

Clinical Image

Hot Cross Bun Sign in Multisystematized Atrophy: The Cerebellar form “HCB in MSA-c”

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Keywords

Hot Cross Bun sign; Multiple System Atrophy; Ataxia; MRI

Abbreviations

MSA-c: Multisystematized Atrophy-Cerebellar Type; HCB: Hot Cross Bun Sign; SCA: Spinocerebral Ataxia; MRI: Magnetic Resonance Imaging

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Multisystematized Atrophy (MSA) is one of the Neurodegenerative causes generating Parkinson's "plus" syndrome having three clinical forms a cerebellar form (MSA-c) characterized by cerebellar ataxia, a predominantly Parkinson form (MSA-p) and MSA with predominant autonomic signs and symptoms [1].

The cross-shaped aspect in the pons or Hot Cross Bun Sign (HCB) is visible on MRI on the axial T2 and Flair weighted more marked in T2 sequence where it is manifested by the shape of the cross at the level of the pons [1]. This name derives from a sweet spicy bun baked by Christian Church on the last Thursday before Easter and marked with the cross on the top, the four arms of which represent the four quarters of the year.

This pontic cross-like appearance is related to T2-weighted high signal (hypersignal) of the transverse fibers of the bridge, the middle part of the cerebellar peduncles and cerebellum, and to low signal (hypo signal) of the corticospinal bundles and upper cerebellar peduncles [2].

However, in MSA-c, this sign indicates that there is atrophy of the pons and the cerebellum while a normal appearance of the putamen is observed on the T2 and SWI sequences [2]. PET-FDG metabolic imaging notes hypometabolism in the Pontus and cerebellum [1].

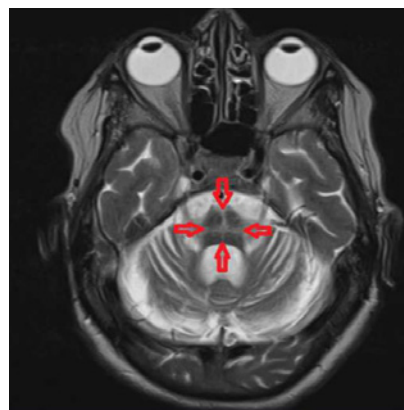


Figure 1: Hot Cross bun sign on the axial T2 sequence MRI.

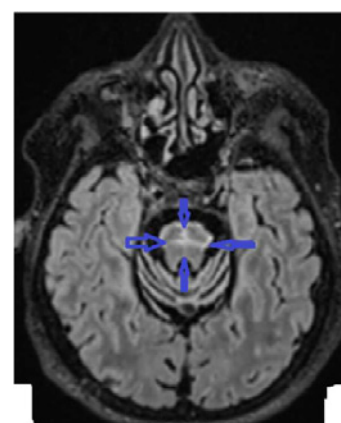


Figure 2: Hot Cross Bun sign on the axial Flair Sequence MRI.

The Figure 1 and 2 illustrate the Hot Cross bun sign respectively on the axial T2 and flair sequences. Recognition of this appearance makes it possible to confirm the diagnosis of cerebellar-type multisystem atrophy (MSA-c) in 76% of cases and will allow it to be dissociated from the MSA-p form.

This sign (HCB) may be present in several differential diagnosis and the context guides the clinicians and radiologists. Then, the following diseases are already reported with Hot Cross Bun sign [3]: bilateral pontine infarction, Cerebrotendinous xanthomatosis, Encephalitis, Fragile X tremor ataxia syndrome, HIV-related progressive multifocal leukoencephalopathy, Leptomeningeal carcinomatosis, Leptomeningeal metastases from breast cancer, Multiple system atrophy, Neurosarcoidosis, Paraneoplastic cerebellar degeneration from testicular tumor, Parkinsonism secondary to presumed vasculitis, Probable Lewy body dementia. Spinocerebellar ataxia (SCA) type 1,2,3(Machado Joseph Disease),7,8 and 34; Variant

Creutzfeldt-Jakob disease. Among these, MSA and SCA were the most common etiologies [4].

References

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