening and other tics in addition to left hemidystonia.

## REFERENCES

- King RA, Scahill L. Emotional and behavioral difficulties associated with Tourette syndrome. In: Cohen D, Jankovic J, Goetz C, editors. *Tourette syndrome*. Advances in neurology. Vol. 85. Philadelphia: Lippincott Williams and Wilkins; 2001. p 79-88.
- Sacks OW. Acquired tourettism in adult life. In: Friedhoff AJ, Chase TN, editors. *Gilles de la Tourette syndrome*. Advances in neurology. New York: Raven Press; 1982. p 89-92.
- Jankovic J. Differential diagnosis and etiology of tics. In: Cohen D, Jankovic J, Goetz C, editors. *Advances in neurology*. Philadelphia: Lippincott Williams and Wilkins; 2001. p 15-30.
- Kumar R, Lang AE. Tourette syndrome. Secondary tic disorders. *Neurol Clin* 1997;15:309-331.
- Lombroso CT. Lamotrigine-induced tourettism. *Neurology* 1999; 52:1191-1194.
- Moshe K, Iulian I, Seth K, Eli L, Joseph Z. Clomipramine-induced tourettism in obsessive-compulsive disorder: clinical and theoretical implications. *Clin Neuropharmacol* 1994;17:338-343.
- Cardoso FE, Jankovic J. Cocaine related movement disorders. *Mov Disord* 1993;8:175-178.
- Bharucha KJ, Sethi KD. Tardive tourettism after exposure to neuroleptic therapy. *Mov Disord* 1995;10:791-793.
- Jankovic J, Ashizawa T. Tourettism associated with Huntington's disease. *Mov Disord* 1995;10:103-105.
- Collacott RA, Ismail IA. Tourettism in a patient with Down's syndrome. *J Ment Defic Res* 1988;32:163-166.
- Nardocci N, Rumi V, Combi ML, Angelini L, Mirabile D, Bruzone MG. Complex tics, stereotypies, and compulsive behavior as clinical presentation of a juvenile progressive dystonia suggestive of Hallervorden-Spatz disease. *Mov Disord* 1994;9:369-371.
- Spitz MC, Jankovic J, Killian JM. Familial tic disorder, parkinsonism, motor neuron disease, and acanthocytosis: a new syndrome. *Neurology* 1985;35:366-370.
- Keefover RW, Privette J. Adult-onset tourettism following closed head injury. *J Neuropsychiatry Clin Neurosci* 1989;1:448-449.
- Bleeker HE. Gilles de la Tourette's syndrome with direct evidence of organicity. *Psychiatr Clin* 1978;11:147-154.
- Masso JF, Obeso JA. Coprolalia associated with hemiballismus: response to tetrabenazine. *Clin Neuropharmacol* 1985;8:189-190.
- Ward C. Transient feelings of compulsion caused by hemispheric lesions: three cases. *J Neurol Neurosurg Psychiatry* 1988;51:266-268.
- American Psychiatric Association. *Diagnostic and statistical manual of mental disorders*. DSM-IV. 4th ed. Washington (DC): American Psychiatric Association; 1994.
- Stone LA, Jankovic J. The coexistence of tics and dystonia. *Arch Neurol* 1991;48:862-865.
- Nemeth AH, Mills KR, Elston JS, Williams A, Dunne E, Hyman NM. Do the same genes predispose to Gilles de la Tourette syndrome and dystonia? Report of a new family and review of the literature. *Mov Disord* 1999;14:826-831.
- Beckson M, Cummings JL. Neuropsychiatric aspects of stroke. *Int J Psychiatry Med* 1991;21:1-15.
- Leonard HL, Swedo SE, Lenane MC, Rettew DC, Hamburger SD, Barto JJ, Rapoport JL. A 2- to 7-year follow-up study of 54 obsessive-compulsive children and adolescents. *Arch Gen Psychiatry* 1993;50:429-439.
- Swoboda KJ, Jenike MA. Frontal abnormalities in a patient with obsessive-compulsive disorder: the role of structural lesions in obsessive-compulsive behavior. *Neurology* 1995;45: 2130-4.
- Rodrigo P, Adair JC, Roberts BB, Graeber DA. Obsessive-compulsive disorder following bilateral globus pallidus infarction. *Biol Psychiatry* 1997;42:410-412.
- Simpson S, Baldwin B. Neuropsychiatry and SPECT of an acute obsessive-compulsive syndrome patient. *Br J Psychiatry* 1995; 166:390-392.
- Hugo F, van Heerden B, Zungu-Dirwayi N, Stein DJ. Functional brain imaging in obsessive-compulsive disorder secondary to neurological lesions. *Depress Anxiety* 1999;10:129-136.
- Berthier ML, Kulisevsky J, Gironell A, Heras JA. Obsessive-compulsive disorder associated with brain lesions: clinical phenomenology, cognitive function, and anatomic correlates. *Neurology* 1996;47:353-361.
- Max JE, Smith WL Jr, Lindgren SD, Robin DA, Mattheis P, Stierwalt J, Morrissey M. Case study: obsessive-compulsive disorder

