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Case Report

ALS in association or component of the clinical picture of sjögren's syndrome: a case report

ELA em associação ou componente do quadro clínico da síndrome de sjögren: um relato de caso

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ABSTRACT: Introduction: Sjögren's Syndrome (SS) is an immune-mediated systemic inflammatory disease related to the involvement of the exocrine glands. However, its heterogeneity also allows extraglandular involvement, and changes of neurological origin may occur, such as motor neuron involvement, which mimics the symptoms of Amyotrophic Lateral Sclerosis (ALS). **Objectives:** Describe and discuss a case of a patient with SS with neurological impairment, indicating systemic impairment with possible association with Aminotrophic Lateral Sclerosis. **Methodology:** A retrospective study of medical records and exams was carried out, after approval by the Research Ethics Committee. A systematic literature search was carried out in the databases PUBMED/ MEDLINE, LILACS and Cochrane Central using the search strategy "(Sjogren's syndrome) AND (motor neuron disease)" and "(amyotrophic lateral sclerosis) AND (Sjogren's syndrome) Sjogren)" with "full text" filter. **Case Report and Discussion:** Patient reported loss of strength in the right hand for a year, xerophthalmia and xerostomia, and presented positive Anti-SSB/La and ANA with positive glandular biopsy for SS. Initial nerve conduction examination showed a decreased action potential amplitude and increased F waves in the right ulnar nerve. It evolved with impairment of the lower limbs, without significant complaints, just a progressive loss of strength. In the literature consulted, we found that only 11 articles with a clinical profile compatible with isolated motor impairment in SS and only 2 other cases showed the possibility of an association between ALS and the syndrome, describing very compatible conditions, but with an unfavorable evolution. **Conclusion:** The patient presents a rare condition, with few reports in the literature, which, given the symptomatic progression, considered ALS as a component of the clinical picture or associated pathology. The scarcity of information raises doubts about the most appropriate diagnosis and treatment. The patient remains unresponsive to the treatments used and the condition has been evolving progressively.

KEYWORDS: Sjogren's Syndrome; Motor Neurons; Amyotrophic Lateral Sclerosis; Case Reports; Electromyography.

RESUMO: Introdução: A Síndrome de Sjögren (SS) é uma doença sistêmica inflamatória crônica, imunomediada, caracterizada pelo envolvimento das glândulas exócrinas, principalmente salivares e lacrimais. Entretanto, sua heterogeneidade permite também o acometimento extraglandular, podendo ocorrer alterações de origem neurológica, como o acometimento do neurônio motor, que mimetiza os sintomas da Esclerose Lateral Amiotrófica (ELA) dificultando o diagnóstico. **Objetivos:** Descrever e discutir um caso de uma paciente diagnosticada com SS com um comprometimento neurológico sugerindo-se um comprometimento sistêmico com possível associação a Esclerose Lateral Aminiotrófica. **Metodologia:** Estudo retrospectivo de prontuários e exames, após a aprovação do Comitê de Ética em Pesquisa. Foi feita uma busca sistematizada na literatura nas principais bases de dados PUBMED/ MEDLINE, LILACS e Cochrane Central utilizando a estratégia de busca "(Sjogren's syndrome) AND (motor neuron disease)" e "(amyotrophic lateral sclerosis) AND (Sjogren syndrome)" com filtro "full text". **Relato de Caso e Discussão:** Paciente relatou perda da força na mão direita há um ano, xeroftalmia e xerostomia, e na investigação apresentou Anti-SSB/La e FAN positivos com biópsia glandular positiva para SS. Exame de condução nervosa inicial mostrou uma amplitude do potencial de ação diminuída e ondas F aumentadas no nervo ulnar direito. Evoluiu com comprometimento de membros inferiores, sem queixas sensitivas, apenas perda progressiva da força. Na literatura consultada, encontrou-se que apenas 11 artigos apresentaram perfil clínico compatível ao comprometimento motor isolado na SS e apenas outros 2 casos mostravam a possibilidade de associação da ELA com a síndrome, descrevendo quadros muito compatíveis, mas com desfecho desfavorável. **Conclusão:** A paciente apresenta um quadro raro, com poucos relatos na literatura, que dado a progressão sintomática, cogitou-se a ELA como componente do quadro clínico ou patologia associada. A escassez de informações gera dúvidas acerca do diagnóstico e tratamento mais adequado. Paciente segue irresponsiva aos tratamentos utilizados e o quadro vem evoluindo progressivamente.

