ORIGINAL PAPER/*ARTYKUŁ ORYGINALNY*

Tremor associated with focal and segmental dystonia

Drżenie towarzyszące dystonii ogniskowej i segmentarnej

Monika Rudzińska, Małgorzata Krawczyk, Magdalena Wójcik-Pędziwiatr, Andrzej Szczudlik, Anna Wasielewska

Department of Neurology, Jagiellonian University Medical College, Krakow, Poland

Neurologia i Neurochirurgia Polska 2013; 47, 3: 223-231
DOI: 10.5114/ninp.2013.35584

Abstract

Background and purpose: Tremor occurs in 10-85% of patients with focal dystonia as so-called dystonic tremor or tremor associated with dystonia. The aim of this study was to assess the incidence and to characterize parameters of tremor accompanying focal and segmental dystonia.

Material and methods: One hundred and twenty-three patients with diagnosis of focal and segmental dystonia together with 51 healthy controls were included in the study. For each participant, clinical examination and objective assessment (accelerometer, electromyography, graphic tablet) of hand tremor was performed. Frequency and severity of tremor were assessed in three positions: at rest (rest tremor); with hands extended (postural tremor); during 'finger-to-nose' test and during Archimedes spiral drawing (kinetic tremor). Based on the mass load test, type of tremor was determined as essential tremor type or enhanced physiological type.

Results: The incidence of tremor was significantly higher in dystonic patients as compared to controls ($p = 0.0001$). In clinical examination, tremor was found in 50% of dystonic patients, and in instrumental assessment in an additional 10-20%. The most frequent type of tremor was postural and kinetic tremor with 7 Hz frequency and featured essential tremor type. In the control group, tremor was detected in about 10% of subjects as 9-Hz postural tremor of enhanced physiological tremor type. No differences were found between patients with different types of dystonia with respect to the tremor incidence, type and parameters (frequency and severity). No correlations between tremor severity and dystonia severity were found either.

Key words: focal dystonia, accompanying tremor, tremor parameters.

Streszczenie

Wstęp i cel pracy: Drżenie stwierdzane jest u 10–85% chorych z dystonią ogniskową jako tzw. drżenie dystoniczne lub drżenie towarzyszące dystonii. Celem badania była ocena częstości występowania i charakterystyka parametrów drżenia towarzyszącego dystonii ogniskowej i segmentalnej.

Materiał i metody: Do badania włączono 123 chorych z rozpoznaniem dystonii ogniskowej lub segmentalnej oraz 51 osób zdrowych stanowiących grupę kontrolną. U każdego badanego obecność drżenia kończyn górnych oceniano klinicznie i aparaturowo (akcelerometr, elektromiografia i tablet graficzny). Częstotliwość i nasilenie drżenia badano w trzech pozycjach: w spoczynku (drżenie spoczynkowe), w kończynach górnych wyciągniętych do przodu (drżenie pozycyjne) oraz podczas próby „palec–nos" i rysowania spirali Archimedesa (drżenie kinetyczne). Na podstawie testu obciążenia określano także rodzaj drżenia: typu drżenia samoistnego lub nasilonego drżenia fizjologicznego.

Wyniki: Drżenie u chorych z dystonią występowało znacznie częściej niż w grupie kontrolnej ($p = 0{,}0001$). W badaniu klinicznym stwierdzano je u ok. 50% chorych, a w badaniu aparaturowym u ok. 10–20% więcej. Najczęściej było to drżenie pozycyjne i kinetyczne o częstotliwości ok. 7 Hz, typu drżenia samoistnego. W grupie kontrolnej drżenie wykryto jedynie metodami aparaturowymi u ok. 10% badanych i było to drżenie pozycyjne o częstotliwości 9 Hz, typu nasilonego drżenia fizjologicznego. Nie stwierdzono różnic pomiędzy występowaniem drżenia i jego parametrami (częstością i nasileniem) u chorych z poszczególnymi rodzajami dystonii ani też zależności między nasileniem drżenia a nasileniem objawów dystonii.

Słowa kluczowe: dystonia ogniskowa, drżenie towarzyszące, parametry drżenia.

Correspondence address: dr n. med. Magdalena Wójcik-Pędziwiatr, Department of Neurology, Jagiellonian University Medical College, 3 Botaniczna St, 31-503 Kraków, Poland, e-mail: stokrotka283@tlen.pl
Received: 13.05.2012; accepted: 2.11.2012

Introduction

Tremor in patients with focal or segmental dystonia is not uncommonly seen in clinical practice but the results of the studies aimed at the assessment of tremor in these types of dystonia are very divergent as they report presence of tremor in 10-85% of the studied population [1-7]. Two types of such tremor could be discerned, i.e. dystonic tremor (DT), which is observed in the part of the body affected with dystonia, and tremor associated with dystonia (TAD), which occurs in the part of the body not involved in dystonic posturing or dystonic movement.

