

## Secondary Cervical Dystonia Associated with Structural Lesions of the Central Nervous System

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**Abstract:** We tested the hypothesis that structural lesions of the central nervous system (CNS) associated with cervical dystonia more commonly involve the cerebellum and its primary afferent pathways than basal ganglia structures. Cervical dystonia is the most common focal dystonia, the majority of cases are idiopathic, and only a small percentage of patients have a family history of dystonia or other movement disorders. Pathophysiological mechanisms operative in solely or predominantly appendicular dystonias such as writer's cramp and Oppenheim's dystonia, respectively, may not be directly applicable to axial dystonias. The localization of structural lesions of the CNS associated with secondary cervical dystonia may provide some insight into the neural structures potentially involved in primary cervical dystonia. The National Library of Medicine Gateway (from 1960) and a clinical database maintained by the senior author (from 1999) were searched for cases of secondary cervical dystonia associated with structural lesions of the CNS. Search terms included one or more of the

following: dystonia, torticollis, cervical, secondary, and symptomatic. Lesion localization and type, patient age, patient gender, head position, occurrence of sensory tricks, and associated neurological findings were tabulated for each case. Structural lesions associated with cervical dystonia were most commonly localized to the brainstem and cerebellum. The remaining cases were equally divided between the cervical spinal cord and basal ganglia. Although inconsistent, head rotation tended to be contralateral to lesion localization. Additional neurological abnormalities were present in the majority of patients with secondary cervical dystonia. The relative paucity of basal ganglia pathology and concentration of lesions in the brainstem, cerebellum, and cervical spinal cord in patients with secondary cervical dystonia suggests that dysfunction of cerebellar afferent pathways may be important to the pathophysiology of primary cervical dystonia. © 2002 Movement Disorder Society

**Key words:** dystonia; olivocerebellar; cervical; torticollis; stroke

Cervical dystonia is the most common of the focal dystonias.<sup>1,2</sup> It is characterized by sustained, involuntary, active contractions of cervical muscles resulting in abnormal head movements or postures.<sup>3</sup> The vast majority of cases are idiopathic. Secondary cervical dystonia is uncommon, even in movement disorders clinics. Secondary dystonia, in general, has been associated with a wide variety of vascular, traumatic, infectious, and toxic processes affecting the central or peripheral nervous systems.<sup>4</sup> Secondary dystonia is also a common accompaniment of hereditary degenerative diseases such as Wilson's

disease, progressive supranuclear palsy, and cortical-basal ganglionic degeneration.

The network abnormalities associated with both primary and secondary dystonias are poorly understood. Functional imaging,<sup>5–7</sup> intraoperative recordings,<sup>8,9</sup> physiological studies in humans with focal and generalized primary dystonia, animal models of both focal and generalized dystonia,<sup>10,11</sup> and immunohistochemical localization of torsinA in human brain<sup>12</sup> have failed to generate a cohesive pathophysiological model of dystonia. Work to date suggests, instead, that dystonia may be caused by a variety of local network abnormalities with similar effects on motor output. Along these lines, cases of secondary dystonia associated with discrete structural lesions of the central nervous system (CNS) may be important for understanding the pathophysiology of the different focal dystonias. Clinical–pathological correlation of this type has been critical for understanding the oculomotor and vestibular systems.

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Hemidystonia, hand dystonia, and blepharospasm are most commonly associated with lesions of the corpus striatum, thalamus, and rostral brainstem/mesencephalon, respectively.<sup>13–15</sup> The importance of the cerebellum, particularly the vermis and fastigial nucleus, in the control of head position suggests that the cerebellum may play a role in cervical dystonia.<sup>16–18</sup> The vermis and fastigial nucleus receive projections from climbing fibers that originate in the inferior olive and either direct (fastigial nucleus) or indirect (vermis) projections from a wide-variety of pre-cerebellar cell groups. Cases of secondary cervical dystonia associated with discrete structural lesions of the CNS were collected and characterized to test the hypothesis that structural lesions of the CNS associated with cervical dystonia are concentrated in olivocerebellar and pre-olivary structures.

