

THE HOT CROSS BUN SIGN: A CASE SERIES

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ABSTRACT

Multiple system atrophy (MSA) is an adult-onset, sporadic, progressive neurodegenerative disorder. It is an alpha synucleinopathy and the presentations include parkinsonism, cerebellar ataxia, and autonomic symptoms in varying combinations. The two clinical phenotypes recognized are the parkinsonian variant (MSA-P) and the cerebellar variant (MSA-C). Since many of the clinical features overlap with Parkinson's disease, it is often misdiagnosed as Parkinson's disease in the early stages. Since MSA is a progressive disorder with a poor prognosis, the treating physician needs to recognize this entity and provide prognostication. MRI brain may help in differentiating between these two and an important radiological sign in MSA-c is the 'Hot cross bun' sign. It refers to the cruciform T2 hyperintense signal on axial magnetic resonance imaging (MRI) of the pons. We hereby report two patients who presented with progressive slowness in walking, ataxia, pyramidal signs, and orthostatic hypotension, whose MRI brain showed the characteristic hot cross bun sign.

Key Messages:

Multiple system atrophy is a progressive neurodegenerative disorder with a poor prognosis, which may be erroneously diagnosed as Parkinson's disease in the early stages. The 'Hot cross bun' sign on the MRI brain, a characteristic feature of MSA-C helps in the diagnosis, though it is not pathognomonic for the same.

Keywords: *Multiple system atrophy; Synucleinopathies; Neuroimaging; Parkinson's disease; Cerebellar ataxia*

INTRODUCTION

Multiple system atrophy (MSA) is a neurodegenerative disorder with a poor prognosis. The clinical spectrum includes cerebellar ataxia, parkinsonism, and autonomic failure, in varying combinations.¹ The

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presenting motor disorder most commonly consists of parkinsonism with bradykinesia, rigidity, gait instability, and tremor, however in a significant number of cases, cerebellar ataxia is the initial motor disorder.² The two clinical phenotypes are the parkinsonian variant (MSA-P) and the cerebellar variant (MSA-C).^{3,4} Early on in the course of the illness, patients presenting with predominant parkinsonism symptoms may be misdiagnosed as Parkinson's disease. Prompt diagnosis of this fatal disorder can help in prognostication for the treating physician. One radiological sign which points to the diagnosis of MSA-C is the hot cross bun sign on magnetic resonance imaging (MRI).^{5,6} We hereby report two patients who presented with progressive slowness in walking, ataxia pyramidal signs, and orthostatic hypotension, whose MRI brain showed the characteristic hot cross bun sign.

CASE HISTORY

CASE 1

A 53year old lady presented to the Department of Neurology, at our institute with unsteadiness of gait, urinary incontinence, and dizziness on getting up from a supine position for the past year.

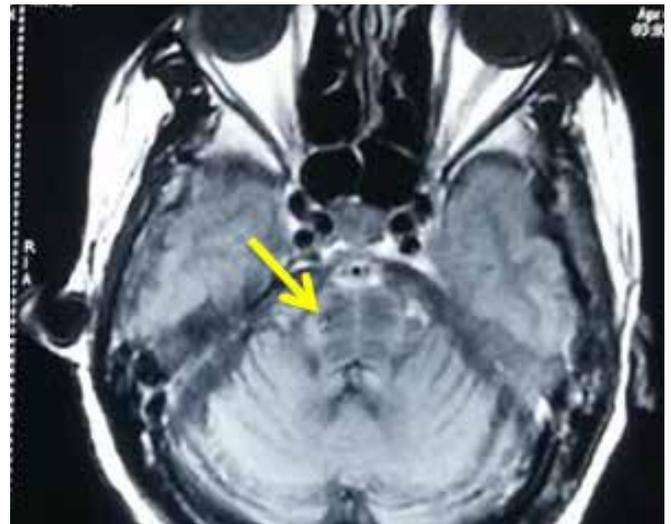
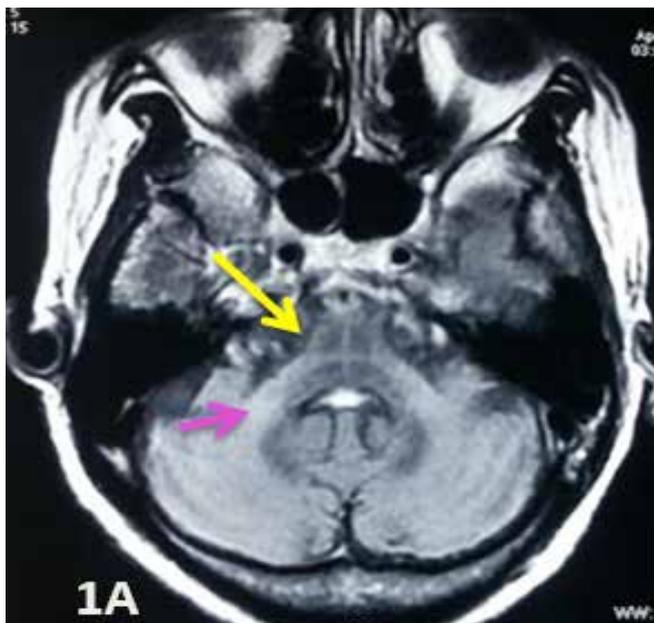