PALAVRAS-CHAVE: Síndrome de Sjogren; Neurônios Motores; Esclerose Amiotrófica Lateral; Relato de Caso. Eletromiografia.

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INTRODUCTION

Primary Sjögren's syndrome (PSS) is a chronic inflammatory systemic disease of an immune-mediated nature with an unknown precise etiology, commonly characterized by the sicca complex (xerostomia and xerophthalmia)[1]. It has an estimated prevalence of 60 cases per 100,000 inhabitants, being more common between 45 and 55 years old, and with a predominant condition in women at a 9:1 (women:men) ratio[1-2]. It is generally possible to perceive pathological findings of dysfunction and lymphocytic infiltration of exocrine glands (salivary and lacrimal) accompanied by a specific profile of autoantibodies[2-3]. However, due to the heterogeneity of the disease, it is also possible to observe extraglandular involvement in different organs in up to 50% of patients, including the skin, joints, lungs, kidneys, gastrointestinal tract, and most notably the nervous system[1,3].

This neurological involvement can be observed in up to 20% of PSS patients and is notable for its difficult diagnosis[4-5]; a retrospective study with 82 patients showed that neurological disorders preceded the onset of symptoms of dry eyes and dry mouth in 47% of patients, which can make the immediate relationship with the syndrome difficult[4].

PSS has the potential to cause damage to the Peripheral Nervous System (PNS), where the sensory nerves are the most affected[4-6]; its pathogenesis may be related to the infiltration of lymphocytes in the dorsal root ganglia[5]. In contrast, involvement of the Central Nervous System (CNS) is relatively rare in patients with PSS (2% to 25%), and symptoms when affected may include cognitive dysfunction, headache, seizures, transverse myelitis, aseptic meningitis, disseminated encephalopathy, neuromyelitis optica, multiple sclerosis and cranial nerve damage[5,7- 9].

Motor neuron involvement is a rare condition that can progressively advance to other systems, causing a loss in muscle strength in addition to muscle spasticity ("claw hand"); other signs would be choking on solids and/or liquids. This progressive condition in the motor neuron can also lead to respiratory failure and consequently to respiratory failure and an increased risk of mortality[9].

In another aspect, Amyotrophic Lateral Sclerosis (ALS) is a neurodegenerative disease manifested by progressive deterioration of the motor nervous system involving the cortex, brain stem and spinal cord. Loss of lower motor neurons leads to muscle weakness, atrophy, cramps, and fasciculations, while loss of upper motor neurons results in spasticity and rapid reflexes. Extramotor systems may also be involved in ALS, such as cognitive and behavioral disorders[10].

The diagnosis of ALS is challenging as the clinical manifestations are diverse and there is only one body fluid biomarker, serum neurofilament light chain (NFL), unlike PSS which has disease markers. Therefore, ALS diagnosis is initially based on the identifying motor neuron involvement and the complete exclusion of other differential diagnoses; furthermore, it is worth highlighting that Sjögren's syndrome (SS) can have the phenotypes of peripheral ALS as its clinical manifestations, and therefore a long screening for PSS should be performed[11].

The onset of sporadic ALS symptoms, which has an unknown cause, appears to be established between the ages of 55 and 65, with an overall average survival rate from the onset of symptoms between 2-3 years for bulbar-onset cases, and 3-5 years for limb-onset ALS cases. Important prognostic indicators of survival are: shorter time from symptom onset to diagnosis, age at symptom onset, site of onset (bulbar vs. limb onset), and El Escorial category at presentation[12]. El Escorial is essential for diagnosis after identifying neuropathy and peripheral impairment, as this score assesses the degree of progressive motor impairment. Furthermore, studies have shown that atrophy of the precentral gyri and changes in the cortico-spinal tract can be identified in the early stages through Magnetic Resonance Imaging (MRI)[11].

