

MRI Features of Multiple System Atrophy - A Case Report

Sujoy Mani¹, Ojaswi Khandediya², Priti Kapoor³

¹JR-1 Radiodiagnosis, MGM Medical College and Hospital, Sector-1, Kamothe, Navi Mumbai-410209.

²JR-3 Radiodiagnosis, MGM Medical College and Hospital, Sector-1, Kamothe, Navi Mumbai-410209.

³Head of Department Radiodiagnosis, MGM Medical College and Hospital, Sector-1, Kamothe, Navi Mumbai-410209

Corresponding Author: Sujoy Mani

ABSTRACT

Multiple system atrophy (MSA) is a synucleinopathy, which is characterized by combined manifestations of Parkinsonian, cerebellar and autonomic features. It is broadly subdivided into two main groups- Parkinsonian MSA (MSA-P) and cerebellar MSA (MSA-C). Magnetic resonance imaging (MRI) of the brain is an essential tool in making a confident diagnosis of this condition. The clinical phenotype usually shows crossover and is at times very complex. On the other hand, Magnetic Resonance Imaging of the Brain may show many characteristic features, such as the 'hot cross bun' sign. This is a cruciform hyperintensity seen at the level of pons in axial T2-weighted MRI images. This sign is characteristically seen in patients with MSA-C. This case report deals with a patient with characteristic brain MRI features of MSA-C type.

Keywords- Multiple system atrophy, Hot-cross bun sign, Magnetic Resonance Imaging, Olivopontocerebellar degeneration

INTRODUCTION

Multiple system atrophy is a relatively uncommon neurodegenerative disorder. It comes under the broad classification of synucleinopathies which are a subgroup of neurodegenerative disorders, characterised by impairment of alpha-synuclein metabolism, resulting in abnormal intracellular deposits. These are further divided into those with and those without the formation of Lewy bodies.^[1,2]

The presentation of MSA is usually complex. It can be characterised by varying degrees of cerebellar ataxia, autonomic dysfunction, Parkinsonism and corticospinal dysfunction. MSA is a sporadic disease, with a prevalence of 4 per 100,000.^[3] The affected age group usually ranges between 40 to 60 years of age.

In cases of early disease of MSA, to make a diagnosis and differentiate it from Parkinson's disease can be very challenging.

Its rarity and mixed clinical phenotype make it a diagnostic dilemma for the concerned physician. Autonomic dysfunction is mandatory in order to make a diagnosis of MSA.^[4]

In its two major subtypes patients having features of Parkinsonism are designated as MSA Parkinsonism type (MSA-P) and those with cerebellar dysfunction are designated as MSA Cerebellar type (MSA-C).

Due to the complex clinical phenotype, Imaging plays a key role in differentiating MSA from Parkinson's especially in the early stages of the disease as well as in characterising and differentiating MSA-P type from MSA-C type. In cases of MSA-P type, imaging features on MRI Brain include putaminal atrophy, putaminal body hypo intensity 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 with brainstem and cerebellar atrophy. These imaging features are key to diagnosis.

This case report deals with a rare case of MSA-C type.

CASE REPORT

58-year-old male patient came to the neurology OPD with chief complaints of loss of bowel and bladder control (autonomic dysfunction) and tingling and numbness in the hands and feet suggestive of peripheral neuropathy.

MRI Brain was suggested for the patient.

Patient underwent an MRI Brain Plain scan and the Imaging Findings were as follows.

Marked atrophy of the brainstem was noted accompanied with prominence of the prepontine cistern and the fourth ventricle. Rest of the ventricular system appeared normal. There was presence of diffuse widening of the folia in bilateral cerebellar hemispheres and the vermis.

Streaks of T2 hyperintensities and corresponding FLAIR hypo intensities are noted in both the cerebellar hemispheres. There is presence of thinning of both the middle cerebellar peduncles. Generalised prominence of the cortical sulci is noted. Axial T2 Weighted and FLAIR Images at the level of the pons showed the presence of linear cruciform hyperintensities, known as the so called ‘hot cross buns’ sign.

Rest of the supratentorial parenchyma appeared normal. There was no evidence of any other focal or diffuse area of altered signal abnormality seen in the cerebral parenchyma. There was no evidence of restricted diffusion on DWI images. There was no evidence of midline shift or intracranial haemorrhage seen.

The clinical features and imaging findings are in keeping with multiple system atrophy. The imaging features are accompanied with autonomic dysfunction. There is presence of cerebellar and brainstem atrophy and the characteristic hot cross buns sign noted in the pons in keeping with MSA Cerebellar type (MSA-C).

