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Challenges in Diagnosing Parkinson's Disease vs. Multiple System Atrophy: A Case Report Highlighting MRI as a Key Diagnostic Tool

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ABSTRACT

We report a case that outlines the diagnostic challenge faced in differentiating between Parkinson's disease (PD) and multiple system atrophy (MSA) in a 58-year-old man with a history of PD. Despite treatment, the patient exhibited limited response and presented with symptoms suggestive of atypical Parkinsonism, prompting a brain MRI. The MRI revealed characteristic features of MSA, showing putaminal atrophy and altered signal intensity. Differential diagnosis between MSA and PD is crucial, with specific MRI findings serving as key differentiators. Early recognition and differentiation are vital for appropriate management.

Keywords: *Atrophy; Magnetic Resonance Imaging; Multiple System Atrophy; Parkinson's Disease*

INTRODUCTION

Multisystem atrophy (MSA) is a rare neurodegenerative disorder classified as a synucleinopathy, marked by abnormal alpha-synuclein deposits. It is divided into MSA-P, with predominant Parkinsonian symptoms, and MSA-C, featuring cerebellar ataxia. Early-stage MSA is often misdiagnosed due to its rarity and overlapping symptoms with Parkinson's disease. Diagnosis relies on clinical criteria but remains challenging, leading to frequent errors. The condition's diverse symptoms and similar disorders further complicate identification. Low early diagnostic sensitivity also limits trial participation for disease-modifying treatments. Thus, brain MRI findings are crucial for confirming MSA.[1,2,7]

CASE REPORT

We present a case concerning a 58-year-old man who presented at the neurology outpatient clinic with complaints of postural imbalance and recurrent falls, particularly when attempting to turn. This patient had been diagnosed with Parkinson's disease five years prior and had been undergoing treatment, with limited response.

The patient displayed a range of signs and symptoms which included tremors, bradykinesia,

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rigidity in the limbs, involuntary movements of the jaw, constipation, slurred speech, and difficulty in looking upward. Additionally, there was a history of speaking and movement during sleep, suggestive of rapid eye movement sleep behavior disorder.

Given the inadequate response to prescribed medications and the observation of upward gaze restriction, the possibility of an atypical Parkinsonism was considered. Consequently, an MRI scan of the brain was done for further evaluation.

The MRI results revealed significant findings. There was a reduction in the size of the putamen and globus pallidus, along with decreased signal intensity observed in the putamen relative to globus pallidus on both susceptibility-weighted imaging (SWI) (Figure 1a) and T2 sequences (Figure 1b). Additionally, a high T2 signal linear rim was detected along the lateral edges of the putamen, with reduced signal intensity on SWI sequences (Figure. 2) The patient also showed diffuse cerebral, cerebellar, and brainstem atrophy without any signal changes.

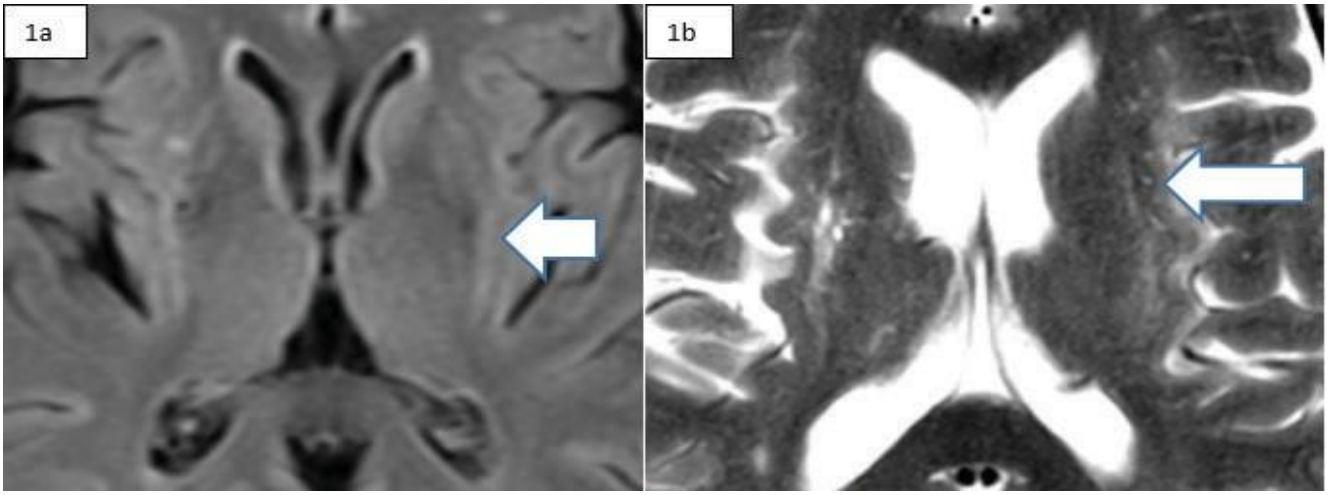

Figure 1: 1a. *Reduction in the size of the putamen and globus pallidus, along with decreased signal intensity observed in the putamen relative to globus pallidus in susceptibility-weighted imaging (SWI) axial section.* ***2a.*** *Arrow showing T2WI hyperintense linear rim detected along the lateral edges of the putamen*

