Case 8862

The hot cross bun sign
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Case Type: Clinical Cases
Authors: Arora A, Kapoor A, Singh S, Puri
Department of Radiodiagnosis, G.B. Pant Hospital and associated Maulana Azad Medical College, New Delhi, India.
Patient: 55 years, male

Clinical History:
A 55-year-old man was being evaluated for worsening problems with balance and difficulty in walking. On examination he had a broad-based cerebellar ataxic gait, slurred speech, and dysmetria on finger-to-nose testing.

Imaging Findings:
Axial T2-w MRI demonstrated the characteristic "hot cross bun sign" i.e. a cruciform hyperintensity in an atrophied pons. The cerebellum and middle cerebellar peduncles were atrophic. Sagittal T1-weighted MRI confirmed the atrophy of the olivary eminences of medulla oblongata resulting in straightening of the angle usually present on the ventral frontier between the pons and medulla oblongata. Cerebellar white matter hyperintensity was noted on the FLAIR images which were extending into the middle cerebellar peduncles. These MR imaging findings were consistent with olivopontocerebellar atrophy of multiple system atrophy-cerebellar type (MSA-c).

Discussion:
Hot cross bun sign refers to the cruciform-shaped hyperintensity in the pons on axial T2-weighted MR images seen in multisystem atrophy (MSA). MSA encompasses the disorders striatonigral degeneration, olivoponto cerebellar atrophy (OPCA), and the Shy-Drager syndrome. MSA is considered synonymous with striatonigral degeneration when parkinsonian symptoms predominate (MSA-p), with olivoponto cerebellar atrophy (OPCA) when cerebellar signs predominate (MSA-c), and with Shy-Drager syndrome when autonomic symptoms are dominant.

The hot cross bun sign is noted in MSA-c which is characterised by atrophy of the infratentorial structures, especially the inferior olivary nucleus, pons and the cerebellum (OPCA). Hot cross bun sign in MSA-c occurs secondary to selective loss of myelinated transverse pontocerebellar fibres and neurons in the pontine raphe with sparing of the pontine tegmentum and corticospinal tracts. The name derives from a sweet spiced bun baked by the Christian church on the last Thursday before Easter and marked with a cross on the top, with the four quarters representing the four quarters of the year (Fig. 4). Although hot cross bun sign has been said to be specific for multiple system atrophy, it has been reported in Parkinson's disease presumed to be secondary to vasculitis.

Differential Diagnosis List: Olivopontocerebellar atrophy in multiple system atrophy (MSA-c)

Final Diagnosis: Olivopontocerebellar atrophy in multiple system atrophy (MSA-c)
References:

Description: Axial T2-w MRI shows the hot cross bun sign as a cruciform hyperintensity in an atrophied pons. The cerebellum and middle cerebellar peduncles are also atrophied. Origin:
Description: The hot cross bun sign. Origin:
Figure 2

Description: Sagittal T1-weighted MRI shows atrophy of the olivary eminences of the medulla oblongata resulting in straightening of the angle usually present on the ventral frontier between the pons and medulla oblongata. Associated cerebellar atrophy is evident. Origin:
Description: Marked cerebellar atrophy is seen with relative sparing of the supra-tentorial structures. Cerebellar white matter hyperintensity is also present. Origin:
**Description:** Sweet spiced buns marked with a cross on the top, with the four quarters representing the four quarters of the year. **Origin:**