Genetic tests can also assist in diagnosis, as approximately 5% to 15% of ALS cases are familial with autosomal dominant inheritance. According to a study by Wendebourg et al. (2022), the mutations are in the C9 orf 72 gene (Chromosome 9 open reading frame 72; around 30-40%), the SOD1 gene (superoxide dismutase 1, 10-20%), the TDP-43 gene (response transactive DNA binding protein 43; about 5%), and the FUS gene (fused sarcoma around 5%)[11].

In this article, we report a case of a patient diagnosed with PSS with neurological impairment in which the first clinical manifestations were caused by this pathology in a mild form, and showing improvement after hormone therapy. Considering the patient's age and progressive deterioration of the clinical condition even with hormone therapy treatment, the case suggests systemic involvement with a possible association with ALS[4,11].

Faced with a pathology involving the motor neuron, it is necessary to master the investigative process (clinical, laboratory and imaging analysis) so that damage and symptoms can be controlled and minimized through early diagnosis. Knowing the singularities of motor impairment in SS helps in the differential diagnosis of Amyotrophic Lateral Sclerosis, as this is essentially carried out by excluding other mimicking diseases.

There is a gap in the literature regarding research on PSS with a possible association with ALS, and considering that the prognosis of this disease takes into account the diagnosis speed, more studies on this topic would contribute to a more favorable evolution of similar patients.

Therefore, the objective herein is to report a case of a 54-year-old female patient who was initially diagnosed with Sjögren's syndrome with isolated motor impairment, highlighting Amyotrophic Lateral Sclerosis as a possible differential diagnosis and reporting its singularities. It is also intended to identify similar cases and their outcomes in the literature, as well as effective alternative diagnostic and therapeutic methods.

METHOD

The present study consists of a rare case study report based on the clinical history of a patient who is being monitored in the city of São Paulo by the private rheumatology practice of Dr. Virginia Fernandes Moça Trevisani together with the neurologist Dr. André Macedo Serafim Silva. The report consists of organizing complementary exams and clinical

history, and monitoring progress. The case data consists of: anamnesis, review of medical records and complementary exams (Electroneuromyography, MRI of the cervical spine, MRI of the lumbar plexus, MRI of the direct coxofemoral joint, MRI of the skull, Cerebrospinal fluid (CSF) exam, Ultrasound guided (USG) biopsy of salivary glands), after due consent to use the information described herein.

That said, an initial analysis established from the articles found was performed based on the titles and abstracts and exclusion of duplicate articles. After this screening, the relevant articles were separated to be read in full, and finally evaluated for their inclusion or not in this study. Original literature review articles and clinical cases were included, without language limitations, published between 1990 and 2021, which dealt with the research topic. Publications presented in the format of reflections, opinions or comments from experts, as well as duplicate publications in the databases, or those which did not refer to the topic were excluded. Case reports which exclusively described sensory neuropathies, clinical tests that proposed the analysis of specific neuronal antibodies and articles in which access to the full text was impossible were also excluded.

Through a systematic literature search in the main PUBMED/MEDLINE, LILACS and Cochrane Central databases using the search strategy "(Sjogren's syndrome) AND (motor neuron disease)" and "(amyotrophic lateral sclerosis) AND (Sjogren syndrome)" with a "full text" filter, not limited to the publication period or the language of the article, it was found that only 11 presented a clinical report compatible with a neurological impairment in SS, and only 2 other cases showed the possibility of association of ALS with SS, describing conditions which are very compatible with the progression of the patient in the present study.

CASE REPORT AND DISCUSSION

Case description

A 54-year-old woman, pediatrician, arrived at the office in February 2021, complaining of loss of strength in her right hand for a year. She also said she noticed slight dryness in her mouth and eyes' area, which had improved substantially due to recent hormonal treatment due to menopause. She uses risedronate sodium, vitamin D and calcium carbonate, medications indicated for treating osteoporosis. That said, eye tests and salivary flow were normal. Autoantibody tests initially showed a negative Anti-SSA/Ro and a positive Anti-SSB/La and positive ANA.

