Hummingbird sign in progressive supranuclear palsy disease

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Progressive supranuclear palsy (PSP) is characterized by slowness, rigidity, bradykinesia, repeated falls, downgaze limitation and dementia. Midbrain atrophy on magnetic resonance imaging is highly suggestive of PSP and is described as "hummingbird sign". This sign is very helpful in differentiating PSP patients from those with Parkinson's disease. We hereby report a 72-year-old female case of PSP primarily diagnosed with Parkinson's disease.

Key words: Progressive Supranuclear Palsy, Hummingbird Sign, Midbrain.

INTRODUCTION

Progressive supranuclear palsy (PSP) is a neurodegenerative condition. It is an important differential diagnosis of Parkinson's disease (PD). Magnetic resonance imaging (MRI) in PSP patients may reveal significant midbrain atrophy with sparing of pons known as "hummingbird" sign.

CASE

A 72 year-old lady with gradually progressive history of slowness, frequent falls, rigidity and forgetfulness for 2 years was referred to us. She was initially diagnosed as a case of Parkinson's disease and was thus treated with levodopa/carbidopa for 3 months. She did not show any improvements in her symptoms and was referred to our hospital. On examination, she was having predominantly frontal lobe dysfunctions in the form of repetitive failure to perform executive function. She also had difficulty in calculation and recent memory. She had marked limitation in downgaze with slowing of horizontal saccades.

She had foot dystonia and rigidity in all 4 limbs including neck and trunk. Motor power was normal and deep tendon reflexes were within normal limit. Her gait was slow with lack of arm swing on both sides and tendency to fall backward.

Due to early onset of falls, dementia, predominant parkinsonian features and poor response to levodopa/carbidopa, a diagnosis of PSP was made. MRI of brain was conducted to rule out secondary causes of Parkinson's disease. MRI was suggestive of predominant atrophy of midbrain and preservation of pons and medulla (Figure 1).

DISCUSSION

Prominent midbrain atrophy with no pontine atrophy in PSP patients has been referred to as "hummingbird" or "penguin" sign. This sign has been found to be quite useful in differentiating PSP from PD and multisystem atrophy (MSA).1,2 This sign is best seen in midsagittal image showing flat and concave contour of midbrain. It also suggests rostral interstitial nucleus of medial longitudinal fasciculus involvement in PSP.3,4 In one study, abnormal superior profile of midbrain has been found to be more helpful in differentiation PSP from PD patients.5 Maximal diameter of midbrain in midsagittal plane has been compared in PSP, PD and MSA patients. It has been reported to be quite useful in diagnosing PSP patients. Magnetic resonance parkinsonism index, defined as the ratio of midbrain area to pons area, is another helpful way in diagnosis of PSP patients.6

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