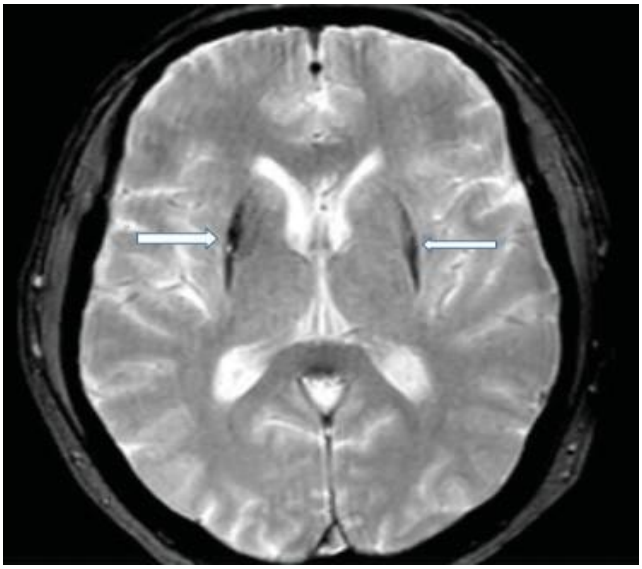

Figure 3: *Arrows showing reduced signal intensity linear rim on SWI sequences along the lateral edges of the putamen*

DISCUSSION

Multisystem atrophy (MSA), an uncommon neurodegenerative disorder, falls under the broad classification of synucleinopathies, a subgroup of neurodegenerative conditions. These disorders involve impairment in the metabolism of alpha-synuclein, resulting in the formation of irregular deposits within the cells. MSA is categorized based on the primary clinical phenotype it presents. MSA characterized primarily by parkinsonian symptoms is labeled MSA-P, while MSA characterized mainly by cerebellar ataxia features is labeled MSA-C.[1,2]

The estimated crude prevalence of MSA ranged from 0.52 - 17 per 100,000 individuals, with a higher occurrence observed in men compared to women (2.75 in men versus 1.19 in women per

100,000 individuals).[3]

Pathologically, MSA is identified by the presence of glial cytoplasmic inclusions and neuronal loss, mainly affecting the olivopontocerebellar systems and striatum. However, discoloration and atrophy of the posterolateral putamen are distinctive characteristics of MSA, especially MSA-P.[4,5]

Parkinsonism, a clinical syndrome is marked by stiffness, tremors, slowed movement, and difficulties with balance. It can caused by idiopathic Parkinson's Disease (PD) or Parkinson-plus syndromes, such as Multiple System Atrophy (MSA), Progressive Supranuclear Palsy (PSP), Corticobasal Degeneration (CBD), and Dementia with Lewy Bodies (DLB).[6]

Diagnosing early-stage MSA and distinguishing it from Parkinson's disease can pose a significant challenge. Given its uncommon occurrence and varied clinical presentation, identifying MSA can be a diagnostic dilemma for physicians. The diagnosis of MSA is primarily clinical and based on the most recent consensus criteria. It has become increasingly apparent that accurately diagnosing MSA based solely on clinical presentation can be challenging, leading to frequent misdiagnoses. The varied clinical manifestations of MSA and the presence of conditions mimicking it, contribute to this difficulty. Due to the initial low sensitivity of MSA diagnosis, many individuals with early-stage MSA are often excluded from clinical trials involving disease-modifying medications. As a result, establishing a definitive clinical diagnosis of MSA typically necessitates identifying specific brain MRI findings indicative of the condition.[4,7]

Due to the complex clinical phenotype, Imaging thus plays a key role in differentiating MSA from Parkinson’s. The most prominent radiological finding indicative of Parkinson's disease is the reduction in thickness of the pars compacta of the substantia nigra, specifically highlighted by the absence of the "swallow-tail sign." This sign refers to the presence of increased signal intensity in the ventrolateral segment of the healthy substantia nigra, flanked by two lines of diminished signal intensity on SWI imaging. With regards to MSA, imaging plays an important role in characterizing and differentiating the MSA-P type from the MSA-C type. In cases of MSA-P type, imaging features on MRI Brain include putaminal atrophy, putaminal hypointensity on T2W image, and hyperintense putaminal rim. On the other hand, in cases of MSA-C type, one can usually appreciate the cruciform linear hyperintensities in the pons (hot-cross bun sign) accompanied by the brainstem and cerebellar atrophy. These imaging features are key to diagnosis.[1,8]

The management of multiple system atrophy (MSA) presents distinct challenges compared to Parkinson's disease (PD) due to its complex symptoms and limited treatment options. While both conditions share sleep disturbances, the approach to treatment may differ, focusing on optimizing PD medications for PD and managing non-motor symptoms for MSA. Overall, MSA requires a comprehensive, multidisciplinary approach due to its unique clinical characteristics as such it is important to confirm the presence of MSA-P in parkinsonism as it helps to tailor management specific to the disease. [9]

CONCLUSION

Diagnosing early-stage MSA is challenging due to its diverse symptoms and frequent misdiagnoses. Clinical criteria alone may be insufficient, making brain MRI crucial for confirmation, with putaminal atrophy and a hyperintense rim indicating MSA-P. Managing MSA-P differs from Parkinson’s, focusing on non-motor symptoms and autonomic dysfunction rather than motor optimization. This case highlights the need for a multidisciplinary approach and the vital role of MRI in ensuring precise diagnosis and targeted treatment.

CONFLICT OF INTEREST

None

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None

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