Parkinsonism and Related Disorders 15 (2009) 758–761

Contents lists available at ScienceDirect

Parkinsonism and Related Disorders

journal homepage: www.elsevier.com/locate/parkreldis

Motor overflow and mirror dystonia☆

Oraporn Sitburana[a,b], Laura Jui Chen Wu[a], James K. Sheffield[a], Anthony Davidson[a], Joseph Jankovic[a,*]

[a] Parkinson's Disease Center and Movement Disorders Clinic, Baylor College of Medicine, Department of Neurology, The Smith Tower, Suite 1801, 6550 Fannin, Houston, TX 77030, USA
[b] Neurology Center, Bumrungrad International Hospital, Bangkok, Thailand

ARTICLE INFO

Article history:
Received 5 January 2009
Received in revised form
7 March 2009
Accepted 9 May 2009

Keywords:
Dystonia
Motor overflow
Mirror dystonia
Focal hand dystonia

ABSTRACT

Motor overflow is an unintentional muscle contraction which accompanies, but is anatomically distinct from the primary dystonic movement. This phenomenological nosology has not been systematically studied in focal hand dystonia (FHD). We conducted a prospective, case-control study to characterize motor overflow and mirror dystonia in patients with FHD. We compared the performance of 30 patients with FHD and 40 healthy controls on a variety of motor tasks, such as writing, drawing a spiral, straight line and a sine wave, repetitive wrist flexion–extension, finger tapping, hand grasping, hand pronation–supination, and a finger-to-nose task with each hand. The assessments were videotaped, the edited video segments were randomized, and an independent investigator who was "blind" to the subject's diagnosis rated the ipsilateral and contralateral overflow and mirror dystonia twice, 6 months apart. Using the mean of the two ratings, ipsilateral overflow was identified in 8.5 ± 2.1 (28%) patients and in 1.5 ± 0.7 (4%) controls ($p < 0.001$), contralateral overflow in 2.5 ± 0.7 (8%) patients and in 1.5 ± 0.7 (4%) of controls ($p = 0.138$), and mirror movement in 20.0 ± 0.0 (67%) of patients and in 15.5 ± 4.9 (39%) of controls ($p = 0.001$). There was a statistically significant correlation of dystonia and overflow score (Pearson's r 0.713, $p < 0.001$). The relatively high frequency of ipsilateral overflow and mirror dystonia in patients with FHD has both pathophysiological and therapeutic implications. In this study, the severity of dystonia was significantly correlated with motor overflow in multiple tasks.

1. Introduction

Focal dystonia is characterized by sustained muscle contractions, frequently causing twisting and repetitive movements, or abnormal posture, which affects only a single body part [1–3]. Task-specific dystonia, most typically represented by focal hand dystonia (FHD), has been classified according to the specific impairment of a particular function, such as writer's cramp, musician's cramp, occupational cramps, or sport performance dystonia [4–7]. FHD was first described in the eighteenth century by Ramazzini after observations of workers who developed disabilities of their hands after continually performing their particular jobs [8]. In some patients the dystonia associated with task specificity may subsequently include other activities by spreading to other anatomical areas, and may even occur at rest [6]. FHD is classified as simple or complex depending on whether symptoms occur only in one specific task or in multiple tasks [9,10]. Mirror movements are defined as involuntary movements occurring on one side of the body, which are same or similar in character, to a voluntary movements performed contralaterally. In addition to their occurrence in otherwise normal individuals [11], mirror movements have been reported in many neurodegenerative disorders including Friedreich's ataxia [12], Huntington's disease [13], corticobasal degeneration [14] and Parkinson's disease [15]. Motor overflow commonly found in dystonia is unintentional muscle contraction which accompanies, but is anatomically distinct from the primary dystonic movement [16]. In our pilot study [17] we identified three different patterns of abnormal muscle activity in patients with FHD: 1. *Ipsilateral overflow*: involuntary contraction of muscles adjacent to those involved in the focal dystonia; 2. *Contralateral overflow*: characterized by an involuntary movement or dystonic posture in the normal, contralateral limb during dystonic movements or posture of the hand primarily affected by FHD, and 3. *Mirror dystonia*: a dystonic movement or a posture in the dystonic limb induced by a specific task such as writing performed by the opposite, normal body part (Fig. 1). We prospectively examined and compared patients with FHD to normal controls and found evidence of substantial motor control abnormality beyond the primary movement disorder.

