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Original Research Article

Tremor beyond Parkinson's disease: role of magnetic resonance imaging

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Abstract

Multiple system atrophy and progressive supranuclear palsy cause tremor, movement disorders and cognitive impairment, are classified as Parkinson-plus syndrome. Magnetic resonance imaging has proved to be a useful investigation in distinguishing benign tremulous Parkinson's disease from atypical tremor syndromes. In this article, we used two case vignettes to illustrate how MR imaging findings can assist in the diagnosis of tremor and various other movement disorders and can change the treatment in some cases. MRI showed selective atrophy of midbrain in progressive supranuclear palsy giving 'hummingbird' appearance and distinctive patterns of atrophy of brainstem and cerebellum with signal changes in pons such as 'hot cross bun' in multiple system atrophy-cerebellar subtype. Thus, neuroimaging gives insight into the pathophysiology of tremor indicating the involvement of the brainstem and cerebellum as the most consistent finding in patients presenting with parkinson-plus syndrome.

Keywords: Tremor; parkinson-plus syndrome; progressive supranuclear palsy; multisystem atrophy-C; hummingbird sign; hot-cross-bun sign

1. Introduction

Movement disorders are the neurological conditions that affect the speed, fluency, quality, and central control of movement and are characterized by either an excess of movement or a paucity of voluntary and automatic movements^[1]. The disease spectrum includes Parkinson's disease and Parkinson-plus syndrome. In Parkinson's disease, conventional MRI may not show any specific finding while in other degenerative forms of parkinsonism, including progressive supranuclear palsy (PSP), multisystem atrophy (MSA), MRI reveals characteristic patterns of regional atrophy combined with signal changes in the basal ganglia, pons, middle and superior cerebellar peduncles^[2]. We report two cases of rare neurodegenerative conditions, i.e. one case each of PSP and MSA-C (cerebellar type). This article will be focusing on the spectrum of variable movement disorders on MRI where tremor and progressive cognitive impairment are the predominant manifestations.

2. Case Reports

A) Case 1

A 55-year-old man had a 4-year history of vertigo, recurrent backward falls while sitting along with onset of abnormal limb movements and resting tremor 1 year back. Symptoms were gradual in onset and progressive. Neurologic examination revealed an expressionless face, absence of blinking of eyes, staring gaze, slurred speech and imbalance. No response to anti-Parkinson drugs was noted. MR imaging of the brain showed atrophy of the midbrain with flattening of its superior surface giving 'Hummingbird' appearance on sagittal T2-weighted images [Figure 1]. Pons was spared. Midbrain to pons area ratio was 0.25 which was reduced [cut-off is 0.52]. Reduction in the AP diameter of the midbrain with thinning of the cerebral peduncles on axial T2W image was noted resembling the 'Mickey mouse.' Focal punctate tegmental hyperintensity was noted in the midbrain on left side [Figure 2]. No altered signal intensity area was seen in putamen. Diffuse cerebral and cerebellar atrophy was also evident.

Based on above clinical and MRI findings, the patient was diagnosed as a case of Progressive supranuclear palsy.



Fig 1: T2 W sagittal image of the brain showing the selective atrophy of midbrain tegmentum with preservation of pons resulting in its flattening forming the silhouette of the head of the 'Hummingbird' [Red arrow]. Also noted was diffuse cerebellar atrophy with a small calcified granuloma in cerebellum as an incidental finding.

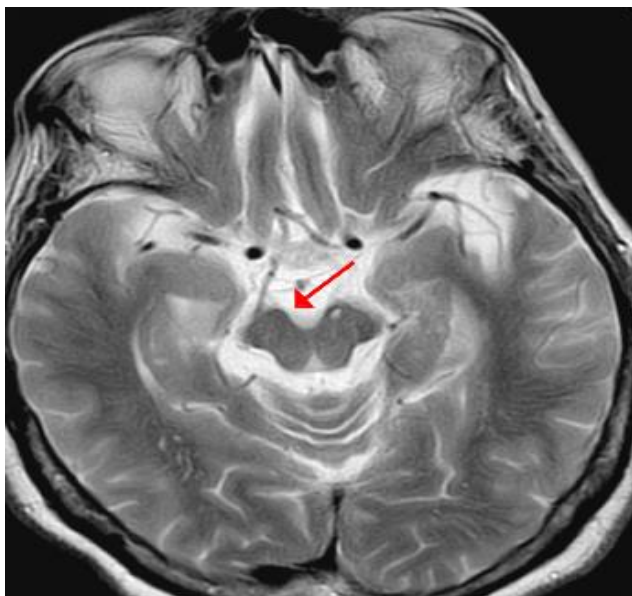


Fig 2: T2-weighted axial image of the brain showing atrophy of midbrain tegmentum and thinning of cerebral peduncles resulting in concavity at the lateral margin of the midbrain, resembling the 'Mickey mouse.' [Red arrow]. Focal punctate hyperintensity was also noted in the tegmentum on left side.

