Open Access Original Research Article

MedPeer Publisher
Abbreviated Key Title: MedPeer
ISSN : 3066-2737
homepage: https://www.medpeerpublishers.com

Progressive Supranuclear Palsy: The Hummingbird and Morning Glory Signs on MRI.

DOI: 10.70780/medpeer.000QGW1

AUTHOR AND AFFILIATION

Kamal Hlioui [1] , Fatma El mabrouk[1], Firdaous Touarsa[1] , Meriem Fikri [1].

Department of neuroradiology , Ibn sina university hospital , university Mohammed V of Rabat , Morocco

Corresponding author: Kamal Hlioui

ABSTRACT

Progressive supranuclear palsy (PSP) is an uncommon neurodegenerative condition marked by a blend of motor, cognitive, and oculomotor dysfunctions. Brain magnetic resonance imaging (MRI) can offer significant supportive insights, especially through the observation of pronounced midbrain atrophy while the pons remains relatively intact. We describe the case of a 62-year-old man who exhibited a gradual decline in psychomotor speed and difficulties with vertical eye movement. Imaging of the brain revealed significant atrophy of the midbrain, with the pons showing relative preservation. On midsagittal T1-weighted scans, the inward curvature of the superior midbrain contour revealed the distinctive “hummingbird sign,” whereas axial images demonstrated midbrain atrophy with a concave appearance of the lateral edges, aligning with the “morning glory sign.” The alignment of these specific MRI characteristics with the patient’s clinical symptoms strongly reinforced the diagnosis of PSP. This case underscores the importance of identifying typical morphological changes in the midbrain on standard MRI scans for patients suspected of having PSP.

KEYWORDS

Progressive supranuclear palsy; Hummingbird sign; Morning glory sign; Midbrain atrophy; Magnetic resonance imaging; Vertical gaze palsy

MAIN ARTICLE

INTRODUCTION:

Progressive supranuclear palsy (PSP) is a rare, progressive neurodegenerative disorder belonging to the spectrum of atypical parkinsonian syndromes. It is a four-repeat tauopathy characterized by the accumulation of abnormal tau protein in several cortical and subcortical structures, particularly the basal ganglia and brainstem. Clinically, PSP encompasses a heterogeneous spectrum of manifestations, with vertical supranuclear gaze palsy representing one of its most characteristic neurological features. Cognitive and behavioral changes, akinesia, axial rigidity, and postural instability may also occur during the course of the disease [1].

Although the diagnosis of PSP remains primarily clinical, magnetic resonance imaging (MRI) provides valuable supportive findings and may help distinguish PSP from other neurodegenerative parkinsonian disorders. Predominant midbrain atrophy with relative preservation of the pons is one of the most characteristic imaging features. On midsagittal MRI, selective atrophy of the rostral midbrain results in flattening or concavity of its superior contour, producing the characteristic “hummingbird sign,” also known as the “penguin silhouette sign” [2,3].

CASE PRESENTATION:

A 62-year-old man presented with progressive psychomotor slowing associated with the development of vertical gaze impairment. Brain magnetic resonance imaging (MRI) was performed as part of the diagnostic work-up.

MRI demonstrated predominant midbrain atrophy with relative preservation of the pons. On midsagittal T1-weighted images, atrophy of the rostral midbrain with concavity of its superior contour produced the characteristic “hummingbird sign” (Figure 1A). Axial T1-weighted images showed marked midbrain atrophy with concavity of its lateral margins, producing the “morning glory sign” (Figure 1B). These characteristic imaging findings, in association with vertical gaze impairment, were highly suggestive of progressive supranuclear palsy.

In the appropriate clinical context, particularly the presence of vertical gaze impairment, these characteristic MRI findings were highly suggestive of progressive supranuclear palsy.

DISCUSSION

Progressive supranuclear palsy is a neurodegenerative tauopathy characterized by considerable clinical heterogeneity. The classic phenotype, historically referred to as Richardson syndrome, is predominantly characterized by early postural instability, axial rigidity, cognitive dysfunction, and supranuclear ocular motor abnormalities. Vertical supranuclear gaze palsy, particularly affecting downward gaze, is one of the most distinctive clinical manifestations and represents a major diagnostic feature in current clinical criteria [1]. In our patient, the progressive psychomotor slowing associated with vertical gaze impairment provided a clinical context strongly suggestive of PSP.

MRI plays an important supportive role in the diagnostic evaluation of PSP, particularly by demonstrating the characteristic pattern of brainstem atrophy. The most recognized finding is disproportionate atrophy of the midbrain with relative preservation of the pons. On midsagittal images, atrophy of the rostral midbrain causes flattening or concavity of its superior contour while the pons remains relatively preserved, resulting in the characteristic hummingbird sign, also known as the penguin silhouette sign [2,3]. This characteristic morphology was clearly demonstrated in our patient.