2. Methods

Thirty consecutive patients with task-specific FHD (mean age 51.0 ± 11.8 years, 46.7% male) and forty normal controls (mean age 58.7 ± 14.2 years, 47.5% male) were

☆ The review of this paper was entirely handled by an Associate Editor, Dr. R.L. Rodnitzkyi.

* Corresponding author. Tel.: +1 713 798 5998; fax: +1 713 798 6808.
E-mail address: josephj@bcm.tmc.edu (J. Jankovic).

1353-8020/$ – see front matter © 2009 Elsevier Ltd. All rights reserved.
doi:10.1016/j.parkreldis.2009.05.003

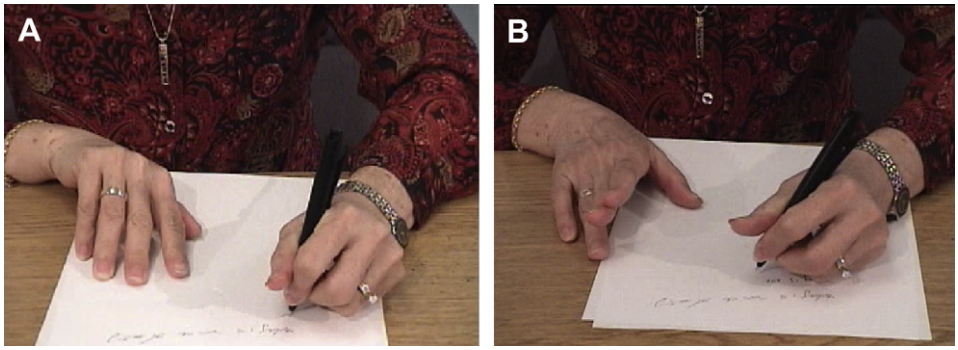

Fig. 1. Mirror dystonia (A) Patient starts writing with the non-affected (left) hand and (B) within a few seconds develops mirror dystonia, manifested by extension of the 2nd and 3rd digits in the right hand.

recruited from the Baylor College of Medicine Movement Disorders Clinic. The control subjects were persons accompanying the patients in the clinic who are not genetically related to the patients. Only patients diagnosed with primary dystonia were included [1]. After signing a consent form to participate in the study and to be videotaped, approved by the Baylor College of Medicine Institutional Review Board, all subjects were instructed to perform multiple tasks described below while being videotaped. Subjects were asked to sit behind a table adjusted to a comfortable height. A video camera was positioned to include the head, the upper part of body, both arms and lap in the frame. All subjects were instructed to perform three iterations of writing a sentence, drawing a spiral, a straight line and a sine wave. While performing writing tasks with one hand, the forearm of the opposite hand rested on a support 3 inches in height, thus allowing the hand to be free and move without constraint. Additionally, subjects were asked to perform five repetitive hand tasks with each hand sequentially for 10 s, including repetitive wrist flexion–extension with the arm in an outstretched position, finger tapping (tapping of the index finger on the thumb), rapid hand opening–closing, hand pronation–supination, and finger-to-nose movements. Subjects were also videotaped in a sitting position with the palms of their hands resting in the lap for 15 s, and while walking approximately 40 ft. All 70 video segments were randomized and rated by a movement disorder neurologist, who was "blind" to the history and diagnosis of the subjects, and who was trained in the Burke–Fahn–Marsden (B–F–M) [18] and motor overflow scale (Appendix).

To assess scoring reliability the same rater completed a second assessment of all 70 video segments six months after the first rating and intra-rater reliability was assessed. Mirror dystonia was identified when the rater observed dystonic postures or movements of the affected limb as subjects performed specific tasks with the non-affected limb. Ipsilateral or contralateral overflow was identified when subjects performed specific tasks with the affected limb. Contralateral overflow is defined as an involuntary movement in the normal (unaffected) limb while tasks are being performed by dystonic (affected) hand. In contrast, mirror dystonia is an involuntary movement in the dystonic (affected) hand while tasks are performed by the normal (unaffected) hand. In calculating values to describe the occurrence of the various types of movements, the number of occurrences of each type in each round of rating was counted, then the two numbers were averaged.

