Progressive supranuclear palsy (Steele-Richardson-Olszewski syndrome) is a neurological disorder characterized by degeneration of the midbrain, basal ganglia and other structures like the frontal lobe, pyramidal tracts and cerebellum. It manifests in middle and late adult age by progressive supranuclear gaze palsy, postural instability with recurrent falls, mainly in a backwards direction, symmetrical rigidity, frontal lobe features, and dementia. It can be confused with other diseases like idiopathic Parkinson disease, multiple-system atrophy and corticobasal degeneration. To date, the diagnosis of progressive supranuclear palsy is based on clinical grounds. No biomarker has been discovered. The neuroimaging characteristics, particularly the hummingbird sign, is considered quite suggestive of progressive supranuclear palsy.

CASE
A 58-year-old male presented with insidious-onset, progressive difficulty in initiation of, and slowness in execution of movements for the last 5 years. The patient had recurrent falls starting 6 months from onset of symptoms. For the same duration, he had impaired gait and a change in speech. The gait was short shuffling gait with hesitation in initiation and difficulty in walking through narrow passages along with freezing episodes. Speech gradually became slow, low in volume and monotonous in character. Family members noticed impaired cognition. He showed a poor response to a trial of levodopa therapy. There was no history of trauma, stroke-like episodes, hallucinations, fluctuations of clinical symptoms or dysautonomia.

His BP was 169/86 mm Hg with no evidence of postural hypotension. He had a staring look with infrequent blinking and presence of neck dystonia. A mini–mental status examination score was 21/30. He had vertical gaze palsy, hypermetric horizontal saccades and hypometric vertical saccades, impaired optokinetic nystagmus and positive square-wave jerks. There was rigidity in both the upper and lower limbs, and the plantars were bilaterally extensor. Other parameters of neurological examination were essentially normal.

The liver function test did not reveal any abnormality. Hemoglobin was 130 g/L, total leukocyte count was 6500 cells/cu. mm, random blood sugar was 126 mg/dL, and serum creatinine was 1.2 mg/dL. Magnetic resonance imaging T2-weighted image showed atrophy of the midbrain tegmentum with relative preservation of the shape of the pons (Figures 1, 2). These images resemble the shape of a hummingbird, and are thus known as the "hummingbird sign."
DISCUSSION

Progressive supranuclear palsy is a neurodegenerative disorder, mainly affecting eye movements, cognition and posture. Close differential diagnoses of this condition are Parkinson disease, multiple-system atrophy, and corticobasal degeneration. MRI findings of progressive supranuclear palsy are midbrain atrophy with dilated cisterns, thinning of the quadrigeminal plate, third ventricular dilatation and the characteristic hummingbird sign. Using mid-sagittal plain MRI, the hummingbird sign was demonstrated in all progressive supranuclear palsy patients but was negative in Parkinson disease and controls. Hummingbird sign in progressive supranuclear palsy patients was due to atrophy of the midbrain and relative lengthening of the interpeduncular fossa and preserved pons. Hummingbird sign on MRI has a sensitivity of around 100%. Hummingbird sign on MRI can be a useful diagnostic clue for physicians for confirmation of diagnosis of progressive supranuclear palsy and can help to differentiate from Parkinson disease, which is a treatable neurological disorder.

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