### MATERIALS AND METHODS

The National Library of Medicine Gateway (from 1960) and a clinical database of over 1200 patients evaluated by M.S.L. from 1999 to 2001 were searched for cases of secondary cervical dystonia associated with structural lesions of the CNS. Search terms included one or more of the following: dystonia, torticollis, cervical, secondary, and symptomatic. Lesion localization, lesion type, patient age, patient gender, head position, occurrence of sensory tricks (i.e., *geste antagoniste*), and associated neurological findings were tabulated for each case. Care was taken to exclude nondystonic causes of abnormal head postures such as bony and congenital abnormalities of the cervical spine or skull base, congenital muscular torticollis, retropharyngeal and paraspinal infections, and reflex cervical spasm secondary to meningeal irritation.<sup>4,19,20</sup> Cases of cervical dystonia associated with isolated compression of the cranial nerves or cervical roots by mass lesions or vascular structures were also excluded from the analysis. Patients with cervical dystonia as part of generalized, cranio-cervical, or multifocal dystonia were not included in the analysis.

### RESULTS

The senior author's database contained 28 new cases of cervical dystonia over a 3-year period. Fourteen of these were cases of secondary cervical dystonia or cervical dystonia associated with hereditary degenerative diseases.<sup>4</sup> Four of the secondary cervical dystonia cases were associated with structural lesions of CNS.

#### Case 1

A 55-year-old man developed the acute onset of left hemiparesis and dysarthria. On initial neurological examination, the patient was noted to have bilateral abdu-

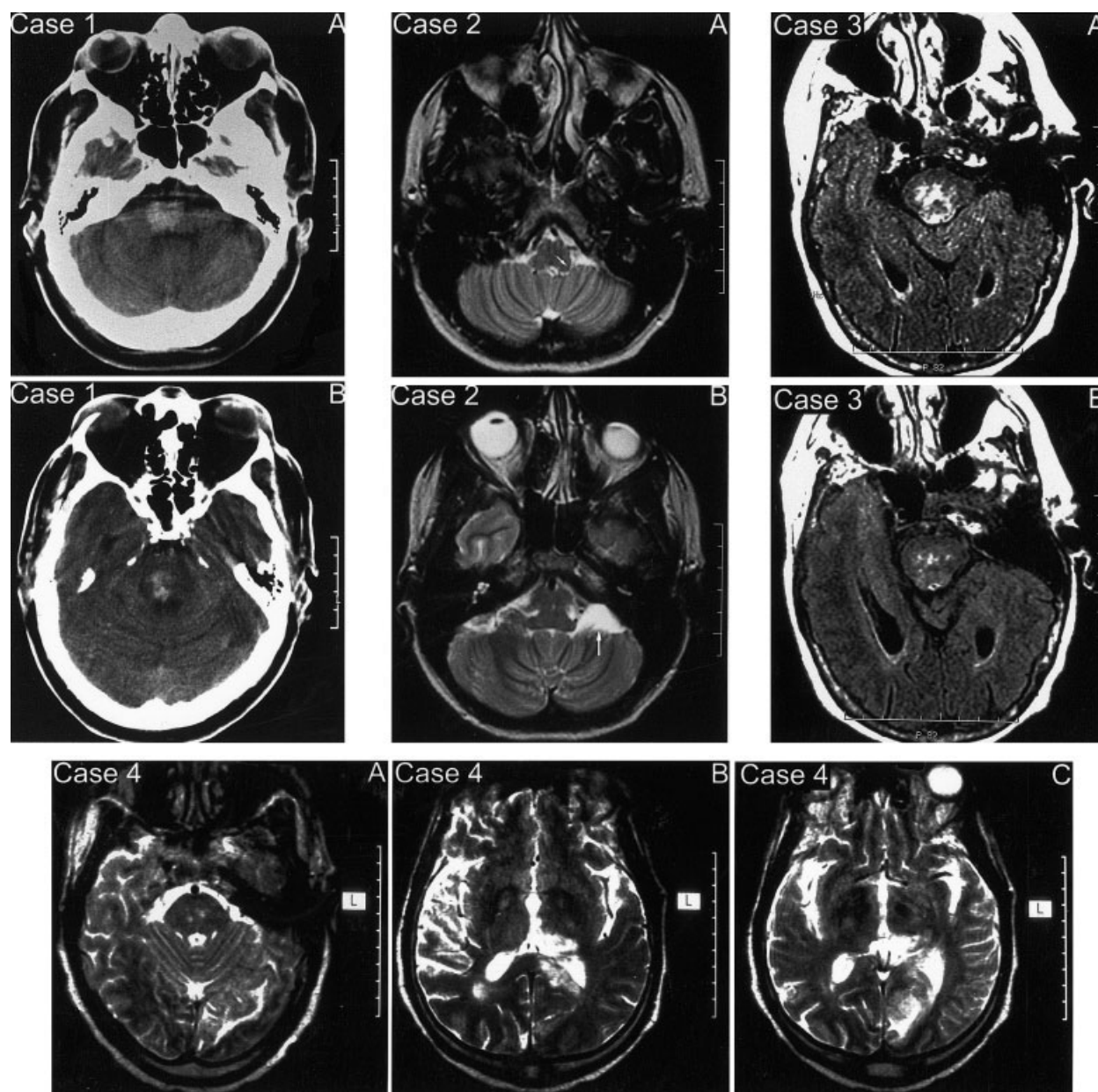
cent paresis, left facial paresis, left hemiparesis, and left hemihypesthesia. He was diffusely hyperreflexic and had bilateral Babinski responses. Computed tomography (CT) scan of the brain demonstrated a 15 mm hemorrhage centered in the right pons with some extension across the midline (Fig. 1, Case 1). Within 12 hours of admission, the patient began complaining of neck pain. One day after admission, the patient continued to complain of neck pain and was noted to have developed cervical dystonia with rotation of his head to the left and abnormal contraction of the right sternocleidomastoid muscle. Initially, the patient was unable to actively move his head to the right. Over the subsequent 4 to 6 weeks, however, he was increasingly able to move his head toward but not across the midline. There was no significant retrocollis/anterocollis, lateral shift, sagittal shift, or shoulder elevation. Cervical dystonia persisted and the patient developed progressive hypertrophy of the right sternocleidomastoid muscle. The patient died shortly after transfer to a nursing home.

