SHORT COMMUNICATION

Prevalence of non-motor symptoms in cervical dystonia

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Introduction

Cervical dystonia (CD) is a movement disorder characterized by sustained or intermittent muscle contractions of cervical muscles leading to abnormal head movements, postures, or both. Dystonic movements are typically patterned and twisting, and may be tremulous. Dystonia is caused by overflow muscle activation, often initiated or worsened by voluntary action, such as gait or holding upright position. Quality and speed of voluntary movements are impaired by interference of abnormal dystonic contractions. Cervical dystonia represents the most common form of focal dystonia with prevalence estimated 28-183 cases/million (Defazio *et al* 2014), more commonly affecting women than men (approx. 2:1).

Despite the pure 'motor' definition of dystonia in common usage, recent studies have revealed that apart from the movement disorder, there are other, non-motor, features in many patients with dystonia, especially depression, anxiety, apathy, fatigue, sleep disturbances, sensory abnormalities and pain (Stamelou *et al* 2012). These papers are suggesting that the non-motor symptoms (NMS) have a larger impact on an individual's quality of life than that of their motor symptoms. In addition, there is growing evidence that NMS belong to the phenotypic spectrum of CD and they are not just the consequence of the motor impairment (Zurowski *et al* 2013, Kuyper *et al* 2011).

Within our ongoing larger study, we are investigating the prevalence of NMS by using an extended version of the NMS rating scale for CD patients as proposed by Kingelhoefer *et al* (2014).

Material and methods

Consecutive patients from specialised dystonia outpatients' clinics in the region of eastern Slovakia with clinically diagnosed cervical dystonia were enrolled in the study. Written informed consent was obtained from all participants. The study was approved by the local ethical committee.

Severity of CD was evaluated by Toronto Western Spasmodic Torticollis Rating Scale –TWSTRS (Consky & Lang 1994) and overall clinical severity of dystonia by using the 7-point Clinical Global Impression Scale – CGI-S dystonia (Guy 1976).

Assessment of NMS prevalence was performed by using the 14 item NMS list as proposed by Klingelhoefer *et al* (2014), with additional question about sexual problems (see Table 1). Furthermore, patients were asked to make a list of their five most bothersome complaints by choosing from the inventory composed of previous 15 NMS supplemented by list of motor symptoms related to CD (dystonic posturing of a head, tremor/jerks, limited movement, limited activities of daily living due to motor impairment).

Statistical analyses were performed by PASW Statistics 22 for Windows (SPSS Statistics, Chicago IL, USA). Correlation analyses were performed with the

Spearman rank correlation coefficient, as data were not normally distributed. A p-value of less than 0.05 was considered to indicate statistical significance.

RESULTS

Fourteen CD patients were included (11 females, 3 males) with a mean age 56.7 years (range 21–86 years). Mean age at onset of CD was 45.6 years (SD=19.5) and a mean disease duration of 9.7 years (SD=5.2). The mean total TWSTRS score was 16.00 (SD=4.9) and the mean CGI-S dystonia score 4.1 (SD=1.0). The prevalence of the NMS is displayed in Table 1. Only one patient didn't suffer from any NMS, the other 13 subjects indicated at least 1 NMS, while 8 patients (57.4%) experienced at least six NMS (out of 15). Average number of reported NMS was 5.9 (range 0–11). Loss of self-confidence (71.4%) and fatigue (64.3%) were reported most frequently. Individual list of the five most bothersome symptoms (NMS together with motor symptoms related to CD – Table 2) revealed again loss of self-confidence as the most frequent complaint (9×), followed by fatigue (8×) and abnormal head posturing (8×). There was no significant correlation of the amount of NMS and dystonia severity (p=0.171 for TWSTRS, resp. p=0.530 for CGI-S), nor of the number of NMS and disease duration (p=0.685).

Tab. 1. Prevalence of non-motor symptoms in CD patients.

