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Case Report

MULTIPLE SYSTEM ATROPHY CAN PRESENT WITH NON SUPPORTING FEATURES

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ABSTRACT
Multiple system atrophy (MSA), despite being common neurodegenerative disorder, remains a diagnostic challenge for the clinician. Extensive research done in last decade has not only added new facets to the already varied clinical picture of MSA but also argue with the diagnostic criteria in current use. We present this case of MSA with Parkinsonism (MSA-P) because it furthers our belief that full clinical features of MSA remains to be known.

***Keywords*:** *Multiple System Atrophy, Dementia in Multiple System Atrophy, MRI and FDG- PET Findings in Multiple System Atrophy*

INTRODUCTION
The key definition of MSA requires presence of extra pyramidal, pyramidal, cerebellar and autonomic dysfunction in any combination (Quinn, 1989). This is interesting to note that the patients with the variable dysfunction of pyramidal, extrapyramidal, cerebellar and autonomic system have been thought to represent different disease entities and named as OPCA, Shy-Drager syndrome, striatonigral degeneration and more recently MSA. Agessandro *et al.,* (2001) Currently described dysfunction of cognition, anterior horn cells and peripheral nerves in MSA, not only complicates the issue further for the physician facing challenge to consider ante mortem diagnosis of MSA but also disputes with the diagnostic criteria in vogue (Quinn, 1989; Abele *et al.,* 2000; Gilman *et al.,* 2008). Despite availability of literature in abundance diagnosing MSA in an individual patient continues to be a challenging task for the clinicians. Our case, although fulfils second consensus criteria for probable MSA-P (Gilman *et al.,* 2008), shows many additional features not commonly reported in MSA.

CASES
Mr. SB was asymptomatic till the age of 52 years, when he started having unexpected backward falls and problems with gait initiation. Almost simultaneously he noted change in handwriting along with dysarthria. Two years after the onset, appeared urinary incontinence, urgency and impotency, indifferent behaviour and fumbling with money matters. Care giver noted repetitive washing of face, fiddling with electrical switches etc. He, at other institute, was diagnosed and treated initially as Parkinson's disease and later as PSP (Progressive supranuclear palsy), however L-dopa did not alter his clinical condition which deteriorated relentlessly. Seven years from the onset, he presented to this institute, his supine blood pressure was 153/90 mmHg which after 5 minutes of standing fell to139/87 mmHg . There was paucity of facial expression and reduced postural adjustments. His voice was monotonous. He scored 28/30 on MMSE. Addenbrookes dementia scores was 52/100 with markedly impaired visuo spatial abilities (0/16). Vision, pupils and ocular fundii were normal. No KF rings were seen. Corrective phase of OKN (optokinetic nystagmus) on upward gaze was severely impaired. Horizontal gaze evoked nystagmus were present on both sides. Marked increased in axial tone and pronounced rigidity in all four limbs was noted. Sensory examination was normal. Deep tendon reflexes in lower limbs were hyper active, plantars were bilateral mute. There was severe difficulty in initiation of gait with preserved ability of imitating "movements like walking" while lying on bed, suggestive of gait apraxia. Kinesogenic dystonic plantar flexion and inversion of right foot, associated with right upper limb dystonic posturing was seen

intermittently, further making it difficult for him to walk. He fulfilled the clinical criteria for probable MSA-P (Quinn, 1989; Gilman *et al.*, 2008) with few additional non supporting features.

FDG PET hypometabolism was seen in left frontal lobe, bilateral temporal lobes and heads of caudate nuclei (Figure 1*)*. MRI Brain showed bilateral Fronto temporal atrophy with atrophy of caudate heads (Figure 2). Huntington's gene PCR in serum was negative Electromyography revealed renervation-denervation pattern with fasciculation's in tibialis anterior, vastus medialis and external anal sphincter.

DISCUSSION

MSA is commonly encountered by the clinical neurologist. Available diagnostic criteria seem to fall short in taking care of full spectrum of variable manifestations of multisystem degeneration. Our patient fulfils the criteria of probable MSA –P. Review of literature suggest that, presence of non supporting features proposed by Gilmann and colleague (2008) are rather common in MSA. This patient has odd features like eye movement abnormality, cognitive impairment, absence of putaminal atrophy in MRI and presence of fronto-temporal hypo-metabolism in FDG- PET. The involvement of cognitive function has been recently investigated and is reported in 14-16% patients of MSA (Wennin *et al.*, 2000; O'Sullivan *et al.*, 2008). Further, in MSA-P the neuropsychological impairment significantly correlates with prefrontal hypoperfusion (Agessandro *et al.*, 2001). The ocular abnormalities have been described in MSA in-fact presence of moderate hypometria of saccades and impaired VOR suppression are not the exception but clue to the presence of MSA (Anderson *et al.*, 2008). Classically MRI has been described to show putaminal atrophy. Paviour etal have described progressive pontine and cerebellar atrophy on MRI. The atrophy of caudate nucleus has not been described earlier however Rutledge *et al.*, 1987 have reported lower caudate uptake in MSA (Quinn, 1989).

