# Myelitis and Sjogren's Syndrome

Nigar Dargah-zada MD, Dewitt Pyburn MD
Department of Neurology, Mount Sinai Beth Israel

## OBJECTIVE

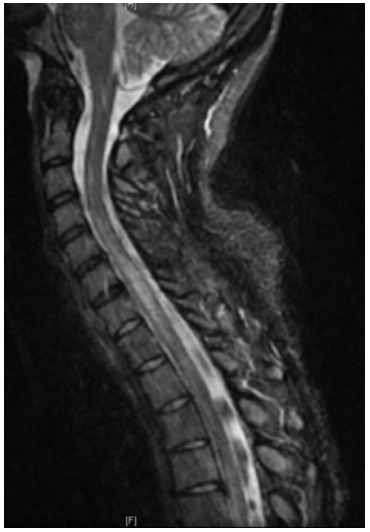
- To present a case of longitudinal transverse myelitis in the absence of typical antibodies and elevation of SSA-B Sjogren

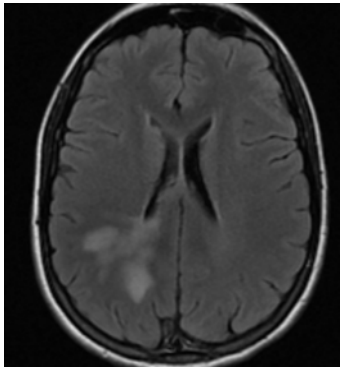
## BACKGROUND

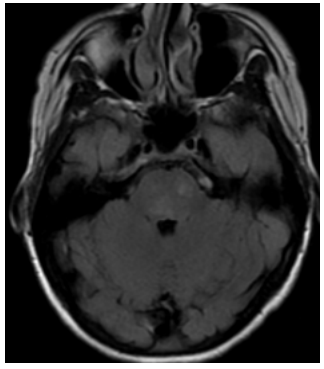
- Sjogren's syndrome is a rare cause of myelitis

## METHODS

- 33 years old Female with history of Hashimoto thyroiditis and shingles in childhood presenting with bilateral legs numbness and tingling, and subsequent bilateral lower extremities weakness and urinary retention. Neurological exam revealed symmetric proximal weakness of both legs with preserved reflexes and sensory level to mid-thoracic spine. Magnetic Resonance Imaging (MRI) of the spine showed hyperintense spinal cord lesion from C4-T1 without enhancement post-gadolinium administration. MRI brain showed demyelinating lesions perpendicular to right lateral ventricle and within the pons and ventral medulla, and an enhancing lesion in white matter of right parietal lobe. CSF showed lymphocytic pleocytosis and elevated protein. Patient was started on antibiotics/antiviral and IVIG; high-dose Solumedrol was then added. She was transitioned to oral steroids and azathioprine after completion of five days course of pulse-therapy with steroids and IVIG. The extensive laboratory workup showed negative Aquaporine-4 antibodies (Aquap-4 Ab), no oligoclonal bands, normal IgG index, negative angiotensin-converting enzyme, negative human T-lymphotropic viruses I/II antibodies, negative antinuclear antibody (ANA), no antibodies to myeloperoxidase, and elevated SSA- Sjogren antibodies, myelin basic protein and anti-TPO and anti-TG Abs. Aquap-4 Ab was rechecked on outpatient basis, and was again negative. Patient continued with clinical improvement. Repeat MRI of C/T spine and brain showed resolution of all lesions. We suggest that LETM in this case was associated with Sjogren's syndrome (SS) without systemic manifestations.







## RESULTS

- The prevalence of myelitis in patients with Sjogren's syndrome is unknown, but is possibly less than 1% (Vincent,2003). We searched relevant literature of the reported myelitis cases associated with Sjogren's syndrome, and found several cases, presenting with acute myelitis with or without systemic manifestations of SS and lack of Aquap-4 antibodies, similarly to our patient. Two cases of chronic progressive myelitis with neurological manifestations of SS were described in series of 93 patients (Moreira, 2015), one case of LETM and evidence of leukoencephalopathic changes in brain with subsequently biopsy proven SS (Cheah, 2011), one case in series of three without Aquap-4 antibodies (Almagro, 2014), one case of relapsing myelopathy (Chahin, 2009),a case of acute transverse myelitis (TM) and polyneuropathy in absence of Sicca symptoms (Tristano, 2006) a case of seronegative patient without Sicca symptoms, and SS confirmed by the biopsy (Tilki, 2017), a pediatric case of TM and concurrent optic neuritis secondary to Sjogren's syndrome (Lim) and a prospective study with report of four patients in series of 6, who had fulfilled the histological or serological criteria of the American European Consensus Group for SS. (Yadav, 2011). Our case is another example of an acute LETM and evidence of demyelinating brain lesions in the absence of Aquap-4 antibodies and oligoclonal bands, and in presence of SSA- Sjogren antibodies, with good response to immunosuppressive therapy.

## CONCLUSION

- Sjogren's syndrome is a rare cause of myelitis, that needs to be further investigated.

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