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Tremor and Other Hyperkinetic Movements

Video Abstracts

Tremor Associated with Chronic Inflammatory Demyelinating Polyneuropathy and Anti-Neurofascin-155 Antibodies

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Abstract

Background: Tremor is an underrecognized feature in certain neuropathy subtypes.

Phenomenology shown: We show a patient with a disabling neuropathic tremor and mild cerebellar syndrome associated with chronic inflammatory demyelinating polyneuropathy (CIDP) and anti-neurofascin-155 (NF155) antibodies.

Educational value: Anti-NF155 testing should be considered in patients with CIDP and disabling tremor because of therapeutic implications.

Keywords: Neuropathic tremor, neurofascin-155, chronic inflammatory demyelinating polyneuropathy

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

We report the case of a 64-year-old female with type 2 diabetes and a longstanding history of bipolar disorder treated with valproate and venlafaxine on stable doses. She presented with gait ataxia and distal limb paresthesia that had gradually worsened over 2 months. She then noticed bilateral upper limb tremor and dysarthria. Clinical examination showed a mild proximal and distal quadriparesis, absent deep tendon reflexes, dysmetria, ataxic dysarthria, and mixed proprioceptive and cerebellar gait ataxia. She had a predominantly action tremor (kinetic, postural, and intentional) with proximal and distal involvement and a mild rest tremor (Video 1). Electroneuromyography showed motor and sensory demyelinating neuropathy in the upper and lower limbs with proximal and distal involvement, consistent with CIDP. Cerebrospinal fluid examination showed an increased protein level (0.67 g/L) with a normal cell count. Polymyographic recording revealed a rest and action tremor with a 5-Hz frequency intermingled with occasional subcortical myoclonus. Brain magnetic resonance imaging and dopamine transporter scan results were normal. Antibody testing was positive for anti-neurofascin-155 (NF155) antibodies. We made the diagnosis of CIDP associated with anti-NF155 antibodies causing a pronounced neuropathic tremor and cerebellar syndrome. The subacute presentation while on stable doses argues against roles of sodium valproate and venlafaxine in the tremor and myoclonic jerks, but a mild effect cannot be excluded. She did not respond to intravenous immunoglobulins (IVIg). Corticosteroids given with rituximab and plasma exchange allowed partial improvement of sensory and motor manifestations, tremor severity, and neurophysiological parameters. NF155 is an adhesion molecule expressed at paranodes at the terminal loops of myelin, where it plays a key role in promoting rapid nerve impulse propagation.[1] CIDP with anti-NF155 antibodies is frequently associated with disabling neuropathic tremor. It is described

Video 1. **Tremor Associated with Chronic Inflammatory Demyelinating Polyneuropathy and Anti-neurofascin-155 Antibodies in a 64-year-old Patient.** Action and rest tremor, predominating on the right side with an occasional myoclonic component. Ataxic dysarthria and gait ataxia are also observed.

as an action tremor with low frequency, high amplitude, and marked postural and intention components.[2] Mild rest component and jerks could also be a feature of neuropathic tremor. In a recently described cohort, some patients with NF-155 antibodies (5/38) had an additional cerebellar syndrome with gait ataxia, dysarthria, and nystagmus. In addition, some patients also had central nervous system (CNS) demyelination (3/38), which may be related to the presence of an NF155 antigen in the CNS.[2] Disabling action tremor in the context of a demyelinating neuropathy is a clue for anti-NF155 antibody-associated CIDP. This has important therapeutic implications as this condition does not usually respond to IVIg but may improve with plasmapheresis and rituximab.[2]

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