Non-motor symptom	Prevalence
Loss of self-confidence due to stigma of visible head/neck dystonia	71.4 %
Fatigue or lack of energy which limits daytime activities	64.3 %
Any walking difficulty or balance problem	57.1 %
Difficulties falling or staying asleep	50.0 %
Feeling not refreshed after an overnight sleep	42.9 %
Feeling sad or depressed	42.9 %
Feeling nervous, worried or frightened for no apparent reason	42.9 %
Problems with or less interested in sexual activities	35.7 %
Experience of unpleasant sensation such as numbness, tingling or pins and needles	35.7 %
Pain, not explained by other conditions	28.6 %
Experience of light headedness or dizziness	28.6 %
Flat moods without the normal 'highs' and 'lows'	28.6 %
Dystonia affecting vision	21.9 %
Any speech problems	21.4 %
Difficulties while eating such as chewing or swallowing	14.3 %

Discussion

Most of the patients reported a presence of NMS, even 57.4% of the subjects indicated 6 or more complaints which might be regarded as a considerable amount of NMS and which is likely to interfere with activities of daily living (ADL) and health related quality of life (HRQoL) and what is in concordance with previous research in this field (Klingelhoefer *et al* 2014). We did not find a significant correlation of the number of NMS with disease severity or duration, although study of a larger sample will be needed to clarify this potential relationship.

Among the most frequently reported NMS of CD patients belong loss of self-confidence (71.4%), fatigue (64.3%), walking/balance difficulties (57.1%), insomnia (50.0%), not refreshing sleep (42.9%), depression (42.9%) and anxiety (42.9%). Remarkable finding

Tab. 2. Overview of the most bothersome symptoms in CD patients – both motor (distinguished by italics) and non-motor symptoms

Non-motor symptom	times listed in top 5 bothersome symptoms
Loss of self-confidence due to stigma of visible head/neck dystonia	9x
Fatigue or lack of energy which limits daytime activities	8x
Dystonic posturing of a head	8x
Any walking difficulty or balance problem	7x
Tremor/jerks	7x
Limited movement	5x
Pain, not explained by other conditions	4x
Limited activities of daily living due to motor impairment	4x
Feeling sad or depressed	3x
Feeling nervous, worried or frightened for no apparent reason	3x
Experience of unpleasant sensation such as numbness, tingling or pins and needles	3x
Difficulties falling or staying asleep	2x
Flat moods without the normal 'highs' and 'lows'	2x
Any speech problems	2x
Difficulties while eating such as chewing or swallowing	2x
Dystonia affecting vision	1x
Problems with or less interested in sexual activities	1x
Experience of light headedness or dizziness	1x
Feeling not refreshed after an overnight sleep	0x

was that from 5 most bothersome complaints chosen from the list of both motor and non-motor symptoms only 2 symptoms were related to the motor features of dystonia (dystonic posturing of a head – 8/14 pts and tremor/jerks – 7/14 pts), while the others were NMS as displayed in Table 2. It is not surprising that loss of self-confidence was the most annoying symptom (9/14 pts), given that CD commonly affect relatively younger people, many still working, while they have to deal with visible movement disorder. Stigma of visible head/neck dystonia was also reported in a study of 289 CD patients (Camfield *et al* 2002) showing that these patients have a disproportionate negative impact on physical role limitation, despite good physical function, compared to patients with other neurological disorders such as multiple sclerosis, Parkinson's disease and stroke. From neuropsychiatric NMS, fatigue was reported most commonly. Considering the smaller sample group, we were not able to distinguish whether this was central fatigue or secondary to a sleep disturbances which were also frequently indicated NMS – especially insomnia (in 50.0% pts) and feeling not refreshed after an overnight sleep (in 42.9% pts).

Since the burden of NMS was proven to play a key role in the determination of HRQoL in other movement disorders such as Parkinson's disease (Martinez-Martin *et al* 2012), similar impact of NMS on HRQoL and ADL could be also expected in CD patients. More robust studies on larger samples will be needed to evaluate the significance of NMS in CD.

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