#### Case 2

A 42-year-old woman presented for neurological evaluation after experiencing two brief episodes of ataxia and tinnitus 3 to 4 months earlier. Each episode lasted for approximately 20 minutes and was followed by a migraine headache. There was no interictal ataxia. The patient had a long history of infrequent migraine headaches and the paroxysms that she described were believed to be complicated migraine headaches. Neurological examination showed mild head rotation to the left and right laterocollis with subtle hypertrophy of the right sternocleidomastoid muscle. There was mild right shoulder elevation but no significant anterocollis/retrocollis, lateral shift, or sagittal shift. There were no other neurological abnormalities. There was no family history of movement disorders. The patient had never been treated with neuroleptics or anti-emetics. Audiometry was normal. A magnetic resonance image (MRI) of the brain demonstrated a small cystic lesion in the left cerebellopontine angle just posterior to the left facial/vestibulocochlear nerve complex with mild distortion of the rostral medulla (Fig. 1, Case 2). The MR lesion was interpreted to be an arachnoid cyst by a neuroradiologist. The patient refused both oral medications and botulinum toxin for treatment of her dystonia.

#### Case 3

A 67-year-old, right-handed woman fractured her sternum and multiple ribs after falling down her stairs at home. There was no cervical fracture or instability. She developed abnormal head posturing several days after



**FIG. 1.** **Case 1:** Non-contrasted CT images show a 15-mm hemorrhage centered in the right pontine tegmentum. **Case 2:** T2-weighted magnetic resonance images (MRI) through the posterior fossa show a 2-cm arachnoid cyst (large arrow) in the left cerebellopontine angle immediately caudal to the left facial/vestibulocochlear nerve complex with mild distortion of the rostral medulla (smaller arrow). **Case 3:** Magnetic resonance fluid-attenuated inversion-recovery (FLAIR) sequences show multiple high-signal abnormalities in the pontine tegmentum. **Case 4-A:** T2-weighted MRI shows bilateral ischemic infarctions localized to the pontine tegmentum. **Case 4-B:** T2-weighted MRI shows an ischemic infarction of the left thalamus. **Case 4-C:** T2-weighted MRI demonstrates a posterior cerebral artery distribution ischemic infarction involving the left occipital lobe.

