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Head Tremor in Essential Tremor: "Yes-Yes", "No-No", or "Round and Round"?

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Abstract

Introduction—Essential tremor (ET) is a common yet frequently misdiagnosed movement disorder. One contributing factor may be the dearth of studies that focus on the nuances of clinical phenomenology. A clinical feature that has received relatively little attention is head tremor. Indeed, there is no consensus regarding the predominant direction of head tremor in ET, and no study has examined the clinical correlates of directionality.

Methods—We identified 51 ET cases with head tremor enrolled in a clinical-epidemiological study of ET at Columbia University. Each had a videotaped neurological examination. Videotapes were viewed and coded by a movement disorders neurologist for head tremor direction ("no-no", "yes-yes", or mixed) and continuity (continuous, intermittent, or rare). Direction was correlated with a wide range of clinical features.

Results—Fourteen cases (27.5%) had "no-no" tremor, 9 (17.6%) had "yes-yes" tremor, and 28 (54.9%) had a mixed tremor. Mixed and "yes-yes" cases were older ($p = 0.004$) and had a longer tremor duration ($p = 0.018$) than "no-no" cases. Tremor severity (arms) was higher for mixed cases than for "yes-yes" and "no-no" cases ($p = 0.04$). More mixed cases had continuously present tremor while more "no-no" cases had rare head tremor ($p<0.001$).

Conclusions—Head tremor in ET seems to start as an infrequent tremor in one direction (esp. "no-no") and becomes more frequent while acquiring additional directionality and a mixed phenotype as the disease progresses. These findings add to our understanding of the clinical spectrum of ET.

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Keywords

tremor; essential tremor; clinical characteristics; neurological examination

Introduction

Essential tremor (ET) is one of the most common movement disorders; however, according to some estimates, it is also misdiagnosed as much as 30 – 50% of the time [1]. One contributing factor is the surprising dearth of studies that focus on the nuances of clinical phenomenology.

Head tremor affects many ET patients, and there is a female preponderance [2]. Even though directionality (i.e., horizontal vs. vertical) was observed in early descriptions of ET head tremor, there are few detailed studies of and no consensus regarding the predominant direction of head tremor in ET. Furthermore, the existing reports addressing head tremor direction must be interpreted within the context of several limitations, including small sample sizes (e.g., n = 5, 16, 23), restriction of analyses to familial cases, or the fact that they were conducted prior to modern diagnostic distinctions between ET and dystonic head tremor [3–7]. The most complete description characterized head tremor in 16 familial ET cases; of these, 12 (75.0%) had “no-no” (i.e., horizontal) tremor, 1 (6.25%) had “yes-yes” (i.e., vertical) tremor, and 3 (18.75%) had mixed tremor (i.e., more than one direction) [7]. In another series of patients with mixed movement disorders (23 ET, 7 cervical dystonia), 13 (56.5%) of 23 ET cases had “no-no” tremor, 4 (17.4%) of 23 ET cases had “yes-yes” tremor, and 6 (26.1%) of 23 ET cases had mixed tremor [5].

In this study, we characterize the direction of head tremor and examine the clinical correlates of directionality in a large series of 51 patients with ET who had head tremor. As the largest study to date to determine head tremor directionality and the first to correlate clinical features with tremor direction, we hope to add to our clinical understanding of this very common neurological disease.

Methods

Description of Study

Participants were prospectively enrolled in a clinical-epidemiological study of ET, (2009 – 2014) [8]. ET cases were identified from a computerized billing database at the Center for Parkinson’s Disease and Other Movement Disorders, Neurological Institute, Columbia University Medical Center (CUMC), with a search conducted of all patients seen within the three years prior to enrollment. Each of the enrolled ET cases had received a diagnosis of ET from their treating neurologist at the Institute and lived within two hours driving distance of CUMC. One of the authors (E.D.L.) reviewed the office records of all selected cases; those with diagnoses or physical signs consistent with other movement disorders were excluded. The CUMC Internal Review Board approved of all study procedures. Written informed consent was obtained upon enrollment. Analysis of data was also approved by the Internal Review Board at Yale Medical School.

Evaluation and Diagnostic Confirmation

A trained research assistant conducted an in-person evaluation, administering demographic and medical history questionnaires. During the in-person assessment, a videotaped neurological examination was performed. This included one test for postural tremor and five for kinetic tremor (12 tests total) as well as assessments of speech and voice (sustained phonation). A neurologist specializing in movement disorders (E.D.L.) used a reliable and valid clinical rating scale, the Washington Heights-Inwood Genetic Study of ET (WHIGET) tremor rating scale, to rate postural and kinetic tremor during each test: 0 (none), 1 (mild), 2 (moderate), 3 (severe), resulting in a total tremor score (range = 0 – 36) [9]. Diagnoses of ET were re-confirmed by E.D.L. using the videotaped neurological examination and WHIGET diagnostic criteria (moderate or greater amplitude kinetic tremor [tremor rating ≥ 2] during three or more tests or a head tremor, in the absence of Parkinson's disease, dystonia or another cause) [9].

