Abstract

Neuromyelitis Optica spectrum disorder (NMOSD) is a rare demyelinating condition of the central nervous system characterized by optic neuritis and long segment transverse myelitis. Previously thought to be related to multiple sclerosis, it is now identified as a distinct entity with seropositivity for AQP4 antibodies although seronegative forms are also present. Systemic sclerosis (SS) may have associated neurology but demyelination has not been reported. The disease association with NMOSD is not well established despite Sjogren syndrome and SLE having a positive association with NMOSD

We report a case of NMOSD with limited cutaneous systemic sclerosis (LcSS) in a Sri Lankan female with recurrent episodes of optic neuritis and transverse myelitis since the age of 32 years with negative cerebrospinal fluid (CSF) AQP4/ Anti-MOG antibody levels. CSF showed elevated proteins with negative oligoclonal bands. Visual evoked potentials (VEP) were delayed on the affected eyes and MRI showed T2 high intensity short segment lesions in cervical and dorsal spines consistent with demyelinating plaques. Subsequently the patient developed clinical features of LcSS and Sjogren syndrome with supportive histology. Her ANA, Anti-Scl70 and anti-centromere antibody levels were all negative. She was treated with methyl prednisolone pulses and Intravenous rituximab and continued on HCQ and oral steroids with favorable response.

NMOSD is known to have an association with other autoimmune diseases such as SLE and Sjogren syndrome but SS is seldom reported. The relationship between NMOSD and SS is not well understood although predisposition to the diseases in the background of an autoimmune milieu is a possibility. Antibody positivity has been a hallmark in the reported cases but antibody negativity can occur in each of the diseases. The positive relationship between the diseases in the context of negative serology needs better understanding and may help in future therapeutics.