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PRIMARY SJOGREN'S SYNDROME: SEVERE NEUROLOGICAL MANIFESTATION WITH ASSOCIATION OF ANTIBODIES ANTI-RO AND ANTI-AQUAPORIN 4.

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BACKGROUND

Sjögren's syndrome (SS) is an immune-mediated systemic disease, characterized by an organ-specific lymphocytic infiltrate in the epithelium of exocrine glands and extraglandular systemic manifestations. Transverse myelitis (TM) in the SS is often extensive longitudinal involving more than three vertebral segments, most common in the cervical medula.

CASE REPORT

G.G.C, female, 27 years old, diagnosis of SS (xerostomia, xerophthalmia, anti-Ro> 240, Schirmer and green Lysamine positive, ANA HEp2 1:1280, fine dotted nuclear pattern, anti-SM and anti-DNAn negatives). It reports intense pruritus in the upper limbs and trunk and traumatic excoriations in the affected sites. After one week it evolved with moderate-intensity bilateral fronto-temporal headache, associated with episodes of vomiting, progressing with right hemiparesis and decreased peripheral sensitivity in face, upper left limb and lower limbs. With resolution of the headache and a slight improvement in hemiparesis after two days of use of Prednisone (1.7mg/kg/day). The Nuclear magnetic resonance (NMR) of spine evidenced spinal cord injury compatible with extensive TM of the bulb region until T11 and there was no change in skull image. Eye fundoscopy: normal. Anti-aquaporin 4 (AQP4) antibody is positive. Infectious diseases were ruled out and methylprednisolone (1g/day) during 5 days was started, with partial improvement of the neurological signs and the pruritus ceased. Rituximab infusion was considered, but three days after methylprednisolone, neurological symptoms worsened and she presented a respiratory tract infection, and so, plasmapheresis and immunoglobulin were given, with no improvement of neurological symptoms. After resolution of infection Rituximab was started and there was clinical and image improvement. Past four months of infusion it persists only with tonic spasms in upper limbs and allodynia in T11 dermatome.

CONCLUSION

Young patient, with TM seropositive for AQP4 in the context of the primary SS of recent diagnosis. Term of spectrum disorders optic neuromyelitis includes positive syndromes for AQP4 that may only partially meet the contemporary criteria for the diagnosis of optic neuromyelitis (prominent involvement of the optic nerve and spinal cord), the patient did not has ocular involvement. Association between positivity anti-Ro and AQP4 is already described in the literature, in a series of cases with good response to immunosuppressive agents.