Furthermore, pain was also noted in the thumb region, showing possible rhizarthrosis with muscle atrophy in the right hand. Thus, a nerve conduction exam was performed which showed decreased action potential amplitude in the right ulnar nerve and borderline in the right median nerve, and increased F waves in the right ulnar nerve. Given the findings, the patient underwent a surgical procedure to decompress the ulnar nerve, from which she did not improve.

Then in March 2021, tne salivary gland biopsy was reevaluated which was positive for SS, containing more than one focus of lymphoplasmacytic infiltrate in an area of 4 mm^2, and a new evaluation of autoantibodies using special techniques was requested. The findings in repeating the exams were surprising, with a positive Anti-SSA/Ro and a negative Anti-SSB/La and negative ANA, contrasting with the initial findings.

Rapid worsening in the strength of the hand was reported at the return appointment in June 2021, now in a claw-like appearance, and discomfort in the right hip and paresthesia in the thigh region were also described. A new request for electroneuromyography (ENGM) demonstrated signs of active denervation affecting the cervical, thoracic (C8-T1) and lumbosacral (left L5 and right L5-S1) regions, indicating motor neuron disease. Considering that Sjögren's syndrome can lead to neuropathy, an MRI of the right brachial plexus was performed, detecting hypersignal and thickening of the right C8 root, suggesting inflammatory involvement at this level, which is the segment verified as abnormal by ENGM. Normal cervical and shoulder magnetic resonance imaging (MRI) and other tests: ESR 27mm/hour, liver enzymes (TGO 80 u/l, TGP 132 u/l).

The condition now affects multiple nerves without any sign of sensory involvement. That said, the following approach was followed to treat systemic manifestations with involvement of the CNS and PNS in pSS: combined use of methylprednisolone sodium succinate 3g and Human Immunoglobulin 90g per month in pulse therapy in the form of induction, followed by Human Immunoglobulin 45g per month in pulse therapy for six months. Azathioprine 100mg/day was added, as the patient did not show any improvement in her upper limbs and new impairments appeared, with no response to previous treatments.

A clinical reassessment of the patient in 2022 revealed an approximately two-year history of slowly progressive asymmetric weakness initially in the upper extremities, but now with mild weakness in the lower extremities and potentially some bulbar involvement. Upper motor neuron findings included hyperreflexia in both upper and lower extremities. Lower motor neuron findings included asymmetric muscle atrophy, muscle fasciculations, and lower motor neuron weakness. Considering this combination of findings in at least two regions, there was room for another possible diagnosis of Amyotrophic Lateral Sclerosis. Electromyography and nerve conduction studies were then performed and showed evidence of a diffuse denervating disorder. Thus, it was advised by the Neurologist to continue with the initial immunosuppressive therapy as treatment for this new possible condition, and add therapies with potential benefit for ALS, including Riluzole (which modulates glutamatergic activity, thereby suppressing excitotoxicity) and Edaravone (most recently approved).

DISCUSSION

A case report study of a 54-year-old female patient who was diagnosed with Sjögren's syndrome with an isolated and progressive motor impairment for more than a year was presented. Repeat ENMG showed findings compatible with lower motor neuron disease, without sensory signs or symptoms. The patient had already received 4 monthly pulses of corticosteroids and intravenous immunoglobulins and immunosuppressive treatment, in addition to treatment with calcium replacement and

a surgical procedure to decompress the ulnar nerve. However, these treatments proved to be inefficient, as no improvement was observed in the upper limbs and new impairments appeared, making the course of diagnostic hypothesis to a possible association of SS with Amyotrophic Lateral Sclerosis rather than an association with the impairment of the motor neuron, as described during the report.

A literature review was conducted in search of published reports with a similar theme to that of the present study. Finally, a total of 36 articles were found, of which only 11 presented a clinical situation compatible with neurological impairment in SS, and only two cases came close to the panorama we addressed.