admission. MRI of the brain obtained during hospitalization showed signal abnormalities in the pons and caudal midbrain consistent with multifocal ischemic lesions (Fig. 1, Case 3). On magnetic resonance angiography of the head and neck vessels, there were no focal areas of stenosis and no evidence for vertebral artery dissection. On outpatient neurological examination approximately

one year later, the patient was noted to have a right conjugate horizontal gaze deficit. She had cervical dystonia with severe right laterocollis and anterocollis along with moderate elevation and anterior displacement of the right shoulder. Her head was mildly rotated to the left. When the patient was awake, her head posture was almost fixed and could not be significantly altered either

actively or passively. The patient's head posture did not normalize, according to her husband, when she was asleep. The patient was mildly dysarthric. She had a very mild right hemiparesis. The patient had increased tone in the right upper extremity with cogwheeling at the wrist. There was a parkinsonian-type resting tremor of the distal right upper extremity. There was also a mild action tremor in the right upper extremity with both postural and kinetic components. There was no significant appendicular ataxia. Reflexes were brisk bilaterally in both the arms and legs. Plantar reflexes were equivocal bilaterally. The patient was ambulatory with a four-legged walker. Her gait was both spastic and ataxic. She had marked postural instability.

The patient's parkinsonism was treated with carbidopa-levodopa (25/100). At a dosage of one carbidopa-levodopa (25/100) p.o. three times daily, both her tremor and right upper extremity hypertonicity resolved. In addition, there was modest improvement in her postural instability. Her cervical dystonia did not improve with carbidopa-levodopa. With electromyographically guided botulinum toxin injections there was moderate improvement in her neck posture and pain.

#### Case 4

A 72-year-old man with a history of hypertension and gout presented to the emergency room with a 2-day history of altered mental status. On initial neurological examination, the patient was awake and alert but disoriented in all spheres. He had moderately severe anomia, right homonymous hemianopsia, mild perseveration, and right hemihypesthesia. As shown in Figure 1 (Case 4A–C), MRI of the brain demonstrated subacute infarctions of the pons, left posterior thalamus, and left occipital lobe. One day after presentation, the patient was noted to have developed cervical dystonia with moderately severe head rotation to the left. There was mild retrocollis. There was no significant laterocollis, lateral shift, sagittal shift, or shoulder elevation. Over the course of the patient's month-long hospital stay, his cervical dystonia slowly improved but was still present at the time of discharge to a nursing home. He died, apparently from sepsis, before his return clinic visit.

#### RESULTS

As shown in Table 1, critical clinical features (lesion localization, lesion type, patient age, patient gender, occurrence of sensory tricks, head position, and associated neurological findings) were tabulated for a total of 25 cases of secondary cervical dystonia. Four of the cases were from the senior author's database and 21 were published previously. Thirteen of the 25 patients were

female (52%). Age at presentation ranged from 9 months to 72 years with a mean of 38 years.

The largest percentage of cases of secondary cervical dystonia was associated with structural lesions of the brainstem (medulla, pons) and/or cerebellum ( $n = 11$ , 44%).<sup>21–24</sup> Of these 11 lesions, three were isolated to the pons and two were isolated to the cerebellum. The tegmentum was involved by all of the pontine lesions. There were single midbrain<sup>25</sup> and frontal lobe<sup>26</sup> lesions. The remaining lesions were equally divided between the cervical spinal cord ( $n = 6$ , 24%)<sup>27–30</sup> and basal ganglia ( $n = 6$ , 24%).<sup>14,31–34</sup> All but one of the spinal cord lesions had bilateral components. Three of the basal ganglia lesions were isolated to the putamen.<sup>31,32</sup> The other three basal ganglia lesions involved the head of the caudate nucleus and adjacent white matter.<sup>14,33,34</sup>