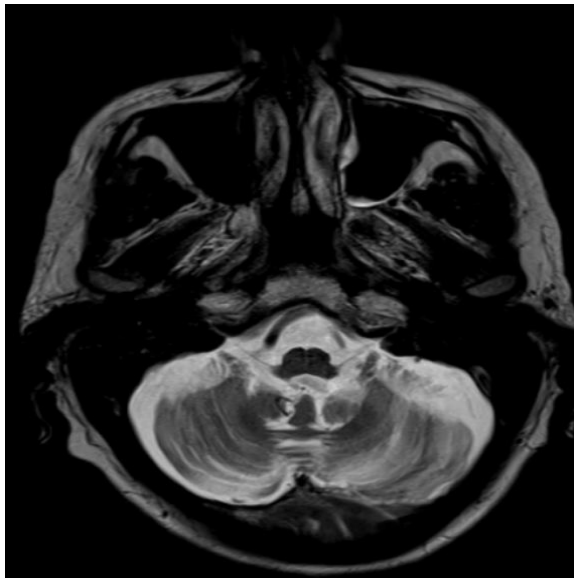


Figure 1. Axial T2W images at the level of cerebellum showing T2 hyperintensities and cerebellar atrophy

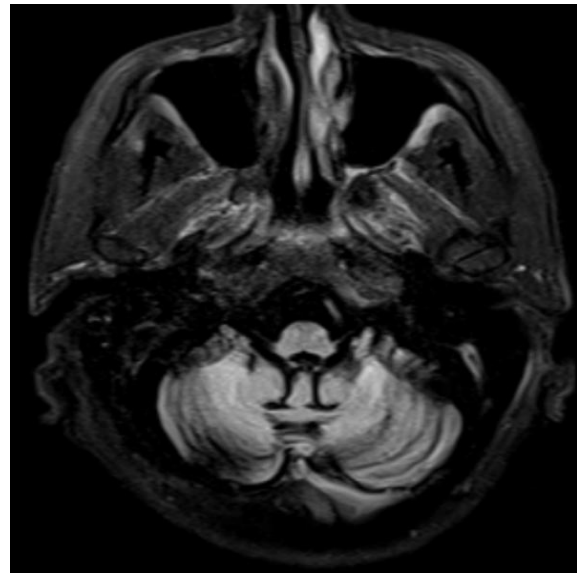


Figure 2. Axial FLAIR images at the level of cerebellum showing corresponding FLAIR hypo intensities.

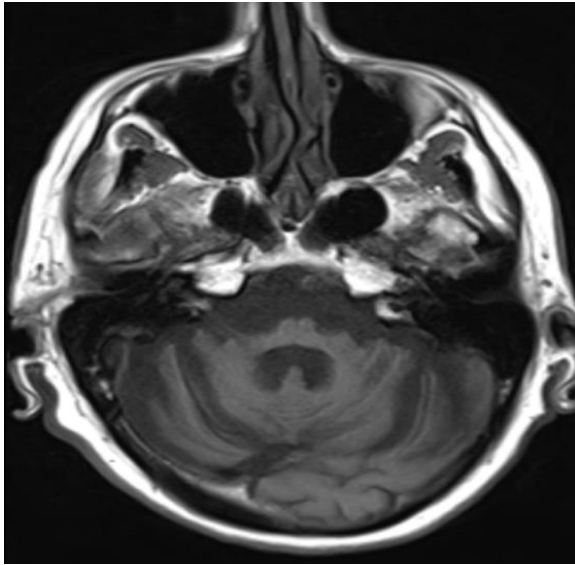


Figure 3. Axial T1 weighted Images showing cerebellar atrophy in bilateral cerebellar hemispheres.

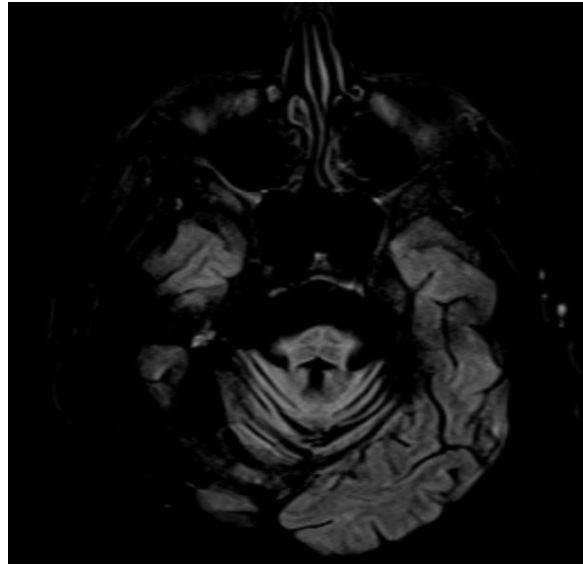


Figure 4. Axial FLAIR images at the level of the pons showing the presence of linear cruciform hyperintensities in the pons demonstrating the characteristic 'hot cross buns' sign.

