

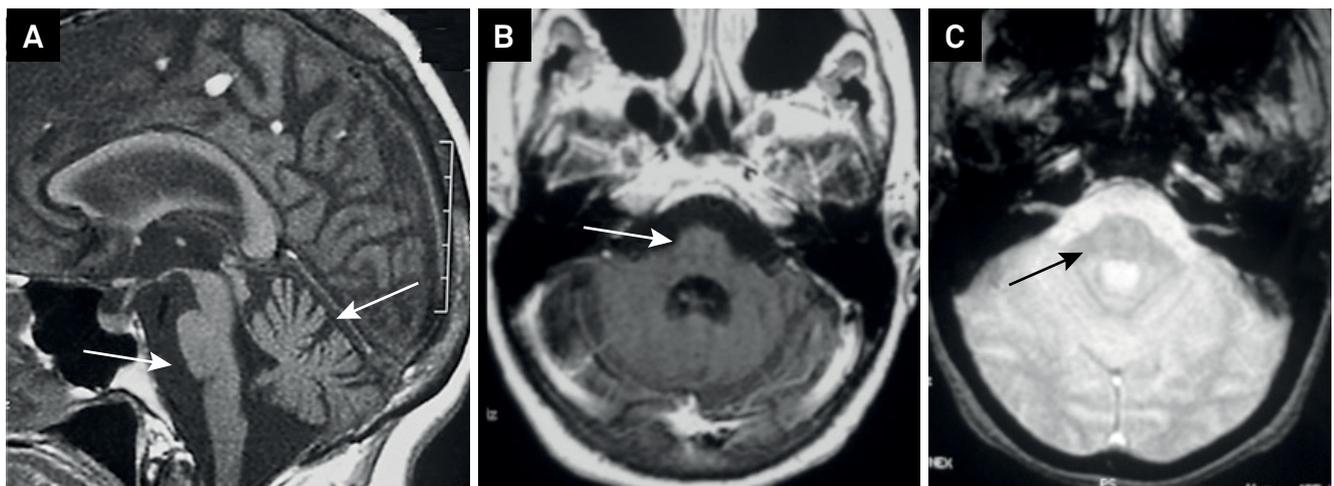
# Pontine calcification in late stage cerebellar multiple system atrophy: a marker of synucleinopathy neurodegeneration?

Calcificação da ponte na atrofia de múltiplos sistemas forma cerebelar avançada: um marcador de neurodegeneração para as sinucleinopatias?

José Luiz Pedroso<sup>1,2</sup>, Orlando G. Barsottini<sup>1,2</sup>, Marcos Knobel<sup>2</sup>, Benjamin Wolf Handfas<sup>2</sup>, Antonio J. Rocha<sup>3</sup>, Elias Knobel<sup>2</sup>

A 78-year-old woman presented with progressive ataxia, dysautonomia (hypotension and urinary incontinence), and pyramidal signs that started 17 years ago. Brain MRI performed 15 years ago showed pontocerebellar atrophy and the “hot cross bun” sign (Figure 1). Multiple system atrophy, cerebellar form (MSA-C) was diagnosed. A routine brain CT scan performed two years ago disclosed pontine calcification (Figure 2).

Multiple system atrophy is a neurodegenerative disorder included in the synucleinopathies group<sup>1</sup>. The cerebellar form, MSA-C, usually causes pontocerebellar atrophy and a “hot cross bun” sign in the pons, due to selective loss of myelinated transverse pontocerebellar fibers and raphe<sup>1,2</sup>. This prospective observation demonstrates that pontine calcification may be part of the neurodegeneration process in MSA-C.



**Figure 1.** MRI performed two years after disease onset. (A) Sagittal T1-weighted shows pontocerebellar atrophy; (B) Axial T1-weighted depicted the “hot cross bun” sign; (C) Axial gradient-echo showed no evidence of calcification in the pons.

