

Post-traumatic Cervical Dystonia: A Distinct Entity?

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ABSTRACT: Background/Objective: The incidence of head/neck trauma preceding cervical dystonia (CD) has been reported to be 5-21%. There are few reports comparing the clinical characteristics of patients with and without a history of injury. Our aim was to compare the clinical characteristics of idiopathic CD (CD-I) to those with onset precipitated by trauma (CD-T). **Methods:** We evaluated 114 consecutive patients with CD over a 9-month period. All patients were interviewed using a detailed questionnaire and had a neurological examination. Their clinical charts were also reviewed. **Results:** Fourteen patients (12%) had mild head/neck injury within a year preceding the onset of CD. Between the two groups (CD-I and CD-T), the gender distribution (F:M of 3:2), family history of movement disorders (32% vs. 29%), the prevalence of gestes antagonistes (65% vs. 64%), and response to botulinum toxin were similar. There were non-specific trends, including an earlier age of onset (mean ages 43.3 vs. 37.6), higher prevalence of neck pain (86% vs. 100%), head tremor (67% vs. 79%), and dystonia in other body parts (23% vs. 36%) in CD-T. **Conclusions:** CD-I and CD-T are clinically similar. Trauma may be a triggering factor in CD but this was only supported by non-significant trends in its earlier age of onset.

RÉSUMÉ: La dystonie cervicale post-traumatique est-elle une entité distincte? Introduction/Objectif: L'incidence d'un traumatisme de la tête ou du cou précédant la dystonie cervicale (DC) serait de 5 à 21%. Il existe peu de publications comparant les caractéristiques cliniques des patients avec et sans histoire de traumatisme. Le but de cette étude était de comparer les caractéristiques cliniques de la DC idiopathique (DC-I) à celles de la DC précipitée par un traumatisme (DC-T). **Méthodes:** 114 patients consécutifs présentant une DC ont été évalués sur une période de 9 mois. On leur a administré un questionnaire détaillé lors d'une entrevue et tous ont subi un examen neurologique. Tous les dossiers cliniques ont également été révisés. **Résultats:** Quatorze patients (12%) avaient subi un traumatisme léger au niveau de la tête ou du cou dans l'année précédant le début de la DC. La distribution quant au genre (F:M 3:2), à l'histoire familiale de désordre du mouvement (32% vs 37.6%), la prévalence de gestes antagonistes (65% vs 64%) et la réponse à la toxine botulique étaient semblables. Il y avait des tendances non spécifiques, dont un âge de début plus précoce (âge moyen 43.3 vs 37.6 ans), une prévalence plus élevée de douleurs cervicales (86% vs 100%), de tremblements de la tête (67% vs 79%) et de dystonie localisée à d'autres parties du corps (23% vs 36%) dans la DC-T. **Conclusions:** La DC-I et la DC-T sont semblables au point de vue clinique. Un traumatisme peut être le facteur déclenchant de la DC, mais ceci n'est supporté que par une tendance non significative à un âge de début plus précoce chez les patients de cette étude.

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Dystonia is a syndrome characterized by sustained muscle contractions causing twisting or abnormal posturing, with fast movements sometimes superimposed.^{1,2} Cervical dystonia (CD) is a focal dystonia characterized by contractions of the neck muscles, resulting in forceful turning or pulling of the head, sometimes associated with shoulder elevation or head tremor.³⁻⁸ An etiologic factor is rarely found in focal dystonia, but post-traumatic cases have been described.⁹⁻¹⁴ Structural damage to the basal ganglia by trauma or cerebrovascular disease may cause hemidystonia.^{10,11} Peripheral injuries to the limbs or the neck, on the other hand, have been implicated in many cases of focal or segmental dystonia.^{11,12} A few patients with cervical dystonia have reported mild trauma to the neck (such as whiplash injury) in the weeks to months before symptom onset.^{9,13}

Several investigators have claimed that post-traumatic cervical dystonia (CD-T) has clinical features that distinguish it from idiopathic CD (CD-I).^{9,13,14} These features include: marked limitation of neck range of motion, the failure of gestes antagonistes (sensory tricks used to alleviate dystonia), the absence of transient symptomatic relief after sleep, tonic rather than phasic neck muscle contractions, and relatively poor

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response to treatment. In this study, we compared the clinical features of CD-I and CD-T in our clinic patients who regularly receive botulinum toxin (BT) injections.

METHODS

Patients

All patients were selected from our clinic population of CD patients at the University of British Columbia Health Sciences Centre. At the time of the interview, all but two patients were returning for injections of BT. These two patients had not yet received injections of BT as part of their treatment, but were scheduled to begin this treatment soon. All patients were informed that the data would be utilized for research and gave verbal consent to participate in the study.

