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Tremor and Other Hyperkinetic Movements

Video Abstracts

Spontaneous Intracranial Hypotension Associated with Kinetic Tremor and Ataxia

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Abstract

Background: Spontaneous intracranial hypotension (SIH) is a clinically variable syndrome caused by low cerebrospinal fluid (CSF) pressure due to a non-traumatic CSF leak.

Phenomenology Shown: This case describes a 68-year-old gentleman who presents with chronic and slightly progressive kinetic tremor of bilateral hands associated with gait ataxia and gait start hesitation.

Educational Value: This case underscores the importance of having a high index of suspicion for the diagnosis of SIH when encountering a patient presenting with late-onset progressive kinetic tremor and gait ataxia syndrome.

Keywords: Spontaneous Intracranial Hypotension, Ataxia, Tremor

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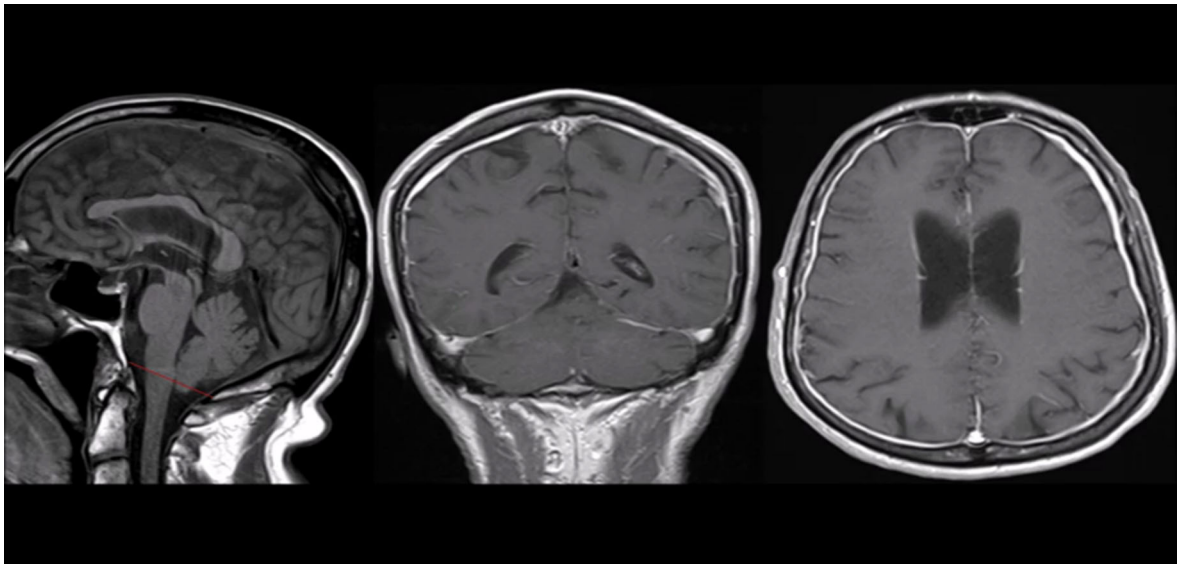
Ethics Statement: All patients that appear on the video have provided written informed consent; authorization for the video-taping and for publication of the videotape was provided.

A 68-year-old male presented with a 2-year history of slightly progressive tremor of bilateral hands associated with an unsteady gait. The tremor was elicited by purposeful movements of the hands. Drinking alcohol did not improve the tremor. He reported mild staggering gait with intermittent hesitation upon gait initiation. He denied bulbar symptoms or deafness. There was no family history of mental retardation. On examination, he displayed a mild, fast, and distal kinetic tremor of bilateral hands without a postural or a resting component (Video segment 1). The Archimedes spiral test was normal. There were no signs of Parkinsonism with the exception of mild gait start hesitation. Cerebellar function was abnormal for mild upper limb dysmetria and trunk titubation. The ambulatory examination showed mild broad base gait associated with impaired tandem gait (Video segment 1). The remainder of the neurological examination, including his mental examination, was normal. A thorough work-up ensued, which included brain magnetic resonance imaging (MRI) studies followed by cerebrospinal fluid (CSF) and serum analysis. Contrast-enhanced brain MRI revealed diffuse, linear, and thick pachymeningeal enhancement of the infra- and supratentorial compartments without involvement of the leptomeninges, and a sagittal T1 slice showed mild brain sagging (Video segment 2). Following the MRI findings, a lumbar puncture identified low CSF pressure of 60 mmH_2O with normal CSF analysis, which included tuberculosis/fungal cultures and cytology. Upon further questioning, he admitted to having occasional orthostatic headaches. On the basis of the clinical presentation and work-up, the diagnosis of spontaneous intracranial hypotension (SIH) was confirmed. Unfortunately, the computed tomography (CT) myelogram failed to show the site of the CSF leak. Later on, the patient decided not to pursue any further work-up, but instead proceeded with conservative management (i.e., increased caffeine intake) with partial response. SIH was first described by Schaltenbrand in 1938. SIH is a syndrome caused by low CSF pressure due to leakage of CSF that mostly occurs at the cervicothoracic or thoracolumbar junction of the spine. Dural weakness could be secondary to an underlying connective tissue disorder, meningeal diverticula, or because of mechanical disruption of the dura,



Video 1. **Movement disorders associated with spontaneous intracranial hypotension.** This video displays mild kinetic tremor of bilateral hands without postural or resting components. The ambulatory pattern is characterized by gait start hesitation in addition to a mild ataxic component and an abnormal tandem gait (**segment 1**). A non-contrast brain magnetic resonance imaging (MRI) T1 sagittal slice shows minimal downward displacement of brain structures. Contrast-enhanced brain MRI shows diffuse and linear meningeal enhancement that involves the pachymeninges of both the infra- and supratentorial compartments without evidence of involvement of the leptomeninges (i.e., no abnormal enhancement around the brainstem, within the depth of the cerebral sulci or the Sylvian fissures) (**segment 2**).

as in the case of craniotomy, spinal surgery, craniospinal trauma, or placement of a ventriculoperitoneal shunt. Medical causes of SIH include dehydration, diabetic coma, uremia, and severe systemic illness. SIH is recognized as a clinically variable syndrome where movement disorders could represent the main presentation.[1,2] In addition to orthostatic headaches, a wide variety of neurological manifestations have been described, such as ataxia, vertigo, blurred vision, cranial nerve palsies, bulbar dysfunction, chorea, parkinsonism, and tremor.[2,3] Unfortunately, the pathophysiology behind these neurological symptoms is not well understood. Traditionally, the work-up consists of contrast-enhanced brain MRI and lumbar puncture followed by CT/magnetic resonance myelogram or radioisotope cisternography to localize the site of the CSF leak. However, the sensitivity of CT myelogram has not been well studied, and many CSF leaks remain occult on all types of spinal imaging studies, as in this case.[2] Brain MRI studies in patients with SIH usually show diffuse thickening of the pachymeninges with gadolinium enhancement, engorgement of venous sinuses, subdural fluid collections, enlargement of the pituitary gland, and brain sagging. The resolution of these abnormalities on magnetic resonance images correlates with improvement in clinical symptoms. The available therapeutic options include conservative management, epidural blood patch, and epidural injection of fibrin glue; surgical repair may be necessary in certain cases. Outcomes are usually favorable but sometimes not long-lasting.[2] This case highlights the broad clinical spectrum of this still under-recognized condition that at times presents with movement disorders as the main manifestation.

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