BRIEF COMMUNICATION

Orthostatic Tremor Inducing Instability

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Abstract Orthostatic tremor (OT) is a neurological disease of unknown aetiology. It is defined by the presence of a 10–20 Hz tremor in the legs while standing still. Symptoms described are dizziness and instability that diminish if the patient sits down or leans on something; drinking small amounts of alcohol significantly reduces OT. Due to the dizziness and/or unsteadiness, these patients are usually referred to the neuro-otology department. We report 4 cases diagnosed with OT. The diagnosis of OT should be considered for patients with instability. The clinical history is a key factor to suspect this entity, and the diagnosis is given by the register of 10–20 Hz contractions on limb electromyography. Treatment for this disease consists of medical treatment; the first option is clonazepam.

PALABRAS CLAVE

Temblor ortostático como causa de inestabilidad

Resumen El temblor ortostático (TO) es una enfermedad neurológica de origen desconocido caracterizada por un temblor de 10-20 Hz en las piernas en bipedestación. Se manifiesta por mareo e inestabilidad, que tipicamente mejoran al apoyarse o sentarse y la ingesta de pequeñas cantidades de alcohol lo reduce de manera significativa. Se muestran 4 casos clínicos atendidos en nuestra consulta cuyo diagnóstico sugiere ser el de TO. Consideramos que ante un paciente con inestabilidad, es preciso plantearlo como diagnóstico diferencial un TO. La historia clínica nos orienta hacia esta entidad y en caso de sospecha, el diagnóstico definitivo viene dado por el registro de la electromiografía en las extremidades inferiores en condición de reposo sentado y en ortostatismo donde se registra un temblor de 10-20 Hz. El tratamiento es médico y, se emplea el clonazepam como primera opción terapéutica.

Introduction

Orthostatic tremor (OT) was first described by Heilman.1 It is a rare entity and its prevalence is unknown due to the lack of epidemiological data.2 It is characterised by presenting an intense and regular electromyographic activity of 10–20 Hz in the muscles of the lower limbs. However, a characteristic pattern suggestive of this entity in electromyographic records is one of 6–20 Hz bursts.3 This generates a sensation

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of instability, dizziness and/or discomfort in the back of the legs during sitting. Precisely this high frequency of contraction causes the tremor not to be felt, only its consequences, with the main one being instability. Typically, this sensation becomes intensified in a standing position, whereas sitting, holding onto something or walking relieves it. An isometric contraction of the lower muscles is necessary to trigger the tremor. The cause is unknown and appears to originate in a supraspinal trigger, located in the brainstem and influenced by uncontrolled activity of the thalamocortical circuit. In general, 75% of cases are idiopathic and 25% are associated with additional neurological disorders. The latter, ordered by frequency, include: Parkinsonism, restless leg syndrome, clonus, myoclonus and cerebellar ataxia. It usually begins at age 50, with a mean diagnostic delay of 5–8 years, and in most cases the symptoms remain unchanged: in 15% of patients the symptoms spread to the thorax and arms.

Vestibular function studies are generally normal or non-specific, with no locator value. Posturography (both static and dynamic) is useful as an initial screening tool, whilst the definitive diagnosis is obtained by an electromyographic (EMG) study of gravity and antigravity leg muscle activity.

Despite being a little known entity for otolaryngologists, it should be included in the differential diagnosis of patients with chronic instability, which is common in otoneurology consultations. We report 4 cases which illustrate different forms of presentation of this process.

### Methods and Results

We present a series of 4 clinical cases (Table 1) of patients attending otolaryngology consultation at a tertiary centre due to instability, and whose medical histories led us to suspect OT.

#### Clinical Case 1

The patient was a healthy, 65-year old woman, who attended consultation due to instability, sensation of nausea and general malaise, along with heaviness of the lower limbs when standing, which was relieved by leg movement. These symptoms had been appearing episodically for 3–4 h, once per week, for the past 2 years.

Both the otological and otoneurological examinations were normal (eye-tracking and saccades, spontaneous evoked nystagmus with and without visual fixation, vestibulo-oculomotor reflex, head impulse manoeuvre towards the right and left sides, dynamic visual acuity, nystagmus after cephalic shaking, Dix-Hallpike manoeuvre, positional nystagmus). The Romberg and Fukuda manoeuvres were normal. Tandem gait was normal. A neurological examination confirmed the absence of associated neurological signs.

Tonal audiometry revealed a symmetrical, mild, bilateral sensorineural hearing loss. Caloric testing (with water, bithermal, monaural and alternating) revealed 35% right canalicual paresis. The result of the study of cervical vestibular evoked myogenic potentials with acoustic stimulation (CvEMpac) was normal (Table 1). A PET was requested, which revealed no pathological findings.

<table>
<thead>
<tr>
<th>Patient</th>
<th>Gender</th>
<th>Age</th>
<th>Duration, Years</th>
<th>Physical Exploration</th>
<th>Caloric Test</th>
<th>Audiometry</th>
<th>Posturography</th>
<th>MRI</th>
<th>Frequency of Tremor</th>
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<td>65</td>
<td>1.5</td>
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<td>Normal</td>
<td>Normal</td>
<td>ND</td>
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<td>14–16 Hz</td>
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<td>1</td>
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<td>Normal</td>
<td>Normal</td>
<td>Pathological</td>
<td>ND</td>
<td>2–9 Hz</td>
</tr>
<tr>
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<td>Female</td>
<td>45</td>
<td>2</td>
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<td>Normal</td>
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<td>Pathological</td>
<td>ND</td>
<td>10 Hz</td>
</tr>
<tr>
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<td>63</td>
<td>10</td>
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<td>Normal</td>
<td>Pathological</td>
<td>Pathological</td>
<td>ND</td>
<td>8 Hz</td>
</tr>
</tbody>
</table>

MRI: magnetic resonance imaging; ND: no data; SNH: sensorineural hearing loss.

