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Case Report

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MR Imaging in Multiple System Atrophy-Cerebellar Type

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Abstract

We present a case of 54-year-old man with resented with two years' history of gait unsteadiness associated with dysarthria and bladder incontinence of one-year duration. MRI of brain of the patient was done which shows "Hot cross bun" sign in the form of cruciform hyper intensity in the pons on T2W images characteristic of MSA-C (Multiple systemic atrophy-Cerellar type). Cerebellum and middle cerebellar peduncles are is atrophied which again favoured MSA-C.

Keywords: *Multiple system atrophy; Cerebellar*

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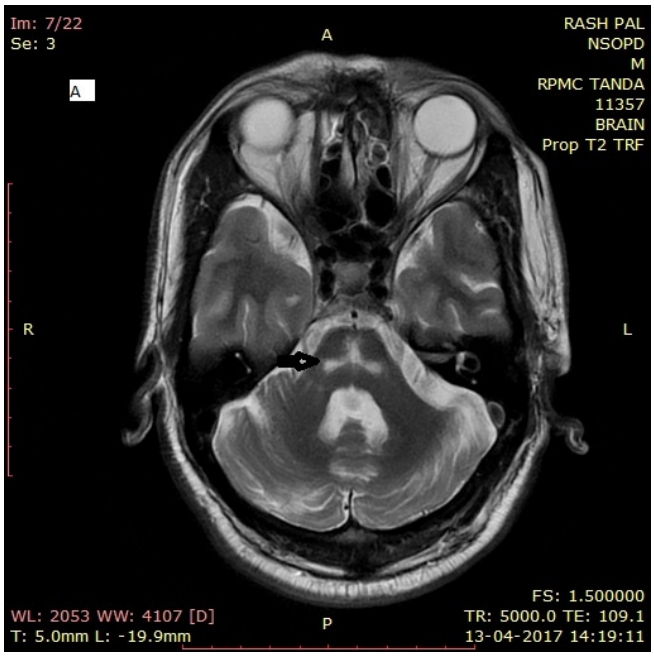
Introduction

Multiple system atrophy is a sporadic neurodegenerative disease characterised by varying degrees of cerebellar ataxia, autonomic dysfunction, parkinsonism and corticospinal dysfunction and MSA-C predominance of cerebellar symptoms olivopontocerebellar atrophy [1,2,6]. Key Diagnostic Features of MSA-C shows Cruciform shape of hyperintense signal in pons on T2WI and atrophy of pons, inferior olives, and cerebellum is also seen [3].

