Parkinsonism and Related Disorders xxx (2016) 1–7

Contents lists available at ScienceDirect

Parkinsonism and Related Disorders

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

Review article

Tremor in dystonia

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ARTICLE INFO

Article history:
Received 28 December 2015
Received in revised form
29 February 2016
Accepted 31 March 2016

Keywords:
Dystonia
Essential tremor
Neurophysiology

ABSTRACT

Tremor has been recognized as an important clinical feature in dystonia. Tremor in dystonia may occur in the body part affected by dystonia known as dystonic tremor or unaffected body regions known as tremor associated with dystonia. The most common type of tremor seen in dystonia patients is postural and kinetic which may be mistaken for familial essential tremor. Similarly familial essential tremor patients may have associated dystonia leading to diagnostic uncertainties. The pathogenesis of tremor in dystonia remains speculative, but its neurophysiological features are similar to dystonia which helps in differentiating it from essential tremor patients. Treatment of tremor in dystonia depends upon the site of involvement. Dystonic hand tremor is treated with oral pharmacological therapy and dystonic head, jaw and voice tremor is treated with injection botulinum toxin. Neurosurgical interventions such as deep brain stimulation and lesion surgery should be an option in patients not responding to the pharmacological treatment.

1. Introduction

Hermann Oppenheim described “dystonia musculorum deformans” in 1911, but detailed description of tremor in dystonia was reported by Larsson and Sjogren in 1966 [1,2]. Subsequently, more studies (Yanagisawa and Goto in 1971 [3] and Marsden and Harrisson in 1974 [4]) reported tremor in dystonia patients, but there was no consensus regarding its definition and classification. In 1984 dystonia medical research foundation provided the first consensus definition of dystonia but tremor was not part of that [5]. In 1998 a consensus statement of the movement disorder society (MDS) on tremor proposed to define dystonic tremor syndromes under two broad groups, dystonic tremor (DT) and tremor associated with dystonia (TAD) [6]. DT was defined as tremor in a body part affected by dystonia, classically seen in spasmodic torticollis patients. TAD was defined as tremor occurring in a body part not affected by dystonia like the upper limb postural tremor in cervical dystonia (CD) patients. In 2013 a consensus committee of MDS came out with several shortcomings of dystonia definition published in 1984 and proposed a new definition which stated that dystonic movements may also be tremulous [7].

But still there are many uncertainties regarding tremor in dystonia patients [8]. One of the most controversial and difficult situation is to differentiate postural and kinetic tremor in TAD from familial essential tremor (ET) patients as they look similar clinically leading to misdiagnosis [9]. Similarly dystonia has been reported in ET patients and relationship among these tremor disorders have long been debated [8].

We reviewed tremor in dystonia with an aim to understand its epidemiology, clinical features including overlap with ET, pathogenesis, and current treatment strategy.

1.1. Search strategy and selection criteria

Relevant studies on tremor in dystonia were reviewed using the PubMed search from 1st January 1960 to 3rd November 2015. A total of 365 articles on ‘Tremor associated with dystonia’, 439 articles on ‘dystonic temor’, 438 articles on ‘dystonia in ET’, 9 articles on ‘neurophysiology of dystonic tremor’, 22 articles on ‘neurophysiology of tremor in dystonia’ 19 articles on ‘neuroimaging of dystonic tremor’, 79 articles on ‘neuroimaging of tremor in dystonia’ 216 articles on ‘treatment of dystonic tremor’ and 181 articles on “treatment of tremor associated with dystonia” were found. The final reference list was based on the relevance to the topic of review.

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http://dx.doi.org/10.1016/j.parkreldis.2016.03.024
1353-8020/