In the majority of cases, cervical dystonia included significant components of laterocollis, anterocollis–retrocollis, or shoulder elevation in addition to head rotation. The clinical information was inadequate to completely characterize head posture in several reports. In cases with both head rotation and a unilateral structural lesion ( $n = 13$ ), the direction of head rotation was contralateral to lesion localization in 9 cases and ipsilateral in 4 cases. Head rotation was contralateral to basal ganglia and cerebral lesions in all but one case. With cerebellar, brainstem (medulla, pons), and spinal cord lesions, head rotation was ipsilateral to lesion localization in 3 cases and contralateral in 4 cases.

Additional neurological abnormalities were present in 18 of the 25 cases of secondary cervical dystonia. Of the 7 cases with isolated cervical dystonia, 3 were associated with lesions of the putamen, 2 with posterior fossa masses, and 2 with tumors of the basal ganglia and adjacent white matter. Sensory tricks were reported in 5 of the 25 cases. The presence or absence of sensory tricks was not reported in 15 of the 25 cases. None of the patients with sensory tricks had basal ganglia lesions.

The temporal relationship between the onset of cervical dystonia and associated structural lesions was impossible to determine in the majority of the cases. Clearly, this relationship could not be determined in patients with tumors such as gliomas and schwannomas or other mass lesions like arachnoid cysts. In cases of cervical dystonia associated with strokes, however, onset was usually within a few days of the ictus. Clinical follow-up was available for 17 of the 25 cases. Nine of these patients had significant improvement or resolution of their cervical dystonia with surgery ( $n = 4$ ), botulinum toxin injections ( $n = 4$ ), or a combination of the two ( $n = 1$ ). Three patients showed some benefit from oral medications.

TABLE 1. Twenty-five cases of secondary cervical dystonia

Case no.	Reference	Age (yr)/gender	Lesion type	Lesion localization	Head position	Sensory trick	Other neurological abnormalities	Follow-up
1	LeDoux and Brady	55/M	Hemorrhage	R pons	Rotation-L	No	Bilateral abducens palsy, left hemiparesis	Mild improvement without treatment
2	LeDoux and Brady	42/F	Arachnoid cyst	L cerebello-pontine angle	Rotation-L, R laterocollis	No	None	NA
3	LeDoux and Brady	67/F	Ischemic infarctions	Bilateral pontine tegmentum	R laterocollis, anterocollis	No	Dysarthria, hyperreflexia, impaired conjugate gaze to R	Moderate improvement with botulinum toxin
4	LeDoux and Brady	72/M	Ischemic infarctions	Bilateral pontine tegmentum, L thalamus, L occipital lobe	Rotation-L	No	R homonymous hemianopsia, anomia, R hemi-hypesthesia	Moderate improvement without treatment
5	Boisen 1979	39/M	Hemangioblastoma	L cerebellar hemisphere	L laterocollis, anterocollis	NA	Nystagmus	NA
6	Boisen 1979	32/F	Ependymoma	Midline between cerebellar tonsils and medulla	Rotation-R	NA	None	NA
7	Kiwak et al. 1983	10/M	Intramedullary glioma with syrinx	Cervical cord to conus medullaris	Rotation-L, R laterocollis	NA	Paraparesis	NA
8	Kiwak et al. 1983	3/M	Grade II astrocytoma with syrinx	Medulla to conus medullaris	Anterocollis, R laterocollis	NA	Urinary incontinence	NA
9	Kiwak et al. 1983	0.75/F	Low-grade glioma	Cervical and thoracic cord	laterocollis	NA	Shoulder girdle muscle wasting, hyperreflexia	NA
10	Marsden et al. 1985	38/M	Calcified angioma	R caudate nucleus and adjacent white matter	Rotation L	NA	None	Temporary improvement after resection
11	Isaac and Cohen 1989	28/M	Hemorrhage	R putamen	Rotation-L, R shoulder elevation	NA	None	Some benefit from trihexyphenidyl
12	Plant et al. 1989	30/F	Multiple sclerosis plaques	R midbrain, R cerebellar hemisphere	Rotation-L	Yes	Gait ataxia, optic atrophy, spastic paraparesis	Persisted for 1 yr and then improved over weeks
13	Tranchant et al. 1991	53/F	Cavernous angioma	R cerebellar hemisphere	Rotation L, anterocollis, laterocollis	NA	R hemihypotonia	NA
14	Klostermann et al. 1993	34/F	Multiple sclerosis plaque	Posterolateral R C2	Rotation-L	Yes	Urinary incontinence, hyperreflexia, optic atrophy L, mild ataxia	Marked improvement with botulinum toxin