Dystonic tremor is mostly postural and kinetic, featuring irregular amplitude and variable frequency, usually about 7 Hz [6-10]. Typical examples of DT include head tremor in torticollis or upper limb tremor in task-specific dystonia, e.g. writer's cramp. Dystonic tremor may precede the fully blown clinical picture of dystonia for several years or may accompany dystonic movements for all years of the disease.

Tremor associated with dystonia could be exemplified by the hand tremor in torticollis. According to the reports published so far, that type of tremor may have features typical for enhanced physiological tremor (EPT) [11] or even fulfil the diagnostic criteria of essential tremor (ET) [4,5]. In many cases, tremor does not fulfil all ET criteria and is then called ET-like tremor [1,3]. Coexistence of dystonia, especially focal dystonia, and ET is known but estimates of its frequency vary. According to the available literature, ET was diagnosed in 3-23% of patients with dystonia, and dystonic features were diagnosed in 0.6-47% of patients with ET [12-17].

The aim of this study was to assess incidence and to characterize parameters of tremor accompanying focal and segmental dystonia.

Material and methods

Participation in this study was offered to all patients diagnosed with focal or segmental dystonia who were treated either in an out-patient clinic or in the Department of Neurology, University Hospital of Krakow between 2005 and 2009 as well as to the sex- and age-matched healthy persons who accompanied or visited patients in that department.

Inclusion criteria consisted of: age between 18 and 85 and the diagnosis of dystonia according to Fahn criteria [18].

Exclusion criteria comprised: (1) alcohol abuse, either current or in the past; (2) diagnosis of concurrent schizophrenia, depression, dementia or other severe psychiatric disturbances; (3) presence of uncontrolled hyperthyroidism; (4) presence of coexistent serious systemic and metabolic disorders leading to the insufficiency of related organs; (5) history of intoxication with medications or heavy metals, and exposure to toxins; (6) presence of neoplastic or autoimmune diseases; (7) intake of medications possibly related to tremor, such as neuroleptics, glucocorticoids or β-mimetics; (8) history of stroke; (9) Parkinson disease and other neurological disorders that may feature tremor; (10) severe vascular lesions in central nervous system (CNS); (11) post-traumatic lesions or other kinds of lesions that may preclude testing of tremor; (12) acute infectious illness.

The exclusion criterion for controls was a history of any neurological disorder or its signs found in neurological examination.

The protocol of the study was approved by the local Bioethical Committee and all subjects included in the study provided their informed consent to participate.

In all patients and controls, the evaluation included interview, physical neurological examination and the studies of tremor. The interview included demographic data (age and education), previous and present illnesses (including diabetes, arterial hypertension, ischaemic heart disease, hypercholesterolaemia, disease of the liver or kidneys, and malignancies, among others), medications used and their adverse effects, exposure to toxins, cigarette smoking, and genetic predispositions, i.e. family history of degenerative disorders and occurrence of tremor. Patients were subjected to an additional detailed interview related to the data on the underlying disorder, including: age at onset, course of the disease, localization of symptoms – those were the basis for the determination of the form of dystonia (e.g. torticollis, blepharospasm).

In addition to the standard neurological examination, the severity of dystonia was assessed with appropriate scales: (1) The Toronto Western Spasmodic Torticollis Rating Scale (TWSTRS) for the assessment of the severity and functional disturbances caused by torticollis; (2) Jankovic Rating Scale for the assessment of the severity of blepharospasm; (3) Function Scale for Patients with Blepharospasm for the assessment of the level of functional disturbances resulting from blepharospasm; and (4) Burke-Fahn-Marsden Evaluation Scale for Dystonia for the assessment of the severity of dystonias other than blepharospasm or torticollis.

Clinical and instrumental assessment of upper limb tremor was performed consecutively in both hands with the use of instruments (accelerometer, electromyography [EMG], graphic tablet) and appropriate software analysing tremor in the Tremor Assessment Laboratory of the Department of Neurology, Jagiellonian University College of Medicine in Krakow.

Clinical evaluation of tremor severity was made using the Simple Tremor Severity Scale (0-4 pts).