Case Report

A 54-year-old man presented with two years' history of gait unsteadiness associated with dysarthria and bladder incontinence of one-year duration. After clinical assessment the patient was referred to our department for MRI brain and find out cause of the above mentioned clinical symptoms. We did routine MRI scan in GE 1.5 Tesla machine which included axial T1W, FLAIR, DWI, axial and coronal T2W andT1W post contrast sequences. MRI of brain of the patient showed as in Figures (1) T2WI(A) shows "Hot cross bun" sign in the form of cruciform hyperintensity in the pons which is seen as hypointensity on T1W(C) and FLAIR(B) axial sequences (Thin arrow). Cerebellum(Thick arrow) as seen on T2W saggital images and midle cerebellar peduncles are is atrophied. On the basis characteristic findings of Cruciform shape of hyperintense signal in pons on T2WI and atrophy of pons, inferior olives, and cerebellum diagnosis of MSA-C was made.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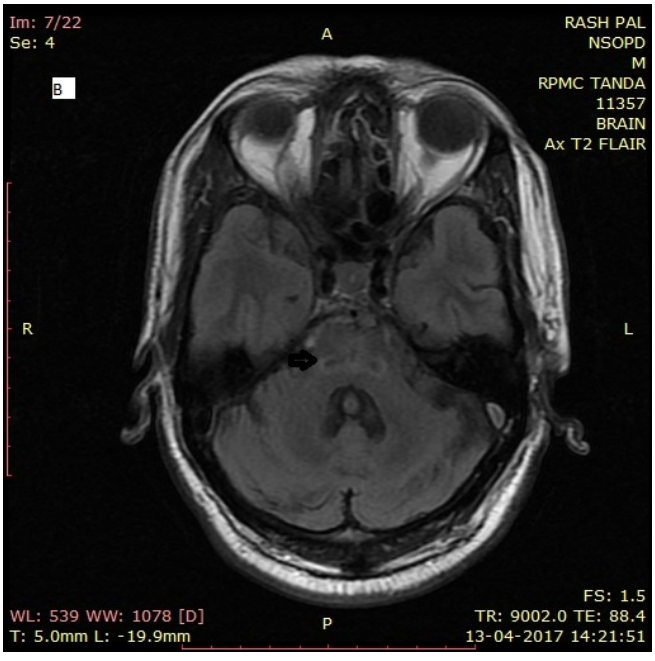
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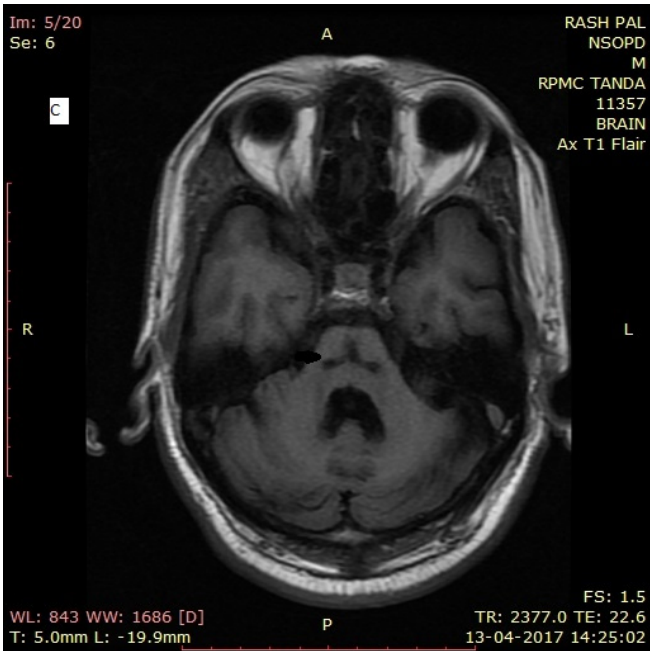
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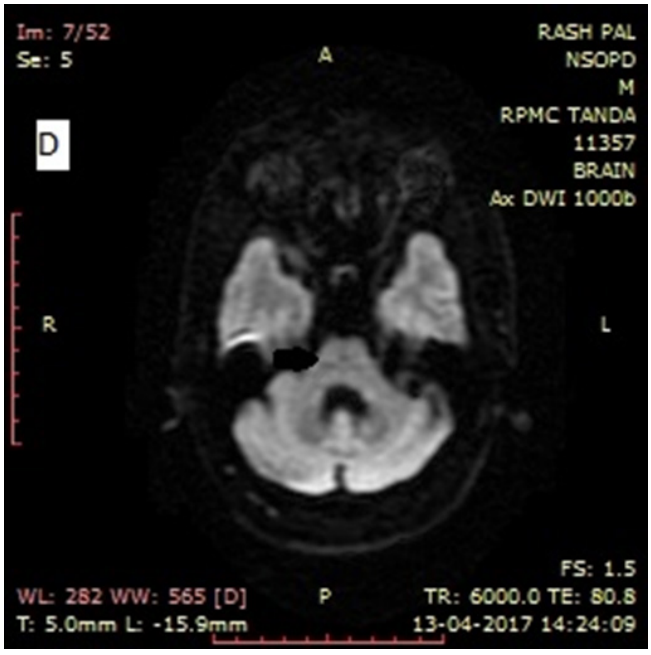
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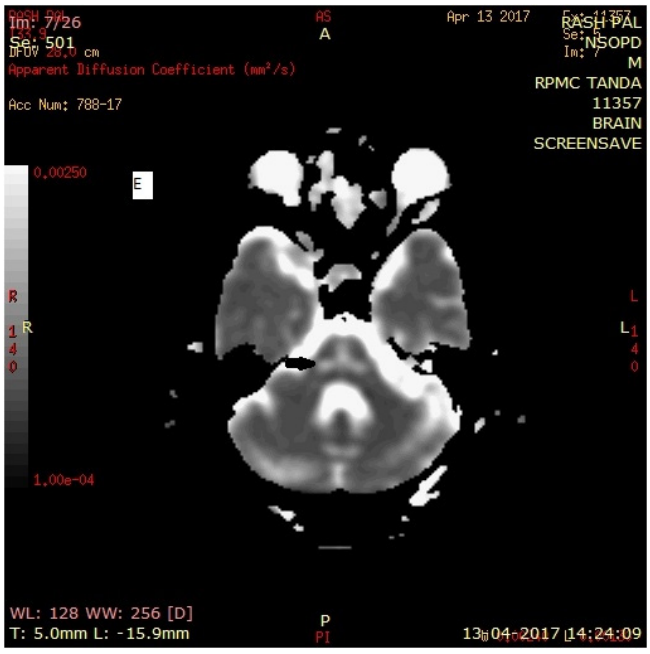