Conclusion

It seems that story of MSA is yet to unfold completely and in subject who otherwise fulfil criteria of MSA, is may not be justifiable to negate the possibility of MSA based on the presence of non supporting features.

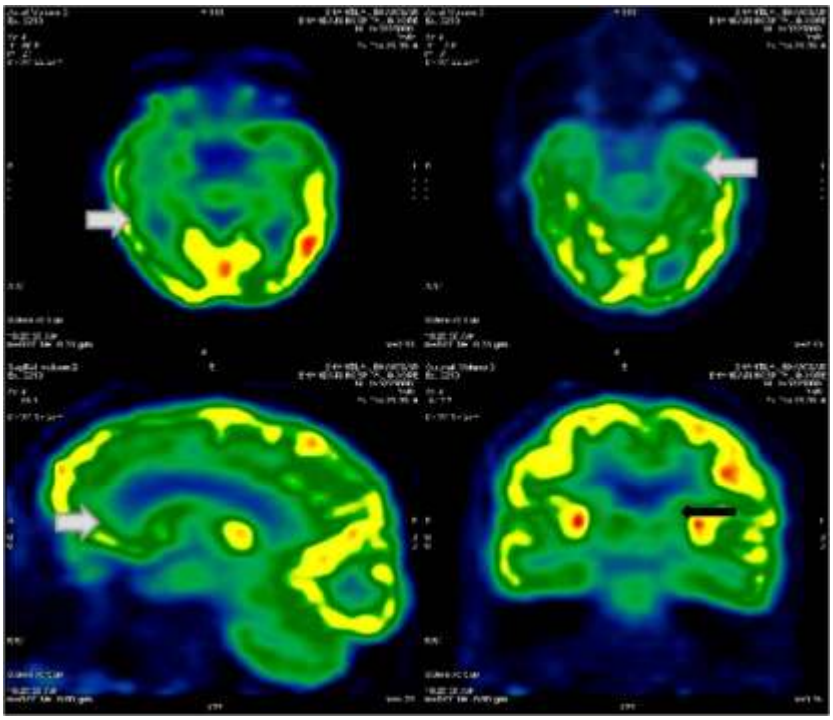

Figure 1: Hypo metabolism in left frontal lobe, bilateral temporal lobes (white arrow) and heads of caudate nuclei (black arrow)

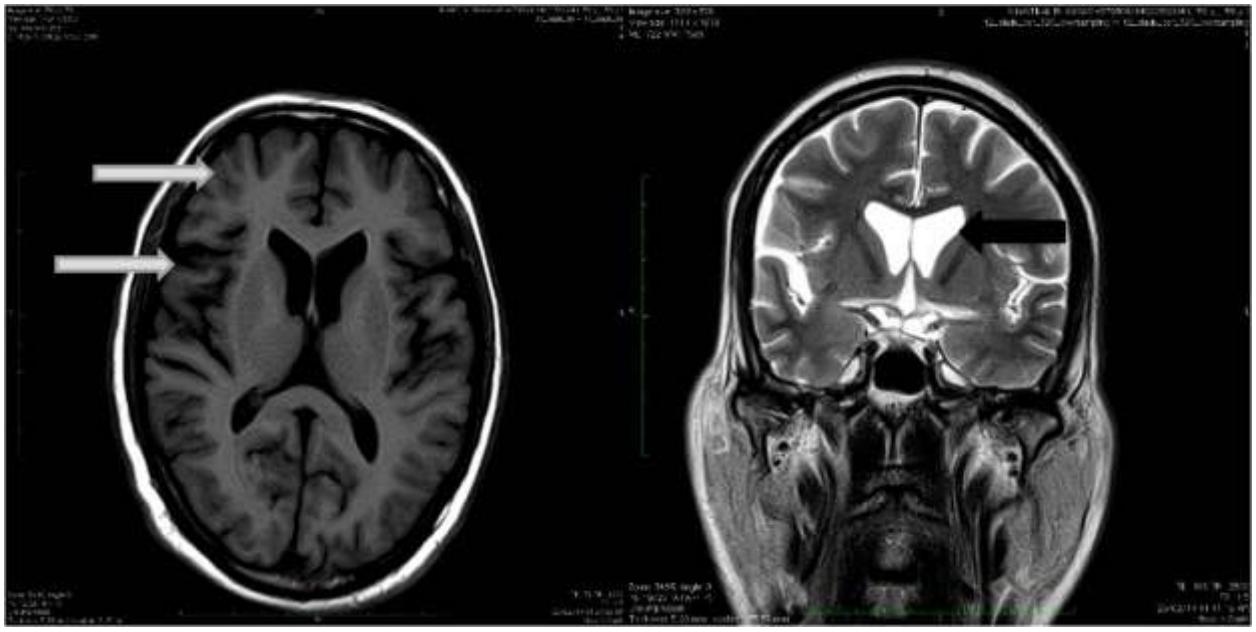

Figure 2: Fronto temporal atrophy with mild atrophy of caudate heads

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