The presence and severity of rest tremor was assessed with the observation of hands placed comfortably on thighs with the subject in a sitting position; postural tremor was assessed in the upper limb outstretched in front of the subject and the kinetic tremor was evaluated during the 'finger-nose' manoeuvre and during the drawing of an Archimedes spiral. The frequency of tremor (in Hz) was assessed according to the results of accelerometric testing, and the severity of tremor was evaluated according to the recordings made with a graphic tablet, as the tremor severity index (0-10 scale). Based on the study with an accelerometer, three types of tremor were also discerned: EPT, ET-type, and tremor of another type. Enhanced physiological tremor and ET-type tremor were diagnosed if the tremor frequency spectrum had apparent domination of one peak only. In case of a greater number of peaks, or if the predominant peak could not be identified, tremor of another type was diagnosed. Enhanced physiological tremor and ET-type of tremor were differentiated according to the assessment of the tremor frequency with the accelerometer and simultaneous EMG recording in the outstretched hand before and after the load with a 500-g weight mounted at the level of the wrist. Enhanced physiological tremor was diagnosed if the frequency of tremor in the loaded hand decreased by more than 1 Hz, and ET-type of tremor was diagnosed if no difference in frequency was noted or if the decrease in tremor frequency was smaller than 1 Hz [19].

Synchronicity of tremor was analysed with the observation of 10-s EMG registration from agonist and antagonist muscles of the forearm. Asynchronicity of agonist and antagonist muscle contraction responsible for the tremor of the hand was diagnosed in case of a marked shift of the bioelectrical activity registered in agonist muscles in relation to the same bioelectric activity of the antagonist muscles in more than 90% of recorded cycles of bioelectric activity of the studied muscles.

Analysis of tremor with the graphic tablet was performed during Archimedes spiral drawing with an electronic pen according to the pattern provided on the tablet. Lateral deviations from the spiral line in X and Y axis, as well as the pressure of the pen (Z axis) were registered and analysed. Presence of upper limb tremor was established according to the presence of the characteristic peak dominant frequency of the spectrum. This method enables the calculation of the dominant frequency of tremor, amplitude of tremor and the coefficient of tremor intensity; it also provides a computerized assessment of tremor severity on a 0-10 scale, where 10 indicates the greatest severity of tremor [20].

Qualitative tremor analysis with the accelerometer enables the assessment of tremor in three-dimensional space. This part of the study was performed using a three-axial accelerometer (BIOPACK). The accelerometer was mounted on the proximal phalanx of the third finger with a Velcro band. Registration of tremor took 1.5 minutes. Presence of tremor was established according to the occurrence of the characteristic peak dominant frequency of the spectrum. This method was used to assess frequency and severity of tremor.

Electromyography testing used superficial electrodes mounted in standard leads over wrist flexors and extensors (extensor carpi radialis brevis muscle and flexor carpi radialis muscle) [21]. Registration of tremor took 1.5 minutes. The use of superficial EMG enabled the assessment of synchronicity of the contractions within two antagonist muscle groups, i.e. wrist flexors and extensors. Visual assessment of 10-second EMG recording was done independently by two investigators.

Among patients with writer's cramp, tremor evaluation was performed both in the limb affected with dystonia (writer's tremor) and in the contralateral limb (TAD).

All patients included in the study who were diagnosed with blepharospasm, Meige syndrome or torticollis were treated with botulinum toxin type A (BTX-A, Botox) at least for one year before this study. Doses of 30-60 U were used to treat patients with blepharospasm or Meige syndrome; doses of 100-200 U were used to treat patients with torticollis, according to the number and size of affected muscles and according to the severity of dystonia. Injections were repeated after the disappearance of the clinical improvement related to the previous injections, not more often than every 12 weeks. All patients included in the study benefited from the treatment with BTX-A. Tremor registration was performed after at least 12 weeks following the last BTX-A injection, directly before the injection of another dose of the medication. Patients with writer's cramp or with lower limb dystonia were not treated with BTX-A. Patients

did not use any other treatment for dystonia, e.g. anticholinergics (biperiden), baclofen, benzodiazepines, or levodopa preparations. No surgery was performed to treat dystonia. Patients did not use any medication to treat tremor.

Laboratory tests were performed in all patients to exclude other causes of tremor; they included complete blood count and blood biochemistry, thyroid gland hormones (TSH, T3, T4) and ceruloplasmin concentration.

Statistical methods

Results are provided as means and standard deviations for the given variable. The results were entered into the database, and their analysis used the statistical package Statistica. Student *t*-test was used for parametric variables, and χ^2 test was used to test categorical variables. *P*-value < 0.05 was considered statistically significant.

Results

The study involved 123 patients with dystonia, including 76 women and 47 men, aged between 22 and 85 (mean age 54 ± 16 years), with the disease duration ranging from 1 to 45 years (mean 10 ± 8 years). Cervical dystonia was diagnosed in 52 patients (42.3%), blepharospasm was found in 41 patients (33.3%), upper limb dystonia (mainly writer's cramp) was diagnosed in 14 patients (11.4%), lower limb dystonia (genetically confirmed Segawa dystonia) was found in 5 patients (4.0%), and Meige syndrome was diagnosed in 11 patients (9.0%). For the purpose of statistical analyses, patients with Meige syndrome were included in the group of patients with blepharospasm because of the dominant blepharospasm-related signs. Severity of particular movement disorders, assessed with appropriate scales, is shown in Table 1.