### Table 1

Demographic Data, Physical Examination and Complementary Tests.
The vestibular rehabilitation suggested initially, worsened the symptoms.

The EMG showed rhythmic muscle activity at 14–16 Hz, with alternating, high-frequency bursts (Fig. 1).

We confirmed the suspected diagnosis of orthostatic tremor and referred the patient to the Neurology Department, to start treatment with clonazepam.

Clinical Case 2

The patient was a 69-year old woman with a history of arterial hypertension (treated with amlodipine and valsartan), dyslipidemia, hyperglycaemia and excess weight, who was referred to the otoneurology consultation due to a "feeling of rocking from front to back" in orthostatism, which improved with walking and the intake of small amounts of alcoholic beverages. She was diagnosed with psychogenic dizziness.

The otological, otoneurological and neurological examinations were normal. The tone audiometry, videonystagmography (VNG: recording of nystagmus and caloric test), CVEMPac and magnetic resonance imaging (MRI) tests were all normal. The posturography study indicated a visual preference pattern (Table 1).

The EMG in standing position showed rhythmic, high-frequency muscular discharges of 2–9 Hz (Fig. 1).

The patient was referred to the Neurology Department for further study and to initiate treatment with clonazepam.

Clinical Case 3

The patient was a 45-year old woman with no history of interest, who had suffered a continuous sensation of instability for 2 years. The patient referred that these symptoms worsened when standing and did not vary in bright or dark conditions or when the surface was less firm. She explained that standing still did not help her to improve when she felt worse, and she only achieved relief by sitting or holding onto something. Discomfort was initially perceived in the legs and improved with occasional alcohol intake. She reported bilateral tinnitus, which increased with instability, but no episodes of vertigo or hearing loss. She was referred to the psychiatrist, who did not identify significant alterations.

The otological and otoneurological examinations were normal. The audiometry, VNG, CVEMPac and neurological examinations were normal.

Posturography indicated an inadequate use of sensory information for balance, with a vestibular deficit pattern (Table 1).

Diffusion MRI sequences showed an asymmetry in the assessment of the corticospinal pathway and a volume reduction of the right cerebral peduncle.

The EMG showed rhythmic, high-frequency discharges at 10 Hz (Fig. 1).

We requested an assessment by the Neurology Department, which established a diagnosis of OT, as well as a mild, vascular encephalopathy secondary to congenital thrombophilia, heterozygous for factor V Leiden. The patient started treatment with clonazepam.

Clinical Case 4

The patient was a 63-year old woman with a history of migraine with aura, who presented instability and vertigo.

The first episode of vertigo dated back 10 years. Since then, the patient had suffered several episodes lasting between hours and days, with associated tinnitus in the left ear. In addition, the patient reported a feeling of instability when walking, which became more intense when standing.
still without support, and which was the most uncomfortable symptom due to the limitations it imposed.

The otological examination was normal. The otoneurological examination highlighted an upwards vertical nystagmus, and clockwise in the left Dix-Hallpike manoeuvre, of very low intensity. The neurological examination reported no pathological findings of interest.

Tone audiometry showed a loss of 30 dB in the left ear. The caloric test (with water, bithermal, monaural and alternating) reported 23% right canalicular paresis. The CVEMPac were normal (Table 1).

Palpation of the lower extremities found signs of fasciculation in orthostatism. The EMG revealed rhythmic, alternating bursts of 8 Hz (Fig. 1).

The patient was diagnosed with benign paroxysmal positional vertigo of the left, posterior, semicircular canal with residual instability. She was treated with clonazepam due to the suspicion of OT.

Discussion

The present study details 4 cases of OT, whose reason for consultation was instability.

The clinical approach for patients with chronic instability (>3 months) should aim to: (1) identify syndromic forms, (2) obtain a good retrospective diagnosis, (3) rule out associated disorders, (4) ensure that there is no gait alteration, and (5) bear in mind that the treatment will be multidisciplinary. The differential diagnosis must include various entities but the clinical history should provide important clues for diagnosis. In the clinical cases presented, the symptom which was "different" from the usual was worsening in orthostatic position and improvement with ambulation, which was contrary to the usual reports from patients with chronic instability secondary to vestibulopathy. The need for further investigation generally stems from the fact that vestibular examination tends to be normal except in those cases with comorbidities (as in cases 1 and 4 presented here).

The most common misdiagnosis may be due to unfamiliarity with this process. Given its atypical features, it is common to diagnose patients with psychogenic instability after ruling out vestibular or systemic disease. From a neurological point of view, the differential diagnosis should be carried out with other diseases and movement disorders that can generate tremor (Table 2).

Therefore, a specific electromyographic study that allows accurate diagnosis should be requested whenever there is suspicion. The presence of brief muscle discharges triggered by orthostatism in both lower limbs is a characteristic finding in OT. Unlike other authors, we have not observed a high degree of correlation between static posturography and EMG in our cases. Perhaps this is because we employed a dynamic posturography device.

Treatment options are numerous and should be indicated by the neurologist. Generally, clonazepam is the initial drug of choice. The 4 patients followed a favourable course with a clear improvement. Side effects of medication were controlled by changing the dosage and intake when necessary.

In conclusion, OT is a rare clinical entity in which the medical history is vital for its suspicion. A posturography study analysing the response in the frequency domain (FFT) can guide the diagnosis, although the definitive diagnosis is determined by the EMG results.

Conflict of Interests

The authors have no conflicts of interest to declare.

References


