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MRI imaging signs of Multiple System Atrophy (MSA-C)

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ABSTRACT

Multiple Systems Atrophy (MSA) is a neurologic disorder clinically characterized by parkinsonism, cerebellar ataxia, and autonomic failure. This disorder was previously known as "striatonigral degeneration sporadic olivopontocerebellar atrophy and "Shy-Drager syndrome Presence of neurogenic bladder is a characteristic feature of MSA. It is regarded as a sporadic disease with a prevalence of 4 per 1,00,000. Typically the symptoms begin between 40 and 60 years of age. It has two phenotypes: parkinsonian (MSA-P) and cerebellar (MSA-C). MRI brain acts as a diagnostic tool for MSA with the characteristic feature of "Hot cross bun' sign, seen as cruciform hyperintensity at the level of the pons in axial T2-weighted images in patients with MSA-C. We report here a case of a 55-year-old female patient who presented with complaint of gait instability, intention tremor, and orthostatic dizziness for the last 1 year. She also developed urinary incontinence from last 4 months.

Keywords— *Hot cross bun sign, Striatonigral degeneration, Parkinson, Sporadic olivopontocerebellar atrophy, Shy-Drager*

1. INTRODUCTION

Multiple System Atrophy (MSA) is rare neurodegenerative disorder with complex presentation and overlapping features of many neurological diseases. This makes the diagnosis of MSA a challenging job for the treating doctor. Routine laboratory and CSF studies are not much helpful in its diagnosis. Neuroimaging with Magnetic Resonance Imaging (MRI) can be helpful for its correct diagnosis. In our case the 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).

2. CASE HISTORY

A 55-year-old Indian female presented to our hospital OPD with complaint of gait instability, intention tremor, and orthostatic hypotension since last 1 year. She also developed urinary incontinence from last 4 months. Her symptoms were gradually progressive. She also had three attacks of orthostatic faintness within last 4 months. She was no diabetic. She had a blood pressure of 142/86 mmHg with orthostatic hypotension. CNS 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 were also evident. Autonomic instability was present. Combinations of autonomic, cerebellar, and mild parkinsonian features led us toward a provisional diagnosis of MSA. Thus patient was referred to our department for MRI of the brain. It showed moderate prominence of bilateral cerebellar foliae suggestive of cerebellar atrophy. Axial T2-weighted MRI showed mild thinning of the pons and medulla with cruciate appearance of the pons on T2W images (Hot cross bun appearance. Patient's clinical profile and typical MRI features thus established our final diagnosis of MSA-C.

3. DISCUSSION

This disorder was previously known as "striatonigral degeneration (disorder by disruption in connection between -the striatum and the substantia nigra. These two areas work together to enable balance and movement)", sporadic olivopontocerebellar atrophy(progressive condition characterized by the degeneration of neurons in specific areas of the brain.), and "Shy-Drager syndrome(disorder of the central and sympathetic nervous systems, with postural hypotension. The presence of autonomic failure is a required clinical criterion for its diagnosis. It has two phenotypes:

(a) Parkinsonian (MSA-P)
(b) Cerebellar (MSA-C)

- MSA-P: Has predominance of parkinsonian signs and symptoms. Presents with progressive bradykinesia, rigidity, postural instability, and, jerky postural tremor.
- MSA-C: predominance of cerebellar symptoms (olivopontocerebellar atrophy Presents with gait and limb ataxia, scanning speech, and cerebellar oculomotor disturbances.

MSA is characterized pathologically by cytoplasmic inclusions in oligodendroglial cells (Papp- Lantos bodies) and the absence of Lewy bodies.

In patients with (MSA-P), MRI shows putaminal atrophy, hypointensity of putaminal body, and markedly hyperintense putaminal rim. However, hypointensity in the putamen may also be seen in other conditions like progressive supranuclear palsy and atypical Parkinsonism. Other features consists of atrophy of the cerebellum, middle cerebellar peduncles, pons, and midbrain, and signal change of pons, and middle cerebellar peduncles. The 'hot cross bun' sign is characteristically seen in patients with MSA-C. and seen in 60%. The sign is typical, but not pathognomonic to MSA. It is also seen in patients of spinocerebellar ataxia and in Parkinsonism probably secondary to vasculitis.

4. CONCLUSION

In our patient, MRI showed atrophy of the brainstem and cerebellum and also the cruciform hyperintensity at the level of pons, knows as 'Hot Cross Bun' sign, characteristic of MSA-C. The specificity of MRI to differentiate MSA from idiopathic Parkinson's disease is almost 93%.