The following comorbidities were found among the studied patients: arterial hypertension – 37 (30.1%), ischaemic heart disease – 20 (16.3%), diabetes mellitus – 3 (2.4%), arrhythmia – 4 (3.2%), hypercholesterolaemia – 2 (1.6%), hyperthyroidism in euthyroid subjects at the time of study – 2 (1.6%), Crohn disease – 1 (0.8%), osteoporosis – 1 (0.8%), migraine – 1 (0.8%), peptic ulcer of the stomach – 1 (0.8%), hypothyroidism in euthyroid subjects at the time of study – 1 (0.8%), epilepsy – 1 (0.8%), congenital valvular heart disease – 1 (0.8%), Barlow syndrome – 1 (0.8%), type B viral

Table 1. Results of scales used to assess clinical severity of primary disease in patients with dystonia

Scales (range)		Severity	
		Mean	Range
The Toronto Western Spasmodic Torticollis Rating Scale	Severity (0-35)	17.7	8-26
	Pain (0-20)	12.3	6-17
	Disability (0-32)	15.6	6-25
Jankovic Rating Scale	Severity (0-4)	3.0	1-4
	Frequency (0-4)	2.7	1-4
Function Scale for Patients with Blepharospasm (0-24)		11	6-20
Burke-Fahn-Marsden Evaluation Scale for Dystonia (0-120)		10.8	4-45

Table 2. Prevalence of upper limb tremor according to the clinical and instrumental assessment among studied patients with dystonia

Method of assessment	Diagnosis					
	All patients	Torticollis	Blepharospasm	Meige syndrome	Upper limb dystonia	Lower limb dystonia
Clinical	60 (48.8%)	25 (48.1%)	24 (58.5%)	6 (54.5%)	8 (57.1%)	2 (40.0%)
Accelerometric	80 (65.0%)	44 (84.6 %)	26 (63.4%)	7 (63.6%)	11 (78.6%)	3 (60%)
Graphic tablet	69 (56.1%)	33 (63.5%)	25 (60.9%)	7 (63.6%)	10 (71.4%)	2 (40%)

hepatitis – 1 (0.8%), and type C viral hepatitis – 1 (0.8%). Above-mentioned disorders do not affect the development of dystonia or tremor.

According to the clinical assessment (with neurological examination), tremor was present in 60 patients (48.8%) with various clinical forms of dystonia. Tremor was present in similar percentages of patients with all forms of dystonia (see Table 2 for details). Postural tremor was noted in 41 (33.3%) patients, kinetic tremor in 13 (10.6%) patients, and resting tremor in 6 patients (4.9%). Six patients had both postural and kinetic tremor; four patients had both postural and resting tremor (Fig. 1).

The severity of tremor, assessed clinically with the 4-point Simple Tremor Severity Scale, was as follows: mild tremor (1 point) in 44 (35.8%) patients, moderate tremor (2 points) in 12 (9.8%) patients, and severe tremor (3 points) in 4 (3.3%) patients.

Instrumental assessment of tremor in patients with various clinical forms of dystonia revealed presence of tremor in 80 patients (65.0%) according to the accelerometric evaluation and in 69 patients (56.1%) in patients according to the evaluation with the graphic tablet. Tremor was present in similar proportion in all forms of dystonia (see Table 2). Among patients with writer's cramp, writer's tremor was found clinically in 7 (50.0%) patients, while tremor accompanying dystonia in contralateral limb was noted in 1 patient (7.1%). In all those patients, presence of tremor was confirmed with instrumental evaluation (EMG, accelerometer, graphic tablet); additionally, tremor accompanying dystonia was recorded in 3 patients (21.4%) in the limb not affected with writer's tremor.

The control group consisted of 51 healthy volunteers (including 19 women and 32 men) aged between 21 and 79 (mean age 55 ± 17); no visible upper limb tremor was found in any of those subjects in neurological examination. Instrumental assessment of tremor among controls revealed presence of tremor in 5 subjects (9.8%) according to accelerometric evaluation and in 4 (7.8%) subjects evaluated with the graphic tablet. Tremor was significantly more common in patients than in controls ($p = 0.0001$).

