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Tremor and Other Hyperkinetic Movements

Brief Reports

Four Essential Tremor Cases with Moderately Impaired Gait: How Impaired can Gait be in this Disease?

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Abstract

Background: A body of literature is emerging regarding gait/balance impairments observed in essential tremor (ET) patients. Although impairment is generally mild, the full extent of the spectrum remains undefined. We present four ET cases with more severe gait/balance impairment.

Methods: A battery of subjective and objective gait/balance assessments was performed: the Activities-specific Balance Confidence Scale, the Berg Balance Scale, and the Tinetti Performance Oriented Mobility Assessment (POMA). Tandem missteps during 10 steps were counted. Quantitative gait testing was performed (GAITRite) to quantify gait speed, dynamic balance, gait symmetry, and gait variability.

Results: Two patients were middle-aged (38 and 52 years) and two were older (70 and 79 years). All had longstanding classic ET (duration 22–60 years). The mean POMA score was 21.5, which is indicative of moderate fall risk. On average, there were five missteps during tandem gait, which was higher than observed in substantially older ET cases (age 86.0±4.6 years), and four times higher than seen in ET patients of comparable age. On quantitative gait analysis, patients demonstrated significant balance impairment.

Discussion: We present a sample of ET patients with a level of gait difficulty that would not be characterized as mild. The existence of such cases raises a number of questions, one of which is how impaired can gait be in ET?

Keywords: Essential tremor, gait, ataxia, cerebellum

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Introduction

A growing number of case-control studies have demonstrated that gait and balance impairments occur in patients with essential tremor (ET). Impairments have been observed both on tandem walk and on standardized clinical assessments of balance.[1] Gait impairments include decreased velocity and cadence, increased time in double support, and step time asymmetry.[2,3] In general, the problem, often referred to as "ataxia," is regarded as mild. Yet the prevalence of gait/balance problems in ET and the full spectrum of deficits are unknown. What proportion of community and/or clinic-ascertained patients with ET has mild ataxia? Moreover, how impaired can gait be in this disease?

The Tremor Task Force of the Movement Disorders Society recently launched an initiative to re-assess the definitions and nosology of tremor disorders. The notion that patients with ET may have physical signs aside from tremor (e.g., ataxia, dystonia) is being incorporated in some form in the revised schema. Yet, the extent to which these physical signs may be present in patients with ET is unclear and remains controversial.

Here, we present four ET patients who each complained of gait/balance difficulty during their routine office visits. They spontaneously brought up the issue as one of their main complaints, aside from arm tremor. Our primary aim was to document their gait/balance impairments, and the overarching aim was to advance discussion about the boundaries of what is clinically observable in ET.

Methods

Four patients with ET were recruited during their regularly scheduled outpatient visits with a movement disorder neurologist (E.D.L.) at Columbia University Medical Center (CUMC). These visits took place in 2012. During their visits, these patients spontaneously raised the issue of gait and balance difficulty as one of their main complaints aside from tremor (e.g., "my walking is getting worse," "I feel unsteady"). Within 6 months, they underwent a standardized study visit designed to more fully assess their gait and balance. At that time, each enrollee signed a study-specific CUMC Institutional Review Board consent form.

The diagnosis of ET was initially assigned clinically and then reconfirmed in each case using published diagnostic criteria,[4] which required the presence of moderate or greater amplitude kinetic tremor in the arms or head in the absence of another known cause (e.g., medications, dystonia, Parkinson's disease [PD], or another neurodegenerative disease).

Kinetic tremor was assessed during a variety of maneuvers, including the drawing of an Archimedes spiral with each hand, which was rated from 0–3 (most severe).[4] An International Co-operative Ataxia Rating Scale (ICARS) score (0–100 [most impaired])[5] was assigned to each subject.