1.2. Epidemiological studies and clinical features

There is a wide variability in studies reporting the prevalence of tremor in dystonia patients ranging from 14% to 86.67%, which may be due to the type of dystonia patients selected in different studies [10–19] (Table 1). These studies suggest that tremor in dystonia is usually postural or kinetic (see video) and rest tremor is rare. The demographic data from studies done by Defazio et al. and Erro et al. indicate higher female to male preponderance in dystonic tremor and tremor associated with dystonia [10,19]. Defazio et al. also reported slightly higher ($p = 0.13$) age at dystonia onset for patients with tremor (52.9 ± 14.9) than patients without tremor (50.2 ± 15) [10]. Similarly, less frequent tremor has been reported by Koukouni et al. who published a detailed clinical description of 76 young onset (<28 years) cervical dystonia patients [20]. Head tremor was present in 13 patients (17.1%) and arm tremor in 10 patients (13.20%) only [20]. Primary dystonia patients are more likely to have tremor than secondary dystonia patients including tardive dystonia. In a retrospective study Molho et al. compared the clinical details of 102 idiopathic cervical dystonia patients and 20 tardive dystonia patients [21]. Head tremor was present in 43 and hand tremor was present in 16 idiopathic cervical dystonia patients, but none of the tardive dystonia patients had a head tremor and only 2 had hand tremor. The authors concluded that dystonic head tremor was strongly suggestive of idiopathic form. The body distribution of tremor has been recently reviewed by Defazzio et al. [10]. They summarized the different studies reporting dystonic tremor in different body parts. Head tremor was most common, followed by upper limb tremor, whereas voice and leg tremor were rare. Tremor is also more common in certain types of dystonia. Erro et al. reported tremor to be more frequent in segmental and multifocal dystonia compared to focal dystonia ($p < 0.01$). Among focal dystonia, head and arm tremor were more frequent in cervical dystonia patients than cranial dystonia and writer's cramp patients [19]. Similarly Defazio et al. also reported that cervical dystonia was more common in the DT and DT + TAD groups. DT alone was 3–4 times more common in cervical dystonia than upper limb dystonia ($p < 0.0001$) on comparison within the group, but TAD was more common in cranial dystonia than neck, larynx and upper limb dystonia ($p < 0.0001$). However, these findings are not consistent and Rudzinska et al. reported almost similar percentage of hand tremor with all forms of dystonia [18]. Dystonic head tremor has also been described as a dominant clinical feature of DYT24 (ANO3 mutation) and DYT 25 (GNAL gene mutation) patients [22,23]. Rarely tremor can occur without overt dystonia in DYT24 patients [22].

Case 1. Video still case 1: Right torticollis with horizontal tremulous and jerky head movements.

Case 2. Video still case 2: Retrocollis with yes-yes type of dystonic head tremor.

Table 1
Clinical studies of tremor in dystonia patients.

Study	Number of patients	Details of tremor
Marsdon 1971	42 Idiopathic torsion dystonia patients	Postural hand tremor in 6 patients (14%)
Couch 1976	30 Patients of torticollis	26 Patients (86.67%) had tremor (postural in all and rest in 2) which was mild in 12, moderate in 11, and severe in three
Chan 1991	266 Patients of idiopathic cervical dystonia	23% Patients had postural or kinetic hand tremor and 28% had dystonic head tremor
Jankovic 1991	300 Patients with cervical dystonia	71% Patients had tremor which was present in head-neck region in 60% and other body regions in 32%. Head-neck tremor was classified as dystonic (37%), essential (30%) and combination of dystonic and essential (8%). Other body region tremor was postural hand tremor (23%), trunk tremor (5%) and tremor involving voice, lips or legs (4%)
Jedynak 1991	45 Idiopathic dystonia patients with tremor	26 Patients had hand tremor (including writer's tremor), 13 patients had neck tremor, 5 patients had head tremor, and 1 patient had lower limb tremor
Deuschl 1997	55 Patients of idiopathic dystonia	33 Patients (60%) had tremor which was present in hand (22), head (22) and combination of hand and head (11).
Pal 2000	114 Patients with cervical dystonia	78 Patients (68.4%) had head tremor and 8.3% patients had hand tremor. 40% of patients with head tremor also had hand tremor
Defazio 2013	429 Patients of dystonia	72 Patients (16.7%) had tremor. 43 patients had dystonic tremor (DT), 23 had tremor associated with dystonia (TAD) while 6 had combination of DT and TAD. All had postural or postural/kinetic tremor and none had rest tremor
Rudzinska 2013	123 Patients of focal and segmental	60 Patients (48.8%) had tremors, which were postural in 41 (33.3%), kinetic in 13 (10.6%), and rest in 6 (4.9%). Tremor was mild in 44 (35.8%), moderate in 12 (9.8%), and severe in 4 (3.3%) patients.
Erro 2013	473 Patients of adult onset primary dystonia	262 Patients (55.4%) had head (196 patients, 74.80%) and arm (140 patients, 53.43%) tremor. Arm tremor was postural in all, kinetic in 103 (73.6%) and rest in 57 patients (40.7%).

Please cite this article in press as: S. Pandey, N. Sarma, Tremor in dystonia, Parkinsonism and Related Disorders (2016), http://dx.doi.org/10.1016/j.parkreldis.2016.03.024