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IMAGE OF THE MONTH

Sign of the cross and MSA-C

Signo de la cruz y AMS-C

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We present the case of a 50-year-old male who consulted in 2005 because of dif f culty in speaking, clumsiness in his movements, gait instability and impotenica coeundi. In the following months, he quickly and progressively developed an involvement of the different neurological systems, where we saw cerebellar syndrome, symmetric parkinsonism, autonomic disturbances and dystonia. A therapeutic trial of levodopa and dopamine agonists was undertaken, with little response. The examination showed: absence of cognitive alteration, severe dysarthria, dysphonia and velopalatine hypokinesia. There was no oculomotor palsy . Cervical dystonia with anterocollis. Bilateral mixed (rest and kinetic) tremors. Plastic rigidity with moderate, symmetric bradykinesia. Bilateral plantar cutaneous extensor response. Bilateral f nger-to-nose and heel-knee dysmetria. Ataxic gait and in anterior f exion without arm movement Push test +. Blood pressure 120/80 mmHg supine and 90/70 mmHg standing. In the complementary test study: general analysis, including ceruloplasmin, Cu in blood and urine within normal ranges. DaT scan with markedly low uptake in both striata. Normal cardiac MIBG uptake. Cranial MR with cerebellar atrophy (f g. 1), hot cross bun sign on the bulge, T2 hyperintense rim in the lateral edge of the putamen and hypoattenuation of the signal in T1-T2 sequences of the posterior putamen (f g. 2).

With this data and following the criteria of the second consensus for the diagnosis of multiple system atrophy (MSA), Neurology 2008, [1] we diagnosed MSA-C (cerebellar form of MSA).

Currently, we have a complete battery of complementary tests that are more or less accessible in clinical practice, and with greater or lesser sensitivity and specif city, but that are undoubtedly useful when pro f ling the diagnosis of atypical parkinsonisms. Among these we can highlight: *a)* smell test (altered α-synucleinopathies); *b)* polysomnography study (REM behaviour disorder in α-synucleinopathies); *c)* transcranial ultrasound (hyperechogenicity of the substantia nigra in P arkinson's disease [PD] and hyperechogenicity of the putamen in MSA); *d)* Radioisotope studies of the striatal dopaminergic pathway, both presynaptic (DaTscan) (differential diagnosis of tremor and pharmacological and vascular parkinsonism versus degenerative) and postsynaptic (IBZM) (differentiating PD from other degenerative parkinsonisms); *e)* cardiac MIBG (altered in α-synucleinopathies, and early and severe in PD); and *f)* cranial MRI.

We would like to point out the importance of MRI as a diagnostic tool in atypical parkinsonisms and especially in MSA. Schrag et al [2,3] described their diagnostic use, establishing a high speci f city but low sensitivity . The characteristic signs are: *a)* at supratentorial level: T2 hyperintensive putaminal lateral rim or edge, T1 hypointensity and atrophy of the putamen, and *b)* on infratentorial level: cerebellar , pontine and middle cerebellar peduncle (MCP) atrophy , and T2 hyperintensity in bulge cruciform, known as hot cross bun sign (due to its similarity to a typical bun eaten at Easter in the Anglo-Saxon

