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Case Report

Multiple System Atrophy-Cerebellar Type (MSA-C): A Case Report

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ABSTRACT

Introduction: Multiple system atrophy (MSA) is a sporadic, progressive neurodegenerative disorder has clinical symptoms characterized by variable combination of Parkinsonism disease like symptoms, cerebellar ataxia & / or autonomic failure.
Presentation of Case: We describe one patient having cerebellar signs with MRI features predominant of MSA-C or olivopontocerebellar atrophy showing pontine "Hot cross Bun sign" with having unusual focal hemorrhages in bilateral globus pallidus.
Discussion: Striatonigral degeneration (MSA-P), characterized clinically by parkinsonian symptoms with degenerative changes predominantly affect basal ganglia, particularly the putamen. Shy-Drager syndrome (MSA-SDS) characterized by autonomic nervous system failure with somewhat variable pathologic changes, characterize by neuronal loss in the substantia nigra and the intermediolateral cell column of the spinal cord. Olivopontocerebellar atrophy (OPCA or MSA-C) characterized by predominantly by cerebellar signs, with predominant olivopontocerebellar atrophy.
Conclusion: We mainly draw attention to an uncommon case with predominant MSA-C signs& findings showing pontine "Hot cross Bun sign" having having unusual focal hemorrhages in bilateral globus pallidus.

Key words: Multiple system atrophy; olivopontocerebellar atrophy; Striatonigral degeneration; Shy-Dager Syndrome.

INTRODUCTION

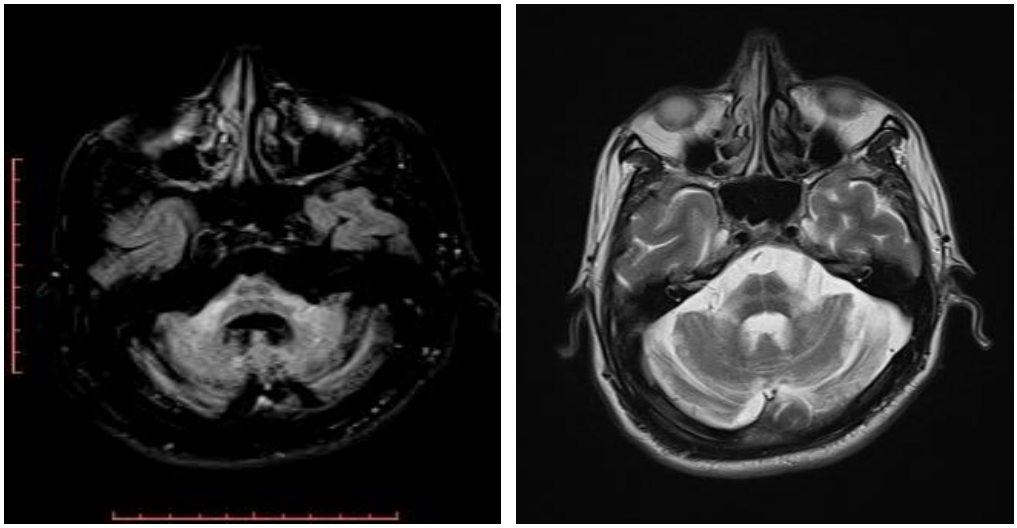
Multiple system atrophy (MSA) is a sporadic, adult onset, progressive neurodegenerative disorder that involves, to varying degrees, the basal ganglia, the olivopontocerebellar complex and the autonomic system. [1,2] The term MSA was first proposed in 1969 by Graham and Oppenheimer based on their observations that Striatonigral degeneration, olivopontocerebellar complex, and Shy-Drager syndrome commonly coexist clinically and pathologically. [3]

MSA-P is synonymous with Striatonigral degeneration when parkinsonism features predominates, with MSA-C when olivopontocerebellar atrophy (OPCA) or cerebellar signs predominates, and MSA –SDS(Shy-Drager syndrome) when autonomic failure is dominant. [1,4-7]

