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Antisynthetase-like dermatomyositis related to anti-pm/scl-75: Case report

Dermatomiositis tipo antisintetasa relacionada con anti-pm/scl-75: Reporte de caso

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RESUMEN

Este artículo describe el caso de una paciente de 29 años que presentó una miopatía inflamatoria grave relacionada con anticuerpos anti-PM/SCL75, con una presentación clínica tipo antisintetasa.

Palabras clave:
antisintetasa,
dermatomiositis,
miopatía.

ABSTRACT

This article reports the case of a 29-year-old female patient who presented with a severe inflammatory myopathy related to anti-PM/SCL75 antibodies, with an antisynthetase-like clinical presentation

Keywords:
antisynthetase,
dermatomyositis,
myopathy.

Introduction

Dermatomyositis is a rare inflammatory myopathy with diverse phenotypic presentations, which vary widely depending on the specific autoantibody present in each patient[1]. Given the broad variability in clinical manifestations, some authors have proposed a new classification of inflammatory myopathies, one of which includes the term *"scleromyositis"* - referring to diseases that show overlapping features of inflammatory myopathies and systemic sclerosis[2].

Not uncommonly, the phenotype is highly suggestive of a specific myopathy. A good example is the antisynthetase-like clinical presentation, characterized by joint involvement, interstitial lung disease, cutaneous manifestations, and the presence of anti-aminoacyl-tRNA synthetase antibodies. However, in some cases, these antibodies test negative. This highlights the importance of having the full myositis-specific autoantibody panel for each patient, which is relevant for accurate diagnosis, prognosis estimation, and predicting the clinical course[3,4].

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Case description

A 29-year-old previously healthy female was referred to dermatology for suspected psoriasis, with scaly lesions on her feet, palms, and hand joints for the past 5 months. During the evaluation, she showed significant decline in general condition and proximal muscle weakness, prompting an urgent referral to rheumatology for suspected inflammatory myopathy.

Upon hospital admission, the patient was bedridden, unable to walk, with grade 2 strength in the upper and lower limbs and cervical muscles, scoring 85/150 on Manual Muscle Testing (MMT-8). She also had dysphonia, dysphagia, bilateral scaly lesions on the palms and soles, and erythematous lesions on the dorsum of the interphalangeal and metacarpophalangeal joints (Figure 1).

A new anamnesis revealed progressive proximal weakness over 30 days, starting in the lower limbs and progressing to the upper limbs and neck. The patient also reported dysphagia, coughing when lying down, and a 20 kg weight loss during this period.

Initial laboratory tests showed: Creatine phosphokinase (CPK) 4736 U/L, lactate dehydrogenase (LDH) 1640 U/L, aldolase 122 U/L, transaminase oxaloacetic (TGO) 150 U/L, glutamic pyruvic transaminase (TGP) 126 U/ with antinuclear antibodies (ANA) 1/640, fine speckled nuclear pattern. Serologies for HIV, hepatitis B, hepatitis C and syphilis were negative. A chest tomography scan was performed, which demonstrated interstitial lung involvement, with ground-glass opacities at the lung bases (Figure 2).

The clinical presentation, along with complementary exams, strongly suggested inflammatory myopathy, specifically AS, with severity markers including severe muscle involvement, pulmonary involvement, and dysphagia.

Treatment was started with methylprednisone 1 g/day for 3 days, followed by intravenous immunoglobulin (IVIG) 400 mg/kg/day for 5 days, and a muscle biopsy was performed. Significant improvement in symptoms and laboratory tests was observed within 48 hours of treatment. The patient was discharged after 8 days of hospitalization with prednisone 1 mg/kg/day.

Two weeks after pulse therapy, the patient was able to stand on her own, with partial improvement in dysphagia, skin lesions, and MMT-8 (106/150). Oral methotrexate was initiated, and five weeks later, the patient showed further improvement in weakness (MMT-8: 126/150), dysphagia, skin lesions (Figure 3), and laboratory parameters.

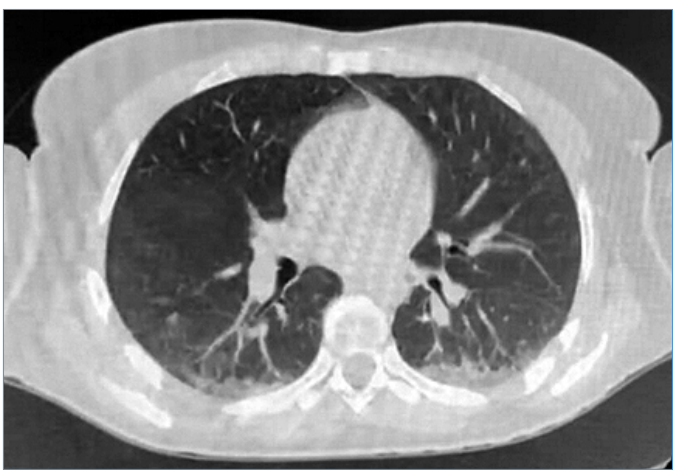

Figure 2. Legend: chest tomografy scan showing basal ground glass changes, suggesting interstitial lung involvement.