Final Sample with Head Tremor and Re-review of Videotaped Examination

Among the 141 enrollees, we identified 51 (36.2%) with head tremor on videotaped neurological examination. These 51 were selected for additional analyses. The videotaped neurological examination was re-reviewed by a movement disorders neurologist (D.R.) who coded clinical features. At the start of the re-review, a 25% sample of these ET cases was co-reviewed with the senior movement disorders neurologist (E.D.L.) to ensure agreement.

During the videotaped neurological examination, the patient's head was visible during the following segments: 1) sitting at rest with arms in lap, 2) sitting with arms outstretched, 3) pouring water from one cup into another, 4) bringing a spoon to the mouth, 5) finger-to-nose testing, 6) speech and sustained vocalization, 7) rapid alternating movements, 8) walking normally and tandem gait, 9) writing, and 10) reading a passage out loud.

Head tremor was coded as horizontal ("no-no"), vertical ("yes-yes"), or mixed. The mixed category included (1) tremor that was sometimes "yes-yes" and sometimes "no-no" and (2) tremor in an axis other than horizontal or vertical (e.g., diagonal). In the former, a certain percentage of head tremors were predominantly "yes-yes" or predominantly "no-no" (i.e., a direction that was present for more than half the time the tremor was visualized), and this was also noted. Tremor was also categorized according to constancy and was labeled as continuously present (i.e., present whenever the head was visible during the videotaped neurological examination), intermittently present, or rarely present (i.e., present during few segments of the videotape).

Data Analyses

Data were analyzed in SPSS (Version 21.0), comparing groups using Student t tests or chi-square tests. For some analyses (e.g., age category, tremor duration, total tremor score), data were stratified into categories.

Results

Fifty-one ET cases had head tremor (Table 1). The group included a preponderance of women (68.6%, Table 1). Head tremor was continuous in 39.2% and intermittent or rare in the remainder (Table 1).

In terms of directionality, 14 cases (27.5%) had “no-no” head tremor, 9 (17.6%) had “yes-yes” head tremor, and 28 (54.9%) were of the mixed type. The mixed types were as follows: 6 of 28 were predominantly “no-no”, 3 of 28 were predominantly “yes-yes”, 10 alternated between “yes-yes” and “no-no” to an equal extent, and 9 involved axes other than horizontal.

We examined the clinical factors that correlated with tremor direction (Table 2). Mixed and “yes-yes” cases tended to be older ($p = 0.004$) and have a longer tremor duration ($p = 0.018$) than cases with “no-no” tremor. More than half the “no-no” cases had a tremor duration of less than 20 years, where only 11.1% of the mixed and none of the “yes-yes” cases had such short tremor duration. We also found an association between total tremor score and directionality ($p = 0.04$); scores tended to be higher for mixed cases than for “yes-yes” and “no-no” cases. The continuity of the tremor was correlated with directionality ($p<0.001$): more mixed cases had continuously present tremor while more “no-no” cases had rare head tremor, and “yes-yes” cases displayed an intermediate distribution. A marginally higher proportion of the mixed tremor cases were female (82.1%) ($p = 0.058$). The proportion of mixed and “yes-yes” tremor cases who had voice tremor was higher than those with “no-no” tremor, a difference that was marginally significant ($p = 0.07$, Table 2).

Discussion

While patients are often unaware of their head shaking, head tremor is nevertheless an embarrassing and, in severe cases, functionally impairing disease manifestation that may have significant social and occupational costs. In the current sample of ET cases, a mixed directionality head tremor was more common than either a purely vertical or horizontal tremor. This is a different finding than that of two previous studies, which reported that horizontal tremor was the most common. However, in one of those studies, the mean tremor duration was only 13.5 years, compared to 38 years in ours. The tremor duration in those with head tremor was not reported in the other study, although the tremor duration in the entire sample was only 26.5 years (index cases) and 19.5 years (relatives with definite ET) [7]. The longer tremor duration of our sample might account for the difference in findings. Indeed, among our 10 ET cases with tremor duration <20 years, 7 (70.0%) had “no-no” tremor and only 3 (30.0%) had mixed tremor.

Another factor contributing to the difference between our findings and those of previous studies may be inter-study differences in how directionality was defined. We defined directionality with strict precision, resulting in multiple categories rather than collapsing our observations into a small number of categories based on the predominant direction. If we were to have categorized according to the predominant axis, 19 would have been mixed, 20 predominantly “no-no”, and 12 predominantly “yes-yes”, with “no-no” thus being the most

common. Even with this, the proportion of mixed tremor remains high in our cohort, possibly for reasons discussed above. Furthermore, we believe a more finely-graded approach yields data that are more granular and of greatest potential utility.

In examining the clinical correlates of head tremor, we discovered an association between head tremor directionality and tremor duration, tremor severity (total tremor score), continuity of tremor, and age. Older patients and those with tremor for over 20 years appeared to be more likely to have a mixed or “yes-yes” tremor type, and mixed tremor was more likely to be continuously present in the head than “yes-yes” and “no-no” tremor. Taken together, these findings suggest that unidirectional tremor may represent a precursor phenotype to mixed tremor. One possibility is that head tremor in ET starts as an infrequent tremor in one direction (esp. “no-no”) and becomes more frequent while acquiring additional directionality and a mixed phenotype as the disease progresses. This is further bolstered by the finding that the total tremor score was higher amongst the mixed tremor cases, and is consistent with our current understanding of ET as a slowly worsening, if not neurodegenerative, disease [10].