So in a 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 diagnosis of MSA-C

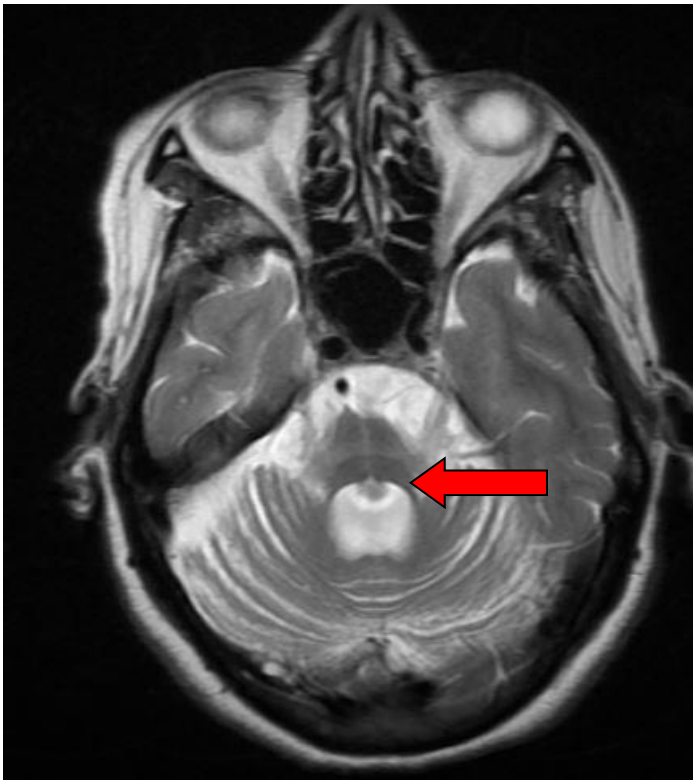

Fig. 1: Axial T2W image of the brain showing mild thinning of the pons and medulla with cruciate appearance of the pons on T2W images (Hot cross bun appearance)

Fig. 2: T2W images (Hot cross bun appearance)

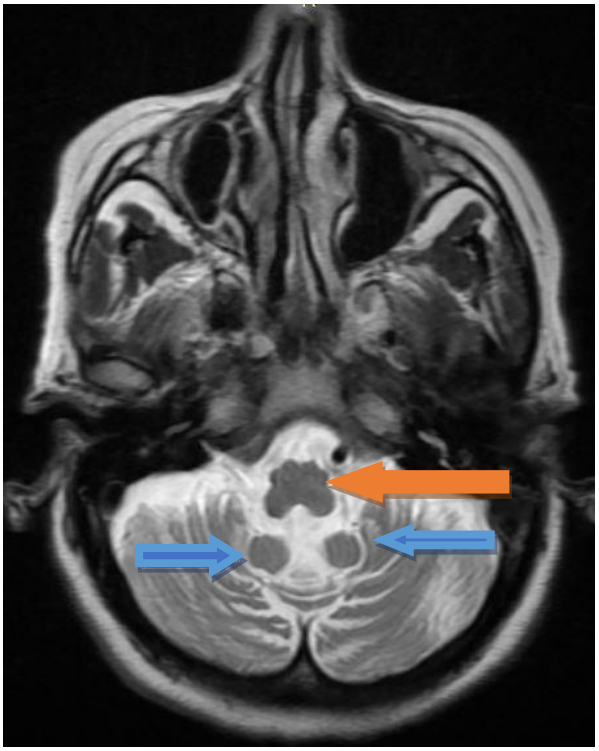

Fig. 3: Axial T2W image of the brain showing moderate prominence of bilateral cerebellar foliae (Blue arrow) suggestive of cerebellar atrophy

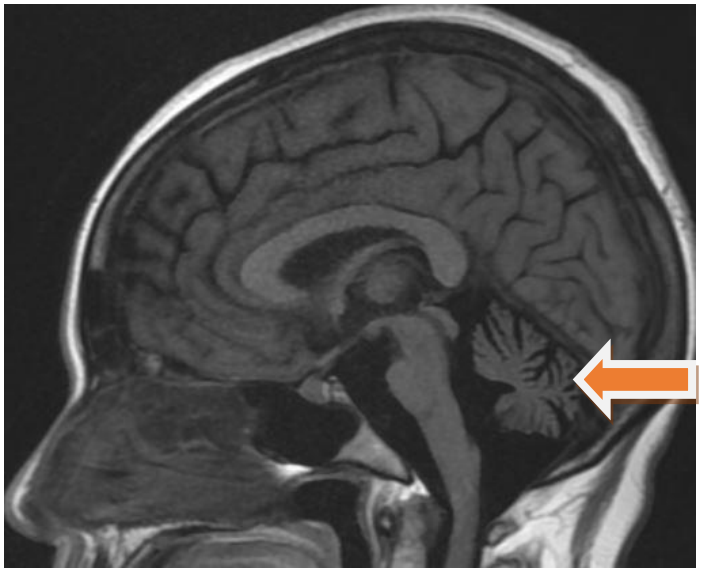

Fig. 4: Sagital T1W image of the brain showing cerebellar atrophy (Orange arrow)

5. REFERENCES

[1] https://jnnp.bmj.com/content/87/4/443
[2] http://www.wajradiology.org/article.asp?issn=1115-3474;year=2014;volume=21;issue=1;spage=35;epage=37;aulast=Mondal
[3] http://www.journalagent.com/tjn/pdfs/TJN_19_1_28_30%5BA%5D.pdf
[4] http://www.jnmjournal.org/journal/view.html?uid=741&vmd=Full
[5] https://www.mdsabstracts.org/abstract/a-young-man-with-multiple-system-atrophy-a-case-report/
[6] https://www.dovepress.com/diagnostic-challenges-in-multiple-system-atrophy-peer-reviewed-fulltext-article-NDT