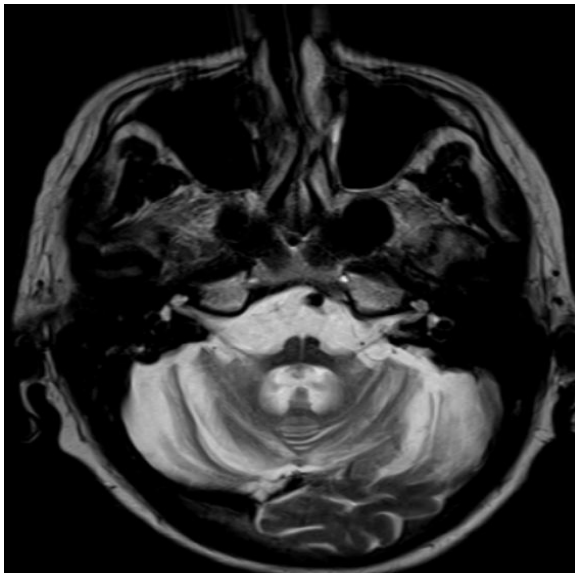


Figure 5. Axial T2 weighted images showing prominent 4th ventricle and pre-pontine cistern

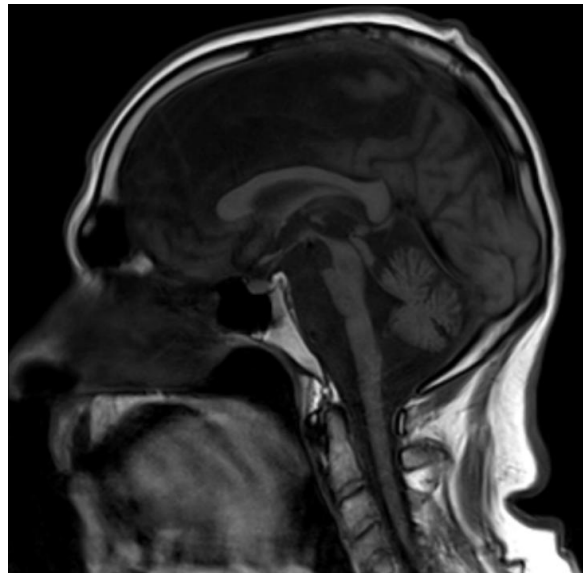


Figure 6. Sagittal T1 weighted Images showing atrophy of the brainstem and the cerebellum

DISCUSSION

Multiple system atrophy (MSA) is defined as a rare, sporadic, adult-onset neurodegenerative disorder. The term MSA was initially proposed to encompass three diseases, that is Shy-Drager syndrome (SDS), Striatonigral degenerative disease (SND) and olivopontocerebellar atrophy (OPCA). MSA commonly occurs within the age group ranging from 40 to 60 years. Our patient in this case report falls within this age group.

Magnetic Resonance Imaging of the Brain remains the gold standard

investigation for the evaluation of Parkinsonian and cerebellar syndromes, including MSA. Every patient suspected of having MSA should undergo an MRI scan including all the standard sequences as well as diffusion weighted imaging (DWI).

MRI helps in differentiating between the two types of MSA, that is MSA-P and MSA-C. MSA-P type shows brain imaging features that include putaminal atrophy, relative hypo intensity of the putamen with respect to the rest of the basal ganglia and hyperintensity surrounding the putamen known as the hyperintense putaminal rim

sign. However, the hyperintense putaminal rim sign may be a normal observation on a 3T MRI brain study.

MSA-C type on the other hand shows distinguishing features with the help of which we can make a confident diagnosis. Autonomic dysfunction is a mandatory criterion and the patient shows cerebellar symptoms as well. MRI Brain findings include atrophy; sometimes gross involving the brainstem and the cerebellum. There may be diffuse widening of the folia observed in both the cerebellar hemispheres. Another key imaging finding which can be pathognomonic for MSA-C type is the presence of linear cruciform T2/FLAIR hyperintensities in the pons known as the 'hot cross buns' sign. It is formed due to the selective loss of the pontocerebellar fibres and pontine raphe neurons with the preservation of pontine tegmentum and corticospinal tracts.

In our case report the patient's MRI Brain plain study showed atrophy of the brainstem and cerebellum, and cruciform hyperintensity at the level of pons, forming 'hot cross bun' sign, characteristic of MSA-C. The specificity of MRI to differentiate MSA from idiopathic Parkinson's disease and controls is 93.3 and 90.6%, respectively. Therefore, in a patient in which MSA is clinically suspected, MRI is an essential diagnostic modality to come up with a conclusive diagnosis. The typical hot cross bun sign as found in our patient also helps to establish a confident diagnosis of MSA-C.

CONCLUSION

Being a rare neurodegenerative disease, which is relatively difficult to diagnose due to the complicated clinical phenotype, imaging features tend to play a key role in differentiating MSA from other neurodegenerative disorders. MSA masquerades as cerebellar, Parkinsonian and autonomic disorders due to which MRI imaging features are key to establish a definitive diagnosis. Imaging can also be used in identifying at-risk individuals and help in monitoring the progression of disease. MRI can help in early diagnosis of MSA and can also be used in monitoring the response to several potential disease-modifying therapies.

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