Midbrain atrophy can also be appreciated on axial images. Concavity of the lateral margins of the midbrain tegmentum produces the morning glory sign. Adachi et al. described this finding in four of five patients with PSP and demonstrated its close association with vertical supranuclear gaze palsy [4]. In our case, axial MRI similarly demonstrated marked midbrain atrophy with concavity of its lateral contours, complementing the characteristic sagittal findings.

Although these qualitative signs are highly suggestive of PSP, their absence does not exclude the diagnosis, and visual assessment may be influenced by imaging technique, slice thickness, and disease stage. Therefore, quantitative MRI measurements have been developed to improve diagnostic accuracy. These include measurement of the midbrain area, the midbrain-to-pons area ratio, and the Magnetic Resonance Parkinsonism Index (MRPI). The MRPI combines the pons-to-midbrain area ratio with measurements of the middle and

superior cerebellar peduncles and has shown utility in differentiating PSP from Parkinson disease and the parkinsonian variant of multiple system atrophy [5]. More recently, MRPI 2.0, which incorporates measurements related to third ventricular enlargement, has further improved discrimination between PSP-parkinsonism and Parkinson disease, particularly at earlier disease stages [6].

The main differential diagnoses include idiopathic Parkinson disease, multiple system atrophy, and corticobasal degeneration. Conventional MRI may be normal or show nonspecific changes in Parkinson disease, whereas multiple system atrophy may demonstrate predominant pontocerebellar or putaminal abnormalities. In PSP, the disproportionate involvement of the midbrain, particularly when associated with characteristic ocular motor abnormalities, strongly favors the diagnosis. Nevertheless, the hummingbird and morning glory signs should be regarded as supportive rather than isolated diagnostic findings, and their interpretation must always be integrated with the clinical presentation.

Our case illustrates the strong clinicoradiological correlation between progressive vertical gaze impairment and selective midbrain atrophy. Recognition of the hummingbird sign on sagittal MRI, together with midbrain atrophy and the morning glory appearance on axial images, can provide an important diagnostic clue and facilitate recognition of PSP in an appropriate clinical setting.

CONCLUSION

Progressive supranuclear palsy is a challenging neurodegenerative disorder in which MRI provides valuable supportive diagnostic findings. Selective midbrain atrophy with relative preservation of the pons produces the characteristic **hummingbird sign** on sagittal images, while axial midbrain atrophy may result in the **morning glory sign**. Recognition of these characteristic imaging features, particularly in patients presenting with vertical gaze impairment, can strongly support the diagnosis of PSP and help differentiate it from other atypical parkinsonian syndromes.

