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Nonmotor symptoms and focal cervical dystonia: Observations from 102 patients

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ABSTRACT

Introduction: Evidence is emerging that nonmotor symptoms (NMS) such as neuropsychiatric, sensory and sleep disturbances play an important role in dystonia.
Methods: In this observation, 102 patients with cervical dystonia (CD) were included. We evaluated the complaints about NMS with an adapted NMS Questionnaire and motor severity measured with Unified Dystonia Rating Scale.
Results: 95% of CD patients experienced NMS with 36% presenting with at least seven NMS. Mean UDRS score was 5.09 points (standard deviation (SD) ± 2.64 points, range 0.5–14.0). 42% patients had additional head tremor. The total number of experienced NMS was not significantly correlated with the age of the patients and disease duration of dystonia. There was also no significant association between number of NMS and sex. The correlation between number of NMS and motor severity assessed with UDRS was weak ($r_S = 0.23$; $p = 0.02$).
Conclusion: The number of NMS as a whole has been shown to be a key determinant of health related quality of live in patients with Parkinson's disease and we would assume that can influence this construct in patients with dystonia, too. As 95% of our patients with focal CD present with NMS, especially loss of self-confidence, insomnia, fatigue and pain, higher awareness for NMS needs to be raised in the context of routine clinical consultations in patients with focal dystonia.

Introduction

Dystonia represents a spectrum of movement disorders typically characterised by sustained or intermittent muscle contractions causing abnormal postures and often repetitive movements [1]. There is emerging evidence that nonmotor symptoms (NMS) may complicate motor presentation in dystonia although the pattern of NMS in specific focal dystonia is unclear [2]. NMS may occur in dystonic patients related to the pathophysiology as different networks are involved in dystonia (cortico-striato-thalamo-cortical and cortico-cerebellar) which may modulate NMS such as sensory, emotional, autonomic, cognitive and sleep/wake cycle related domains [3]. Furthermore, secondary mechanical consequences of focal dystonia, such as cervical dystonia may also lead to problems like local pain and sleep disruption.

At King's College Hospital in London, United Kingdom historical motor and nonmotor information from patients with dystonia as part of routine clinical care has been collected since the year 2000. In this report, we describe relevant clinical data from 102 patients with focal cervical dystonia (CD) with a particular focus on NMS which have been collected during clinical consultations. Furthermore, based on these results we discuss the feasibility of the future development of a dystonia specific questionnaire for screening of NMS.

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Methods

Patients

The King's College Hospital dystonia clinic serves a population of over 500 dystonia patients with generalised dystonia and focal dystonia such as limb, cervical and oro-facial dystonia. For this observation we only included patients with a confirmed diagnosis of cervical dystonia with sufficient historical information documented in the notes. All included patients are treated with botulinum toxin (BoNT) injections and patients undergoing deep brain stimulation were excluded from this report.

The clinical consultation included enquiry regarding specific NMS and acquisition of NMS related queries were aided by the supplementary use of specific questions from the NMSQuest, validated for use in Parkinson's disease (PD) [4]. The range of NMS related questions asked during clinical consultation were gleaned from an international advisory group (co-authors in this paper) and also patient experience. The use of NMSQuest in clinic was approved by the ethical committee of the relevant institution and also is part of a natural history study addressing NMS in PD (NILS: UKCRN Number 10084).

Additional assessments

As part of routine clinical care patients are assessed with the Unified Dystonia Rating Scale (UDRS) [5] and NMS were noted in the NMSQuest [4]. The use of an adapted version of the NMSQuest for use in patients with dystonia has been reported previously [6,7].

The UDRS includes ratings for 14 body areas of motor severity and duration of dystonia, maximal total score is 112. As we focused on patients with focal CD only patients with dystonia affecting the neck domain and shoulder/proximal arm domain have been included in this survey resulting in a maximal total score of 24 in the UDRS.