TABLE 1. (Continued)

Case no.	Reference	Age (yr)/gender	Lesion type	Lesion localization	Head position	Sensory trick	Other neurological abnormalities	Follow-up
15	Molho and Factor 1993	68/F	Ischemic infarction	L putamen	Rotation-R, mild L laterocollis & L shoulder elevation	NA	None	Successfully treated with botulinum toxin
16	Molho and Factor 1993	41/F	Ischemic infarction	L putamen	Rotation-R, R laterocollis, mild antero-collis	NA	None	Successfully treated with botulinum toxin
17	Cammerota et al. 1995	23/M	Ependymoma	C2 to C5	Rotation-R, L laterocollis	Yes	Gait ataxia, paraparesis	Resolution with surgical removal of tumor
18	Schulz-Bunhage and Ferbert 1995	40/M	Mixed grade III glioma	R basal ganglia and frontoparietal white matter	R laterocollis	NA	None	Dystonia resolved after tumor extended into the internal capsule
19	Schwartz et al. 1995	63/M	Ischemic infarction	R caudate putamen, and anterior limb internal capsule	Rotation-R, L laterocollis, R shoulder elevation	NA	Hyperreflexia	None
20	Caress et al. 1996	4/M	Gangliocytoma	R cerebellum, medulla, pons, cervical spinal cord	Rotation-L	NA	Gait ataxia, hyperreflexia	Improvement after subtotal resection of tumor
21	Soland et al. 1996	47/F	Angioblastic meningioma	R frontal	Rotation-L	Yes	Head tremor, poor short-term memory, reduced attention	Improvement after resection of tumor
22	Krause et al. 1997	42/M	Schwannoma adherent to CN IX, CN X, & CN XI	L cerebello-pontine angle with compression of cervical cord, brainstem, and cerebellum	Rotation-L, L laterocollis, L shoulder elevation	NA	L hypacusis	Mild improvement with surgery, marked improvement with botulinum toxin
23	Krause et al. 1997	13/F	Schwannoma adherent to CN IX, CN X	L cerebello-pontine angle with compression of brainstem	Rotation-R	NA	L hypacusis, ataxia, dysarthria, nystagmus	Resolution at an interval after surgery
24	Krause et al. 1997	52/F	Meningioma	R cerebello-pontine angle with compression of brainstem	Rotation-L, L laterocollis, antero-collis, L shoulder elevation, head tremor	Yes	R facial paresis with synkinesis	Some improvement with botulinum toxin and medications
25	Hill et al. 1999	48/F	Syrinx	C2 to thoracic cord	Rotation-L, L laterocollis, retro-collis	NA	Postural instability, reduced vibratory and proprioception sensation in feet	Mild improvement with botulinum toxin and medications

Roman numerals refer to respective cranial nerves.  
C, cervical; CN, cranial nerve; NA, not available.

## DISCUSSION

Four new cases of secondary cervical dystonia associated with structural lesions of the CNS have been described and cases reported previously have been reviewed to determine those neural networks potentially involved in cervical dystonia. The cases that we have presented underscore the difficulties in establishing causal relationships between CNS lesions and focal dystonias and highlight the complex and, possibly, heterogeneous pathophysiology of cervical dystonia. Three of the 4 cases either had multiple lesions or more than one potential etiological mechanism. For example, in Case 4 the initial MRI demonstrated lesions of the pons, thalamus, and occipital lobe. In this case, was cervical dystonia caused by the pontine lesions or by the combination of pontine, thalamic, and occipital lesions? Because head rotation was ipsilateral to the thalamic lesion, it may be that the pontine lesions were causally related to the patient's cervical dystonia.