2.1. Statistical analysis

Statistical analysis was performed using Stata V 5.0 (Stata Corporation, College Station, TX). Descriptive analyses included mean and standard deviation for age at onset and at enrollment, duration of dystonia, and proportion of sex for both patients and controls. The different patterns of motor overflow and the validity of each task including the sensitivity, specificity and positive predictive value (PPV) for mirror dystonia were calculated and averaged between first and second ratings. Summary statistics were presented as mean ± standard deviation. Intra-rater reliability was calculated using unweighted Cohen's kappa (κ) statistic for the categorization of motor overflow movements, mirror dystonia in each specific task, dystonia and overflow scores. The comparison of different group means was analyzed with Student's *t*-test with unequal variance. Results were considered to be significant if the *p*-value was 0.05 or less ($p < 0.05$). The correlation of dystonia and overflow scores was calculated using the Pearson product-moment correlation coefficient, *r*.

3. Results

The mean age at disease onset of the 30 FHD patients (48% male) was 41.2 ± 11.9 years with a mean duration of 9.7 ± 7.4 years. Right-hand dominance was reported by 90% of the 30 FHD patients and 95% of the 40 normal controls. There were 26 (86.7%) patients with writer's cramp and 4 with task-specific FHD: 2 pianists, 1 drum player and 1 professional pistol shooter. In FHD patients, there was low-to-moderate intra-rater agreement of both ipsilateral overflow and mirror dystonia ($\kappa = 0.27$–0.55) but no agreement for contralateral overflow in controls (Table 1). The rating consistency of each specific task for mirror dystonia also showed low-to-moderate agreement. ($\kappa = 0.22$–0.52) (Table 2).

Based on the average of the two ratings performed, motor overflow, including ipsilateral overflow, was identified in 8.5 ± 2.1 (28%) FHD patients and 1.5 ± 0.7 (4%) controls ($p < 0.001$), contralateral overflow in 2.5 ± 0.7 (8%) patients and 1.5 ± 0.7 (4%) controls ($p = 0.138$), and mirror dystonia in 20.0 ± 0.0(67%) of patients and in 15.5 ± 4.9 (39%) controls ($p = 0.001$) (Table 1, Fig. 2). Writing with the non-affected hand had a sensitivity of 53%, a specificity of 61% and a PPV of 51% to detect mirror dystonia ($p = 0.017$). Finger tapping, hand grasping and hand pronation–supination had the highest specificity and PPV with statistical significance compared to the other tasks (Table 2). Based on the two separate assessments of the FHD patients, the average dystonia score was 1.4 ± 1.3 and the mean overflow score was 1.0 ± 0.8 ($\kappa = 0.31$, $p < 0.001$). The correlation

Table 1
The different patterns of motor overflows.

	Ipsilateral overflow	κ[a]	Contralateral overflow	κ[a]	Mirror dystonia	κ[a]
Patients (*n*)						
R1st:R2nd[b]	7:10	0.27	3:2	−0.09	20:20	0.55
Average[c]	8.5 ± 2.1 (28%)		2.5 ± 0.7 (8%)		20.0 ± 0.0 (67%)	
Controls (*n*)						
R1st:R2nd[b]	2:1	−0.04	2:1	0.66	12:19	0.34
Average[c]	1.5 ± 0.7 (4%)		1.5 ± 0.7 (4%)		15.5 ± 4.9 (39%)	
p value[d]	<0.001		0.138		0.001	

[a] κ = Cohen's kappa for intra-rater agreement.
[b] R1st = the first rating (*n* = number of subjects positive for trait); R2nd = the second rating (*n* = number of subjects positive for trait).
[c] Average number of subjects between first and second ratings, mean ± SD (%).
[d] *p* Value between patients and controls.