A range of subjective and objective gait and balance assessments were performed. The Activities-specific Balance Confidence Scale (ABC, range=0 [most impaired]–100%)[6] asks the patient to self-assess confidence during 16 activities (e.g., walking around the house, walking on icy sidewalks). The Berg Balance Scale (total score=0 [most impaired]–56)[7] is a performance-based test during which patients are rated on their ability to maintain balance while performing 14 tasks (e.g., retrieving an object from the floor, standing on one foot). The Tinetti Performance Oriented Mobility Assessment (POMA) was administered, which included assessments of both balance and gait.[8] POMA scores <19, 19–24, and 25–28 are associated with high, medium, and low fall risk, respectively. Finally, each patient was asked to walk tandem (place one foot in front of the other, touching toe to heel), and the number of missteps during 10 steps was counted.

Quantitative gait testing was performed using the GAITRite system, a 4.6 m long computerized mat (CIR Systems, Havertown, PA) placed in the middle of a quiet hallway to collect gait data. The mat registers the location and timing of each footfall. Subjects began walking 3 m from the beginning of the mat and stopped 3 m beyond the end of the mat to record steady-state gait on the mat without the influence of gait initiation and termination. For further analysis, we computed: 1) measures of gait speed (velocity, cadence, and step length), 2) dynamic balance (double support percent of gait cycle, support base, and tandem missteps [the latter was measured only during tandem walk]), 3) gait symmetry (step time difference), and 4) gait variability (coefficient of variation [CV] in swing time).

We compared gait performance of participants in the present study with normative data for healthy participants aged 70–79 years, and where available, with normative data for healthy participants aged 40–49 years.[9] We also used published data on ET cases as additional comparison/reference points, including our own published data on a sample of 122 ET cases with a mean age of 64.9 ± 15.4 years (similar to the mean age of the four ET patients),[10] data from other research groups on ET cases with ages similar to that which we report,[1,11] and a sample of 104 ET cases with a mean age that was considerably older (86.0 ± 4.6 years) than patients in this study.[2]

Results

Two of four patients with ET were middle-aged (38 and 52 years) and two were older (70 and 79 years); the mean age was 59.8 years. All had longstanding (duration=22–60 years) classic ET, a syndrome characterized mainly by kinetic and postural tremors (Table). All had a clear family history of ET. Three patients were currently taking ET medications, and none had undergone deep brain stimulation surgery. None had primary or gaze-evoked nystagmus, dysarthria, scanning speech, peripheral neuropathy, or a family history of a progressive ataxia syndrome.

The ABC scores of our patients (Table, mean=74.2) were considerably lower than typically reported in ET (~90 in ET cases with a mean age of ~58 years[1] and 96.4 in ET cases with a mean age of 55.8 years),[11] indicating relatively low confidence in balance. The mean POMA score was 21.5, which is indicative of moderate fall risk, although in one patient the score was <19, indicating a high fall risk. Moreover, our participants had, on average, 5 missteps during tandem gait, which was higher than the mean number of missteps (4.3 ± 4.7) seen in ET cases whose mean age was >25 years older (86.0 ± 4.6 years)[2] and four times higher than the mean number of missteps (1.3 ± 1.8) seen in ET cases of a comparable mean age (64.9 ± 15.4 years).[10]

Quantitative gait testing indicated that gait velocity for our participants was well below the age-normative values, especially for the two older patients (Table). Dynamic balance (measured by percent time spent in double support) was impaired in all four patients; indeed, even the two younger patients with ET had higher scores (27.5 and 32.4, indicative of poorer performance) than the normative data for 70–79 year olds (double support percent=26.3). Gait symmetry (measured by step time difference) was even more impaired in our four ET patients than in older ET cases (age=86.0 ± 4.6 years).[2] Gait variability (measured by swing time CV) was higher in our participants compared with older ET cases (age=86.0 ± 4.6 years)[2] and almost double that observed in healthy older controls (Table).

Discussion

We present a sample of ET patients with a level of gait difficulty that would not be characterized as mild. During their regularly scheduled outpatient visits, these four patients spontaneously raised the issue of gait and balance difficulty as one of their main complaints aside from tremor. Indeed, balance impairment was greater than that observed in a group of significantly older ET cases. Our participants had impairment in dynamic balance, gait symmetry, and gait variability, all of which are related to increased risk for falls. In fact, scores on clinical tests indicate that most of our participants were at a moderate or

