“Hot Cross Bun” Sign Associated with SCA1

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The “hot cross bun” sign (HCBS) is not pathognomonic for multiple system atrophy (MSA), although it is occasionally seen in patients with hereditary ataxia (1). A search of the literature, however, failed to identify any cases of SCA1 associated with HCBS. We herein present the first cases of SCA1 presenting with HCBS.

A 37-year-old man with 52 CAG repeats, whose mother had genetically confirmed SCA1 (2), had a five-year history of progressive gait ataxia, nystagmus, slurred speech, pyramidal tract signs and slight euphoric dementia. Brain MRI showed HCBS in the lower pons (Picture A), but not in the middle or upper pons, with severe atrophy in both the cerebellum and brainstem (Picture B). His 35-year-old brother, also with 52 CAG repeats, exhibited similar clinical and radiological features (Picture C).

The slight HCBS observed in these patients compared with the typical HCBS seen in cases of MSA may be due to the fact that the patients were in the early stage of the disease course and that the pontocerebellar system is less impaired in the setting of SCA1 than in MSA (1).

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References

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