Clinical assessment

Three neurologists experienced in movement disorders (A.S., P.K.P, and J.K.C.T.) conducted an interview (using a detailed questionnaire), the neurological examination, and the chart review. All patients had undergone a complete neurological examination on at least one occasion before being enrolled in the study. At the time of the interview, the neurological examination was focused to determine the direction of head deviation, the presence of tremor in the head, hands, or voice, and the presence of dystonia in other body parts.

Appendix 1 summarizes the items in the questionnaire pertinent to the present study. Age at onset, the interval between symptom onset and the diagnosis of CD by a neurologist, and the interval between symptom onset and first treatment (either systemic or injection of BT) were among the questions. We inquired about the family history of movement disorders, defined as the presence of any form of dystonia, tremor, or parkinsonism in first (parents, siblings, children) or second degree (grandparents, grandchildren, siblings of parents) relatives. History of trauma related to CD was defined as the occurrence of head/neck injury within one year of symptom onset. The severity of head/neck trauma was rated using a scale of 1-4 (Appendix 1).

We also inquired about the presence of pre-morbid and post-morbid psychiatric disorders requiring treatment, the presence of pain, head tremor, gestes antagonistes, hand tremor, and dystonia in other body parts. Since the main clinical symptoms of CD include head deviation, head tremor, and cervical pain, the order in which these three features occurred was documented as best as possible using the history provided by patients and a detailed review of medical records. Cervical pain was rated using a scale of 1-4 (Appendix 1). Spontaneous remission was defined as three months of complete symptom relief before BT treatment.

The response to BT injections was rated using a scale of 0-3 (Appendix 1). Since most patients had numerous injections of BT, we asked the patients to provide us with the average response to BT injections, once the optimal dose and target muscles were established (usually by the third injection). If the response fluctuated from injection to injection, the average response was taken for analysis.

Statistical analysis

The patients were divided into CD-I and CD-T groups depending on the presence of preceding head/neck trauma. The percentages of various characteristics in each of the two groups

Table 1: The clinical characteristics of patients with CD-I and CD-T.

Clinical characteristics	CD-I (n=100)	CD-T (n=14)
Female : male ratio	59 : 41	8 : 6
Age at onset	43.3 ± 13.6	37.6 ± 10.2
Time from onset to diagnosis (years)	6.2 ± 7.8	8.2 ± 13.0
Time from onset to 1st BT injection (years)	9.4 ± 10.0	10.2 ± 12.7
Duration of treatment with BT (years)	5.9 ± 3.6	6.0 ± 3.2
Interval between BT injections (months)	3.8 ± 5.0	3.1 ± 0.3
Family history of movement disorders	32 (32%)	4 (29%)
Pain associated with dystonia	86 (86%)	14 (100%)
Presence of head tremor	67 (67%)	11 (79%)
Presence of hand tremor	31 (31%)	3 (21%)
Presence of gestes antagonistes	65 (65%)	9 (64%)
Presence of extranuchal dystonia	23 (23%)	5 (36%)
History of pre-morbid psychiatric disease	23 (23%)	2 (14%)
History of post-morbid psychiatric disease	31 (31%)	5 (36%)

(CD-I and CD-T) were determined. Unpaired t-tests and chi-square tests were used to compare the characteristics of the two groups.

Results

Three patients with CD as part of a more extensive neurologic picture (one with cerebral palsy, one with multiple sclerosis, and one with generalized tardive dyskinesia) were excluded from the study. This left 114 patients (47 men and 67 women) whose data were analyzed for this study. Fourteen patients (six men and eight women) reported a history of head/neck injury within a year of torticollis onset.

The clinical characteristics of CD-I and CD-T patients are shown in Table 1, and the characteristics of individual CD-T patients are shown in Table 2. The age of onset was lower in CD-T (37.6 ± 10.2) than in CD-I (43.3 ± 13.6), but this difference was not statistically significant. Between the two groups, the gender distribution (F:M of approximately 3:2), a family history of movement disorders (32% vs. 29%), and the prevalence of gestes antagonistes (65% vs. 64%) were almost identical. The interval between symptom onset and the first BT injection, the duration of BT treatment, the presence of spontaneous remissions, as well as the response to BT (Table 3) were also quite similar between the two groups. In both groups, the direction of head deviation was fairly evenly divided between rotation to the left or the right. The same was true for anterocollis and retrocollis.

Cervical pain was more prevalent in CD-T, but the severity of pain in the two groups was comparable (Table 4). All CD-T patients had cervical pain, while 86% of CD-I patients had cervical pain associated with their torticollis. In CD-I patients, 11% had head tremor and 12% had cervical pain as their initial symptom, and the rest (77%) had head turning with or without cervical pain as their initial symptom. Half of the patients with

Table 2: Characteristics of individual CD-T patients. In three patients, the onset of symptoms occurred within 4 weeks of trauma. The characteristics of these patients are italicized.