We adapted the original NMSQuest which is validated for PD only to suit dystonia related NMS in a pragmatic manner. 14 items from the original NMSQuest were used asking about the presence of NMS during the past month. However, as the NMSQuest is self completed and the wording of the questionnaire is adapted to lay expressions and the response option is binary "Yes" or "No", it could easily be incorporated in clinical assessment of patients particularly as many NMS listed in the NMSQuest overlap with the ones experienced by CD patients. A high total number of NMS experienced by the patient can be rated as a considerable amount of NMS, although information on symptom severity and frequency is not obtained.

Statistical analyses were performed using *t*-test for normal distributed values and Mann–Whitney test for not normal distributed values. Correlation analyses were performed with the Spearman rank correlation coefficient. A p value of less than 0.05 was considered to indicate statistical significance.

Results

We were able to audit 102 focal CD cases with documented NMSQuest data. Patient demographics and clinical characteristics are described in Table 1.

Mean UDRS score was 5.09 points (standard deviation (SD) ± 2.64 points, range 0.5–14.0). 42% (43/102) patients had additional head tremor. Reported NMS aided by the use of the adapted NMSQuest are shown in the Table 2.

In total 14 questions on NMS were asked in a "yes"/"no" format with therefore a range from 0 to 14 possible "yes" answers with higher numbers meaning presentation of more NMS. Only 5% (5/102) of all patients with focal CD reported no NMS, 36% (37/102) presented with at least seven NMS which might be regarded as a considerable amount of NMS. When the total number of "yes" answers was correlated with the age of the patients ($r_S = -0.03$; $p = 0.75$) and disease duration of the dystonia ($r_S = 0.01$; $p = 0.89$), no significance was found. Three patients with a disease duration shorter then one year presented with 6 and 8 NMS, respectively. Furthermore no significant difference in the number of NMS by sex was found ($p = 0.78$). On the other hand, we found a significant weak correlation between the number of NMS and motor severity assessed by UDRS ($r_S = 0.23$; $p = 0.02$).

Table 1
Characteristics of patients with focal cervical dystonia.

Characteristics	Patients with focal cervical dystonia ($N = 102$)
Age {mean ± standard error, in years}	59.19 ± 1.21
Male sex – no. (%)	33 (32.4)
Ethnicity white – no. (%)	96 (94.1)
Duration of disease {mean ± standard deviation, in years}	10.99 ± 7.10
Range of duration of disease {minimum–maximum, in years}	0–44

Discussion

Our report is a clinical observational survey and provides a "snapshot" summary of the range of NMS which could be experienced in a common variant of focal dystonia such as cervical dystonia. However, this was neither a study to validate a new instrument for dystonia nor to investigate the pathophysiological basis of the reported symptoms. Therefore the discussion will focus on the clinical characteristics of the reported NMS.

Firstly we wish to emphasise that the chosen NMS were based on a set of questions which was derived from clinical opinions from a group of clinicians dealing with day to day assessment of dystonia patients and incorporated to clinical assessment. Such assessment was aided by the use of the NMSQuest, a self completed tool which was developed and validated from the

Table 2
Nonmotor symptoms reported by patients with focal cervical dystonia, responses (percentages reporting "yes" to questions, presented from most frequent to least) based on the use of the adapted version of the NMSQuest.

Nonmotor symptoms reported by patients with focal cervical dystonia	Percentage of "yes" answers ($N = 102$) {%}
Loss of self-confidence due to stigma of visible head/neck dystonia	61.8
Difficulties falling or staying asleep	59.8
Fatigue (tiredness) or lack of energy which limits daytime activities	51.0
Any walking difficulty or balance problem	49.0
Experience of light headedness or dizziness	47.1
Pain not explained by other conditions	43.1
Feeling not refreshed after an overnight sleep	40.2
Feeling nervous, worried or frightened for no apparent reason	30.4
Feeling sad or depressed	30.4
Experience of unpleasant sensation such as numbness, tingling or pins and needles	30.4
Difficulties while eating such as chewing or swallowing	25.5
Flat moods without the normal "highs" and "lows"	23.5
Dystonia affecting vision	21.6
Any speech problems	13.7