Abstract

Progressive supranuclear palsy (PSP) is a rare but not uncommon neurodegenerative disorder that belongs to the category of atypical parkinsonism carrying a relentless progression resulting in significant morbidity and mortality. Depending on the individual and the phenotype, the early presentation would vary making the diagnosis more difficult. In this particular case scenario, the patient presented years after the initial onset of symptoms including gait disturbance with early-onset recurrent falls, bradykinesia and cognitive impairment. By the time of presentation, her clinical condition was complicated with near-total immobilization, speech perseverance and most importantly vertical supranuclear gaze palsy. Among the few differential diagnosis made, PSP, idiopathic Parkinson's disease (IPD), multiple system atrophy (MSA) and Alzheimer's disease got highlighted. The presence of core clinical features of Parkinsonism including bradykinesia, rigidity, postural instability and unilateral resting tremors pointed the clinical picture towards idiopathic Parkinson's disease. However, lack of response to levodopa and the diagnosis of vertical supranuclear gaze palsy turns the direction of diagnosis towards PSP rather than IPD. Furthermore, MRI findings suggestive of significant midbrain atrophy without significant pontine atrophy provided essential supportive criteria to establish a unifying diagnosis of PSP which could explain the overall clinical scenario. Surprisingly, unlike in typical PSP, this patient had developed falling tendency towards forward instead of backwards. And also, she had not only axial rigidity but also peripheral limb rigidity whereas more prominent axial rigidity is appreciated in classical PSP. Unfortunately, the neuropathologic examination for neurofibrillary tangles which is the gold standard for diagnosis of PSP could not be performed due to certain restrictions. Consequently, this case report emphasizes the importance of early application of invaluable non-pharmacological measures including physiotherapy, speech therapy, timely swallowing assessments, occupational therapy and nutritional support to improve the quality of life and to hinder the rapid progression of PSP despite the fact of unavailability of a definitive treatment option.