The incidence is 0.6 cases/ 100000/ year and the prevalence ranges from 1.86 to 4.9 cases/100000. [8]

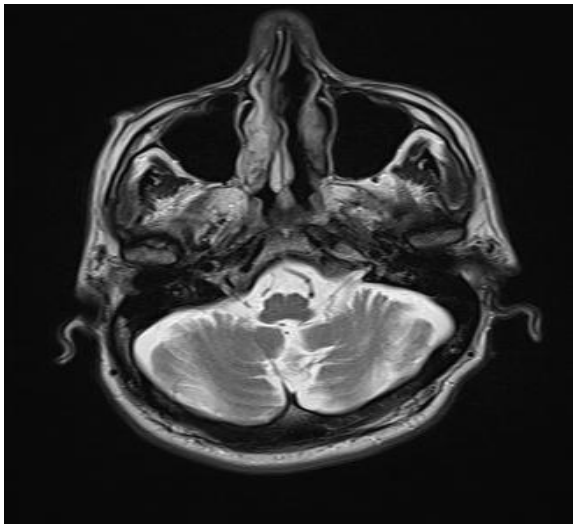
Pathologically, presence of Glial cytoplasmic inclusions containing alpha synuclein clump aggregates is a criterion for definite diagnosis of MSA. [9-11] Alpha synuclein, which is also a major component of the Lewy body, which is the pathologic hallmark lesion in Parkinson disease and dementia with Lewy bodies Recently the term "synucleinopathy" has been proposed to encompass a presumed common

pathogenic process shared by these neurodegenerative disorders. [12,13]



Axial FLAIR **Axial T2 WI**

Figure 1: Cruciform hyperintensity in inferior pons "Hot Cross Bun Sign"



Axial T2WI

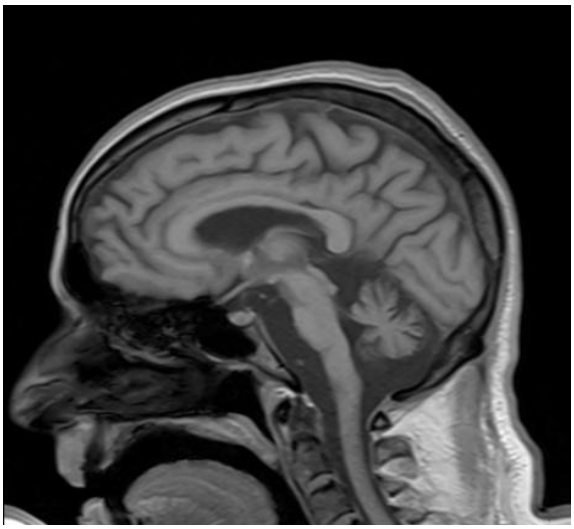
Figure 2: T2WI- upper medulla- loss of normal olivary bulge on anterolateral aspect of medulla

PRESENTATION OF CASE

58 year old normotensive, non-diabetic male came with case of progressive ataxia and right sided weakness since 1 year with single episode of history of fall and unconsciousness having signs of bilateral finger nose ataxia, ataxic gate, brisk exaggerated tendon reflexes & plantar extensors.

