MRI Features of Multiple System Atrophy

Sumantro Mondal, Sumit Chakraborty[1], Atanu Chakraborty, Debanjali Sinha, Tony Ete, Arijit Nag

Departments of Medicine and [1]Radiology and Imaging, Institute of Postgraduate Medical Education and Research, Kolkata, West Bengal, India

Correspondence: Dr. Sumantro Mondal, Doctor's Hostel, Room No.-427, 242 AJC Bose Road, Kolkata - 700 020, West Bengal, India.
E-mail: drmsumantro@gmail.com

ABSTRACT

Multiple system atrophy (MSA) is a synucleinopathy, characterized by combined manifestations of cerebellar, parkinsonian, and autonomic features. It is subdivided into two main types-parkinsonian MSA (MSA-P) and cerebellar MSA (MSA-C). Magnetic resonance imaging (MRI) of the brain is a valuable tool in diagnosing this condition as it shows many characteristic features, such as the 'hot cross bun' sign. This is a cruciform hyperintensity imaging feature, seen at the level of pons in axial T2-weighted MRI images. This sign is typically seen in patients with MSA-C. This reported case describes a patient with MSA-C with characteristics brain MRI features.

Key words: Hot cross bun sign; multiple system atrophy; magnetic resonance imaging synucleinopathy; pons

Introduction

Multiple system atrophy (MSA) is an uncommon neurodegenerative disorder. The presentation is complex with overlapping features of cerebellar dysfunction, Parkinsonism, and autonomic disturbance. This combination of rarity and mixed clinical phenotype makes the diagnosis of MSA a challenging job for the physician. Neurodegenerative disorders also offer significant morbidity to the patients. Appropriate treatment strategies demand accurate diagnosis as the treatment protocols differs significantly in various conditions which have overlapping symptomatology. Routine laboratory investigations are inconclusive in these neurodegenerative disorders, neither is there any role of cerebrospinal fluid (CSF) study. Functional imaging of brain, like positron emission tomography can show abnormalities in Parkinson's disease, but it is still used for research purpose and their widespread applicability is limited by economic constraints. In this scenario, neuroimaging, especially magnetic resonance imaging (MRI) can be helpful to some extent to achieve a correct diagnosis. Our patient presented with mixed features of cerebellar, Parkinsonian's dysfunctions, and autonomic dysfunction and MRI of the brain showed the classical signs to have made a final diagnosis of cerebellar MSA (MSA-C). This report is concerned with the complex presentation of a rare disease (MSA-C) and the important role of MRI to diagnose this disease.

Access this article online	
Quick Response Code:	Website: www.wajradiology.org
	DOI: 10.4103/1115-1474.128086

Case Report

A 58-year-old female attended medical outpatient department of an Institute of Post Graduate Medical Education and Research with history of gait instability, intention tremor, and orthostatic dizziness for the last 1 year duration. She also developed urinary incontinence and impaired sweating of her acral regions for 4 months. Her symptoms were gradually progressive. She had two attacks of orthostatic faintness each lasting for up to 2-3 min within last 3 months. She was nondiabetic, but recently diagnosed hypertensive and presently on amlodipin (5 mg daily) medication since last 1 year. Examination revealed mild pallor. She had a blood pressure of 140/86 mmHg with orthostatic hypotension. Neurological examination showed intention tremor with a frequency 10-12 Hz and moderate degree of rigidity in her lower limbs. There was cerebellar type of ataxia with impaired tandem gait. Some features of cerebellar dysfunction like dysmetria, past pointing, and impairment of synergistic movements were also evident. Autonomic instability was present. Quantitative sudomotor axon reflex testing (QSART) revealed diminution of sweating. Combinations of autonomic, cerebellar, and mild parkinsonian features led us toward a provisional diagnosis of MSA. MRI of the brain was performed. It showed gross atrophy of the infratentorial structures especially brain stem and cerebellum [Figure 1]. Axial T2-weighted MRI at the level of pons showed cross-shaped

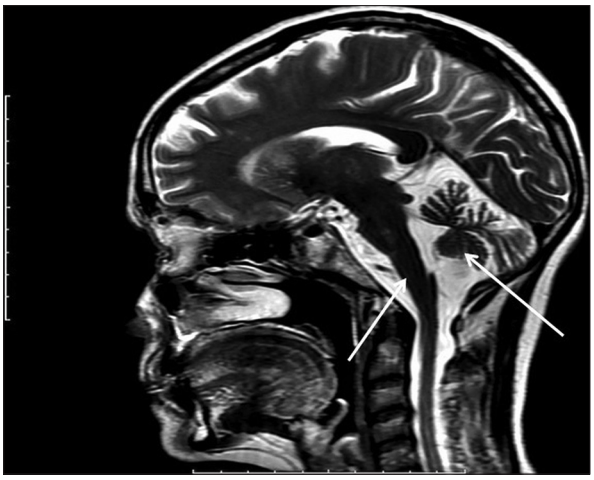

Figure 1: Sagittal T2-weighted magnetic resonance imaging (MRI) showing gross atrophy of brainstem and cerebellum with widening of extra-axial cerebrospinal fluid (CSF) space (white arrows)

