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A patient with limb apraxia and myoclonic tremor. Two possible culprits: venous drainage anomaly or cortico-basal syndrome?

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Introduction: Unilateral apraxia is often evocative of corticobasal syndrome (CBS). It is a neurodegenerative disorder that poses a significant diagnostic challenge given the wide variety of presentations: from early behavioral or cognitive impairment to classic asymmetric akinetic-rigid motor syndrome and apraxia.

Objective: In the present work, we describe the case of a 78-year-old female with a three-year history of myoclonic tremor and left arm apraxia not responsive to L-dopa and biperiden.

Methods: The patient was admitted to our department for clinical-diagnostic assessment.

Results: Clinical and neurophysiological evaluations were performed. Left hand tremor presented an irregular and high frequency myoclonic pattern at the electromyographic examination. Left hand ideomotor apraxia was confirmed at a neuropsychological assessment, and it was associated with a mild cognitive impairment with short-term memory and executive functions involvement. Single-photon emission computerized tomography (SPECT) with DaTscan showed a bilateral reduction of putaminal uptake, predominantly on the left. Surprisingly, brain MRI highlighted a venous drainage anomaly in the right posterolateral thalamic area, associated with potential paramagnetic deposits in the putamen and substantia nigra bilaterally and fronto-parietal cortical sulci enlargement. The patient was discharged at home with the indication to start clonazepam for the myoclonic tremor, which was only partially effective. Five months later, she underwent a Positron Emission Tomography (PET), which showed moderate bilateral FDG hypofixation in the sensorimotor cortex, predominant on the right.

Conclusions: Asymmetric presentation of myoclonic tremor and apraxia can pose a diagnostic challenge. In our case, SPECT radiological features showed reduced basal ganglia uptake while MRI detected a venous anomaly, which only partially fitted the clinical picture. Based on the clinical presentation, evocative of cortico-basal syndrome, a PET was performed and was diagnosed as cortico-basal degeneration. The clinical presentation should guide the choice of the right diagnostic test.