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Pontine hot cross bun sign in spinocerebellar ataxia type 2

"Sinal da cruz" na ponte na ataxia espinocerebelar tipo 2

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A 27-year-old man presenting with 11 years of progressive gait ataxia, imbalance, hand dexterity impairment, dysarthria, and urinary frequency.

Examination showed slow saccades, dysarthria, head and upper extremities tremor, dysmetria, and gait ataxia.

Brain MRI (Figure 1) showed severe pontocerebellar atrophy with hot cross bun sign.

A genetic study revealed a heterozygous 43 CAG repeat expansion of the ATXN2 gene compatible with spinocerebellar ataxia type 2 (SCA2).

The hot cross bun sign was classically regarded as a typical neuroimaging finding of the cerebellar variant of multiple system atrophy. However, it is not pathognomonic, having been described in other degenerative disorders manifesting ataxia[1,2,3].

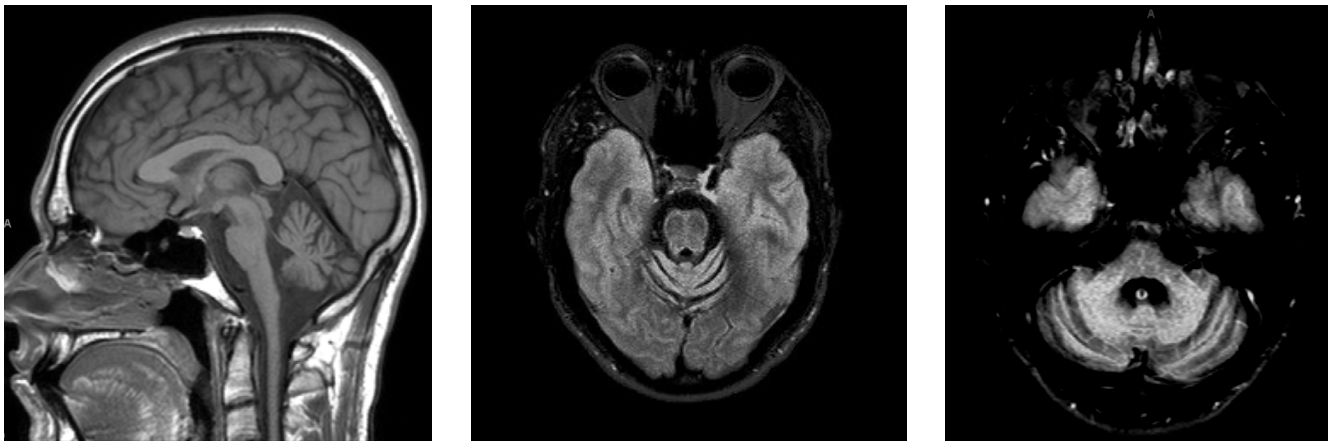

Figure 1. (A) Brain MRI showing severe pontocerebellar atrophy in sagittal T1-weighted imaging; (B and C) cruciform hyperintensity in the pons, the hot cross bun sign, in axial T2 FLAIR imaging reflecting selective degeneration of pontocerebellar tracts.

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