B) Case 2

A 64-year-old male presented with one year history of progressive cognitive impairment. The patient's memory and ability of judgment had been impaired for 6 months. He had impairments in retention and retrieval of verbal memory and retrieval of visual memory. In addition, he had suffered from the tremor, recurrent falls with postural imbalance, and difficulties in swallowing and articulation. Neurological examination revealed mild dysarthria and mild dysphagia.

Autonomic function tests revealed significant postural hypotension, mild detrusor over activity with significant residual urine.

MRI brain showed disproportionate atrophy of midbrain & crura cerebri [Figure 3]. High signal intensity in cerebral peduncles and pons on axial T2 W images giving the classical 'hot cross bun sign' suggested the diagnosis of Multisystem Atrophy-C [Figure 4].

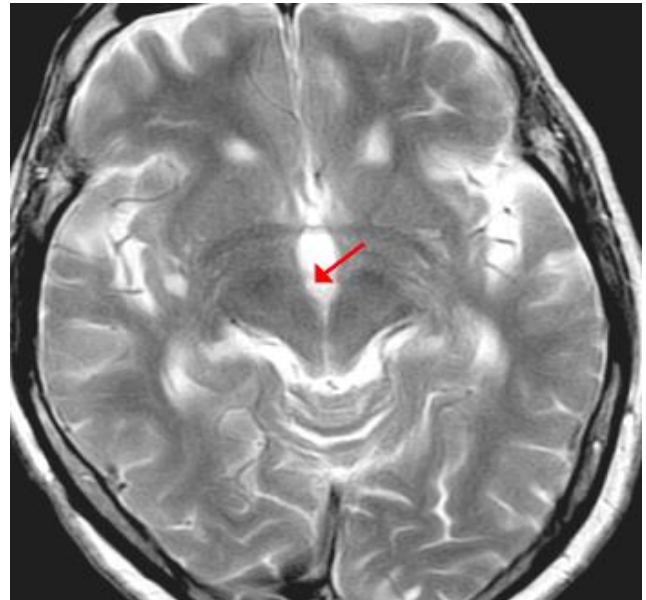


Fig 3: Axial T2-weighted MR image at the level of the midbrain showed atrophy of the midbrain, bilateral cerebral crura and mild diffuse cortical atrophy. [Red arrow]

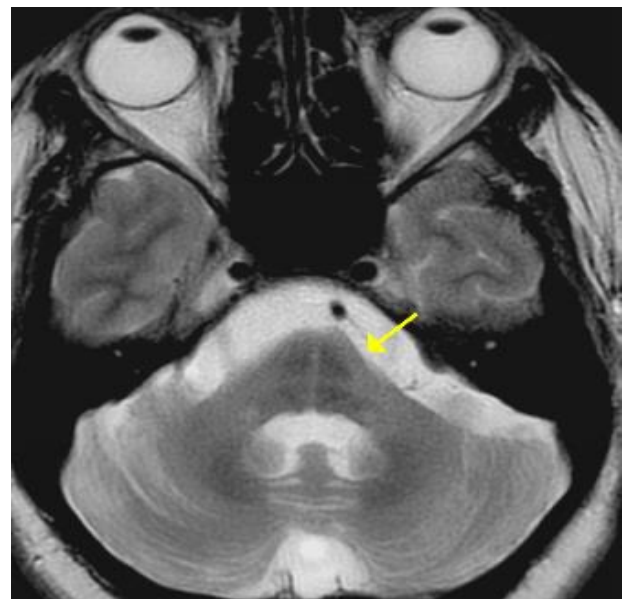


Fig 4: Axial T2-weighted MR image of the brain showed the 'hot cross bun sign' as a cruciform hyperintensity in an atrophied pons [Yellow arrow]. Cerebellum and middle cerebellar peduncles were also atrophied.

