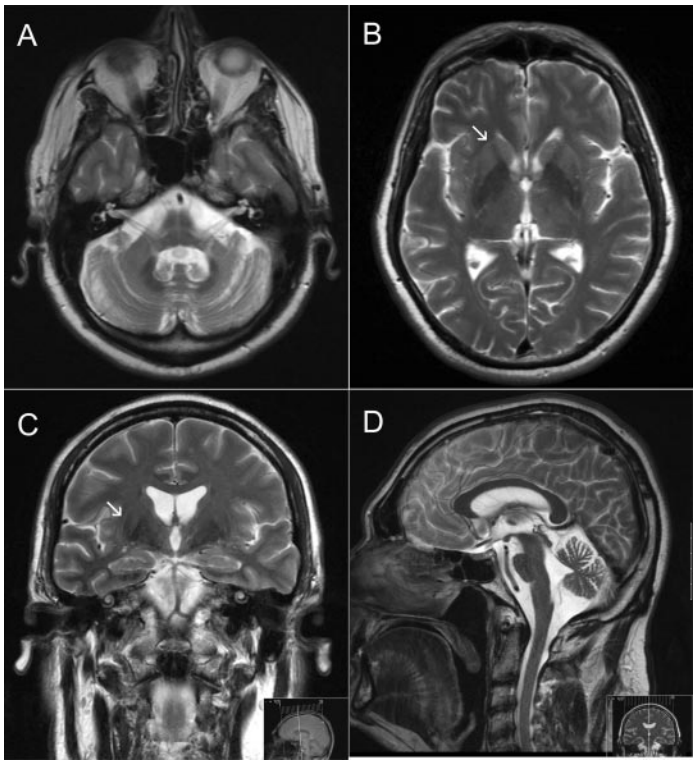
Teaching NeuroImage: MRI in multiple system atrophy

"Hot cross bun" sign and hyperintense rim bordering the putamina

João Massano, MD
Fernando Costa, MD
Goreti Nadais, MD

Address correspondence and reprint requests to Dr. João Massano, Department of Neurology, Hospital de São João, Alameda Prof. Hernani Monteiro, 4200-319 Porto, Portugal
jmassano@med.up.pt

Figure Brain MRI (1.5 T)



(A) Axial T2 image showing the classic "hot cross bun" sign in the pons; axial (B) and coronal (C) T2 images depicting a hyperintense rim bordering the lateral margin of both putamina (white arrows); (D) sagittal T2 sequence disclosing pontine and cerebellar atrophy, the latter also evident in A. Right side of images is left side of patient in A, B, and C.

A 59-year-old man developed progressive gait ataxia, dysarthria, sexual dysfunction, and Raynaud phenomenon over 2 years. On examination he also manifested bradykinesia and rigidity, postural and action tremor of the upper limbs, and orthostatic hypotension. Family history was negative. Brain MRI disclosed the "hot cross bun" sign typical of multiple system atrophy (MSA) (figure).[1,2] This sign is probably related to degeneration of transverse pontocerebellar fibers, whereas the hyperintense rim bordering the putamina has been associated with neuronal loss, reactive microgliosis, and astrogliosis.[2] Glial cytoplasmic inclusions, neuronal inclusions, and demyelination have been linked to MSA, but the mechanisms involved require further clarification.[2] MRI has diagnostic potential in parkinsonism-plus syndromes; several typical findings like those depicted here have been described.[1,2]

REFERENCES

1. Schrag A, Good CD, Miszkiel K, et al. Differentiation of atypical parkinsonian syndromes with routine MRI. Neurology 2000;54:697–702.
2. Wenning GK, Colosimo C, Geser F, Poewe W. Multiple system atrophy. Lancet Neurol 2004;3:93–103.

From the Departments of Neurology (J.M., G.N.) and Neuroradiology (F.C.), Hospital de São João; and Faculty of Medicine (J.M.), University of Porto, Portugal.

Disclosure: The authors report no disclosures.

Teaching Neuro*Image*: MRI in multiple system atrophy: "Hot cross bun" sign and hyperintense rim bordering the putamina

João Massano, Fernando Costa and Goreti Nadais

Neurology 2008;71;e38
DOI 10.1212/01.wnl.0000327520.99034.28