<sup>1</sup>Universidade Federal de São Paulo, Unidade de Ataxia, Departamento de Neurologia, São Paulo SP, Brasil;

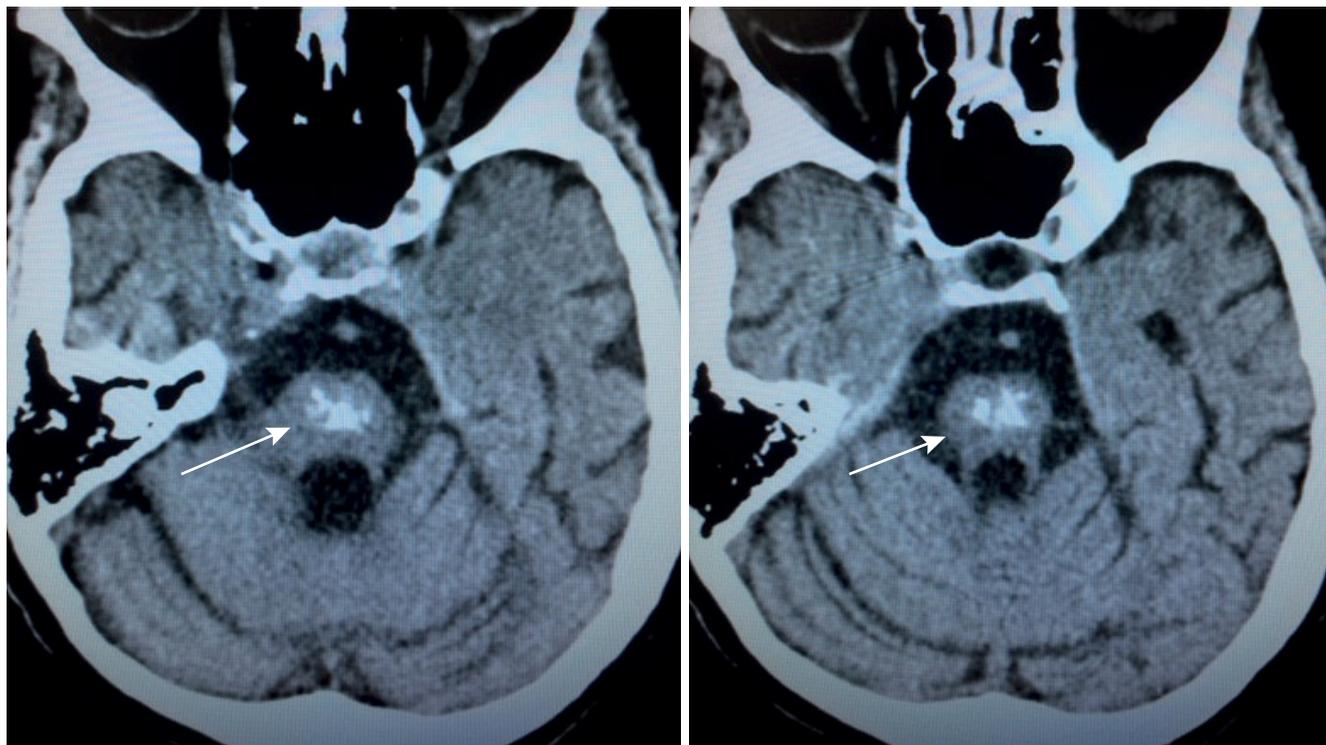
<sup>2</sup>Hospital Israelita Albert Einstein, São Paulo SP, Brasil;

<sup>3</sup>Faculdade de Ciências Médicas da Santa Casa de São Paulo, Divisão de Radiologia, São Paulo SP, Brasil.

**Correspondence:** José Luiz Pedroso; Departamento de Neurologia da UNIFESP; Rua Pedro de Toledo, 650, CEP 04039-002; São Paulo SP, Brasil. E-mail: jlpedroso.neuro@gmail.com

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

Received 10 December 2016; Accepted 12 July 2017.



**Figure 2.** Brain CT performed 15 years after disease onset. Marked pontine calcification, centered on transverse pontocerebellar fibers and raphe.

## References

1. Gilman S, Low PA, Quinn N, Albanese A, Ben-Shlomo Y, Fowler CJ et al. Consensus statement on the diagnosis of multiple system atrophy. *J Neurol Sci.* 1999;163(1):94-8. <https://doi.org/10.1007/BF02309628>
2. Nemoto H, Konno S, Nakazora H, Wakata N, Kurihara T. Pontine calcification in a case with cerebellar type of multiple system atrophy: incidental or related to synucleinopathy. *Intern Med.* 2006;45(9):667-8. <https://doi.org/10.2169/internalmedicine.45.1706>