

Letters to the Editor Related to Published Article

Distinguishing Scan Without Evidence of Dopaminergic Depletion Patients with Asymmetric Resting Tremor from Parkinson's Disease: A Clinical Diagnosis of Dystonia is Required

Video 

We read with interest the article by Schwingenschuh et al.,¹ who used blinded clinical assessments and electrophysiology to provide evidence to support the hypothesis that patients diagnosed with Parkinson's disease (PD), who have an asymmetric resting tremor with normal dopamine transporter imaging scan without evidence of dopaminergic depletion (SWEDD), may actually suffer from primary dystonia. We have recently reviewed the broad issue of misdiagnosis of dystonia² and highlighted the importance of a thorough clinical assessment aimed to detect features of dystonia.

The article by Schwingenschuh compared PD-like SWEDD patients with PD patients who had abnormal DAT scan. The SWEDD status is an incidental negative finding in patients diagnosed to have PD³ that per se does not provide any alternative diagnostic category. The SWEDD status encompasses a variety of different movement disorders, including dystonia, essential tremor, dopa-responsive dystonia, vascular parkinsonism, drug induced, or psychogenic conditions.⁴ This negative finding requires searching for an alternative diagnosis in presence of classical PD hallmarks, particularly tremor and bradykinesia, which can also be observed in dystonia.²

A possible pitfall is that readers may be lead to consider that the SWEDD condition corresponds to dystonia. The diagnosis of dystonia should be made, instead, by applying specific diagnostic criteria, such as the ones that have been recently proposed,⁵ which have not been used in Schwingenschuh's article. By using such criteria, dystonia can be distinguished from PD, avoiding misdiagnosis, and further ancillary confirmation can be provided by a normal DAT scan.

The relevance of clinical engagement in the diagnosis of dystonia is highlighted by the videocase reported here. This patient's phenotype resembles that of Schwingenschuh's patient 3 with asymmetric dystonic tremor (see Supporting Information Video). The 41-year-old man shown in the video received a diagnosis of PD after a 1-year history of increasing resting and postural tremor in the right hand. Neurological examination showed right rest, postural, and kinetic tremor of the right hand, reduced right arm swing with tremor during gait. Repetitive finger movements were slowed on the right hand. The tremor did not respond to alcohol, primidone, propranolol, and levodopa. Psychiatric evaluation

resulted normal. Brain MRI, routine blood tests, including copper studies and protein electrophoresis, DYT1 testing were all normal. EMG of the right arm demonstrated 7 Hz rhythmic activity at rest and on posture with asynchronous activity in agonist/antagonist pairs. DAT scan was performed twice and resulted normal, indicating SWEDD status.

In this case, the overall appearance was that of PD, but expert neurological examination revealed, in addition to dystonic tremor, abnormal posturing, and a geste antagoniste leading to a clinical diagnosis of primary dystonia. This patient fulfilled the diagnostic algorithm for dystonia⁵; DAT provided a confirmatory test. In our view, all SWEDD patients should receive a clinical diagnosis if not before, at least after that of PD has been ruled out by DAT scan.

LEGENDS TO VIDEO

Segment 1. This patient has unilateral right hand resting and postural tremor. During movement with the right fingers, a slight tremor occurs in left hand (overflow). Reduced right arm swing during gait is associated with right hand tremor, abnormal posturing, and extension of the right wrist.

Segment 2. There is tremor during writing, which is reduced when the patient holds the wrist with the left hand. This geste antagoniste also improves handwriting, as shown by the comparison of writing without or with the geste (arrow).

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Additional Supporting Information may be found in the online version of this article.

Potential conflict of interest: Nothing to report.

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