Three types of upper limb tremor were identified accelerometrically among patients: postural tremor in 50 (40.7%) patients, kinetic tremor in 19 (15.5%) patients, and resting tremor in 11 (8.9%) patients. Among controls, 4 subjects (7.8%) had postural tremor and 1 person (1.9%) had kinetic tremor; resting tremor was not found in controls. Postural tremor and kinetic tremor

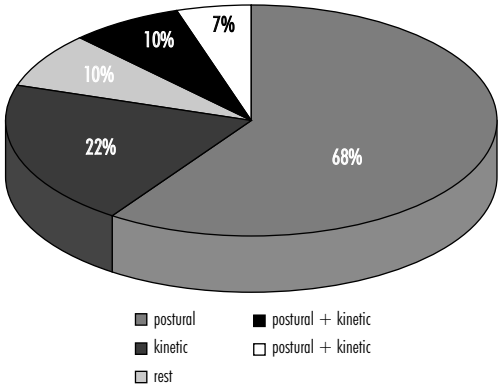

Fig. 1. Proportions of various types of tremor among patients with dystonia

were significantly more common among patients with dystonia than in controls ($p = 0.002$ and $p = 0.04$, respectively).

Essential tremor-like type of tremor was the most common one (43 patients, 35.0%), while increased physiological tremor was rare (5 patients, 4.1%). It was impossible to categorize tremor into one of the above-mentioned types in 32 patients (26.0%) (tremor of other type was diagnosed). Enhanced physiological tremor was diagnosed in 4 controls (7.8%), and in one other person (1.9%) ET-like tremor was diagnosed. Essential tremor-like type of tremor was significantly more common when compared with the control group ($p = 0.0004$).

Tremor due to asynchronous type of contractions of agonist/antagonist muscles was predominant in patients (89.7%). Tremor frequency in those patients ranged from 2.7 to 14.4 Hz (mean 7.7 Hz). The prevalence of upper limb postural tremor with different frequency range in accelerometric assessment is shown in Figure 2. No difference in prevalence or severity of tremor among patients with different forms of dystonia was found ($p > 0.05$).

Severity of TAD, as assessed with the graphic tablet, was mild (range: 1-7, mean: 2.5 [on 1-10 scale]). The distribution of coefficients of dystonia severity is shown in Figure 3. Severity of dystonia on appropriate scales and severity of dysfunction or impairment related to dystonia did not correlate with the severity of tremor ($p > 0.05$).

In controls, mean frequency of tremor was 9.3 Hz (range: 5.8-12.6 Hz), and the mean coefficient of tremor severity, as assessed with the graphic tablet, was 2.0 (range: 1-3). In all studied controls, tremor due to asyn-

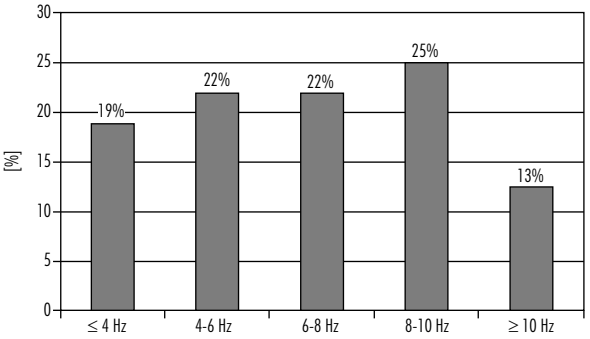

Fig. 2. Prevalence of upper limb postural tremor with different frequency range in accelerometric assessment

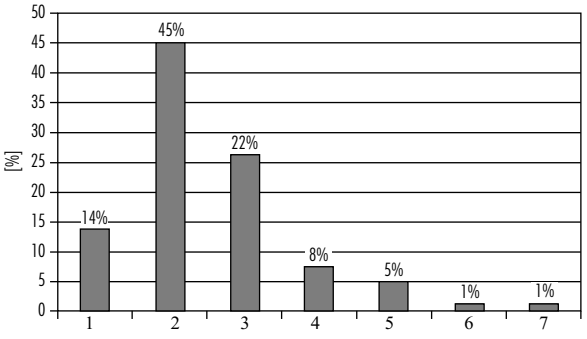

Fig. 3. Distribution of coefficients of dystonia severity

chronous contractions of agonist/antagonist muscles was found.

Discussion

In this study, tremor was a common sign accompanying focal and segmental dystonia. In clinical assessment, tremor was visible in about 50% of patients, and instrumental evaluation revealed its presence in about 60% of patients, i.e. in a much greater proportion than among control subjects. The obtained results are within the broad range (10-85%) of prevalence of upper limb tremor, as reported in various studies among patients with dystonia [1-7]. Our findings are similar to those reported by specialized centres, such as Baylor College of Medicine Movement Disorders Clinic (71% of patients) [4], Christian Albrechts University in Kiel (60%) [11] or the Neurodegenerative Disorders Centre in Vancouver (40% of patients) [22]. In regional centres, however, the proportion of patients with TAD was 22% [23]. Our patients were recruited from a specialized outpatient clinic for extrapyramidal disorders which gathers patients with a complex clinical picture of the disease (e.g. associated with tremor) or with atypical course of the disease.