The clinical similarities between secondary and primary cervical dystonia suggests that pinpointing the networks involved in the former may be applicable to the latter. Like patients with primary cervical dystonia, the majority of patients with secondary cervical dystonia exhibited head postures that were not limited to pure rotation. Patients with secondary cervical dystonia responded to treatments such as botulinum toxin injections and anticholinergics that are used for patients with primary cervical dystonia. Sensory tricks (i.e., *geste antagoniste*) have been reported in several patients with secondary cervical dystonia. Finally, some patients with secondary cervical dystonia associated with structural lesions of the CNS had no other abnormalities on neurological examination.

Secondary cervical dystonia must be distinguished from nondystonic causes of torticollis and other abnormal head postures.<sup>4,19,20</sup> Some authors have used the term "pseudodystonia" to classify disorders that are associated with sustained muscle contractions as a reflex mechanism or reaction to some other problem such as Sandifer syndrome or trochlear nerve palsy.<sup>4</sup> Pseudodystonia and nondystonia causes of torticollis would include abnormal head posturing as an attempt to reduce painful meningeal irritation, nausea, or vomiting associated with cervical and posterior fossa mass lesions or infections. Psychiatric, orthopedic, and congenital disorders simulating dystonia must be excluded by historical information, physical examination, and appropriate imaging studies.<sup>20</sup>

Review of Table 1 supports our hypothesis that structural lesions of the CNS associated with cervical dysto-

nia are concentrated in regions immediately afferent to the cerebellum including the pons, medulla, and spinal cord. A much smaller percentage of lesions were localized to the "basal ganglia." In addition, half of these "basal ganglia" lesions appeared to involve the anterior limb of the internal capsule, which contains fronto-pontine and other descending fiber systems. One feature common to most of the lesions listed in Table 1 is disruption of excitatory projections to the inferior olive or cerebellum.

Olivocerebellar dysfunction plays an important role in at least three rodent models of dystonia. First, a defect in transmission of information from the inferior olive to Purkinje cells in cerebellar cortex is central to the pathophysiology of dystonia exhibited by the genetically dystonic (dt) rat.<sup>11,35-38</sup> Second, rats treated with a neurotoxin (3-acetylpyridine) that selectively destroys inferior olivary neurons develop a movement disorder very similar to the dt rat's generalized dystonia.<sup>39</sup> Third, the mutant mouse tottering (tg) exhibits paroxysmal dystonia that is abolished on the Purkinje cell degeneration (pcd) mutant background.<sup>40</sup> One obvious conclusion based on a review of behavioral and electrophysiological studies in these rodent models is that a defect in climbing fiber excitation of Purkinje cells in cerebellar cortex can produce dystonia. Because climbing fibers originate in the inferior olive, higher-order defects in excitatory neurotransmission (i.e., olivary afferents) may also produce dystonia.

The inferior olive receives a wide variety of excitatory and GABAergic projections. Inferior olivary afferents arise from multiple areas including the spinal cord, medulla, pons, midbrain, cerebellar nuclei, and frontal cortex.<sup>41-44</sup> Projections from the red nucleus, periaqueductal gray, and other midbrain nuclei such as the interstitial nucleus of Cajal and nucleus of Darkschewitsch enter the central tegmental tract, descend uncrossed, and terminate in the inferior olive.<sup>45-47</sup> Fibers ascending in the spinal cord reach the inferior olive either directly through the spino-olivary tract that courses through the anterior funiculus or indirectly via the dorsal column nuclei.<sup>42,43</sup>

The specific idea that secondary cervical dystonia may be caused by a defect in inferior olivary afferents receives some indirect support from lesion and pharmacological studies in animals. For example, cervical dystonia has been produced in monkeys by creating electrolytic midbrain lesions<sup>48-50</sup> and, in rats, by injecting sigma receptor agonists into the red nucleus.<sup>51</sup> This evidence is, of course, nonspecific because both the red nucleus and midbrain nuclei send projections to several caudal structures besides the inferior olive.

