YOUNG WOMAN WITH MULTIPLE SCLEROSIS AND SYSTEMIC LUPUS ERYTHEMATOSUS: CONTRIBUTION TO DIFFERENTIAL DIAGNOSIS BY BRAIN MRI USING T2*SEQUENCES.

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Background

The frequency of MRI lesions presenting the "central vein sign", a marker specific to the inflammatory demyelinating lesions and not visible in ischaemic lesions, was established by T2* sequences [1]. This technique can identify the central vein sign in demyelinating lesions distinguishing between MS and CNS inflammatory vasculopathies [2]. The frequency of demyelinating lesions presenting the central vein sign characteristic of MS is at least 70%, whereas in SLE the highest frequency thus far observed is 30% [2].

Aim

To establish by brain MRI scans including T2* sequences, the nature of brain lesions in a rare case of multiple sclerosis (MS) and Systemic Lupus Eritematosus (SLE) comorbidity [3].

Clinical Picture

In 2007, a 25-year-old woman developed unilateral leg weakness and paresthesia. Brain MRI lesions fulfilling the diagnostic criteria for MS were present along with a CSF oligoclonal profile. Thus diagnosis of MS was made and Interferon-beta therapy was started but interrupted in August 2013 following pleuritis. The clinical course was then complicated by pericarditis and renal failure with nephrotic range proteinuria. At that time auto-antibody titer became positive: ANA 1:640, antidsDNA 1:320. High titers of anti-nucleosomal, anti-Sm, anti-histone, anti-RNP antibodies and low levels of complements were also present as well as normocytic anemia and elevated ESR and CRP. Lupus anticoagulant testing and antiphospholipid antibodies were negative. This clinical picture subsided after steroid therapy, even if in December 2013 ANA, antidsDNA and ENA were still positive. A subsequent renal biopsy demonstrating a diffuse proliferative glomerulonephritis (class IV) allowed the diagnosis of SLE. Treatment with azathioprine and oral corticosteroids was started and no further flares were observed although in April 2017, auto-antibodies and proteinuria were still present. In 2015 the onset of ataxia and paraparesis associated with new lesions on brain MRI indicating disease reactivation suggested Rituximab add on, that was administered following the protocol used in MS. Since then the patient has been neurologically stable.

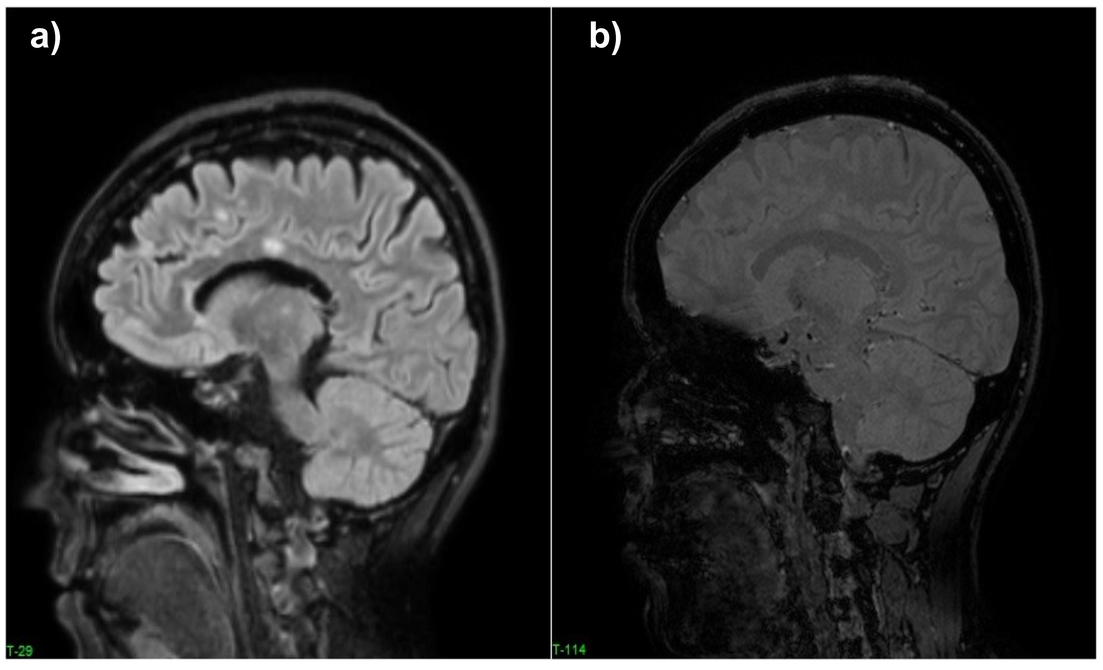

Fig. 1: Lesions hyperintense on FLAIR without central vein sign on SWI. a) Sagittal FLAIR MRI scan showing pericallosal and juxtacortical lesions. b) Sagittal SWI MRI scan showing how these lesions do not present the central vein sign.

