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“Santiaguino” sign in multisystem atrophy

Signo de santiagoño en la atrofia multisistémica

Dear Editor,

Multisystem atrophy (MSA) is an adult sporadic neurodegenerative disease that presents as a variable

combination of autonomic dysfunction symptoms, Parkinsonism, cerebellar ataxia and pyramidalism.¹ It currently falls within the category of synucleinopathies, since its anatomopathological markers are oligodendroglial alpha-synuclein inclusions.² Magnetic resonance imaging (MRI) studies of patients with MSA with a cerebellar origin show a hyperintense lesion in the shape of a cross, called the “hot cross bun” sign, at the centre of the pons. This sign reflects the degeneration of the median raphe and the

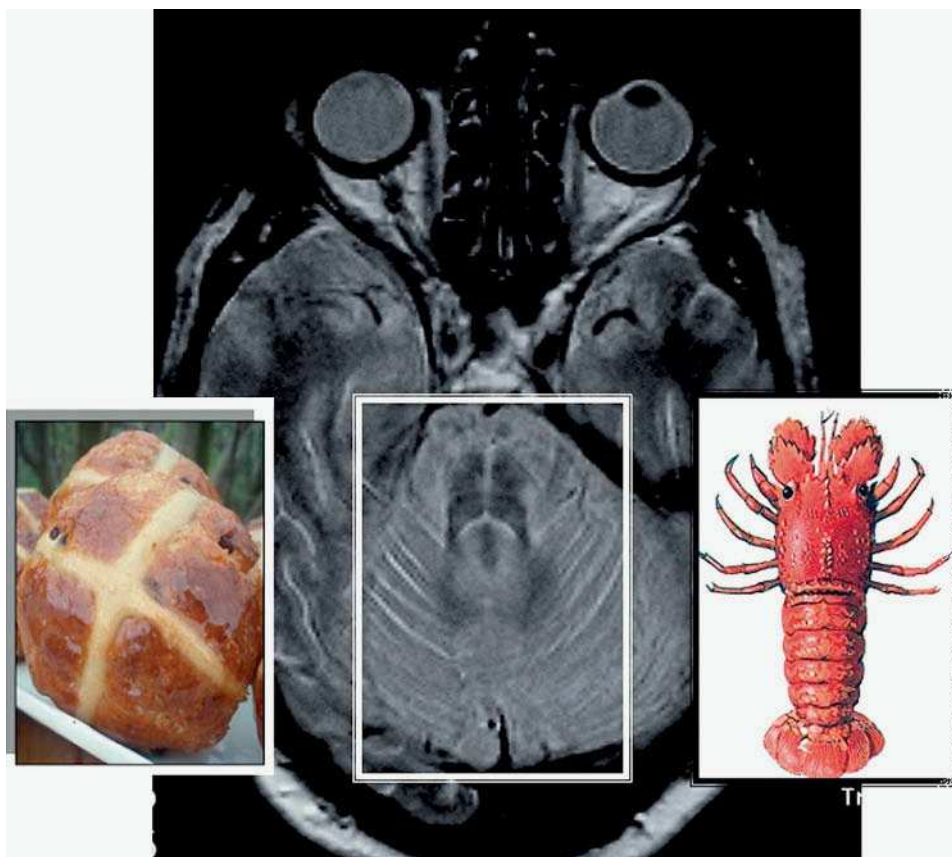


Figure 1 T2 axial MRI section: the pontine hypersignal resembles the *hot cross bun* sign (left), but when observed in association with the atrophy of the cerebellar vermis and hemisphere, it mimics the silhouette of a *Santiagoño* (right).

pontine transverse fibres that interconnect the pontine nuclei and the cerebellum through the medial cerebellar peduncles.^{3,4} The hot cross bun is a classic product of British confectionery, which was traditionally eaten on Good Friday, but its origin may be even pre-Christian. We describe a new sign for MSA with initial and predominant cerebellar dysfunction symptoms (cMSA), which we call “Santiaguíño” sign, a term having both local and universal culinary connotations.

Case history: a 65-year-old male, without personal or family history of interest, attending consultation due to difficulty in walking. He initially presented midline (and, to a lesser degree, appendicular) cerebellar ataxia. Two years later, his condition had worsened and he developed impotence and orthostatic hypotension. Later still, he presented generalised and symmetric Parkinsonism with dystonic postures and bilateral Babinski sign. He died at age 72 after an acute abdomen condition, secondary to intestinal volvulus. In the necropsy, the cerebellum, brain stem and putamen were atrophic and showed significant neuronal loss. Oligodendroglial alpha-synuclein inclusions were also observed. Figure 1 shows an axial section of the T2-weighted MRI study, that presents a cross-shaped central pontine hyperintense signal, with vermian and hemispheric cerebellar atrophy, forming the silhouette of a *Santiaguíño*.

The *Santiaguíño* (*Scyllarus arctus*) it is a very tasty, highly-appreciated variety of seafood, which breeds in the estuaries of Galicia. It is named after its distinctive cross-shaped shell, reminiscent of the cross of Santiago (Saint James). This increasingly rare delicacy undoubtedly lived in the Arosa estuary, which is where, according to tradition, the disciples of Saint James the Elder arrived with the body of their master after he was tortured and beheaded in the Holy Land in 41 BCE and shipped from the port of Haifa in a stone boat. The cross of the pons (shell) in combination

with the vermian (tail) and hemispheric atrophy form the silhouette of a “Santiaguíño” (fig. 1). The “Santiaguíño” sign was present in 4 patients with cMSA in a series of 11 patients with MSA studied at our centre.⁵

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