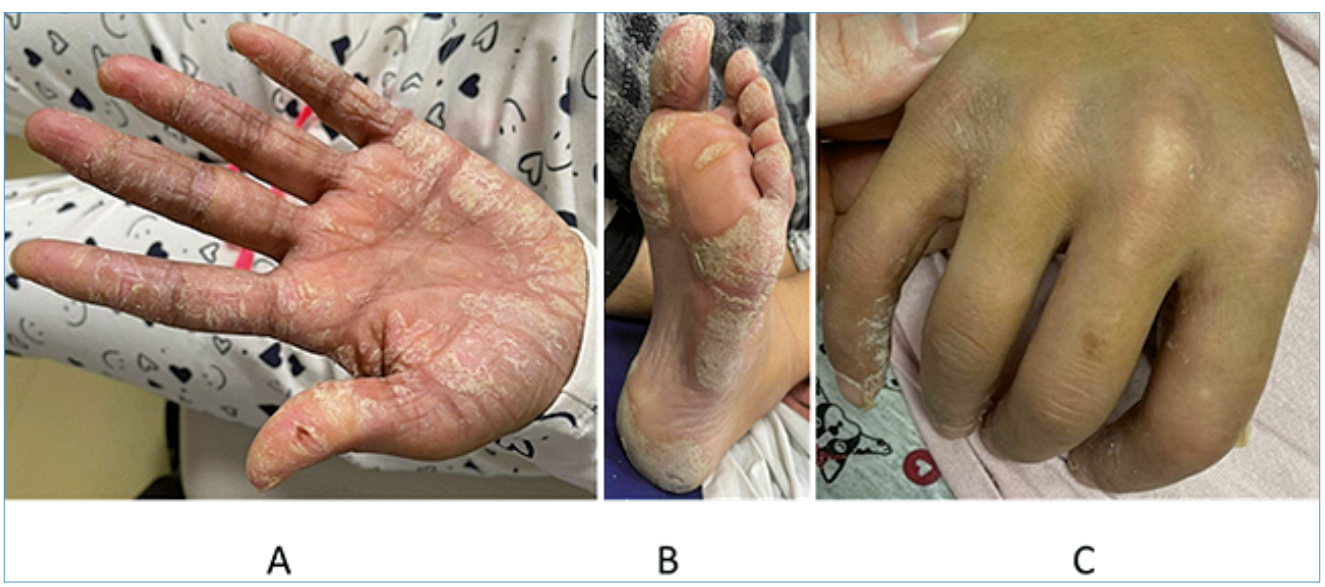

Figure 1. Legend: A: mecanic hands; B: hikers feet; C: scaly lesions on the extensor aspect of the hand joints; mild sclerodactyly may be noted.