That said, little is found in the literature about SS associated with motor neuron disease. Dalalande (2004)[4] presented a comparative study which aimed to highlight the diversity of neurological complications of SS in which this relationship was present in only one patient out of the total of 82 patients studied. He also concluded that the average age of neurological onset was 53 years, interestingly corresponding to the onset of our patient's symptoms. In addition, neurological involvement often preceded the diagnosis of Sjögren's in 81% of patients.

Screening was performed in order to search for new means of treatment. The articles found included a symptomatic approach, immunosuppressive treatments, such as cyclophosphamide and hydroxychloroquine, the use of corticosteroids, such as administration of methylprednisolone, and the use of human immunoglobulin[9,13-17]. Most reports showed significant neurological improvement or recovery of the patient almost immediately after drug therapy, unlike what we observed in this case.

Yang (2020)[9] presented a patient with rapidly progressive motor neuron disease secondary to SS, which was the most similar case to the case in this study. In his study conducted in Changchu (China) between 2019 and 2020, he describes a woman, 42 years old, with a 2-month history of progressive limb weakness, with xerophthalmia and biopsy confirming SS. Extensive neurological damage is observed in the lower and upper limbs, without response to immunosuppressive treatment, use of corticosteroids or immunoglobulins. The patient developed respiratory failure and died. This case concerns us due to its clinical similarity with our patient, best shown in Table 1, who also presented progressive worsening without apparent response to any treatment[9].

A possible differential diagnosis according to a case report study carried out by Suk-Won[18], would be inflammatory polyradiculopathy mimicking motor neuron disease in a patient with SS, whose symptoms were slow progressing weakness and muscular atrophy, a condition confirmed in this study through spinal magnetic resonance imaging and cerebrospinal fluid examination results. However, the results of our patient's spinal MRI and cerebrospinal fluid examination were normal, ruling out this diagnosis.

Table 1 - Comparison of the case of Yang et al. (2020) with the case of the present study

Yang et al.	Case of the present study
Woman, 42 years old	Woman, 54 years old
2 months of progressive limb weakness, with xerophthalmia and biopsy confirming SS	Isolated and progressive motor impairment for more than a year, with xerostomia and xerophthalmia and biopsy confirming SS
Extensive neurological damage in the lower and upper limbs, unresponsive to immunosuppressive treatment, use of corticosteroids or immunoglobulins	No improvement in the upper limbs and appearance of new diffuse and progressive impairments. No response to treatments
She developed respiratory failure and died	Patient is progressively worsening

According to a case report study carried out by Suk-Won[18], a possible differential diagnosis would be inflammatory polyradiculopathy mimicking motor neuron disease in a patient with SS, whose presenting symptoms were slow progressing weakness and muscular atrophy, a condition confirmed in this study through spinal magnetic resonance imaging and cerebrospinal fluid examination results. However, our patient's spinal magnetic resonance imaging and cerebrospinal fluid results were normal, ruling out this diagnosis. Furthermore, another differential diagnosis would be myopathy, whose symptoms also involve muscle weakness in which the skeletal muscle fibers are affected by dysfunction, but considering how in the study carried out by Suk-Won[18], our patient also presented normal creatine phosphokinase levels, making the possibility of myopathy very low.

On the other hand, there are few studies relating SS in association with ALS, the most similar to the report described was that of Wendebourg[11] carried out between 2019 and 2021 in Basel (Switzerland), which addresses the case of a 72-year-old woman who initially presented with mononeuropathy of the right leg and complaints of xerostomia and xerophthalmia. Initial diagnostic workup confirmed a regional sensorimotor neuropathy associated with an SS. However, motor symptoms progressively spread, even with immunosuppressive treatment. This impairment progressed, affecting the diaphragm and generating respiratory discomfort, in addition to affecting arms and legs, leading to the patient's inability to walk[11].