Table. 1 Clinical Data on Four Patients with ET

	Patient 1	Patient 2	Patient 3	Patient 4
Age (years)	79	70	52	38
Gender	Male	Male	Male	Female
Age of tremor onset (years)	35	6	12	16
Tremor duration (years)	44	64	40	22
Family history of ET	Yes (Brother, Sister)	Yes (Sister)	Yes (Grandfather, Nephew)	Yes (Father, Grandmother)
Current ET medications	Pro 10 mg/d, Top 50 mg/d	None	Gab 1600 mg/d, Top 100 mg/d	Pro 80 mg/d
Mean Archimedes spiral score (range=0–3[1])	2.5	2.0	3.0	3.0
International Co-operative Ataxia Rating Scale (range=0–100[1])	24	13	27	37
Posture/Gait (0–34)	9	4	3	10
Kinetic Functions (0–52)	15	9	24	27
Speech (0–8)	0	0	0	0
Oculomotor (0–6)	0	0	0	0
Activities-specific Balance Confidence Scale (ABC, range=0[1]–100)	48.8	78.8	78.4	90.9
Berg Balance Scale (range=0[1]–56)	42	54	54	53
POMA (range=0[1]–28)	18	22	25	21
Number of tandem missteps (range=0–10[1])	10	4	1	5
GAITRite Data[2]				
Velocity (m/s)	0.79 [1.17]	0.90 [1.17]	1.18 [1.28]	1.16 [1.28]
Cadence (steps/min)	85.2 [102.0]	100.1 [102.0]	129.6 [125.1]	87.5 [125.1]
Step length (m)	0.55 [0.69]	0.52 [0.69]	0.60 [0.65]	0.80 [0.65]
Double support time (%)	28.7 [26.3]	40.1 [26.3]	27.5	32.4
Step time difference (s)	0.07 [0.03]	0.04 [0.03]	0.05	0.05
Swing time CV (%)	9.4 [4.5]	6.7 [4.5]	11.8	6.7

[1]Most impaired. [2]Values in brackets are normative data for healthy participants aged 70–79 years, or where available, normative data for healthy participants aged 40–49 years.

Abbreviations: CV, coefficient of variation; Gab, gabapentin; m, meters; mg/d, mg/day; m/s, meters/second; POMA, Tinetti Performance Oriented Mobility Assessment; Pro, propranolol; Top, topiramate.

higher risk for falls. The balance impairment was also evident in the higher number of missteps during the tandem walk. These data raise a number of questions, one of which is how impaired can gait be in ET?

Gait impairment in ET is consistent with an emerging understanding that the cerebellar system is impaired in this disease. This likely accounts for the motor features of ET. That tremor in ET can range broadly from mild to severe is clear, but the extent to which other motor features (e.g., gait ataxia) may be present and the extent to which they may worsen remain unknown. The full spectrum has not been elucidated, but it is conceivable that it is broader than currently envisaged.

The four patients described here spanned a broad age range, and it was not possible to compare them as a group to a single age norm, but it is clear that their level of impairment was abnormal. Indeed, even the younger ET patients performed worse than ET cases who were many decades older.

Could any of these patients have spinocerebellar ataxia? Though this is possible, it is not likely. None of the patients had primary or gaze-evoked nystagmus, dysarthria or scanning speech. None had a family history of a progressive ataxia syndrome, and all had a longstanding history of a kinetic tremor syndrome with later gradual development of gait disturbance. The ICARS scores, although higher than typically reported in ET (0–19),[11] are far lower than typically reported in patients with longstanding (8–9 years) multiple system atrophy (~90) and cortical cerebellar atrophy (~70).[12]

The sample we present is a self-selected group of ET cases and is not representative of all ET. Indeed, this was the point of presenting this small case series. At this time, we do not know the prevalence of individuals with such marked gait difficulty in ET. Determining the prevalence is certainly worthy of additional study but is beyond the scope of the current case series.

In summary, we present a group of ET patients with a level of gait difficulty that would not be characterized as mild. During their regularly scheduled outpatient visits, these four patients had spontaneously raised the issue of gait and balance difficulty as one of their main complaints aside from tremor. These data raise a number of questions, one of which is how impaired can gait be in ET?Further study of the full spectrum of gait/balance difficulty in ET and how it evolves over time is needed.

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