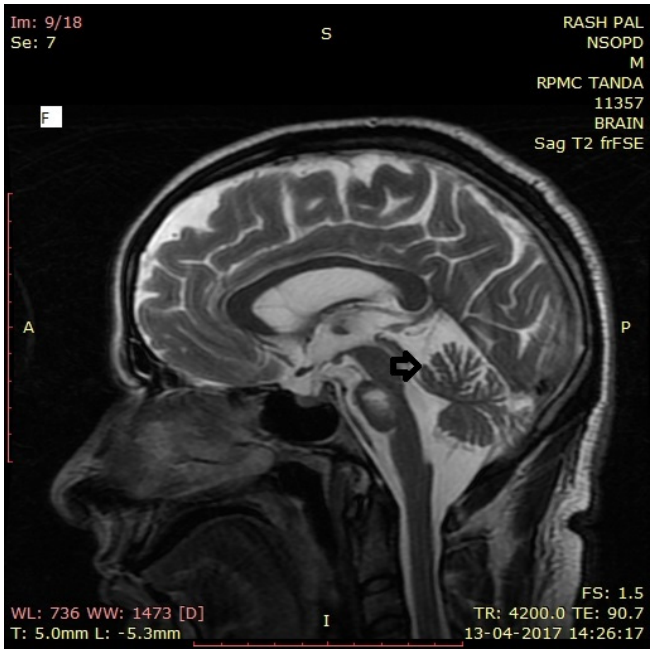

MRI of brain of the patient : T2WI(A) shows "Hot cross bun" sign in the form of cruciform hyperintensity in the pon which is seen as hypointensity on T1W(C) and FLAIR(B) axial sequences(Thin arrow).Cerebellum(Thick arrow) as seen on T2W saggital images and midle cerebellar peduncles are is atrophied.

Discussion

Multiple system atrophy (MSA) is a sporadic neurodegenerative disease characterised by varying degrees of cerebellar ataxia, autonomic dysfunction, parkinsonism and corticospinal dysfunctionand MSA-C predominance of cerebellar symptoms olivopontocerebellar atrophy [1,2,7,8]. Multiple systemic atrophy is a sporadic disease, with a prevalence of 4 per 100,000. Typically symptoms begin in 4th or 5th decade. Clinical presentation is variable, but typically presents in one of three patterns MSA-C demonstrates primarily cerebellar dysfunction Shy-Drager syndromeis used when autonomic symptoms predominate and in striatonigral degeneration shows predominant parkinsonian features. Multiple systemic atrophy results from abnormalities of alpha-synuclein metabolism, resulting in intracellular deposition which are found not only in neurons but also in oligodendroglia [3]. An MRI of the brain in T2/ PD/FLAIR sequences reveals the classical 'hot-cross bun' sign in the pons characterized by cruciate hyperintensity secondary to atrophy of the transverse pontine fibers [4,5].

There was no restricted diffusion of the signal changes on apparent diffusion coefficient (ADC) mapping. These features were seen in our case with atrophy of pons, inferior olives, and cerebellum which is also key diagnostic feature. Differentials are firstly Cerebello-olivary Atrophy which has cortical cerebellar degeneration with Selective atrophy of lateral cerebellum ("fish-mouth deformity" on parasagittal sections) and superior vermis [4,5]. Other differential can be Friedreich Ataxia which shows severe atrophy of spinal cord and medulla oblongata and mild atrophy of vermian and paravermian structures is seen. As far as treatment is concerned unfortunately no effective treatment is currently available and the disease progresses relentlessly culminating in death usually within decade of diagnosis [6-8].

Conclusion

Multiple system atrophy (MSA) is a sporadic neurodegenerative disease with MSA-C predominance of cerebellar symptoms olivopontocerebellar atrophy. Key Diagnostic Features on MRI of MSA-C are Cruciform shape of hyperintense signal in pons giving a characteristic "Hot cross bun" sign along with atrophy of pons, inferior olives, and cerebellum.

Acknowledgments

We are grateful to almighty for his blessings.

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