Shaky legs? Think POT!

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Abstract

Primary orthostatic tremor (POT) may be a cause of postural instability in older people. Though unusual, this condition is relatively easily diagnosed, based in part on typical clinical features, impacts on quality of life, and is amenable to symptomatic treatment (Heilman KM. Orthostatic tremor. Arch Neurol 1984; 41: 880–1). We present three cases seen in a general neurology clinic over a 15-month period to highlight the typical clinical features which should alert clinicians to the possibility of this diagnosis.

Keywords: primary orthostatic tremor, postural instability, diagnosis, elderly

Case 1

A 78-year-old man was referred from primary care with a 5- to 6-year history of a ‘wobbly, shaky feeling’ in his legs when standing. The general practitioner was querying a diagnosis of cerebellar degeneration. Matters had come to a head when the patient was visiting the antiquities of Egypt: standing listening to a tour guide’s lecture, he had to lean against the ancient tomb paintings to relieve his symptoms, for which he was rebuked by the tour guide. On direct questioning, his symptoms were reliably stopped by leaning against a wall and were not present on walking or sitting. On examination, there was a fast tremor of the legs on standing, and a mild postural upper limb tremor. Electromyography (EMG) showed a reproducible 13–14 Hz tremor in all leg muscles sampled with a latency of several seconds after standing, thus confirming the clinical suspicion of POT. He was treated with clonazepam 0.5 mg with symptomatic improvement.

Case 2

A 65-year-old man was referred from primary care with a 4- to 5-year history of shaking in the legs if standing, for example in a queue, at the bar or addressing a golf ball. A working diagnosis of anxiety had been made, leading to therapeutic trials of a beta-blocker, a benzodiazepine (lorazepam), and an antidepressant, all without effect. On direct questioning, neither walking nor sitting was associated with symptoms. A fast tremor of the legs was evident on standing and EMG showed 15–16 Hz rhythmic tremor in both quadriceps on standing. He was treated with clonazepam 0.5 mg with symptomatic improvement.

Case 3

A 65-year-old woman was referred from primary care at the suggestion of an orthopaedic surgeon, with an 18-month history of trembling in the legs when standing in queues, or when attending church; leaning against a pillar would relieve the feelings. She also complained of shakiness in the hands when using them for fine work. She had also been seen by a rheumatologist for these symptoms, who had arranged nerve conduction studies which were unremarkable, aside from evidence of a carpal tunnel syndrome. On examination, she had a postural upper limb tremor, and a fast tremor in the legs on standing. EMG showed reproducible 15 Hz activity in quadriceps on standing, which stopped with leaning against a support. She wanted no drug treatment, being content to learn that there was no other serious underlying diagnosis. Carrying a shooting-stick (a walking stick with a head that opens out into a seat) allowed her to sit to relieve her symptoms whenever required.

Discussion

These cases illustrate the typical symptomatology of POT: shakiness, trembling, wobbliness or a jelly-like feeling of the legs on standing, reliably relieved by leaning, sitting or walking, and sometimes associated with upper limb tremor [1]. It has sometimes been known as the ‘shaky legs syndrome’. Although patients do not fall, their complaint of unsteadiness may prompt referral to falls clinics. Concurrence with Parkinson’s disease has been reported on occasion [2], but this may simply be a chance association with a common disease. Examination reveals a fast tremor of the legs on standing, which may sometimes be palpable as a fine-amplitude
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rippling of leg muscles such as quadriceps with an associated knee tremor, more easily felt than seen because of its high frequency. Auscultation over lower limb muscles using the diaphragm of the stethoscope may reveal a regular thumping sound, likened to distant helicopters [3]. Because of the sparse findings on neurological examination, patients with POT have sometimes been labelled as having psychiatric or psychological problems (e.g. case 2).

The EMG finding of reproducible 14–18 Hz synchronous activity in leg muscles on standing is pathognomonic. Posturography may also be helpful in detecting cases [4]. Pathophysiology is uncertain: prior reports suggesting evidence of a central dopaminergic deficit have not been confirmed in more recent studies using (123)I-FP-CIT-SPECT (DatSCAN) imaging [5].

POT was first described as such some 25 years ago [1], although there may have been earlier descriptions [6]. Although the condition is known to most neurologists, it may be unfamiliar to geriatricians and general physicians. The history and examination findings are very characteristic, and hence POT should not be mistaken for other types of tremor. The differential diagnosis encompasses symptomatic causes of orthostatic tremor, including Parkinson’s disease and other Parkinsonian syndromes, pontine lesions and Graves disease, but these conditions are invariably associated with other neurological symptoms and signs. Mean age at onset in a series of 41 patients was 50 years, but with a broad standard deviation (15 years) [7]. Certain breeds of dogs, such as Great Danes, may also be afflicted with POT [8].

A number of drugs have anecdotally been reported to be helpful in POT, including phenobarbitone, primidone, clonazepam, pramipexole and levodopa, although the only double-blinded placebo-controlled study suggesting efficacy is with gabapentin [9]. This latter study involved six patients titrated to the maximum effective dose of gabapentin who then had medication withdrawn, with increase in symptoms. They underwent studies including posturography a week later, and then a further 2 weeks later after being randomised to either gabapentin or placebo. Posturography and quality of life improved in all patients treated with gabapentin [9].

The evidence for levodopa is based on an open study of eight patients, five of whom elected to remain on medication after a brief (8-week) assessment period [2]. Unlike the situation in essential tremor, propranolol is not helpful. POT strongly impacts on quality of life measures [9] and there may be concurrent depression which merits treatment in its own right [10].

Key points

- Primary orthostatic tremor (POT) may be a cause of postural instability in the elderly which impacts on quality of life.
- “Shaky legs” on standing, relieved by walking, leaning, or sitting, are the typical symptoms of POT.
- The paucity of clinical signs may sometimes prompt consideration of psychiatric diagnoses.
- Electromyography shows pathognomonic fast (14–18 Hz) synchronous activity in leg muscles on standing.
- Symptomatic treatments for POT are available; treatment of concurrent depression may also be required.

Conflicts of interest

No financial disclosures or conflict of interest.

References


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