- after severe traumatic brain injury in an adolescent. *J Am Acad Child Adolesc Psychiatry* 1995;34:45–49.
28. Demirkol A, Erdem H, Inan L, Yigit A, Guney M. Bilateral globus pallidus lesions in a patient with Tourette syndrome and related disorders. *Biol Psychiatry* 1999;46:863–867.
  29. Peterson BS, Bronen RA, Duncan CC. Three cases of symptom change in Tourette's syndrome and obsessive-compulsive disorder associated with pediatric cerebral malignancies. *J Neurol Neurosurg Psychiatry* 1996;61:497–505.
  30. LaPlane D, Levasseur M, Pillon B, DuBois B, Baulac M, Mazoyer B, Tran Dinh S, Sette G, Danze F, Baron JC. Obsessive-compulsive and other behavioral changes with bilateral basal ganglia lesions. A neuropsychological, magnetic resonance imaging and positron tomography study. *Brain* 1989;112:699–725.
  31. Salloway S, Cummings J. Subcortical disease and neuropsychiatric illness. *J Neuropsychiatry Clin Neurosci* 1994;6:93–99.
  32. Kumar R, Lang AE. Coexistence of tics and parkinsonism: evidence for non-dopaminergic mechanisms in tic pathogenesis. *Neurology* 1997;49:1699–1701a.
  33. Baxter LR Jr. Neuroimaging studies of obsessive compulsive disorder. *Psychiatr Clin North Am* 1992;15:871–884.
  34. Strub RL. Frontal lobe syndrome in a patient with bilateral globus pallidus lesions. *Arch Neurol* 1989;46:1024–1027.
  35. Trautner RJ, Cummings JL, Read SL, Benson DF. Idiopathic basal ganglia calcification and organic mood disorder. *Am J Psychiatry* 1988;145:350–353.
  36. Bejjani BP, Damier P, Arnulf I, Thivard L, Bonnet AM, Dormont D, Cornu P, Pidoux B, Samson Y, Agid Y. Transient acute depression induced by high-frequency deep-brain stimulation. *N Engl J Med* 1999;340:1476–1480.
  37. Steffens DC, Helms MJ, Krishnan KR, Burke GL. Cerebrovascular disease and depression symptoms in the cardiovascular health study. *Stroke* 1999;30:2159–2166.
  38. McKee AC, Levine DN, Kowall NW, Richardson EP Jr. Peduncular hallucinosis associated with isolated infarction of the substantia nigra pars reticulata. *Ann Neurol* 1990;27:500–504.
  39. Peterson BS. Neuroimaging studies of Tourette syndrome. A decade of progress. In: Cohen D, Jankovic J, Goetz C, editors. *Tourette syndrome. Advances in Neurology*. Vol. 85. Philadelphia: Lippincott Williams and Wilkins; 2001. p 179–196.
  40. Cummings JL. Frontal-subcortical circuits and human behavior. *Arch Neurol* 1993;50:873–880.
  41. Leckman J, Cohen D, Goetz C, Jankovic J. Tourette syndrome: pieces of the puzzle. In: Cohen D, Jankovic J, Goetz C, editors. *Tourette syndrome. Advances in Neurology*. Vol. 85. Philadelphia: Lippincott Williams and Wilkins; 2001. p 369–390.