3. Discussion

Progressive supranuclear palsy (PSP) is a neurodegenerative tauopathy characterized by parkinsonism like features, tremor, vertical gaze palsy and early falls. It is a disorder of middle or late age, affecting both men and women almost equally after the sixth decade of life, with a prevalence rate of 5-6 per 100,000 and median survival of 7-12 years after

diagnosis [3]. The clinical research criteria given by the National Institute of Neurological Disorders and Stroke for the diagnosis of PSP was used to make a diagnosis in our case [4]. Many clinical features overlap with the Parkinson's disease and differentiation on clinical grounds is difficult.

Classical MRI features of PSP include selective atrophy of midbrain tomentum with relatively preserved pons, gives an appearance of the 'Hummingbird' or the 'King Penguin' on sagittal T2W images [5]. (The hummingbird sign, in which usual upwardly convex outline of the superior aspect of the midbrain is flattened or concave, also well seen in our case. This sign is reported to have a sensitivity of nearly 100% in diagnosing PSP [5].

The reduced midbrain to pons ratio, as in our case was 0.25 [cut-off is 0.52], has been found to have 100% specificity in confirming PSP [6].

Reduction in the AP diameter of the midbrain with thinning of cerebral peduncles on axial T2 MR image giving appearance of the 'Mickey mouse' is very sensitive in establishing the diagnosis of PSP [7]. Other imaging findings in PSP that may be seen are third ventricle dilation, periaqueductal T2 hyper intensities, elevated apparent diffusion coefficient in the putamen, globus pallidus and caudate nucleus. Atrophy of the superior cerebellar peduncles was reported to be an additional distinctive feature of PSP that seems to be related to heavy deposits of tau-positive inclusions in the dentatorubrothalamic WM tracts with subsequent axonal and myelin loss [8].

Multiple system atrophy (MSA) is defined as an adult-onset, sporadic, rapidly progressive, multisystem, neurodegenerative disease of undetermined etiology, characterized clinically by autonomic dysfunction, varying severity of parkinsonian features, cerebellar and urogenital dysfunction, and corticospinal disorders. The incidence rate is about 0.6 cases per 100,000 persons per year [9].

MSA is characterized by the progressive loss of neuronal and oligodendroglial cells in the striatonigral and olivopontocerebellar structures of the brain and spinal cord. American Autonomic Society and American Academy of Neurology in 2007 categorized MSA in MSA-P with predominant parkinsonism and MSA-C with dominant cerebellar features (MSA-C) [10]. The disease manifests in the 4th to 5th decade, and shows fast progression.

In patients with MSA-C, Cerebral MRI reveals mild to moderate cortical atrophy, with disproportionate atrophy of pons and cerebellum. T2/PD/FLAIR sequences give the classical 'hot-cross bun' sign in the pons, characterized by cruciate hyperintensity secondary to atrophy of the transverse pontine fibers, which was also well appreciated in our case [11]. Hyper intensities in the pons, middle cerebellar peduncles, and cerebellum, corresponding to pontocerebellar tract atrophy on T2-weighted images, have been described as characteristic findings on conventional MR images.

The 'putaminal rim sign, also known as the putaminal slit sign, is a relatively specific sign of multiple system atrophy-parkinsonism (MSA-P), and refers to a linear region of high T2 signal surrounding the lateral aspect of the putamen at 1.5T MRI. Importantly this appearance can be seen in normal individuals at 3T MRI [12]. Absence of 'putaminal rim sign' at 1.5 T MR in our cases helped us to narrow our diagnosis to MSA-C.

The conventional MR imaging findings in our cases were typical for PSP and MSA-C.

4. Conclusion

These case reports illustrate how MR imaging can be an indispensable tool in the diagnosis of tremor and other movement disorders induced by the various neurodegenerative conditions. MRI helps in diagnosing and differentiating the various types of MSA. The typical neuroimaging findings in PSP, MSA-C, MSA-P, may be the first diagnostic clue and if, taken in the context of the clinical picture can point the way to correct diagnosis.

5. References

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6. Abbreviations

MRI	: Magnetic resonance imaging
PSP	: Progressive supranuclear palsy
MSA	: Multiple system atrophy
MSA-C	: Multiple system atrophy- Cerebellar
MSA-P	: Multiple system atrophy- Parkinsonism
PD	: Proton density
FLAIR	: Fluid-Attenuated Inversion Recovery