Austin Journal of Radiology

Open Access

Austin Publishing Group

Case Report

# Radiological Manifestations of Progressive Supranuclear Palsy

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**Received:** November 13, 2021; **Accepted:** December 09, 2021; **Published:** December 16, 2021

**Abstract**

Progressive supranuclear palsy is one of the causes of atypical parkinsonian syndrome.

We present the case of an 83-year-old patient with typical symptomatology suggesting progressive supranuclear palsy with characteristic imaging.

**Keywords:** Supranuclear palsy; Radiological aspect; Morocco

## Abbreviations

MRI: Magnetic Resonance Imaging; SPECT: Single-Photon Emission Computed Tomography; PET: Tomography Positron Emission; 18FDG: Fluorodeoxyglucose; 99mTc: Technetium-99m

## Introduction

Progressive supra nuclear palsy is one of the neurodegenerative disorders of the central nervous system responsible for atypical parkinsonian syndromes.

It occurs in the sixth decade and is related to a phosphorylation disorder of the Tau protein, which accumulates in the basal ganglia and brainstem [1].

Imaging through MRI looks for characteristic abnormalities including the colibri and glory morning sign [2], which support the diagnosis of this condition in the face of clinical suspicion.

## Observation

The patient an 83-year-old male, who was being followed for benign prostatic hypertrophy, presented an atypical parkinsonian syndrome with vertical oculomotor disorder. Brain MRI without injection performed to explore this syndrome, found rostral atrophy of the midbrain with flattening of its roof realizing the classic hummingbird or penguin sign on sagittal sections and bindweed <<morning glory sign>> on axial sections. We also noted an enlargement of the interpeduncular cistern with cerebral peduncles in the shape of mickey ears, which is the sign of <<Mickey Mouse>>.

## Discussion

Progressive supranuclear palsy is a tauopathy revealed in the majority of cases following an atypical parkinsonian syndrome. It has a large phenotypic polymorphism depending on the degree of tau protein accumulation. The most common for mis the one described by Richardson and which associates supranuclear gaze palsy, progressive axial rigidity, pseudobulbar palsy and mild dementia [3].

It occurs in the sixth decade and mostly in males, as shown by E. jabbari et al. [4] who found a median age of 68 years in a study of 222 patients with 101 with progressive supranuclear palsy.

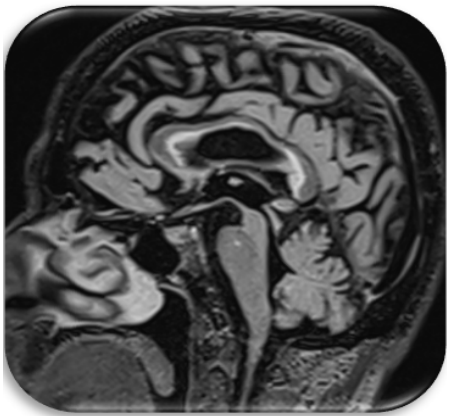

**Figure 1:** Sagittal section in FlAIR showing the hummingbird sign.

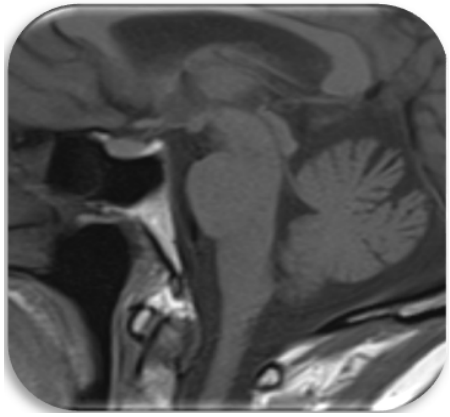

**Figure 2:** Sagittal section in T1 for a normal patient.

From the physiopathological point of view, this affection is linked to the accumulation of a protein associated with microtubules. The tau is known to be found in the subthalamic nucleus, the pallidum, the striatum, the red nucleus, the substantia nigra, the tegmentum ponticis, the oculomotor nucleus, the medulla and the dentate nucleus. It is consecutive to the hyperphosphorylation of this protein,




**Citation:** Diallo ID, EZ-Zaky S, Traore WM, El Kettani NE, Fikri M, Jiddane M, et al. Radiological Manifestations of Progressive Supranuclear Palsy. Austin J Radiol. 2021; 8(13): 1182.

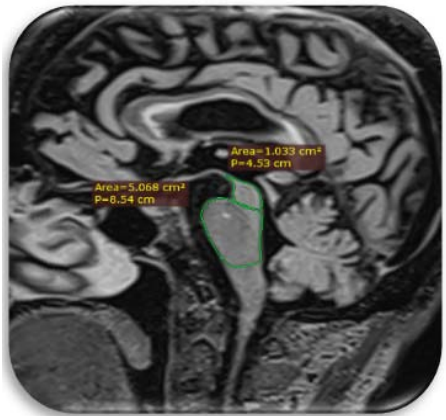


**Figure 3:** Sagittal section in FLAIR showing the mesencephalic atrophy.

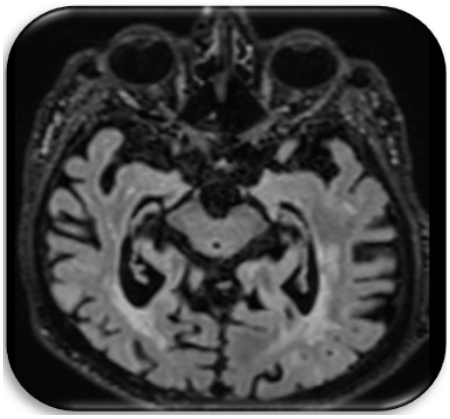

**Figure 4:** Axial section in FLAIR showing the glory morning sign and Mickey Mouse sign.

which thus loses affinity towards the microtubule becoming resistant to proteolysis [5].

Morphological (MRI) and functional (isotopic) imaging show suggestive but non-specific abnormalities. As such, mesencephalic atrophy is found in this condition to be responsible for a hummingbird-like aspect of the midbrain and/or penguin-like aspect of the brainstem on sagittal sections related to flattening of the roof of the midbrain. It is also associated with, the morning glory sign, which is the concave aspect of the posterolateral faces of the midbrain, and a Mickey ears aspect, which is related to tegmental atrophy contrasting with a relative preservation of the tectum and cerebral peduncles on axial sections [3,6].

More specifically, the measurement of the mesencephalic surface (>110mm$^2$), of the mesencephalon/bridge ratio (>0.21) and of the AP diameter of the mesencephalon (>14mm) allows to appreciate this atrophy [7]. Thus, in our patient the mesencephalic surface was 103mm$^2$, confirming this atrophy.

The discriminating functional abnormalities in progressive supranuclear palsy are localized perfusion disorders, namely hypoperfusion in the anterior cingulate, fronto-mesial and prefrontal cortex, which can be well demonstrated by Single-Photon Emission Computed Tomography (SPECT) with 99mTc. 18FDG Positron Emission Tomography (PET) appreciates the consumption of glucose in the hypoperfusion territories, including deep structures, notably the caudate nucleus [8].

There is no curative treatment yet, but several promising neuroprotective treatments are currently being tested [9]. The management of this disease is therefore essentially symptomatic.

## Conclusion

Progressive supranuclear palsy is a pathology of the elderly male subject not to be confused with Parkinson's disease where the parkinsonian syndrome is typical and therefore a good response to L-DOPA. The manifestations on imaging are essentially the signs of hummingbird and morning glory. Its management is essentially symptomatic.

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