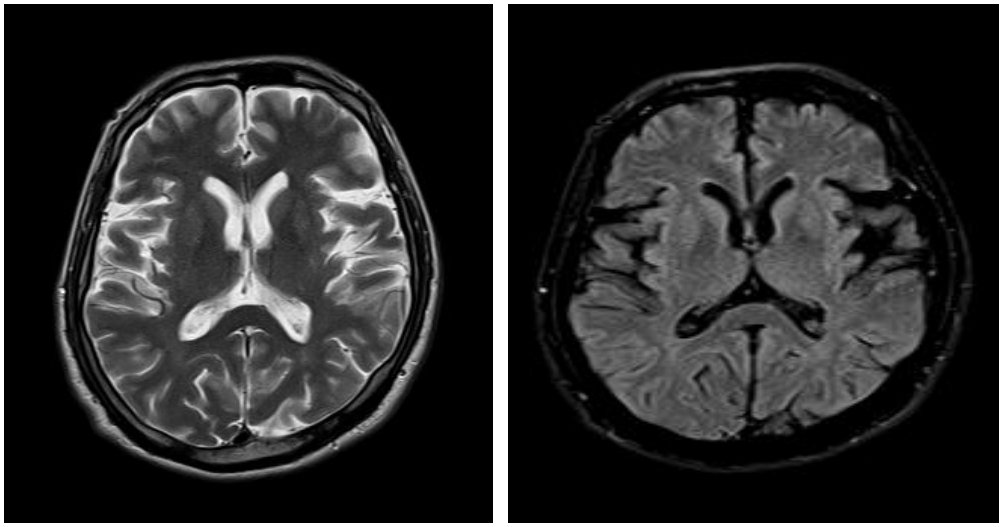
MRI Brain was performed on 1.5 T Siemens Avanto using short TR /TE (450 / 10 ms), Long TR/TE (3600 / 80ms), FLAIR, DWI, ADC, SWI, PHASE.

- On MRI Brain :
  - Cerebellar atrophy
  - Cruciform hyperintensity on T2WI/FLAIR involving inferior pons referred to as "Hot
  - Cross Bun Sign".
  - Small concave appearing bilateral middle cerebellar peduncles.
  - Loss of Bulge / Flattening of inferior surface of pons.
  - Loss of inferior olivary nucleus bulge.
  - Diffuse Cortical Atrophy.



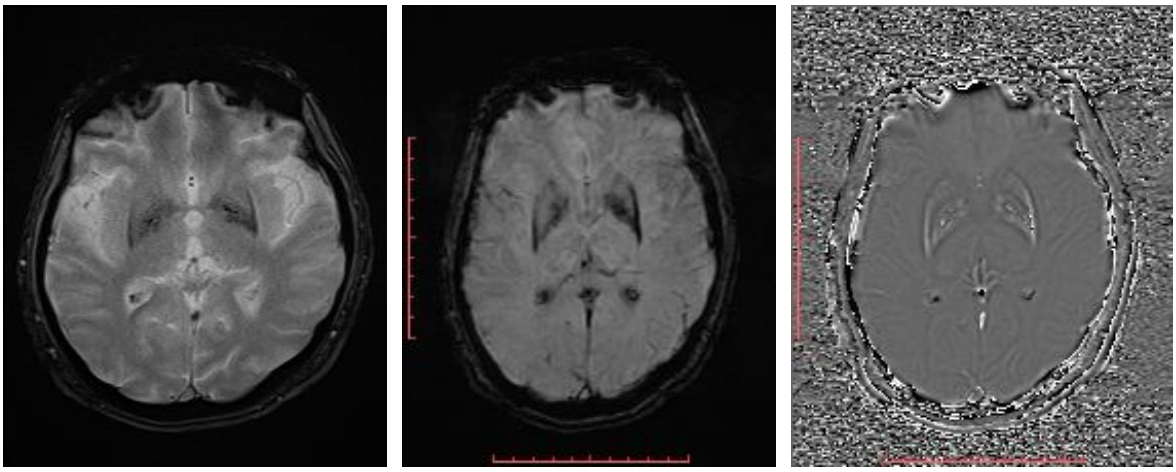
Sag T1WI

Figure 3: Loss of bulge in inferior pons & Cerebellar atrophy



Axial T2WI **Axial FLAIR**

Figure 4: Slight Hyperintensity along lateral margin of putamen bilaterally.



