Radiographic Characteristics of Multi-System Atrophy: "The Hot Cross Bun" Sign

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Purpose
The purpose of this presentation is to describe the neuropathologic findings and radiographic appearance of multisystem atrophy specifically the Shy-Drager syndrome. In 1960, Shy and Drager noted the clinical findings correlated with neuropathologic changes in the basal ganglia and brainstem. Neurologists, consider the Shy-Drager syndrome to be a unique form of multisystem degeneration, which is similar yet, distinctly different from either Parkinson’s disease or olivopontine cerebellar atrophy (OPCA). Although these findings and characteristics are noted in the neurology literature, very little information is noted in the neuroimaging literature.

Materials & Methods
Three patients, two male and one female, with detailed neurologic evaluation for multisystem atrophy, were evaluated with MR imaging. Imaging was obtained on 1.5 T magnet with both standard T1-, proton density and T2-weighted images, with attention to the brainstem.

Results
All patients demonstrated severe volume loss in the pons and cerebellum. In addition, all demonstrated a cruciate pattern of increased signal intensity on T2-weighted images seen within the brainstem, described in the neurologic literature as mimicking the appearance of a “hot cross bun” if viewed from above. Additional findings abnormal foci of increased signal intensity on the T2-weighted images within the branchium pontis and lentiform nuclei.

Conclusion
This presentation reviews the salient neurologic and radiographic findings in multisystem atrophy, specifically the Shy-Drager syndrome seen in patients evaluated at our institution over the past year. Pathologically multisystem atrophy is associated with widespread symmetric neuronal degeneration affecting the caudate nucleus, substantia nigra, locus coeruleus, olivary nuclei, dorsal vagal nuclei, and cerebellum with cell loss often associated with gliosis. Radiographically, a cruciate pattern of increased signal intensity on T2-weighted images is noted within the brainstem, previously referred to as the “hot cross bun sign”.