

A Rare Case of Asymmetric Progressive Supra Nuclear Palsy Diagnosed *In vivo* with Magnetic Resonance/Positron Emission Tomography

Abstract

PSP and CBD are usually multi system sporadic disorders characterized by tau inclusions in neurons and glia. The clinical and neuroimaging features are different. However in some cases overlapping of features are noted. Here we present a case of a 65 years old female patient, presenting a 3 years history of insidious onset of asymmetric right upper and lower limb dystonia, followed by slowness, falls and injuries to the back, Parkinsonism, urinary incontinence and cognitive dysfunction and upward gaze palsy. MRI findings were suggestive of moderate cerebral and cerebellar atrophy with prominent ventricular system, reduced antero-posterior midline midbrain diameter, at the level of superior colliculus on axial imaging (morning glory sign was positive) on the left side. PET showed asymmetric hypo metabolism noted in the left superior and middle frontal gyrus, superior temporal and mid temporal gyrus in addition to striatum and thalamus, as well as midbrain, pons and right cerebellar hemisphere. Overall MR/PET was suggestive of unilateral PSP (left) and it corroborated with clinical history of unilateral dystonia and supranuclear gaze palsy. Based on MRI the differential considered was also CBD, but PET showed metabolic activity in the motor cortex. Additionally based on the hummingbird sign and morning glory sign a rare diagnosis of unilateral PSP could be made which also corroborated with the clinical picture. The case report emphasizes the utility of PETMRI simultaneously in situations like these to pick atypical variants or cases with overlapping pathology to reach a diagnosis with *in vivo* imaging methods.

Keywords: Corticobasal degeneration, F18-fluorodeoxyglucose, progressive supranuclear palsy

Progressive supranuclear palsy (PSP) and corticobasal degeneration (CBD) are usually multisystem sporadic disorders characterized by tau inclusions in neurons and glia.^[1] PSP is characterized by progressive axial rigidity, vertical supranuclear gaze palsy, frontal lobe cognitive decline, and balance issues.^[2] The classic clinical syndrome of CBD is the presentation of an apraxic, dystonic, and rigid limb with asymmetrical cortical signs and distal myoclonus known as CBD.^[3] As noted clinically, PSP predominantly presents with a supranuclear vertical gaze palsy and early postural instability with falls.^[4] Clinical overlap makes it difficult for clinicians to diagnose these properly, and biopsy to characterize the site of tau inclusions is not always feasible.^[5] Here, we present the case of a 65-year-old female patient, presenting a 3-year history of insidious onset of asymmetric right upper and lower limb dystonia, followed by slowness, falls and

injuries to the back, Parkinsonism, urinary incontinence, and cognitive dysfunction and upward gaze palsy.

Magnetic resonance/positron emission tomography (MR/PET) using F18-fluorodeoxyglucose was done with the following clinical history.

MR imaging (MRI) findings [Figure 1a and b] were suggestive of moderate cerebral and cerebellar atrophy with prominent ventricular system, reduced antero-posterior midline midbrain diameter, at the level of superior colliculus on axial imaging (morning glory sign was positive) on the left side as seen typically.^[6]

PET showed asymmetric hypo metabolism noted in the left superior and middle frontal gyrus [Figure 1e and f], superior temporal and middle temporal gyrus in addition to striatum and thalamus, as well as midbrain, pons, and right cerebellar hemisphere [Figure 1c and d].

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**Keerti Sitani,
Sandhya Mangalore,
Nitish Kamble,
Pramod Kumar Pal,
Vikram Venkayappa
Holla**

*NIIR (Neuroimaging and
Interventional Radiology),
National Institute of Mental
Health and Neuro Sciences,
Bengaluru, Karnataka, India*

Address for correspondence:
Dr. Keerti Sitani,
National Institute of Mental
Health and Neuro Sciences,
Bengaluru, Karnataka, India.
E-mail: keerti.sitani@gmail.com

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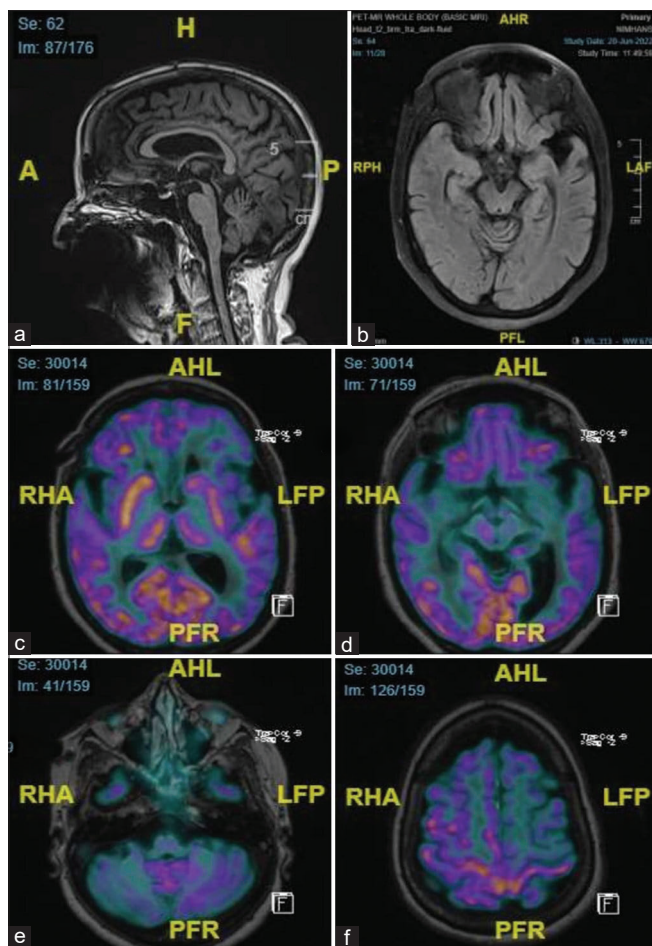


Figure 1: Head T1 MPRAGE sagittal showing the humming bird sign with midbrain atrophy. (b) Head T2 dark fluid transverse showing left side midbrain atrophy. (c) ^{18}F -FDG MR/PET axial fused showing hypometabolism involving the left thalamus, striatum, frontal, and temporal lobes. (d) ^{18}F -FDG MR/PET axial fused showing hypometabolism involving the midbrain left side. (e) ^{18}F -FDG MR/PET axial fused showing hypometabolism involving the right cerebellum. (f) ^{18}F -FDG MR/PET axial fused showing maintained left perirolandic region. ^{18}F -FDG MR/PET: F18-fluorodeoxyglucose magnetic resonance/positron emission tomography

Overall MR/PET was suggestive of asymmetric PSP (left) and it corroborated with clinical history of unilateral dystonia and supranuclear gaze palsy.

Based on MRI, the differential considered was also CBD, but PET showed metabolic activity in the motor cortex. In addition, based on the hummingbird sign and morning glory sign, a rare diagnosis of unilateral PSP could be made which also corroborated with the clinical picture. The case

report emphasizes the utility of PETMRI simultaneously in situations like these to pick atypical variants or cases with overlapping pathology to reach a diagnosis with *in vivo* imaging methods.

Ethical approval

This article does not contain any studies with animals performed by any of the authors.

Informed consent

Informed consent was obtained from all the individual participants included in the study.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given her consent for her images and other clinical information to be reported in the journal. The patient understands that name and initials will not be published and due efforts will be made to conceal identity, but anonymity cannot be guaranteed.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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