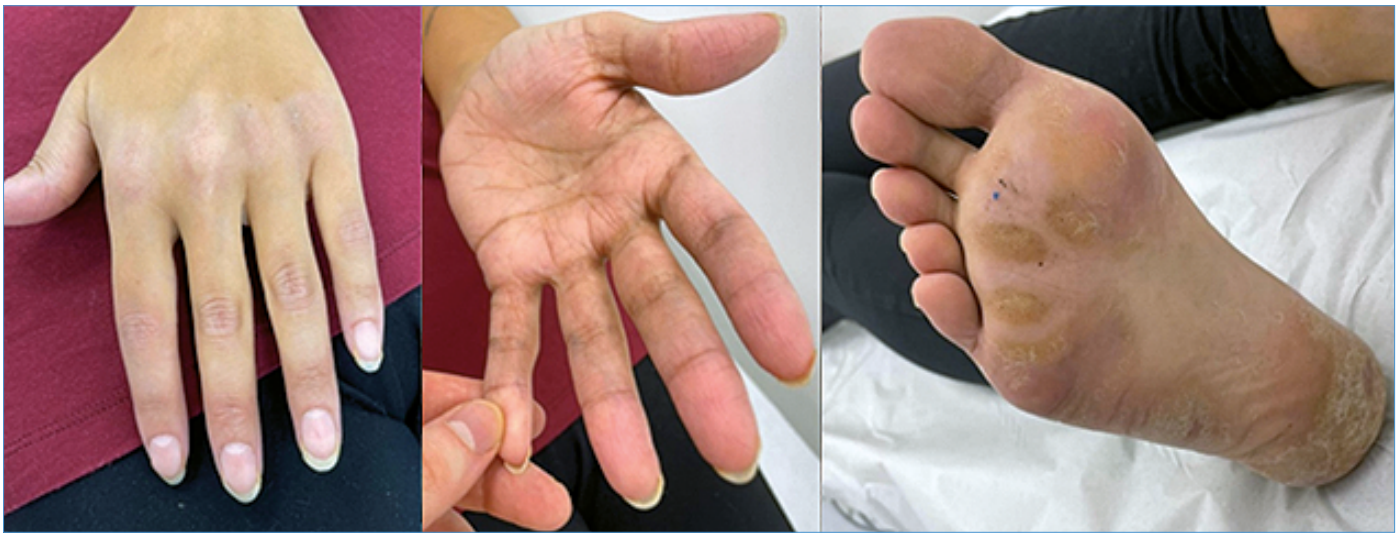

Figure 3. Legend: evolution of skin lesions after 5 weeks of treatment.

Table 1

	Admission	2 weeks from starting treatment	13 weeks from starting treatment
Creatine phosphokinase	4.736 U/L	979 U/L	181 U/L
Lactate dehydrogenase	1.640 U/L	792 U/L	472 U/L
Aldolase	122 U/L	10 U/L	6,6 U/L
Transaminase oxaloacetic	150 U/L	56 U/L	18 U/L
Glutamic pyruvic transaminase	126 U/L	70 U/L	15 U/L
Legend: U/L units per liter.			

The results obtained from the muscle biopsy demonstrated lymphocytic inflammation, necrotic muscle fibers, perifascicular atrophy, and damage to the perimysial tissue. Findings compatible with inflammatory myopathy, more specifically dermatomyositis or AS.

Thirteen weeks after starting treatment, the patient had almost no skin lesions and practically no limitations related to muscle strength (MMT8: 140/150), except for a mild degree of dysphonia. The evolution of laboratory tests can be seen in Table 1.

The final antibody profile at the end of the investigation was:

- Positive tests: PM/SCL75; ANA 1/640, fine speckled nuclear pattern.
- Negative/non-reactive tests: DNA double stranded, RNP, SCL70, centromere, rheumatoid factor, MI-2, TIFI1-Y, MDA-5, NXP2, SAE-1, SRP, JO-1, PL-7, PL-12, EJ, OJ, KU, PM-SCL100, RO-52.

The patient is currently undergoing routine rheumatology consultations and is showing progressive clinical improvement, with a final diagnosis of inflammatory myopathy related to the PM/SCL75 antibody.

Discussion

The case presented clinically as antisynthetase syndrome, however the detection of anti-PM/SCL75 antibodies led to a revised diagnosis of dermatomyositis with overlapping features of systemic sclerosis. This is a presentation characteristically associated with this autoantibody, with the literature describing the frequent misdiagnosis of antisynthetase syndrome in such cases[4]. Accurate diagnosis and performing the specific myositis antibody panel are important due to their prognostic implications, as patients with antisynthetase syndrome have a worse prognosis compared to those with anti-PM/SCL75, especially lung prognosis[5,6].

The findings of predominant weakness in the deltoids,

sclerodactyly, and good therapeutic response are described as being associated with anti-PM/SCL75 and were observed in this case[4]. Due to its unique phenotype, some authors even suggest a different subclassification for myositis: the anti-PM/SCL syndrome[7].

The positivity of this antibody also correlates with a lower prevalence of muscle weakness at disease onset, with weakness developing over time[7]. In this case, the patient presented skin symptoms several months before muscle symptoms, which led her to initially seek consultation at a dermatology outpatient clinic, emphasizing the importance of diagnostic suspicion in atypical presentations.

Initial management included high-dose corticosteroids combined with intravenous immunoglobulin (IVIG), in accordance with guidelines for managing severe inflammatory myopathies, especially with dysphagia and risk of bronchoaspiration[3]. Methotrexate was selected as a corticosteroid-sparing agent, leading to a favorable progression.

Conclusion

This case highlights the importance of recognizing anti-PM/SCL75 as a clinical marker in patients with inflammatory myopathy, particularly due to its ability to mimic an antisynthetase syndrome. Characteristic findings of systemic sclerosis serve as clues to its positivity. The favorable therapeutic response underscores the value of intravenous immunoglobulin (IVIG) in severe presentations and the potential role of methotrexate in the therapeutic approach.

Ethics approval and consent to participate: All procedures involving human subjects are researched in accordance with the institution's ethical standard and the 1964 Helsinki declaration and its subsequent amendments. This study was approved by the Research Ethics Committee of Faculdade Evangélica Mackenzie, Curitiba, PR, under the protocol 86004624.4.0000.0103 of 20/02/25, which can be accessed by telephone +55 (41) 3240-5570 or by e-mail: comite.etica@fempar.edu.br

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