Figure 1: MRI brain: 1A and 1B: axial section showing the hot cross bun sign (yellow arrow) and the bright MCP sign (pink arrow), 1C: sagittal section showing atrophy of pons and cerebellum (arrows)

She would sway to either side on walking and would require support to walk. She denied a history of vertigo, ear symptoms, or paraesthesia/numbness of feet. She did not have worsening of unsteadiness in the dark. She also complained of urinary precipitancy, she would feel bladder sensations normally but would void before reaching the toilet. She had a graying of her vision on getting up from the supine position. She also had intermittent episodes of stridor. On examination, she had inspiratory stridor and was noted to have

postural hypotension (fall in systolic BP by 30 mm Hg and diastolic by 20 mm Hg).

She had mild rigidity in all limbs, bilaterally brisk deep tendon reflexes, and bilateral cerebellar signs. Her MRI brain revealed atrophy of pons and cerebellum, T2 hyperintensity of bilateral middle cerebellar peduncles (bright MCP sign), with characteristic cruciform T2 and FLAIR hyperintensities in pons – ‘The Hot cross bun sign’ (Figure:1). Basic vasculitic work up, and the paraneoplastic panel was negative. She tested negative for HIV. She declined genetic testing for cerebellar ataxias. The diagnosis of probable MSA-C (Multiple system atrophy-cerebellar) was made and she was started on Levodopa. On follow-up 3 months back, she had progression of symptoms and was bed-bound.

CASE 2

A 53-year-old male presented to us with complaints of slowness in walking, reeling of gait, and urinary precipitancy for the past 2 years. He complained of recurrent episodes of lightheadedness and graying of vision for the past several months. He had slurred speech and had difficulty swallowing.

On examination, he had a significant postural fall in BP, with a fall of 40 mm Hg in systolic BP and 30 mm in diastolic BP. He had a scanning speech, with undue separation of syllables. He had a slow saccade and broken pursuit movement of eyes. He had an exaggerated gag reflex. He had rigidity in all limbs, bilaterally brisk deep tendon reflexes, extensor plantar, and bilateral cerebellar signs. MRI brain revealed the ‘hot cross bun’ sign in the pons, with atrophy of pons and cerebellum. (Figure 2)

His ESR was normal, ANA profile, HIV testing, and the paraneoplastic panels were negative. He opted out of genetic testing for cerebellar ataxias. He was diagnosed as a probable case of MSA-C.

