

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?

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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 group¹. 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^{1,2}. This prospective observation demonstrates that pontine calcification may be part of the neurodegeneration process in MSA-C.

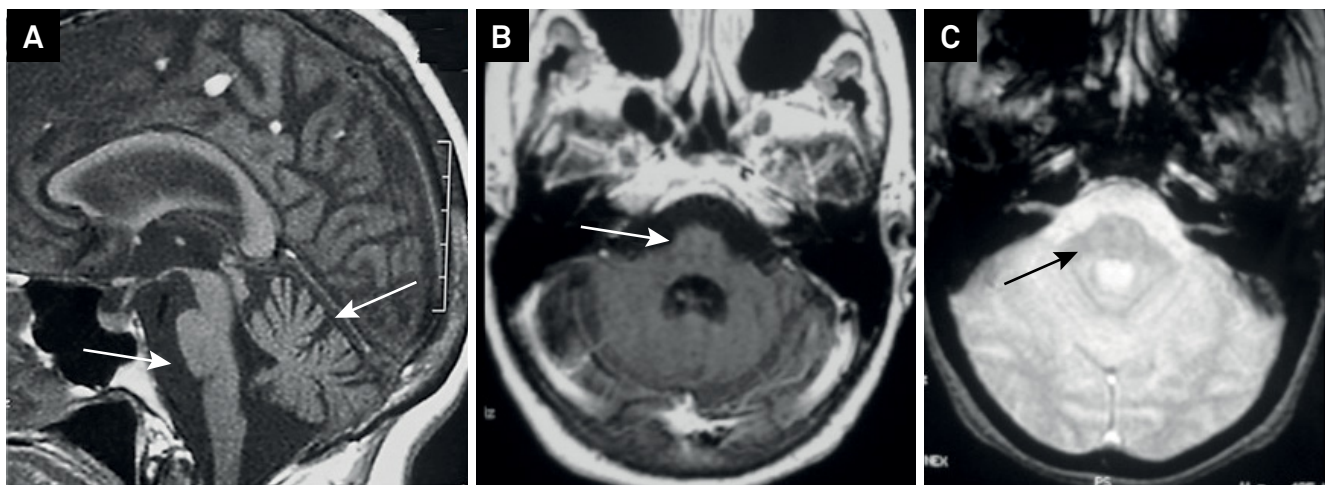


Figure 1. MRI performed two years after disease onset. (A) Sagittal T1-weighted shows pontocerebellar atrophy; (B) Axial T1-weighted depicted the “hot cross bun” sign; (C) Axial gradient-echo showed no evidence of calcification in the pons.

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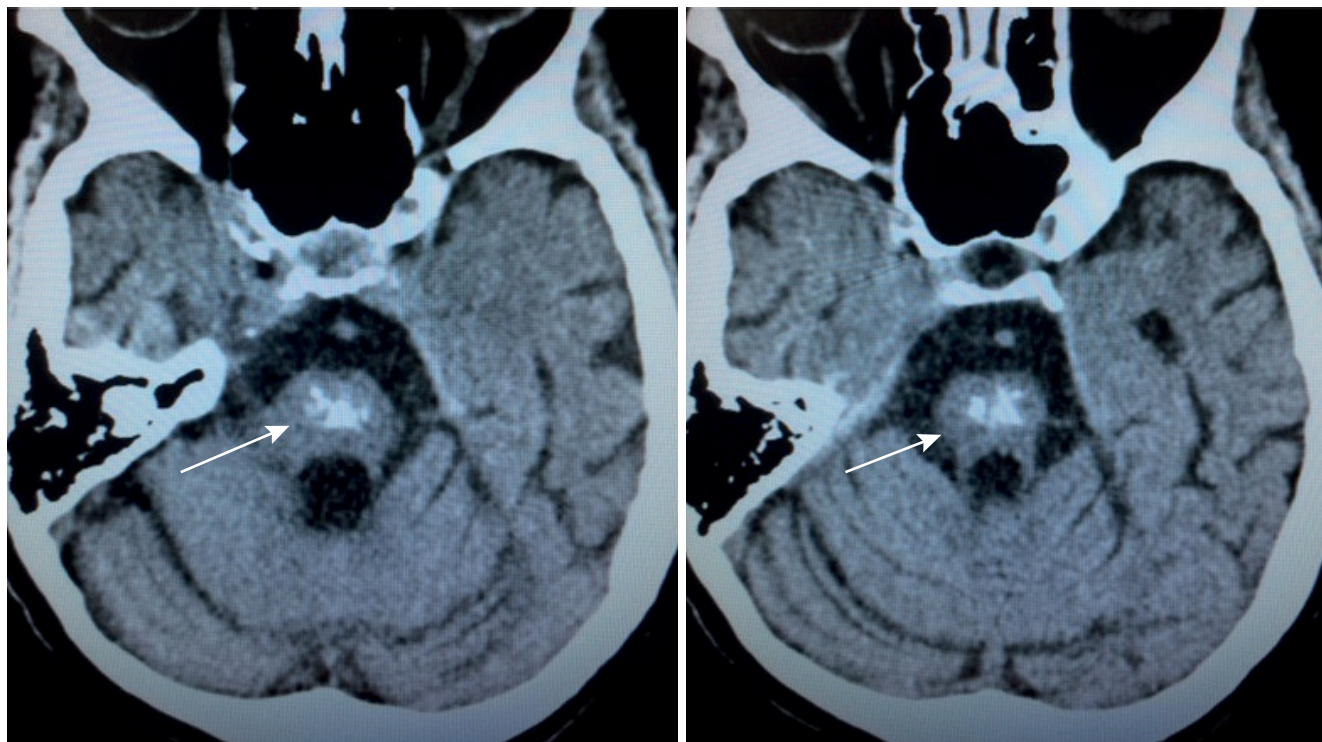


Figure 2. Brain CT performed 15 years after disease onset. Marked pontine calcification, centered on transverse pontocerebellar fibers and raphe.

References

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