DIFUSSE WHITE MATTER ABNORMALITIES AND NEUROLOGIC MANIFESTATIONS IN A PATIENT WITH ANTI-RO POSITIVE ANTIBODIES. CASE REPORT.
Liliana Robles MD1, Leopoldo Dealvare MD2, Pilar Guillermo Prieto MD1, George J. Hutton MD1, Victor M. Rivera MD1
1Baylor College of Medicine, Department of Neurology; 2Neurological Associates of LA

INTRODUCTION
Anti-Ro (SS-A) antibodies are found in 70% to 97% of patients with primary Sjögren syndrome (pSS) and in 10% to 60% of patients with systemic lupus erythematosus (SLE). Patients with secondary Sjögren associated to rheumatoid arthritis may present with SS-A antibodies in up to 10% to 15% of the cases.

Anti-Ro positivity in pSS has been correlated with severe, progressive and large CNS lesions but the role of isolated anti-Ro (SS-A) antibodies in severe multifocal demyelination in absence of rheumatologic conditions is undetermined.

We describe the case of a 32 year old male with anti-Ro antibodies and severe CNS MRI abnormalities without criteria for systemic lupus erythematosus (SLE), primary Sjögren syndrome (pSS) or rheumatoid arthritis. Other demyelinating diseases and leukodystrophies were investigated.

CASE REPORT
A 32 year old man with history of meningitis at 9 months of age without sequelae and one episode of transverse myelitis 10 years ago from which he completely recovered, presented with new neurologic symptoms in 2008.

During the 1999 evaluation, brain MRI showed diffuse signal abnormalities predominantly in the subcortical white matter of both cerebral hemispheres with no contrast enhancement. (Fig. 1 and Fig. 2) The spinal MRI demonstrated high signal on T2 sequences extending from C4-T1. (Fig. 3) He was treated with IVMP and oral taper of prednisone. No definite diagnosis was established.

He remained asymptomatic until June 2008 when he presented with sudden onset of dysarthria, anemia and right sided weakness involving face, arm and leg.

MRI of brain demonstrated extensive T2 periventricular white matter abnormalities with areas of contrast enhancement in the left frontal lobe and right parietal lobe on T1 sequence. (Fig. 4 and Fig. 5) No hypointensities were evident on T1 axial images. (Fig. 6) Cord lesions were appreciated on MRI at C5-C6, C7-T1 and below T5 level (Fig. 7 and Fig. 8).

Further testing included normal results for VEP, BAEP and SSEP, negative serum NMO antibody and normal serum VLCFA levels. The MRI spectroscopy was consistent with demyelination. (Fig. 9) Anti-Ro antibody returned positive. Follow up MR images continued to show significant post-contrast enhancement. (Fig. 10)

He was placed on oral immunosuppressive treatment (azathioprine) and continues follow up.

CONCLUSIONS
The anti-Ro antibody detects an intracellular RNA-protein complex. It is primarily found in patients with SLE and Sjögren syndrome. This autoantibody is described to have a 54% sensitivity and 82% specificity in pSS and only 0.1% to 0.5% of control samples have tested positive.

Approximately 25% of patients with positive SS-A antibodies and undifferentiated connective tissue disease will evolve into a definite disorder. Neurologic involvement occurs in 20% of the patients with pSS and was present prior to diagnosis in 81% of the patients in a small case series.

Anti-Ro positivity in pSS has been correlated with severe, progressive and large CNS lesions. Yet the role of anti-Ro antibodies in CNS involvement in the absence of clinical and laboratory data of connective tissue disease is undetermined.