CD-T patient	Trauma Type	Trauma severity	Interval to symptom	Gestes antagonistes	Response to BT
1	<i>MVA whiplash</i>	<i>Mild</i>	<i><4 weeks</i>	<i>No</i>	<i>No response</i>
2	<i>MVA whiplash</i>	<i>Moderate</i>	<i><4 weeks</i>	<i>No</i>	<i>Excellent</i>
3	<i>MVA whiplash</i>	<i>Minor</i>	<i><4 weeks</i>	<i>No</i>	<i>Excellent</i>
4	MVA whiplash	Mild	>4 weeks	Yes	Excellent
5	Car vs. pedestrian	Moderate	>4 weeks	Yes	Fair
6	Fall from stairs	Minor	>4 weeks	Yes	Fair
7	MVA whiplash	Mild	>4 weeks	Yes	Fair
8	Skating fall	Minor	>4 weeks	Yes	Good
9	Diving	Mild	>4 weeks	Yes	Good
10	Skiing Fall	Minor	>4 weeks	No	Good
11	Skating fall	Minor	>4 weeks	Yes	Good
12	MVA whiplash	Minor	>4 weeks	No	Good
13	MVA whiplash	Minor	>4 weeks	Yes	Good
14	Skiing fall	Minor	>4 weeks	Yes	Excellent

head tremor as the initial symptom had essential tremor involving the hands. In the CD-T group, pain was present at onset in all patients, and head tremor was never the initial symptom.

Hand tremor was more common in CD-I, but head tremor and the presence of dystonia in other body parts were more prevalent in CD-T. The most common sites of dystonia were the face, the eyelids, and the vocal cords. Pre-morbid history of psychiatric disease was more common in CD-I, while psychiatric disease after the onset of dystonia occurred more frequently in CD-T. The most common psychiatric diagnosis was depression, followed by anxiety disorder. None of these differences between CD-I and CD-T were statistically significant.

Table 3: Responses to treatment with botulinum toxin injection. See Appendix for a description of the response scale.

Response to BT treatment	CD-I (n=100)	CD-T (n=14)
No BT injections	2 (2%)	0
Excellent response	22 (22%)	4 (29%)
Good response	49 (49%)	6 (43%)
Fair response	23 (23%)	3 (21%)
No change to minimal improvement	4 (4%)	1 (7%)

Table 4: Severity of neck pain associated with CD. See appendix for a description of the pain scale.

Severity of pain	CD-I (n=86)	CD-T (n=14)
Minimal	21 (24%)	3 (21%)
Mild	36 (42%)	7 (50%)
Moderate	22 (26%)	2 (14%)
Severe	7 (8%)	2 (14%)

DISCUSSION

The epidemiology, clinical characteristics, and natural history of CD have been the subject of numerous reports.^{4,6,7,15-21} With the introduction of BT, the treatment outcome for CD has changed dramatically.²²⁻²⁴ Most reports on CD only discuss head/neck trauma briefly as a risk factor for CD, and studies specifically aimed at comparing CD-I and CD-T are scarce.^{9,13,25} In the present study, we compared the clinical features of CD-I and CD-T.

There were selection biases in our CD patients, most of whom were returning for BT injections. These patients had not experienced sustained remissions and continued to have significant symptoms requiring BT injections every 3-4 months. Indeed, the remission rate in our series was much lower than those from most other series.^{7,16-18,26,27} However, the mean age of onset, the female preponderance, the prevalence of family history and many other clinical characteristics seem to generally correspond to those of other reports,^{5-7,16-19,27,28} implying that our population of CD patients is in many ways similar to populations reported elsewhere.

A number of investigators have reported on the role of brain injury secondary to severe head trauma in the etiology of dystonia.^{10,29,30} Others have reported on the relationship between mild peripheral trauma and focal dystonias.^{9,13,31-33} Koller et al.¹¹ divided trauma into "cranial-cerebral" implying serious head injury, and "peripheral" implying mostly neck injury in relation to CD. Jankovic et al.¹² also suggested "central" and "peripheral" mechanisms for the pathogenesis of dystonia, depending on the site and severity of injury. In our series, none of the patients suffered from serious head injuries requiring hospitalization, and they would therefore be classified as "peripheral" cases. In our population, the prevalence of preceding trauma was 12%, which is in the typical range (5%-21%) reported by others.^{6,16,18,19,25,31,33}

We found that the clinical characteristics of CD-I and CD-T were more similar than different. These findings are similar to

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