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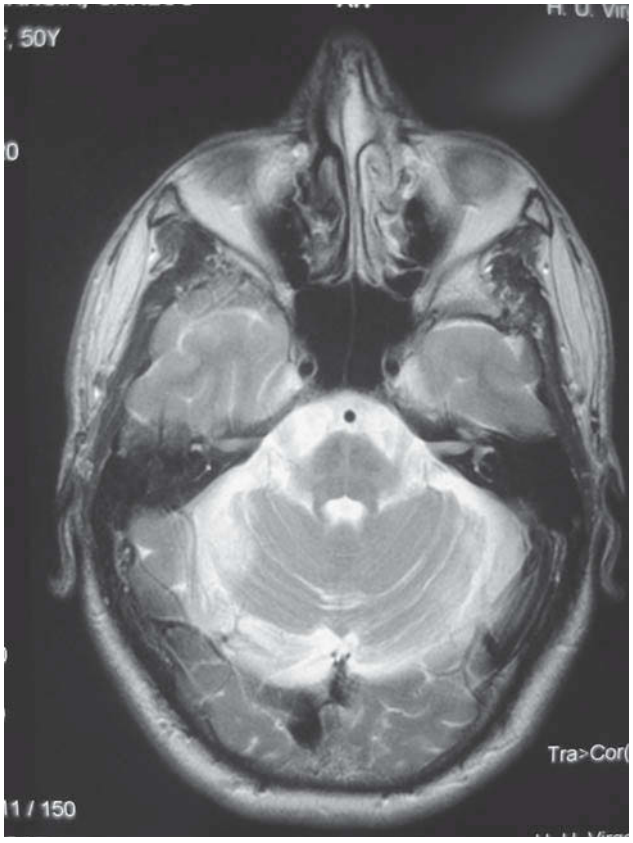

Figure 1 Cranial MRI. T2-weighted sequence, axial section. We can see a cross (hot cross bun sign), with cruciform hyperintensity in the pons. Cerebellar atrophy.

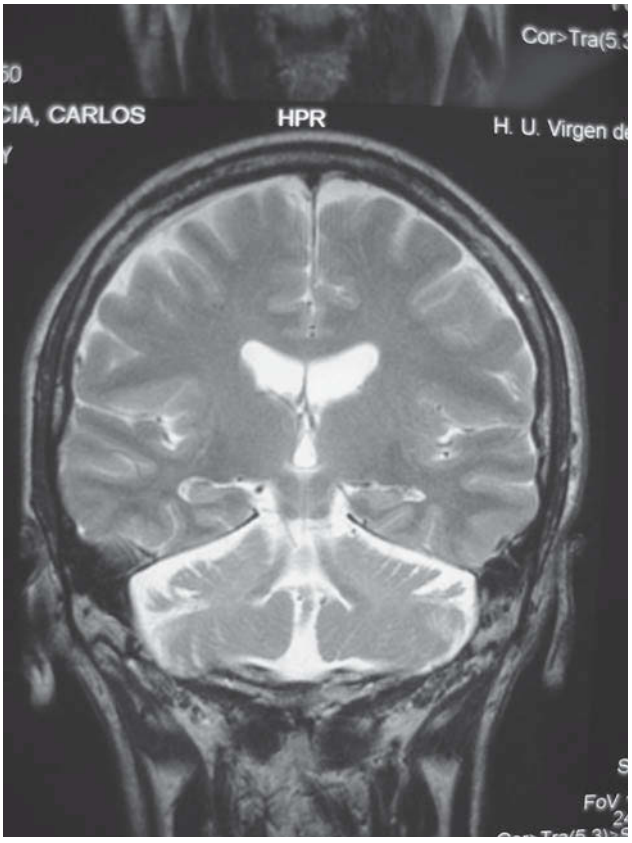

Figure 2 Cranial MRI. T2-weighted sequence, coronal section. Hot cross bun sign in coronal view Hyperintense lateral edge or rim of the putamen, with putaminal hypointensity and atrophy.

world). In a recent review, Sitburana et al[4] found atrophy of the putamen in 100% of MSA cases, putaminal hypointensity in 60%, and hyperintensive putaminal rim in 36%. The frequency with which the cross sign appears in MSA is not well defined, as it is not as common as first thought. Lastly, the new diffusion techniques with an increase in the ADC map in putamen and MCP, and volumetry with atrophy of the putamen allow us to differentiate an MSA from PD.[5,6] The hot cross bun sign or sign of the cross (as it is known in Spanish) is very characteristic of MSA-C, although it has also been exceptionally described in spinocerebellar ataxias and trunk vasculitis. The image of the cross is due to the preservation of the corticospinal pathway and nuclei in the pontine tegmentum, with concomitant degeneration of the pontocerebellar fibres. Although not a pathognomonic sign, if it is found in the context of atypical parkinsonism, with dysautonomia, ataxia and little response to levodopa, it is very suggestive of MSA-C.

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