AQUAPORIN 4 ANTIBODY–NEGATIVE NEUROMYELITIS OPTICA SPECTRUM DISORDER (NMOSD) ASSOCIATED WITH SJÖGREN'S SYNDROME

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BACKGROUND

Longitudinally extensive transverse myelitis (LETM) is defined as a myelitis that extends over at least three vertebral segments in length at spinal cord magnetic resonance imaging (MRI). It is frequently associated with severe clinical presentation. Several conditions can be associated with LETM, although it is classically related to aquaporin (AQP4)-antibodies neuromyelitis optica spectrum disorders (NMOSD) or more recently to myelin oligodendrocyte glycoprotein (MOG)-antibodies disease. Serum anti-AQP4 antibody is a sensitive and highly specific marker for NMOSD. Patients with NMO frequently have an accompanying autoimmune disease, most commonly Sjögren's syndrome (SS) and lupus. AQP4 antibody–negative NMO is rare and coexisting autoimmune disorders are more frequent in AQP4–Ab–positive than negative cases.

CASE REPORT

A 59-year-old female was admitted with a 1-month history of malaise and ascending paresthesia of the lower limbs, urinary and fecal retention, dysphonia and asymmetric tetraparesis with hyperreflexia and tetrahypoesthesia. During hospitalization, she developed hypercapnic respiratory failure and need for orotracheal intubation. Neuroaxis MRI showed hypersignal on T2-weighted extending from the posterior aspect of the medulla to the T2 level in the thoracic cord along with smaller ones at T8 to T10 levels and conus, without supratentorial lesions. Ophthalmologist did not evidence optic neuritis. Cerebral spinal fluid analysis demonstrated 66 leukocytes/mm³ (57% lymphocytes), increased protein (151 mg/dL), normal glucose (66 mg/dL), normal protein electrophoresis, no oligoclonal bands and negative microbiological tests. NMOSD was suspected and intravenous pulse therapy with methylprednisolone (1 g/day for 5 days) plus plasmapheresis were started. Further work-up evidenced positive ANA test (polar cytoplasmic pattern, 1/80), positive anti-SSA/Ro (45 U/mL), negative rheumatoid factor, anti-SSB/La, anti-dsDNA, anti-Sm and antiphospholipid antibodies, normal C3/C4 levels, negative infectious serologies, normal B12 vitamin and normal chest/abdomen CT scans. Moreover, anti-AQP4 was absent. Despite no sicca symptoms, investigation for SS revealed Schirmer's test

CONCLUSION

This is a rare case of AQP4-antibody-negative NMOSD associated with SS. The patient had some features which are more frequent in AQP4-antibody–positive cases (>70%), such as older age onset and coexisting autoimmune disorder. This case draws attention to this association (NMOSD and SS), and necessity for prompt immunosuppressive treatment, given that high rate of relapse in the following 24 months.

KEYWORDS

Neuromyelitis, Aquaporin, Sjögren's syndrome.