The suspicion of SS was only due to a positive Schirmer test; however, a biopsy of the left superficial peroneal nerve was performed due to worsening of the clinical picture with worsening weakness, which revealed a reduction in myelinated fibers without signs of vasculitis and neurofilament light chain (NfL) serum levels were highly elevated. Then, electroneuromyography resulted in progressive signs of severe acute denervation in the

upper limbs and in the sternocleidomastoid muscle, with only the tongue preserved and what permitted a probable diagnosis regarding this clinical picture was via the El Escorial criteria[11].

The numerous converging points between the case of Wendebourg[11] and that of the present study, as illustrated in Table 2, point to the existence of a curious association between SS and ALS, which is rarely described in the literature. That said, the difficulty in diagnosing Amyotrophic Lateral Sclerosis is evident, given the peripheral symptoms and neuropathy that may also be present in Sjögren's syndrome[11].

Table 2 - Comparison of the case of Wendebourg et al. (2022) with the case of the present study

Wendebourg et al.	Case of the present study
Woman, 72 years old	Woman, 54 years old
She initially presented mononeuropathy of the right leg and complaints of xerostomia and xerophthalmia	Isolated and progressive motor impairment for more than a year, with xerostomia and xerophthalmia and biopsy confirming SS
Initial diagnostic workup confirmed a regional sensorimotor neuropathy associated with a SS	Repeat ENMG showed findings compatible with lower motor neuron disease, without sensory signs or symptoms
Motor symptoms spread progressively, even with immunosuppressive treatment. It affected the diaphragm, causing respiratory discomfort, in addition to affecting arms and legs, making the patient unable to walk	Motor symptoms spread progressively and did not respond to immunosuppressive treatment. Upper motor neuron findings included hyperreflexia in both upper and lower extremities. Lower motor neuron findings included asymmetric muscle atrophy, muscle fasciculations, and lower motor neuron weakness
Suspicion of SS was only due to a positive Schirmer test	Salivary gland biopsy positive for SS, containing more than one focus of lymphoplasmacytic infiltrate in an area of 4 mm^2 + Anti-SSA/Ro positive + xerostomia and xerophthalmia
Serum neurofilament light chain (NfL) levels were highly elevated, electroneuromyography resulted in progressive signs of severe acute denervation in the upper limbs and sternocleidomastoid muscle	ENGM demonstrates signs of active denervation affecting the cervical, thoracic and lumbosacral regions, indicating motor neuron disease

CONCLUSION / FINAL CONSIDERATIONS

The purpose of this work was to carry out a case study of a patient who was initially diagnosed with Sjögren's syndrome with an isolated motor impairment, which prompted us to search for similar case reports in the scientific literature.

The first clinical manifestation was an impairment of the ulnar nerve, without complaints of xerostomia and xerophthalmia. It was possible to diagnose Sjögren's syndrome with repeated tests through a salivary gland biopsy, positive ANA and positive Anti-SSA/Ro. The patient does not respond to treatment with a monthly pulse of Methylprednisolone and intravenous Gammaglobulin, in addition to showing involvement of other nerves. Given the patient's evolution, it was noted that there could be an association between SS and Amyotrophic Lateral Sclerosis, proposing an additional treatment for this condition.

When analyzing the literature, it was found that Sjögren's syndrome with motor neuron involvement is rarely reported, and that some patients may progress to a more serious clinical condition and die. Another study showed the difficulties in diagnosing ALS as its signs and symptoms can mimic SS, which can also have a lethal outcome. Further research will continue with the theme presented herein and new scientific results may clarify the subject addressed.

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Authors and participation: Vanessa Furtado do Vale Bento: Medical student responsible for part of the selection of articles and tabulation, the dissertation of the case report, final preparation of the summary, methods and part of the introduction, discussion with the literature and conclusion, as well as structuring the illustrative tables. She also made the necessary corrections and revisions for publication. Fernanda Aparecida Mateus Vieira: Medical student responsible for part of the selection of articles, partial preparation of the introduction and conclusion. Maria Fernanda Reis Nunes da Silva: Medical student responsible for selecting articles and writing the introduction and objectives. Virginia Fernandes Moça Trevisani: Provision of data for formulating the case report, guidance and critical review of the work.

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