Severity of tremor in the studied patients with dystonia was low – in almost one half of the patients it was graded as 2 points on a 10-point scale. Similar low severity of TAD was reported by Deuschl *et al*. [11]. The severity of tremor did not correlate with the severity of dystonia, level of impairment due to dystonia or with the duration of dystonia. Munchau *et al*. [24] in a study that analysed clinical relationships between tremor and dystonia presented similar findings, i.e. severity of tremor in upper limbs did not correlate with severity of dystonia, and duration of tremor did not correlate with duration of dystonia.

In our study, patients with dystonia most often presented postural tremor with moderate frequency of 7.7 Hz and with dominant (90%) asynchronous type of contractions of agonist and antagonist muscles. This is in agreement with the data found in the literature that suggest a moderate range of frequency, about 7-8 Hz, as typical for postural tremor accompanying other movement disorders, including dystonia [3,4,11].

Postural tremor, found clinically or instrumentally (33.3% and 40.7% of patients, respectively), was significantly more common among patients with dystonia than in controls ($p = 0.002$). Kinetic tremor was three times less frequent clinically or instrumentally (10.6% and 15.5%, respectively); resting tremor was the rarest type of tremor (4.9% vs. 8.9%, respectively). Essential tremor-like tremor was found in 54% of patients; EPT was found in 6% of patients only. Among the other 40% of patients, tremor could not be categorized as any of the above-mentioned types. Available literature contains few papers reporting evaluation of the presence of particular types of tremor in dystonia according to the specific analyses, for example using a load test [11,24]. Authors reporting on that topic only stated that tremor could be EPT [11], ET [1,3], dystonic or non-specific tremor [4,11,16,24].

Presence of postural tremor in dystonia is explained with the common coexistence of ET, the most common extrapyramidal disorder, found in 5% of the population. Essential tremor was diagnosed in 3-23% of patients

with dystonia [4,5]. On the other hand, numerous publications confirm the presence of dystonia, especially focal dystonia, in 0.6-30% of patients diagnosed with ET [12-17]. Lou and Jankovic [15] found the coexistence of dystonia in 47.0% of patients out of 350 patients with ET. Baxter and Lal [12] studied 100 patients with ET and found dystonia in only 12 of them. In patients with dystonia, in whom postural tremor of ET-like type is observed commonly also in body parts not affected with dystonia, the prevalence of family history of postural tremor is higher than expected [3,4,11]. Focal dystonia or tremor exists in 26-52% of first-degree relatives of patients with focal dystonia [2-4,25].

Greater prevalence of tremor in dystonia might suggest a pathogenetic association between those two movement disorders. An association between ET and dystonia or parkinsonism could result from common genetic susceptibility. Some members of the families with genetically transmitted generalized dystonia related to the DYT1 mutation present with isolated tremor, so-called tremor associated with dystonia gene, which may be an abortive form of dystonia [26]. Studies by Conway *et al*. [27] using genetic linkage analysis excluded presence of DYT1 mutation on chromosome 1 in familial form of ET. This suggests that the genes responsible for those two clinical entities have separate loci; the association between those two entities is pathophysiological rather than genetic [28].

Many authors stress the clinical difference between typical ET and upper limb tremor seen in patients with dystonia. According to their observations, tremor in patients with dystonia is more often irregular and asymmetric, and may resemble myoclonus [29,30], while ET is usually regular and symmetric [31,32]. It is not known, however, if those differences are due to separate pathomechanisms of those two types of tremor or if they result from the impact of dystonia on the different clinical forms of accompanying tremor. In our study, 40% of patients assessed instrumentally had so-called 'other type of tremor' which was characterized by irregular amplitude and variable frequency. These are typical features of dystonic tremor described by Jankovic and Tolosa [33], Deuschl [8], as well as by Findley and Koller [34]. It might be therefore an abortive form of dystonia clinically invisible as dystonic movement or dystonic tremor. It is further supported by the fact that upper limb tremor observed in patients with upper limb dystonia, e.g. with writer's cramp, was most commonly identified, both clinically (57.1%) and instrumentally, with the accelerometer (78.6%) and with the graphic tablet (71.4%).

