''Hot-cross bun sign'' in multiple system atrophy: A presentation of 3 cases☆

Signo de la cruz *(hot cross bun sign)* en la atrofia multisistémica. A propósito de 3 casos

Dear Editor:

Multiple system atrophy (MSA) is a sporadic neurodegenerative disease characterised by the association of parkinsonism with autonomic failure, cerebellar ataxia and pyramidal signs. It typically begins at about 60 years of age and affects men and women equally. Mean survival is 6—9 years. According to the predominant clinical characteristics, it can be further categorised as MSA-P when parkinsonian clinical features predominate, MSA-C when cerebellar ataxia predominates, and MSA-A when autonomic failure predominates.[1] MSA is mainly diagnosed based on clinical findings. Cranial magnetic resonance images may show putamen atrophy and infratentorial abnormalities such as the hot cross bun sign due to pontocerebellar degeneration.[2] When associated with atrophy of the cerebellar vermis and hemispheres, this image is also known by the name *''signo de santiaguiño''* in Spanish literature, due to its resemblance to a type of crustacean.[3]

In this letter, we describe 3 clinical cases of cerebellar-type MSA (MSA-C) with typical MR images showing a hot cross bun sign. Of these 3 patients, 2 met diagnostic criteria for probable MSA while the third met criteria for possible MSA. The first patient is a 62-year-old woman evaluated due to symptoms of postural instability/falling and difficulty articulating speech which had been evolving over 3 years. At a later point, she also developed urinary urgency with occasional incontinence. Examination revealed hypomimia, dysarthria, persistent horizontal nystagmus, and bilateral bradykinesia along with ataxic gait and pyramidal signs. Results from a genetic study for ataxias were negative. In light of this clinical profile, we established a suspected diagnosis of probable MSA-C. In the second case the patient was a woman aged 74 years being monitored for possible MSA-C. Symptoms initiated as an akinetic-rigid, tremor-free syndrome with poor response to L-DOPA and poor tolerance for dopamine antagonists, instability due to ataxic gait and scanning dysarthria evolving over 6 years. Our third case was that of a male patient aged 85 years who was monitored for symptoms of akinetic-rigid parkinsonism, rapidly evolving postural instability and urinary incontinence, evolving over 3 years. Response to L-DOPA was poor. Examination revealed ataxic gait, horizontal-rotary nystagmus at extreme lateral gaze and bradykinesia. In these 3 cases, neuroimaging studies showed a hot cross bun sign on the pons.

We described 3 patients diagnosed with MSA-C and typical neuroimaging findings. Finding the hot cross bun sign in the imaging study supports our diagnosis of this entity, which is essentially diagnosed on a clinical basis.

In MSA-C (historically known as olivopontocerebellar atrophy), we can observe radiological changes, mainly in T2-weighted sequences in brain MRI (Fig. 1). Changes are mostly infratentorial,[4] and include the typical hyperintense hot cross bun sign in the pons and other anomalies, such as putamen atrophy, putamen hypointensity and hyperintense putaminal rim.[2—5] The hot cross bun sign is the result of the degeneration of pontine neurons and transverse pontocerebellar fibres. This could be considered an expression of degeneration and neuronal loss in the brain stem, and serves as a parameter for measuring disease progression.[6] While the hot cross bun sign is typical, it is not pathognomonic to MSA.[7] It has also been observed in cases of spinocerebellar ataxia (SCA) types 2 and 3, and in parkinsonism with cerebellar and brain stem alterations, presumably secondary to vasculitis.[8] In our opinion, this radiological sign is not specific to MSA, but it does support that diagnosis when clinical symptoms are compatible with that entity.

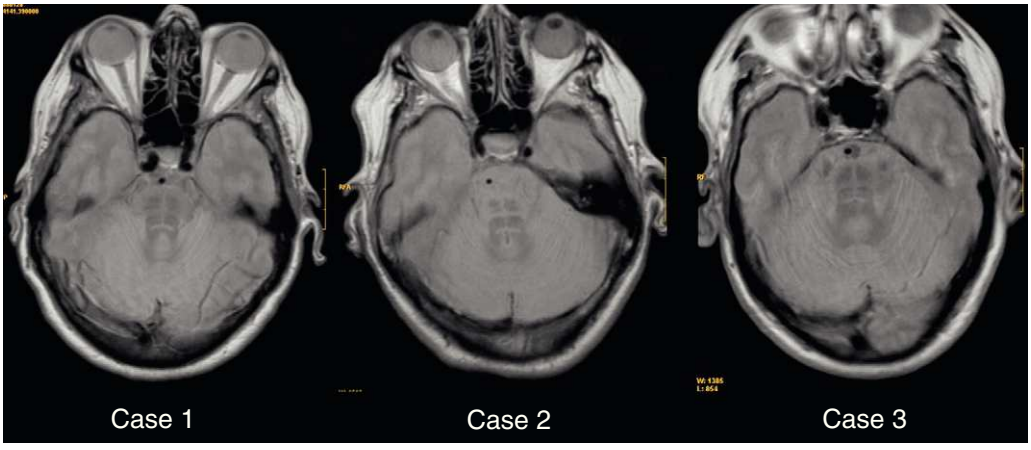

Figure 1 T2-weighted MRI images showing a cross-shaped hyperintensity (hot cross bun sign).

☆ Please cite this article as: Recio Bermejo M, et al. Signo de la cruz (hot cross bun sign) en la atrofia multisistémica. A propósito de 3 casos. Neurología. 2012;27:314—5.

