EXTENSIVE LONGITUDINAL MYELITIS AS THE INITIAL PRESENTATION OF PRIMARY SJÖGREN'S SYNDROME

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BACKGROUND

Sjögren's syndrome (SS) is an autoimmune disease characterized by the presence of inflammatory infiltrate in exocrine glands and involvement of internal organs. It usually starts with symptoms of oral and ocular dryness and the involvement of the central nervous system is uncommon. Devic's disease (DE) is characterized by transverse myelitis and optic neuromyelitis. There are descriptions in the literature of the association between DE and primary SS, making the differential diagnosis difficult.

CASE REPORT

Female, 40 years, previously with hypertension, achalasia and gastric paresis without endoscopic evidence, complaining of paresthesia in the right upper limb (RUL) beginning in February 2021. At the time, she was diagnosed with complex regional pain syndrome and was medicated with gabapentin and duloxetine with symptomatic relief. After 2 months, she started with proximal paresthesia in the left leg and after 15 days with paralysis. She reported a purplish coloration in the fingers, in addition to a previous episode of paresthesia in the chest. In May, she entered the emergency with decreased visual acuity, mobile scotoma of the left eye and a history of urinary retention requiring a urinary catheter. Electroneuromyography exams were normal, ANA + fine speckled nuclear pattern 1/640 and FR 109. The cranial tomography showed no acute alterations. Magnetic resonance of the brain and spine showed cervicothoracic myelitis with an inflammatory pattern. Spinal tap showed a cerebrospinal fluid in rock water with an oligoclonal band and increased protein. Anti-Ro (165), anti-La (55) and anti-aquaporin 4 positive. Left lower lip biopsy with chronic sialadenitis (FS = 1). On physical examination, she presented Raynaud's phenomenon and edematous hands, asymmetric tetraparesis, decreased patellar and Achilles reflexes, in addition to thermal hypoesthesia below the T1/T2 level and unchanged cranial nerves. She underwent pulse therapy with methylprednisolone 1 g associated with plasma exchange, and after, gamma globulin for 5 days. Intravenous cyclophosphamide 1 g monthly for 6 months and rituximab 1 g with an interval of 15 days were also prescribed, and good clinical and laboratory response were obtained.

CONCLUSION

The relation between DE and SS is still unknown, but it can occur in 2 to 30% of cases. The presence of anti-aquaporin-4 may be useful in the differential diagnosis. Our case is characterized by a difficult differential diagnosis and an infrequent initial presentation of SS. In many cases, the start of immunosuppressive treatment can be delayed, leading to irreversible consequences.

KEYWORDS

Longitudinal myelitis, Sjögren's syndrome, Myelitis.