AMYOTROPHIC LATERAL SCLEROSIS IN PATIENT WITH SJÖGREN'S SYNDROME: A CASE REPORT

Cristiane Junqueira de Carvalho^{1,*}, Eduarda Demoner Paseto¹, Arthur Márcio Sabatini Santos¹, Alexandre da Costa Pinto Lopes¹, Sarah Kissia Jorge Ruas¹

1.Universidade Federal de Viçosa, Viçosa (MG), Brazil.

*Corresponding author: cristiane.carvalho@ufv.br

BACKGROUND

Sjögren's syndrome (SS) is a chronic autoimmune disease that affects the exocrine glands, causing mainly symptoms such as xerophthalmia and xerostomia, besides extraglandular manifestations such as pneumopathies and neurological disorders. Amyotrophic lateral sclerosis (ALS) is a neurodegenerative disease that causes progressive and irreversible motor paralysis. The purpose of this report is to present the case of a patient with SS who developed a condition suggestive of ALS. Therefore, it is questionable whether the motor neurological involvement may be secondary to SS.

CASE REPORT

MCFC, 69 years old, female, started in 2014 with xerostomia, xerophthalmia, erythematous skin lesions, dyspnea on moderate effort, chronic cough and Raynaud's phenomenon. Laboratory tests revealed rheumatoid factor of 275 IU/mL, anti-RO 240U, anti-LA < 7U and polyclonal increase in gamma globulins. High-resolution computed tomography chest showed distortion of lung parenchyma architecture, fibrotic reticulation with honeycomb lung, and traction bronchiectasis, evidencing interstitial lung disease. Pulmonary function test revealed moderate mixed ventilatory disorder. Diagnosis of SS was established in 2014 and the treatment started with prednisone, azathioprine, and hydroxychloroquine. Despite the immunosuppressive therapy, in early 2020 she developed dysarthria, sialorrhea, pain and weakness in lower and upper limbs. The physical exam showed flaccid/spastic quadriparesis, dysphagia, steppage gait, fallen foot on the right, spasticity on the left foot, fasciculation in the upper limbs, increased reflexes, absence of clonus, and atrophy of the right arm. Electromyography revealed a diffuse neuropathic process, exclusively motor, asymmetric, diffuse signs of chronic reinnervation, sparse signs of active denervation and motor instability, compatible with involvement of the alpha motoneuron. Diagnosis of ALS was made and the treatment started with riluzole.

CONCLUSION

Sjögren's syndrome can cause sensory neurological disorders by compromising the neuronal body of the sensory ganglion, and its most common neurological complication is peripheral neuropathy, particularly sensory polyneuropathy. However, central nervous system involvement is much less common, as is damage to cranial nerves and the autonomic nervous system. In the researched literature, it was not possible to correlate the pathophysiological mechanisms of SS and ALS. Some case reports describe motor neuron involvement in SS, imitating ALS, but in a sensory and motor polyneuropathy presentation. An English study reported an increased incidence of ALS in patients with autoimmune diseases, including SS, stating that this association is mainly due to shared but unclear genetic and environmental factors.

KEYWORDS

Sjögren's syndrome, Amyotrophic lateral sclerosis, Motor neurons, Nervous system diseases, Autoimmune diseases.