



## Hot cross bun sign

M. Portet<sup>1</sup> · M. Filyridou<sup>1</sup> · D. C. Howlett<sup>1</sup>

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Dear Sirs,

The “hot cross bun” (HCB) sign refers to a cruciform-shaped hyperintensity within the pons found on T2-weighted magnetic resonance imaging (MRI). This is a rare MR finding, and it is most commonly associated with atrophy of the pons, cerebellum, and putamen in cerebellar variant multiple system atrophy (MSA-C). We report a case of (HCB) in an adult male patient with MSA-C, with emphasis on the clinical presentation, and a discussion of the characteristic imaging findings.

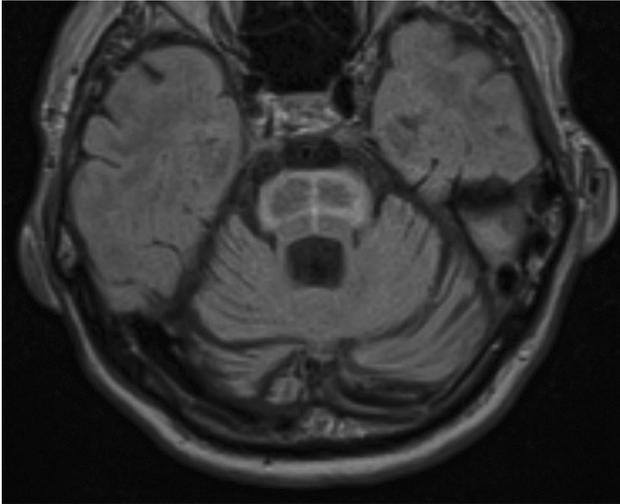
A 76 year old male presented with a 1 year history of slurred speech and a progressive gait disturbance. He had no bradykinesia, rigidity, memory loss or features suggestive of dementia. There was no significant past medical history of note, and no previous family history of similar complaints. The patient was not taking any medications. On examination he was dysarthric, with an ataxic gait. He had mild hyper-tonia and hyperreflexia, the neurological examination was otherwise unremarkable, and he did not describe any autonomic symptoms. As part of his further investigations an MR brain was arranged. This demonstrated prominent cruciform linear hyperintensity in the pons on T2W and FLAIR sequences (Fig. 1), the HCB sign. Pontine and middle cerebellar peduncle atrophy were also noted, as well as high signal in the middle cerebral peduncles, the bright middle cerebellar peduncle (MCP) sign (Fig. 2). Based on the history, clinical examination, and imaging findings, a diagnosis of Multi System Atrophy (MSA-C) was made.

Multiple system atrophy is a neurodegenerative disorder that is classified under Parkinson plus syndromes, with variable involvement of the basal ganglia, pontocerebellar region and the autonomic system. Mean survival from time of diagnosis ranges between 6 and 10 years, and definitive diagnosis

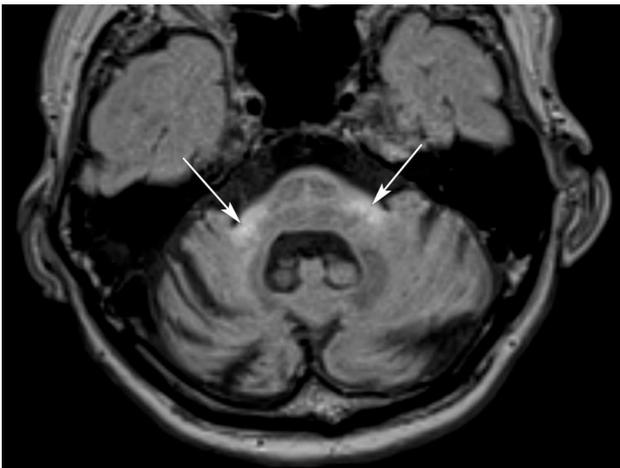
is made on autopsy with demonstration of oligodendroglial cytoplasmic inclusions consisting of fibrillar  $\alpha$ -synuclein [1]. Based on the clinical presentation, MSA can be classified into MSA-P with predominant Parkinsonism symptoms and signs, and MSA-C with predominant cerebellar symptoms and signs, as in our patient [2]. In this case MRI demonstrated characteristic findings of HCB, cruciform T2/FLAIR hyperintensity within the pons, marked pontocerebellar atrophy, and high T2/FLAIR signal in the middle cerebellar peduncles. These findings are thought to be due to the selective loss of myelinated transverse pontocerebellar fibres, and neurons in the pontine raphe. Although there is a strong association, the bright HCB sign is not specific for MSA, and has also been described in a number of other conditions including Parkinsonism secondary to vasculitis, spinocerebellar atrophy, and variant Creutzfeldt-Jakob disease, amongst others. In one recent study looking at the underlying aetiology of the HCB sign, for example, only 55% of patients had MSA [3]. In that particular study, the most common alternate aetiology was an undefined hereditary cerebellar ataxia. In addition, the HCB sign is also a relatively rare finding, and not always seen in MSA. In another recent study reviewing MRI features in MSA, for example, only 24% of patients with MSA had the bright HCB sign [4]. In that study, it was shown that a combination of mid brain atrophy, corpus callosum atrophy and cerebellar atrophy was more commonly observed, and more specific to the diagnosis. The bright HCB sign is nevertheless, a useful imaging characteristic, which can be used in conjunction with other MRI findings, such as the bright MCP sign and pontocerebellar atrophy, alongside the clinical history and examination to support the diagnosis of MSA.

✉ M. Portet  
markportet@doctors.org.uk

<sup>1</sup> Radiology Department Eastbourne DGH, East Sussex Healthcare NHS Trust, Eastbourne, England



**Fig. 1** Axial FLAIR MR image at the level of the upper pons demonstrating cruciform linear pontine hyperintensity, the “hot-cross bun” sign



**Fig. 2** Axial FLAIR MR image at the level of the inferior pons, with arrows demonstrating high signal in the middle cerebellar peduncles, the “bright MCP” sign. Note the middle cerebellar peduncles are atrophied. The HCB sign can also be seen faintly in this image

## Compliance with ethical standards

**Conflicts of interest** On behalf of all authors, the corresponding author states that there is no conflict of interest.

**Ethical standards** This type of article does not require ethical approval in our institution.

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