References

1. Wenning GK, Colosimo G, Geser F, Poewe W. Multiple system atrophy [review]. Lancet Neurol. 2004;3:93—103.
2. Sitburana O, Ondo WG. Brain magnetic resonance imaging (MRI) in parkinsonian disorders. Parkinsonism Relat Disord. 2009;15:165—74.
3. Pereiro I, Arias M, Requena I. Signo de santiaguiño en la atrofia multisistémica. Neurologia. 2010;25:336—7.
4. Shrivastava A. The hot cross bun sign. Radiology. 2007;245:606—7.
5. Massano J, Costa F, Nadais G. Teaching neuroimagen: MRI in multiple system atrophy. Neurology. 2008;71:e38.
6. Abe K, Hikita T, Yokoe M, Mihara M, Sakoda S. The cross signs in patients with multiple system atrophy: a quantitative study. J Neuroimaging. 2006;16:73—7.
7. Srivastava T, Signh S, Goyal V, Shukla G, Behari M. Hot cross bun sign in tow patients with multiple system atrophy-cerebellar. Neurology. 2005;64:128.
8. 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;16:513—6.

M. Recio Bermejo*, S. Navarro Muñoz, B. Espejo Martinez, C. Mangas Aveleira, R. García Ruiz

Servicio de Neurología, Complejo Hospitalario La Mancha Centro, Alcázar de San Juan, Ciudad Real, Spain

* Corresponding author.
E-mail address: marecber@yahoo.es (M. Recio Bermejo).

doi:10.1016/j.nrleng.2011.12.003

Optical neuropathy in Lewis-Summer syndrome: A casual relationship?☆

Neuropatía óptica en un síndrome de Lewis-Sumner: ¿una asociación casual?

Dear Editor:

It was with great interest that we read the clinical case published in your journal under the title ''Optic neuropathy in Lewis-Sumner syndrome: Presentation of a case''.[1] We expected to read about a nosological link between central demyelinating disease in optic neuropathy and the peripheral demyelinating disease that characterises Lewis-Sumner syndrome (LSS). At the very least we thought the authors would have envisaged this possibility, upon finding both pathologies in the same patient. Instead, we found a short (but interesting) review of neuropathies with conduction block mentioning both multifocal motor neuropathy (MMN) and LSS. The differences between the two syndromes may not have been sufficiently emphasised. LSS is currently considered an asymmetric or multifocal type of chronic inflammatory demyelinating polyneuropathy (CIDP). Unlike in MMN, the sensory fibres are affected, finding anti-GM1 antibodies is uncommon, and a significant number of cases of this disease respond favourably to corticosteroid treatment.[2,3] Nevertheless, this therapeutic response has been called into question by a recent study proposing that LSS be clearly distinguished not only from MMN, but also from CIDP.[4] Lastly, regarding the case mentioned in the letter by Sánchez Ferreiro and Barreiro González, impairment of the central nervous system has been described in LSS, but as far as we know, not in MMN.[5—7] In the case recently mentioned in NEUROLOGÍA,[1] the authors describe the co-presence of LSS and retrobulbar optic neuritis in the same patient. However, neither of the pathologies is completely illustrated: no EMG results are given, and the pathogenesis of the optic neuritis is not clearly explained, since the authors list no vascular or autoimmune tests and do not specify the anatomical structure in which the magnetic resonance scan revealed demyelinating lesions. Above all, the authors' intent to establish an aetiopathogenic association between the two illnesses is never fully clarified. We therefore wonder if Sánchez Ferreiro and Barreiro González, in their letter to the editor, meant only to report the presence of two unusual pathologies in the same patient (pathologies which, furthermore, were not correctly described), or if they meant to suggest that the co-presence of two demyelinating diseases might be due to more than a mere chance. If the latter is true, the authors have unfortunately failed to transmit the interesting points of this case and their possible hypothesis.

References

1. Sanchez-Ferreiro AV, Barreiro Gonzalez JR. Neuropatía Óptica en un Síndrome de Lewis-Summer: a propósito de un caso. Neurología. 2011;26:438—9.
2. Rajabally YA, Chavada G. Lewis-Sumner syndrome of pure upper-limb onset: diagnostic, prognostic, and therapeutic features. Muscle Nerve. 2009;39:206—20.
3. Lewis RA. Neuropathies associated with conduction block. Curr Opin Neurol. 2007;20:525—30.
4. Attarian S, Verschueren A, Franques J, Salort-Campana E, Jouve E, Pouget J. Response to treatment in patients with Lewis-Sumner syndrome. Muscle Nerve. 2011;44:179—84.
5. Parry GJ. Are multifocal motor neuropathy and Lewis-Sumner syndrome distinct nosologic entities. Muscle Nerve. 1999;22:557—9.
6. Viala K, Renié L, Maisonobe T, Béhin A, Neil J, Léger JM, et al. Follow-up study and response to treatment in 23 patients with Lewis-Sumner syndrome. Brain. 2004;127:2010—7.
7. Liguori R, Rizzi R, Vetrugno R, Salvi F, Lugaresi A, Cevoli S, et al. Steroid-responsive multifocal demyelinating neuropathy with central involvement. Muscle Nerve. 1999;22:262—5.

☆ Please cite this article as: Segura T, Sánchez-Ayaso PA. Neuropatía óptica en un síndrome de Lewis-Sumner: ¿una asociación casual? Neurología. 2012;27:315—6.