The type of postural tremor observed in dystonia has been examined in a few studies, and their results are equivocal. Jedynak [30] showed that the tremor accompanying dystonia has clinical and electromyographic features that differentiate it from ET. Yanagisawa and Goto [35], on the other hand, described two types of tremor in dystonia according to the findings in EMG with superficial electrodes – rhythmic tremor with the frequency of 5-11 Hz, similar to ET, and non-rhythmic tremor with the frequency of 1-6.5 Hz, that did not fulfil criteria for ET. Many investigators believe that the upper limb tremor observed in dystonia differs substantially from ET, which proves different origin and pathomechanism of that tremor. An electrophysiological study by Münchau *et al*. [24] supports that view. The authors used electrophysiological studies to compare 11 patients diagnosed with ET with 19 patients with dystonia accompanied with upper limb tremor. They found that latency of impulses registered from antagonist muscle during ballistic wrist flexion movements was longer among patients with ET than in patients with cervical dystonia accompanied with upper limb tremor. Moreover, patients with cervical dystonia accompanied with upper limb tremor exhibited greater variability of presynaptic inhibition between agonist and antagonist forearm muscle than patients with ET, as evidenced by EMG findings. Similar abnormalities in reciprocal inhibition were observed in the unaffected upper limb of patients with writer's cramp [37] and in the unaffected upper limb in patients with torticollis [37]; those abnormalities were absent, however, in patients with ET in upper limbs [38].

The pathomechanism of dystonic tremor differs from that involved in TAD. It is believed that the pathomechanism of dystonic tremor is similar to that of dystonia itself – it results from the impaired mechanisms of inhibition at the cortical, subcortical and spinal level, as well as from the impaired sensory perception and integration and from the mechanism of sensorimotor integration. According to Deuschl [39], dysfunction involves pallido-thalamic connections and leads to the secondary disinhibition of connections within the thalamus and premotor cortex. The aetiopathogenesis of TAD is more complex and differs according to the type of the tremor, i.e. EPT, coexistent ET or ET-like tremor. The pathomechanism of TAD, especially of the ET-like type, is poorly understood and certainly warrants further studies.

Conclusions

1. Tremor as an accompanying sign is much more common in patients with dystonia than in control subjects. This tremor has relatively low severity; postural ET-like tremor is the most prevalent one.
2. Instrumental assessment of tremor enables the registration of tremor in an additional 10-20% of patients, in whom tremor is clinically invisible.

Disclosure

Authors report no conflict of interest.

