

Pontine hot cross bun sign in spinocerebellar ataxia type 2

“Sinal da cruz” na ponte na ataxia espino cerebelar tipo 2

María Carolina Sepúlveda SOTO^{1,2,3}, Renato Puppi MUNHOZ^{1,2,3,4}

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}.

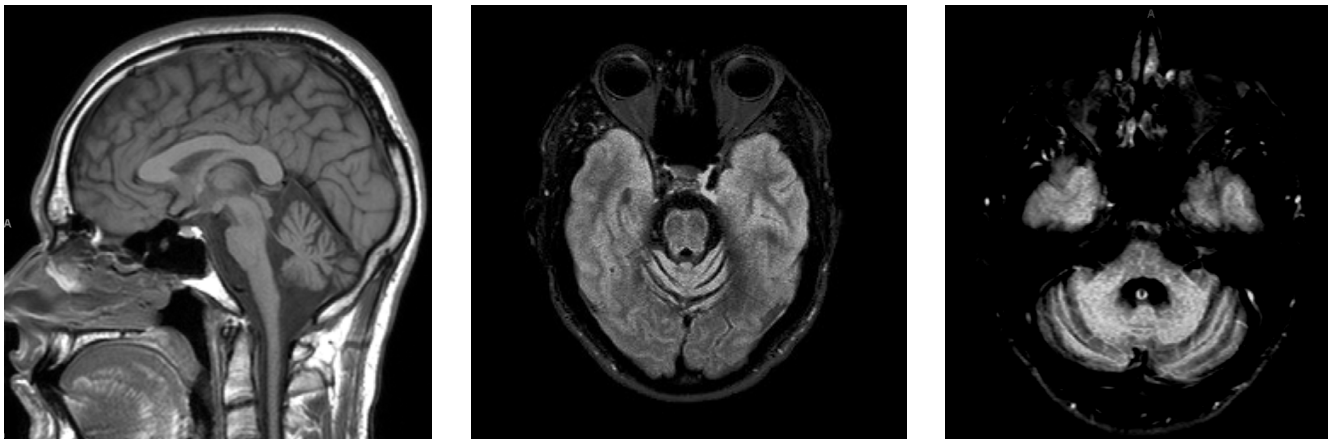


Figure 1. (A) Brain MRI showing severe pontocerebellar atrophy in sagittal T1-weighted imaging; (B and C) cruciform hyperintensity in the pons, the hot cross bun sign, in axial T2 FLAIR imaging reflecting selective degeneration of pontocerebellar tracts.

References


1. Lee YC, Liu CS, Wu HM, Wang PS, Chang MH, Soong BW. The 'hot cross bun' sign in the patients with spinocerebellar ataxia. *Eur J Neurol.* 2009 Apr;16(4):513-6. <https://doi.org/10.1111/j.1468-1331.2008.02524.x>
2. Gooneratne IK, Caldera MC, Perera SP, Gamage R. Hot cross bun sign in a patient with cerebellar ataxia. *Ann Indian Acad Neurol.* 2013;16(3):406. <https://doi.org/10.4103/0972-2327.116973>
3. Meira AT, Arruda WO, Ono SE, Carvalho Neto A, Raskin S, Camargo CH, et al. Neuroradiological findings in the spinocerebellar ataxias. *Tremor Other Hyperkinet Mov (NY).* 2019 Sep;9:10.7916/tohm.v0.682. <https://doi.org/10.7916/tohm.v0.682>

¹University of Toronto, Division of Neurology, Toronto ON, Canada.

²University Health Network, Toronto ON, Canada.

³Toronto Western Hospital, Edmond J. Safra Program in Parkinson's Disease and Morton and Gloria Shulman Movement Disorders Centre, Toronto ON, Canada.

⁴Pontificia Universidad Católica de Chile, School of Medicine, Department of Neurology, Santiago, Chile.

María Carolina Sepúlveda SOTO  <https://orcid.org/0000-0001-9425-7808>; Renato Puppi MUNHOZ  <https://orcid.org/0000-0002-4783-4067>

Correspondence: Renato Puppi Munhoz; E-mail: renato.munhoz@uhn.ca

Conflict of interest: There is no conflict of interest to declare.

Authors' contributions: SS: Design of the work, drafting the article. RPM: Conception of the work, data collection, critical revision of the article, final approval of the version to be published.

Received on April 16, 2020; Received in its final form on May 06, 2020; Accepted on May 25, 2020.