Our data and prior data indicate that it is part of the normal disease spectrum of ET for head tremor to manifest in the vertical and horizontal directions as well. Rivest and Marsden examined 5 patients with isolated dystonic head tremor and noted that the tremor was in the “no-no” direction in all cases [11]; these data have been used in diagnostic settings to argue that “no-no” head tremor is more consistent with dystonia than ET [12]. From our data, we can see that inferences about diagnosis cannot so easily be drawn based on directionality alone and that tremor duration should be factored in to these considerations.

A main limitation of our study was the clinical method used to assess the direction of head tremor. A computer-based system of determining tremor axis would perhaps have been more precise; unfortunately, such software was not available to us. Similarly, accelerometry data would have provided more precise and objective measurements of tremor amplitude than clinical observation. Physiologic data in the form of electromyographic recordings of individual muscles typically involved in head tremor may also have been useful in further excluding any subtle, subclinical dystonic activity in these muscles. In addition, one might argue that a horizontal tremor is easier to detect when observing the patient head-on, whereas a vertical tremor is more easily recognized in profile. Since patients in our videos spent most of the time facing the camera, it is possible that the presence of “no-no” tremor was slightly overestimated. In terms of study design, a cross-sectional study has inherent limitations as it does not allow us to track the progression in the characteristics of head tremor over time in ET. Lastly, the evaluation of patients was based on an in-person neurological examination performed by trained clinical research staff and videotaped for further review by a senior movement disorders neurologist. Some frequency ranges of tremor are less easily detected on videotaped examination.

This report has elucidated an under-studied clinical manifestation of ET, which may be of relevance to practicing physicians. In the diagnostic setting, it is useful to be familiar with the typical patient as well as the range of what is seen across a group of patients. This has the potential to increase diagnostic certainty and limit diagnostic errors. In the future we

may come to a better understanding of the natural history of head tremor in ET and of the factors that affect its presentation and its successful treatment.

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- Head tremor is present in 1/3 of cases of ET remains under-studied.
- A mixed direction is more common than horizontal or vertical head tremor in ET.
- Head tremor direction correlates with duration, severity, and continuity of tremor.

Table 1

Clinical characteristics of 51 ET cases with head tremor on examination

Characteristic	Data
Age (years)	74.5 ± 10.9
Female gender	35 (68.6)
Tremor duration (years)*	38.3 ± 19.3
Reported one or more 1st degree relative with ET	17 (33.3)
Prescribed tremor medications	35 (68.6)
Tremor in voice (examination)	19 (37.3)
On examination, the head tremor is:	
Continuous	20 (39.2)
Intermittent	15 (29.4)
Rare	16 (31.4)
Total tremor score (0–36)**:	
0–12	9 (18.0)
13–24	30 (60.0)
25–36	11 (22.0)

All values represent mean ± standard deviation or number (percentage)

* Data unavailable for 4 ET cases

** Data unavailable for 1 ET case

Table 2

Relationship between direction of head tremor and clinical characteristics in 51 ET cases

	mixed	no-no	yes-yes	chi-square	p-value
N	28	14	9		
Age category (years)				15.45	0.004
<60	0 (0.0)	6 (42.9)	1 (11.1)		
60–74	9 (32.1)	3 (21.4)	4 (44.4)		
≥75	19 (67.9)	5 (35.7)	4 (44.4)		
Female	23 (82.1)	8 (57.1)	4 (44.4)	5.68	0.058
Tremor duration (years)*				18.52	0.018
<20	3 (11.1)	7 (58.3)	0 (0.0)		
20–39	13 (48.1)	1 (8.3)	3 (37.5)		
40–59	6 (22.2)	4 (33.3)	3 (37.5)		
≥60	5 (18.5)	0 (0.0)	2 (25.0)		
Reported one or more 1st degree relative with ET	7 (25.0)	6 (42.9)	4 (44.4)	2.22	0.33
Prescribed tremor medications	18 (64.2)	9 (64.2)	5 (55.6)	1.63	0.80
Tremor in voice (examination)	12 (42.9)	2 (14.3)	5 (55.6)	5.20	0.07
Tremor continuity				20.13	<0.001
Continuous	18 (64.3)	1 (7.1)	1 (11.1)		
Intermittent	6 (21.4)	4 (28.6)	5 (55.5)		
Rare	4 (14.3)	9 (64.3)	3 (33.3)		
Total tremor score (0–36)**:				9.88	0.04
0–12	2 (7.1)	4 (30.8)	3 (33.3)		
13–24	16 (57.6)	8 (61.5)	6 (66.7)		
25–36	10 (35.7)	1 (7.7)	0 (0.0)		

*Data unavailable for 4 ET cases

** Data unavailable for 1 ET case