References

1. Couch J.R. Dystonia and tremor in spasmodic torticolis. In: Eldridge R., Fahn S. [eds.]. Advances in neurology. *Raven Press*, New York 1976; 14: 245-258.
2. Duane D.D. Spasmodic torticollis. *Adv Neurol* 1988; 49: 135-150.
3. Chan J., Brin M.P., Fahn S. Idiopathic cervical dystonia: clinical characteristics. *Mov Discord* 1991; 6: 119-126.
4. Jankovic J., Leder S., Warner D., et al. Cervical dystonia: clinical findings and associated movement disorders. *Neurology* 1991; 41: 1088-1091.
5. ubinsky R.M., Gray C.S., Koller W.C. Essential tremor and dystonia. *Neurology* 1993; 43: 2382-2384.
6. Dubinsky R.M. Tremor and dystonia. In: Findley L.J., Koller W.C. [eds.]. Handbook of tremor disorders. *Marcel Dekker*, New York 1995, pp. 405-410.
7. Manyam B. Uncommon forms of tremor. In: Watts R.L., Koller W.C. [eds.]. Movement disorders. Neurologic principles and practice. 2nd ed. *McGraw-Hill*, New York 2004, pp. 459-480.
8. Deuschl G. Clinical and neurophysiological differential diagnosis of tremor. *J Clin Neurophysiol* 1996; 2: 110-121.
9. Deuschl G., Bain P., Brin M. Consensus statement of the Movement Disorder Society on Tremor. *Mov Disord* 1998; 13 (Suppl 3): 2-23.
10. Deuschl G., Volkmann J., Raethjen J. Tremor: differential diagnosis, phathophysiology, and therapy. In: Jankovic J., Tolosa E. [eds.]. Parkinson's disease and movement disorders. 5th ed. *Lippincott Williams & Wilkins*, Philadelphia 2007, pp. 298-320.
11. Deuschl G., Heinen F., Guschlbauer B., et al. Hand tremor in patients with spasmodic torticollis. *Mov Disord* 1997; 12: 547-552.
12. Baxter D.W., Lal S. Essential tremor and dystonic syndromes. *Adv Neurol* 1979; 24: 373-377.
13. Martinelli P., Gabellini A.S. Essential tremor and buccolinguofacial dyskinesias. *Acta Neurol Scand* 1982; 66: 705-708.
14. Rajput A.H., Offord K.P., Beard C.M., et al. Essential tremor in Rochester, Minnesota: a 45-year study. *J Neurol Neurosurg Psychiatry* 1984; 47: 466-470.
15. Lou J.J., Jankovic J.J. Essential tremor: clinical correlates in 350 patients. *Neurology* 1991; 41: 234-238.
16. Koller W.C., Busenbark K., Miner K., et al. The relationship of essential tremor to other movement disorders: report on 678 patients. *Ann Neurol* 1994; 35: 717-723.
17. Tallon-Barranco A., Vazquez A., Jimenez-Jimenez F., et al. Clinical features of essential tremor seen in neurology practice: a study of 357 patients. *Parkinsonism Relat Disord* 1997; 3: 187-190.
18. Fahn S., Bressman S.B., Marsden C.D. Classification of dystonia. *Adv Neurol* 1998; 78: 1-10.
19. Elble R.J. Mechanism of physiological tremor and relationship to essential tremor. Koller W., Findley L. [eds.]. Handbook of tremor disorders. *Marcel Dekker Inc.*, New York 1995, pp. 51-62
20. Rudzińska M., Izworski A., Banaszkiewicz K., et al. Quantitative tremor analysis with a graphic digitizing tablet. *Neurol Neuroch Pol* 2007; 41: 510-516.
21. Timmer J., Lauk M., Deuschl G. Quantitative analysis of tremor time series. *Electroencephalogr Clin Neurophysiol* 1996; 101: 461-468.
22. Pal P.K., Samii A., Schulzer M., et al. Head tremor in cervical dystonia. *Can J Neurol Sci* 2000; 27: 137-142.
23. Shukla G., Behari M. A clinical study of non-parkinsonian and non-cerebellar tremor at a specialty movement disorders clinic. *Neurology* 2004; 52: 200-202.
24. Münchau A., Schrag A., Chuang C., et al. Arm tremor in cervical dystonia differs from essential tremor and can be classified by onset age and spread of symptoms. *Brain* 2001; 124: 1765-1776.
25. Rondot P., Marchand M.P., Dellatolas G. Spasmodic torticollis – review of 220 patients. *Can J Neurol Sci* 1991; 18: 143-151.
26. Eidelberg D. Metabolic brain networks in idiopathic torsion dystonia. In: Fahn S., De Long M., Marsden C.D. [eds.]. Dystonia 3. Advances in neurology. *Lippincott-Raven*, Philadelphia 1998.
27. Conway D., Bain P.G., Warner T.T., et al. Linkage analysis with chromosome 9 marker in hereditary essential tremor. *Mov Disord* 1993; 8: 374-376.
28. Jankovic J., Beach J., Schwarz K., et al. Tremor and longevity in relatives of patients with Parkinson's disease, essential tremor and control subject. *Neurology* 1995; 45: 645-648.
29. Rivest J., Marsden C.D. Trunk and head tremor as isolated manifestation of dystonia. *Mov Disord* 1990; 5: 60-65.
30. Jedynak C.P., Bonnet A.M., Agid Y. Tremor and idiopathic dystonia. *Mov Disord* 1991; 6: 230-236.
31. Machowska-Majchrzak A., Pierzchała K., Pietraszek S., et al. Przydatność rejestracji akcelerometrycznej z oceną parametrów drżenia i ich symetrii w diagnostyce różnicowej drżenia parkinsonowskiego, samoistnego i móżdżkowego. *Neurol Neurochir Pol* 2012; 46: 145-156.
32. Machowska-Majchrzak A., Pierzchała K., Pietraszek S., et al. Drżenie samoistne – ocena symetrii wskaźników akcelerometrycznych i związku dominacji ręki z nasileniem drżenia. *Neurol Neuroch Pol* 2011; 45: 121-127.
33. Jankovic J., Tolosa E. Parkinson's disease and movement disorders. 3rd ed. *Williams and Wilkins*, Baltimore 1998.
34. Findley L.J., Koller W.C. Definition and behavioral classification. In: Findley L.J., Koller W.C. [eds.]. Handbook of tremor disorders. *Marcel Dekker*, New York 1995, pp. 1-5.

35. Yanagisawa N., Goto A. Dystonia musculorum deformans: analysis with electromyography. *J Neurol Sci* 1971; 13: 39-65.
36. Chen R.S., Tsai C.H., Lu C.S. Reciprocal inhibition in writer's cramp. *Mov Disord* 1995; 10: 556-561.
37. Deuschl G., Heinen F., Kleedorfer B., et al. Clinical and polymyographic investigation of spasmodic torticollis. *J Neurol* 1992; 239: 9-15.
38. Rothwell J. Pathophysiology of essential tremor. In: Findley L.J., Koller W.C. [eds.]. Handbook of tremor disorders. *Marcel Dekker*, New York 1995, pp. 185-193.
39. Deuschl G. Dystonic tremor. *Rev Neurol* 2003; 10: 900-905.