FIGURES

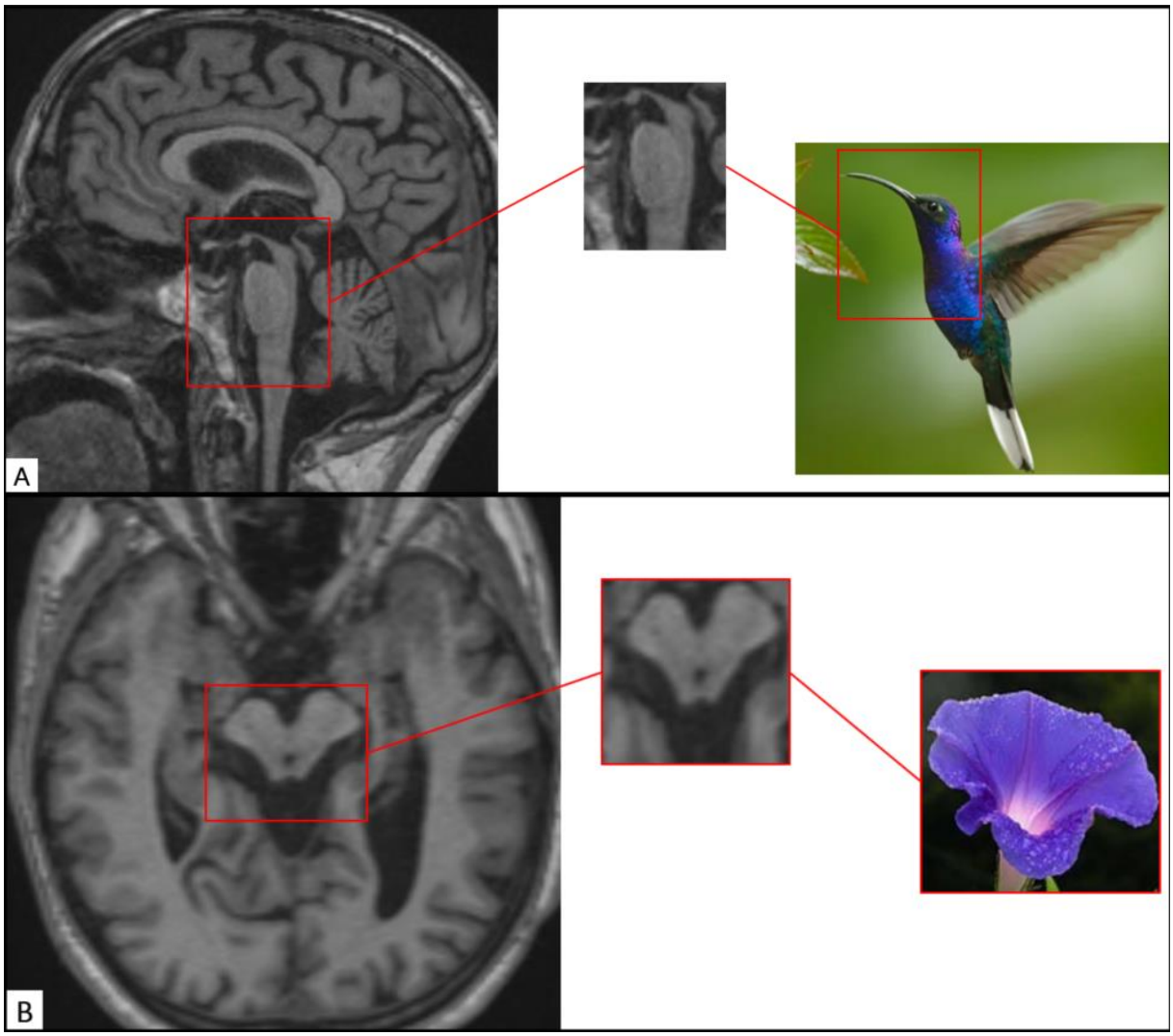

Figure 1. *Characteristic MRI findings of progressive supranuclear palsy. (A) Midsagittal T1-weighted MR image demonstrates marked atrophy of the rostral midbrain with concavity of its superior contour and relative preservation of the pons, producing the characteristic "hummingbird sign." (B) Axial T1-weighted MR image shows midbrain atrophy with concavity of its lateral margins, consistent with the "morning glory sign."*

ACKNOWLEDGEMENTS

Competing interests

The authors declare no competing interests.

Authors' contributions

Dr Fatma El mabrouk contributed to the conception and design of the study, image interpretation, literature review, manuscript drafting, and final approval of the submitted version.

Acknowledgements

The authors would like to thank the Department of Neuroradiology for its support in the diagnosis and management of this case.

REFERENCES

[1] Höglinger GU, Respondek G, Stamelou M, et al. Clinical diagnosis of progressive supranuclear palsy: The Movement Disorder Society criteria. Mov Disord. 2017;32(6):853-864. doi:10.1002/mds.26987.
https://doi.org/10.1002/mds.26987

[2] Kato N, Arai K, Hattori T. Study of the rostral midbrain atrophy in progressive supranuclear palsy. J Neurol Sci. 2003;210(1-2):57-60. doi:10.1016/S0022-510X(03)00014-5.
https://doi.org/10.1016/S0022-510X(03)00014-5

[3] Gröschel K, Kastrup A, Litvan I, Schulz JB. Penguins and hummingbirds: Midbrain atrophy in progressive supranuclear palsy. Neurology. 2006;66(6):949-950. doi:10.1212/01.wnl.0000203342.77115.bf.
https://doi.org/10.1212/01.wnl.0000203342.77115.bf

[4] Adachi M, Kawanami T, Ohshima H, Sugai Y, Hosoya T. Morning glory sign: A particular MR finding in progressive supranuclear palsy. Magn Reson Med Sci. 2004;3(3):125-132. doi:10.2463/mrms.3.125.
https://doi.org/10.2463/mrms.3.125

[5] Quattrone A, Nicoletti G, Messina D, et al. MR imaging index for differentiation of progressive supranuclear palsy from Parkinson disease and the Parkinson variant of multiple system atrophy. Radiology. 2008;246(1):214-221. doi:10.1148/radiol.2453061703.
https://doi.org/10.1148/radiol.2453061703

[6] Quattrone A, Morelli M, Nigro S, et al. A new MR imaging index for differentiation of progressive supranuclear palsy-parkinsonism from Parkinson's disease. Parkinsonism Relat Disord. 2018;54:3-8. doi:10.1016/j.parkreldis.2018.07.016.
https://doi.org/10.1016/j.parkreldis.2018.07.016