GRE **SWI** **PHASE**

Figure 5: Focal hemorrhage in bilateral globus pallidus

DISCUSSION

MSA parkinsonism (MSA-P), formerly called striatonigral degeneration, is characterized clinically by parkinsonian symptoms with prominence of rigidity, with neuroimaging and gross pathology showing atrophy of the striatum as a result of neuronal loss, involving putamen more than caudate. [14,15]

On long TR/TE MR images, a hyperintense rim at the putaminal edge, putaminal atrophy without any intrinsic signal intensity change in putamen, hypointense signal of the putamen, particularly along its posterolateral margin, on T2-weighted images which may be accompanied by a thin rim of hyperintense signal. The hypointense signal is due to abnormal deposition of iron the accompanying hyperintense signal likely reflects increased water associated with cell loss and/or gliosis. [15,16]

In contrast to PD, where well more than 90% of patients show a therapeutic response to levodopa, only 13% to 20% of patients with MSA-P show a therapeutic response. [17]

MSA-SDS is characterized by autonomic nervous system failure (orthostatic hypotension, urinary incontinence, and inability to sweat). Although the pathologic changes are somewhat variable, neuronal loss in the substantia nigra and the intermediolateral cell column of the spinal cord, accompanied by gliosis, commonly occurs. [18]

MSA-SDS may occur alone, although typically it occurs in association

with the clinical, pathologic, and radiologic features of MSA-P and/or MSA-C. [7] On MR, patients with MSA-SDS associated with MSA show typical findings of MSA-P and/or MSA-C. MR exams in pure autonomic failure patients are normal. [7]

MSA-Cerebellum (MSA-C) formerly called as olivopontocerebellar degeneration [1] onset varying between early childhood to old age. The primary degeneration centers in Pons, with a subsequent progressive anterograde degeneration of pontocerebellar fibers and of the cerebellum (hemispheres greater than vermis). [18,19]

Pontocerebellar fibers originate in the pontine nuclei, have a transverse course in the pons (thus they are called transverse pontine fibers), run to the cerebellum through the middle cerebellar peduncles, and terminate in all lobules of the cerebellar hemisphere and cerebellar vermis. The degeneration in MSA-C is characterized by myelin sheath loss and gliosis of this pontocerebellar pathway and neuronal loss of the cerebellar cortex. This in turn leading to retrograde degeneration of the inferior olives, whose fibers originate in the contralateral inferior olive and reach the cerebellum via the inferior cerebellar peduncle. [8,19]

On MR Signal changes & atrophy of Pons, middle cerebellar peduncles, the cerebellum, inferior cerebellar peduncle & olives. On Axial T2WI and FLAIR sequences-typically hyperintensity in cruciform pattern described as "Hot Cross Bun Sign"is seen consisting of the transverse pontine fibers coursing mediolaterally and the pontine raphe coursing anteroposteriorly. [20] Atrophy &hyperintensity involving middle cerebellar peduncles along with atrophy of Cerebellum & loss of bulge of inferior olives. The midline sagittal section shows selective pontine atrophy with flattening / loss of normal inferior pontine bulge. The characteristic T2 hyper intense sign in pons and middle cerebellar peduncle ("cross sign") reflects pontocerebellar fibers degeneration and despite very suggestive of MSA it can be found in other forms of parkinsonism. [21]

CONCLUSION

This is an uncommon case of an MSA C in a 58 year old male with predominant cerebellar signs. MRI revealed Atrophy with abnormal signal intensity of pons, middle cerebellar peduncles, cerebellum & inferior olivary nucleus features indicating atrophy of olivopontocerebellar pathway. Hyper-intensity on T2WI involving the lateral margin of putamen on right side probably due to increase water content without intrinsic putamen signal change, likely representing striatonigral degeneration component. Focal hemorrhages in bilateral globus pallidus remain unexplained. We did not come across this in any literature.

MRI features likely indicate Predominant MSA C type involvement correlating with cerebellar signs. Our case is classified as probable MSA, since the diagnosis of MSA is defined just with pathological analysis. There is no specific treatment to MSA until the present, only symptomatic interventions. [22]

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