In addition to the localization data presented in Table 1, the general idea that idiopathic cervical dystonia may not be due to a primary "basal ganglia" abnormality is suggested by a wide variety of physiological studies in humans. Acoustic reflex,<sup>52</sup> blink reflex, and masseter inhibitory reflex abnormalities in patients with cervical dystonia indicate the presence of abnormally excitable brainstem interneuronal pathways.<sup>53</sup> Impaired vestibulo-collic<sup>54</sup> and vestibulo-ocular<sup>55</sup> reflexes in patients with cervical dystonia and the appearance of cervical dystonia after unilateral vestibular nerve lesions<sup>56</sup> strongly suggest that deficits in vestibulocerebellar function may play a role in this movement disorder. Because the cerebellum is required for adaptive gain modification of both the blink<sup>57</sup> and vestibulo-ocular<sup>58</sup> reflexes, it would be reasonable to assume that olivocerebellar dysfunction could explain the physiological abnormalities detected in patients with primary cervical dystonia. Many investigators, however, have provided practical explanations for these physiological abnormalities based on dysfunction of the basal ganglia.

Rather than implicating any particular local-area network, functional imaging in patients with cervical dystonia, generalized dystonia,<sup>5</sup> and blepharospasm<sup>7</sup> presents, instead, a complex pathophysiological picture of dystonia that presents the possibility of substantial interactions between olivocerebellar and basal ganglia-thalamocortical circuits. In cervical dystonia, hypermetabolism was found in the basal ganglia, thalamus, premotor-motor cortex, and cerebellum.<sup>59</sup> A similar pattern of hypermetabolism was detected in both manifesting and nonmanifesting carriers of DYT1 mutations.<sup>5</sup> In movement-free conditions, both manifesting and nonmanifesting carriers showed increased metabolic activity in the lentiform nuclei, cerebellum, and supplementary motor areas. In movement-related conditions, manifesting carriers showed increased metabolic activity in the midbrain, cerebellum, and thalamus.

Anatomical and physiological interactions between the olivocerebellar and basal ganglia-thalamocortical circuits that have been demonstrated in both rodents and primates provide considerable insight into mechanisms potentially operative in cervical dystonia. For example, it has been shown that primary motor cortex is the transneuronal target of somatotopically organized outputs from both the cerebellum and basal ganglia.<sup>60</sup> Approximately 30% of neurons in the dentate nucleus and 15% of neurons in the internal segment of the globus pallidus project transneuronally to primary motor cortex via the thalamus. These projections terminate in overlapping fields onto specific motor representations within primary motor cortex. The cerebellum also sends projections to

several prefrontal regions via the thalamus.<sup>61</sup> Transneuronal projections from the basal ganglia terminate in some of the same regions as those from the cerebellum.<sup>62</sup> Terminal fields from the basal ganglia and cerebellum also overlap at subcortical sites. For example, the cerebello-tectal projection from the rat interpositus nucleus overlaps with terminals from the nigroreticular pathway originating in the dorsolateral substantia nigra pars reticulata.<sup>63</sup> These results suggest that: 1) olivocerebellar dysfunction can alter the physiology and metabolic topography of the basal ganglia-thalamocortical network<sup>22</sup>; 2) abnormalities of olivocerebellar and basal ganglia-thalamocortical circuits may, in some situations, produce similar movement disorders; and 3) the appearance of cervical dystonia, although due to a primary olivocerebellar abnormality, may require interaction between the olivocerebellar and basal ganglia-thalamocortical networks.

Secondary cervical dystonia associated with structural lesions of the CNS is much less common than nondystonic causes of torticollis. Abnormal head posturing in neonates and children should be considered nondystonic until proven otherwise. Adults with abnormal head postures that present with other neurological abnormalities or features atypical for idiopathic dystonia such as failure to improve with sleep, severe pain, headaches, nausea, or vertigo should undergo imaging of the brain and cervical spine.

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