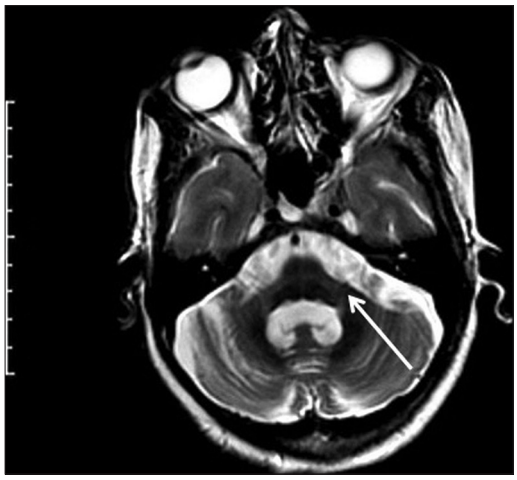

Figure 2: Axial T2-weighted MRI at the level of pons showing cruciform hyperintensity—the "hot cross bun" sign (white arrow)

hyperintensity (hot cross bun sign) [Figure 2]. Patient's clinical profile and typical MRI features established the diagnosis of MSA-C. Till now no therapy is curative for MSA. In our patient, orthostatic hypotension was troublesome for which she was advised for nonpharmacologic measures like leg crossing, squatting, and advised for adequate salt and fluid intake. Urinary incontinence was managed by intermittent self-catheterization of the urinary bladder. The movement disorder was controlled with a combination of levodopa and carbidopa and advised for regular follow-up.

Discussion

MSA is a synucleinopathy like Parkinson's disease and dementia with Lewy bodies.[1] It is an uncommon disease with an annual incidence of 0.7:100,000 and a point prevalence of 3.4:100,000. The disease affects the elderly population. The mean age group for MSA is 55.4 ± 8.3 years.[2] The age of our patient fits within this mean age group. MSA typically manifests as combination of both autonomic and non-autonomic neurological features. Among the non-autonomic features parkinsonian and cerebellar features usually predominates. The prognosis of MSA is less favorable than Parkinson's disease.[2] Our patient presented with both autonomic and cerebellar features, favoring MSA as a better diagnostic consideration than Parkinson's disease. MRI of brain is a very informative diagnostic tool in MSA and it differentiates this condition from idiopathic Parkinson's disease. It shows putaminal atrophy and hypointensity, hyper intense putaminal rim, and infratentorial signal change.[3] In patients with parkinsonian MSA (MSA-P), MRI of the brain shows putaminal atrophy, hypointensity of putaminal body, and markedly hyperintense putaminal rim.[4] However, hypointensity in the putamen may also be seen in progressive supranuclear palsy and atypical parkinsonism.[5] Infratentorial abnormalities in MSA consists of atrophy of the cerebellum, middle cerebellar peduncles, pons, and midbrain, and signal change of pons, and middle cerebellar peduncles.[3] Infratentorial signal changes has a characteristic appearance. The 'hot cross bun' sign is chiefly seen in patients with MSA-C. The cruciform T2 hyper intensity is formed due to selective loss of transverse pontocerebellar fibers and pontine raphe neurons with preservation of pontine tegmentum and corticospinal tracts.[3] It is seen in 60-64% patients with MSA.[2] The sign is typical, but not pathognomonic to MSA.[6] It is also seen in patients of spinocerebellar ataxia types 2 and 3, and in Parkinsonism probably secondary to vasculitis. Histological section at the level of cruciform signal showed gliosis of middle part of the reticular formation, pontocerebellar fiber between the medial lemniscus, and pyramidal tract and the crossing part of the pontocerebellar fibers at the basis pontis. The sign is thought to be produced primarily due to gliosis.[7] In our patient, MRI of the brain showed atrophy of the brainstem and cerebellum and also the 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. So in a patient with clinically suspected MSA, MRI is an essential diagnostic modadility to come at a conclusive diagnosis. The typical hot cross bun sign as found in our patient also helps to establish a confident diagnosis of MSA-C.

Conclusions

1. MSA is a rare neurodegenerative disease which masquerades as cerebellar, parkinsonian, and autonomic disorders.
2. MRI of the brain can be very informative in diagnosing MSA-C.

References

1. Chavarría C, Souza JM. Oxidation and nitration of α-synuclein and their implications in neurodegenerative diseases. Arch Biochem Biophys 2013;533:25-32.
2. Watanabe H, Saito Y, Terao S, Ando T, Kachi T, Mukai E, *et al*. Progression and prognosis in multiple system atrophy: An analysis of 230 Japanese patients. Brain 2002;125:1070-83.

3. Schrag A, Kingsley D, Phatouros C, Mathias CJ, Lees AJ, Daniel SE, *et al*. Clinical usefulness of magnetic resonance imaging in multiple system atrophy. J Neurol Neurosurg Psychiatry 1998;65:65-71.
4. Bhattacharya K, Saadia D, Eisenkraft B, Yahr M, Olanow W, Drayer B, *et al*. Brain magnetic resonance imaging in multiple-system atrophy and Parkinson disease: A diagnostic algorithm. Arch Neurol 2002;59:835-42.
5. Stern MB, Braffman BH, Skolnick BE, Hurtig HI, Grossman RI. Magnetic resonance imaging in Parkinsons disease and parkinsonian syndromes. Neurology 1989;39:1524-6.
6. Srivastava T, Singh S, Goyal V, Shukla G, Behari M. "Hot cross bun" sign in two patients with multiple system atrophy-cerebellar. Neurology 2005;64:128.
7. Takao M, Kadowaki T, Tomita Y, Yoshida Y, Mihara B. "Hot-cross bun sign" of multiple system atrophy. Intern Med 2007;46:1883.

How to cite this article: Mondal S, Chakraborty S, Chakraborty A, Sinha D, Ete T, Nag A. MRI features of multiple system atrophy. West Afr J Radiol 2014;21:35-7.

Source of Support: Nil, **Conflict of Interest:** None declared.