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Case Report

Orthostatic Tremor: A Rare Cause of Tremor

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Abstract

Though tremors are most common of the movement disorders, orthostatic tremor is a rare form of tremor seen commonly in adult between ages of 13 years-85 years, most commonly around 60 years. It is so rare that only case report or some small case series have been reported so far. It is one of the most poorly understood of movement disorders.

Keywords: Electromyography; Orthostatic tremor; Deep brain stimulation

Case Presentation

UD, 47-year-old woman came to out-patients department of Neurology with complaints of unsteadiness on standing. She had no problems on sitting, lying down or walking, but as soon as she stood, within few seconds she felt very unstable and feared she may fall. Careful examination did not reveal any abnormality in cranial nerve, cerebellar, pyramidal and sensory systems. When seated no tremors were observed, but on standing fine rippling movements of both thighs, calves could be palpated and on holding arms straight in front of her similar movements could be felt on arms with fine tremors of hands. Her CBC, LFT, KFT, thyroid function tests were all normal including serum electrophoresis. Non-contrast MRI was also normal. Surface EMG performed on Tibialis anterior, quadriceps on standing and brachioradialis on holding arm up showed bursts of muscle activity at 15 Hz of 40 msec-50 msec duration. There was no rhythmic burst activity when the muscles were at rest. She was prescribed 0.5 mg clonazepam at bed time 0.25 mg. at morning and in the afternoon. She has yet to come for review.

Discussion

Orthostatic Tremor (OT) was first described in to 1970 by Pazzaglia et al. [1,2], who reported on three patients with this disorder occurring only on standing. The term was first coined by Heilman in 1984 who described it as "shaky legs syndrome" [3]. The disorder usually presents as gait disorder, fear of falls and possible cognitive impairment. It greatly affects quality of life and functionality. The age of onset ranges from 13 to 85 years, though it usually occurs around 60 years of age. The symptoms may start acutely or insidiously. The symptoms may start immediately or after standing for some time. Due to feeling of unsteadiness, most patient keep on walking, shifting weight form one leg to other or lean against wall or chair. Progression of symptoms may occur in about 79.4% of patients [4,5], which can occur in terms of severity or progression from leg to trunk and then arms [6]. Progression in severity is measured by the time for which the patient can remain standing without need to walk or sit. OT may be primary or secondary [7]. In patients with primary OT there is no discernible cause or associated neurological features. Primary OT can further be divided as those with arm tremors and those without. Among the secondary causes there are reports of patients with pontine, cerebellar lesion [8,9], parkinsonism (typical, atypical, vascular, drug induced, [9,10]), aqueductal stenosis, Lewy body dementia, cognitive impairment, cerebellar signs and restless leg syndrome. Interestingly, some patients of OT may develop Parkinsonism subsequently [11,12] after several years.

Etiology

Though predominantly a sporadic disorder, some familial cases have been described in monozygotic twins [13], in siblings [14-16] and mother and son [16]. In a large series 184 patients family history of OT was reported in 4.9% of patients [14]. The genes described associated with OT are C10orf2 (Twinkle) and REEP1 mutation (formerly SPG31). REEP1 mutation is also reported with pure hereditary spastic paraparesis phenotype [17]. The role of these mutations as causative of OT remains enigmatic.

Pathophysiology

There is growing evidence that cerebellum and/or its connection are involved in the pathogenesis of OT. Evidences in favour of cerebellum and/or its connections are derived from clinical [18], imaging [19-24], functional imaging and positron emission tomographic studies [16].

Diagnosis

Though the history and examination are very typical, confirmation of diagnosis is by surface Electromyography (EMG) which reveals rhythmic activation of lower limb muscles at frequency of 13 HZ-18 Hz or even higher. Similar activity can be seen even in upper limbs in some cases especially in proximal muscles. The EMG oscillations in various muscles show intermuscular coherence.

Investigations

Since it is idiopathic disorder, investigations should be directed to rule out secondary causes of OT, including thyroid function tests, serum protein electrophoresis to rule out gammopathies, vitamin B12 deficiency, Wilson's disease (serum ceruloplasmin), and dopamine transporter imaging to rule out Parkinson's disease. Brain MRI should be performed to rule out structural causes of OT such as pontine and cerebellar lesions and multiple sclerosis.

Treatment

Since it is an uncommon and poorly understood disorder, it takes several years before it is diagnosed especially by non-movement

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disorders neurologists. Many patients suffer from lot of psychological issues like anxiety and depression. These patients need careful and sympathetic handling. Since it is uncommon disease, no randomised trials are available either. Unlike essential tremor, response to alcohol and beta-adrenergic blocking agents is not rewarding. Pharmacological agents that have been used are: clonazepam, clobazam, gabapentin, levodopa, pramipexole, primidone, phenobarbital, leviteracetam, valproate, Immunoglobulin, beta-blockers with variable results. Best results are seen with clonazepam followed by gabapentin and levodopa [7, 25-27] surgical therapy in the form of Deep Brain Stimulation (DBS) can be offered to medically refractory cases, though no guidelines are presently available. Stimulation of unilateral or bilateral ventral intermediate thalamic nucleus [28,29] and two patients of spinal cord stimulation [30,31] have been offered to patients with mixed results.

Conclusion

We report a rare case of OT in a 47 year old healthy female. This is a poorly understood disorder presenting as unsteadiness on standing, sometimes also with tremulousness of legs as well. The diagnosis is clenched by surface EMG of leg muscles done while standing. Investigations are needed to rule out secondary causes of OT. The treatment is also unsatisfactory, some patients improving with clonazepam, levodopa or gabapentin. Some patients also need psychological support.

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