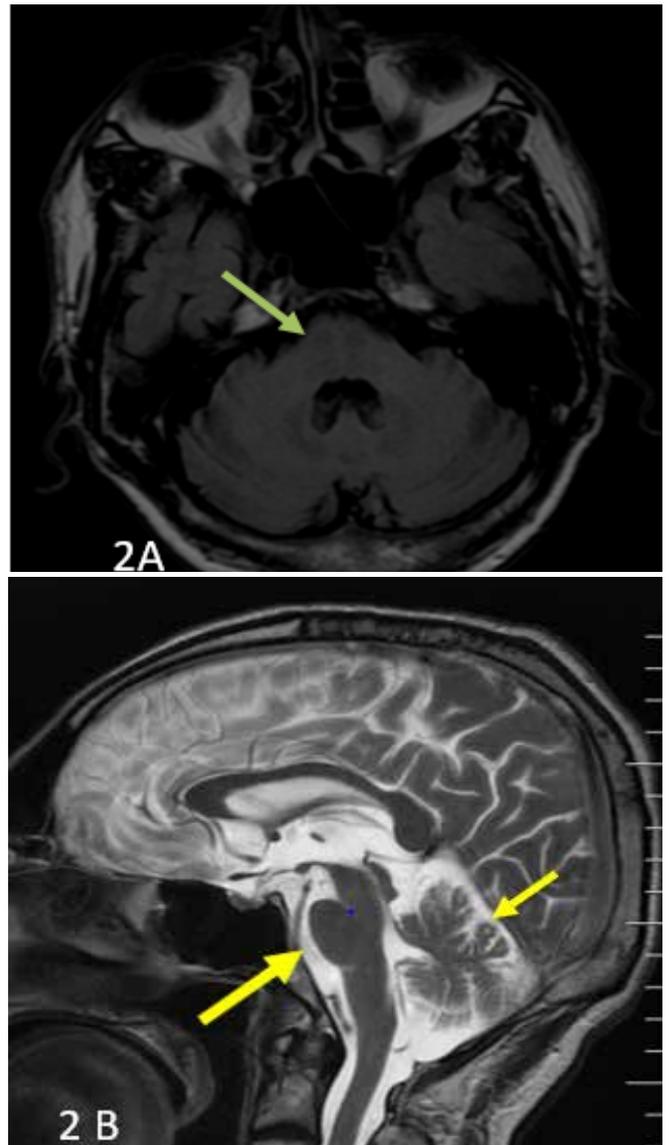


Figure 2: MRI brain: 2A: axial section showing the hot cross bun sign (arrow), 2B: sagittal section showing atrophy of pons and cerebellum (arrows)

DISCUSSION

Multiple system atrophy, an alpha synucleinopathy, is a rapidly progressive neurodegenerative disorder characterized by varying degrees of parkinsonism, cerebellar signs, pyramidal signs, and autonomic dysfunction.¹ The reported mean age of onset is around 55 to 60 years and the median survival is around 8 years.⁷ MSA is classified into either Parkinsonian type (MSA-P) or cerebellar type (MSA-C), based on the predominant motor deficit.

During the initial stage of the disease, it may be difficult to differentiate it from idiopathic Parkinson's disease. The presence of stridor, prominent pyramidal, and cerebellar signs, with a rapid progression and poor responsiveness to levodopa and/or characteristic neuroimaging abnormalities on the MRI help to treat physicians in making an early diagnosis of MSA.⁸

The characteristic neuroimaging abnormalities described in MSA are, the hot cross bun sign, the bright MCP (middle cerebellar peduncle) sign, the putaminal slit sign, and atrophy of the pons and cerebellum. The 'hot cross bun' sign (HCBS) refers to the cruciform T2 hyperintense signal on axial MRI of the pons, resulting from the atrophy and gliosis of the transverse pontocerebellar fibers, with preserved corticospinal tracts, which run craniocaudally.^{9,10} It is most commonly seen in patients with the cerebellar subtype of multiple system atrophy (MSA-C), and is reported to be highly specific for the same.¹⁰ It derives the name from the sweet spiced bun, marked with a cross on the top, traditionally eaten on a 'Good Friday'. It is considered to be a hallmark of the disease progression in the cerebellar type of MSA, though it is not pathognomonic for the same. The "hot cross bun" sign was seen in both of our patients.

Other conditions where the hot cross bun can be seen are bilateral pontine infarction; spinocerebellar ataxias type 1, 2, 3, 7, 8; cerebrotendinous xanthomatosis; paraneoplastic cerebellar degeneration; HIV associated progressive multifocal leukoencephalopathy; neurosarcoidosis, variant Creutzfeldt–Jakob disease; and vasculitis.⁹

The bright MCP sign is another sign reported in MSA-C. It denotes atrophy and T2 hyperintensities of bilateral middle cerebellar peduncles, on MRI.

It was seen in one of our patients. Both of our patients had atrophy of the pons and cerebellum, evident on MRI (Figure 1, 2).⁸ MRI brain is hence an invaluable tool in diagnosing and characterizing multiple system atrophy.

CONFLICT OF INTEREST

None

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