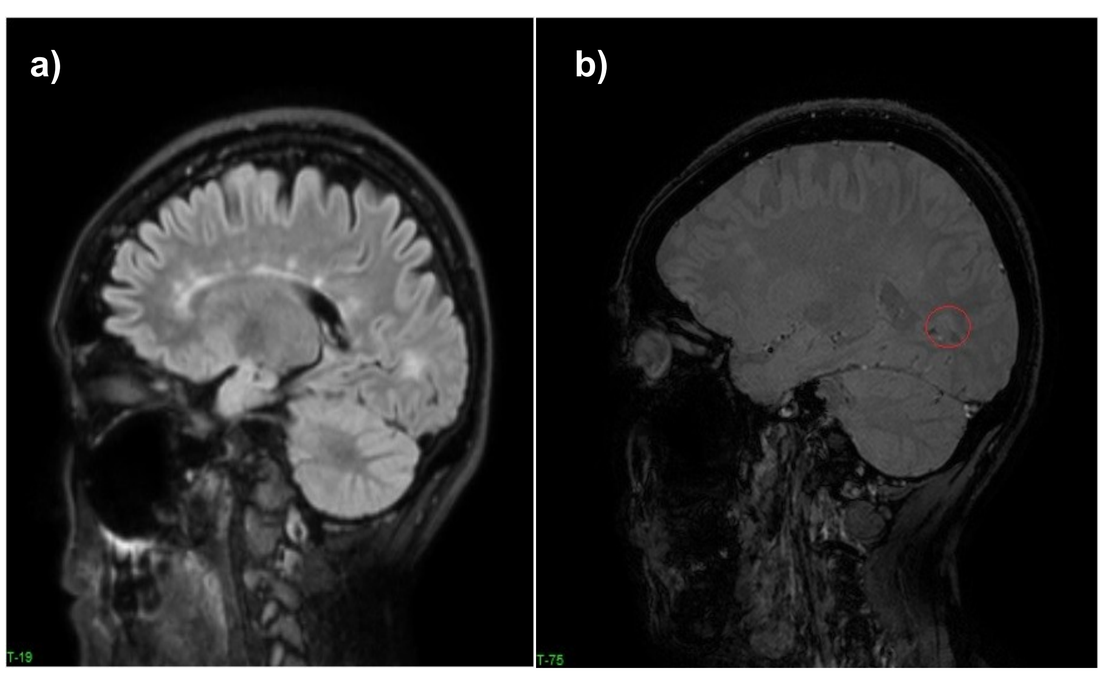

Fig. 2: Lesion hyperintense on FLAIR presenting central vein sign on SWI. a) Sagittal FLAIR MRI scan showing an occipital lesion. b)) Sagittal SWI MRI scan showing how this lesion presents the central vein sign (red circle).

Results

The frequency of demyelinating lesions presenting the central vein sign in our case was 46%.

Discussion and conclusion

In this patient the frequency of the central vein sign observed was remarkably lower than the threshold observed in MS but higher than the frequency thus far observed in SLE. These data suggest that the nature of the brain lesion may be mainly ischemic due to CNS vasculitis secondary to SLE.

Bibliografia

[1] Mistry et al. Central veins in brain lesions visualized with high-field magnetic resonance imaging: a pathologically specific diagnostic biomarker for inflammatory demyelination in the brain. JAMA Neurol. 2013 May; 70(5):623-8.
[2] Vuolo et al. Susceptibility weighted MRI can help differentiate pathogenesis of white matter lesions in MS and CNS inflammatory vasculopathies. Abstract for 2015 EAN congress.
[3] A.Fanouriakis et al. Coexistence of systemic lupus erythematosus and multiple sclerosis: Prevalence, clinical characteristics, and natural history. Seminars in Arthritis and Rheumatism (2013)