PRURITUS IN SYSTEMIC SCLEROSIS, A THERAPEUTIC CHALLENGE: A CASE REPORT

Gabriela Almeida Barbosa^{1,*}, Robson Antônio Gonçalves¹, Valéria Bezerra da Silva¹, Tamara Santos Melo¹, Maria Roberta Melo Pereira Soares¹, Ana Karla Guedes de Melo¹, Danielle Christinne Soares Egypto¹, Sandra Rejane Cabral Batista¹, Alessandra de Sousa Braz¹, Eutília Andrade Medeiros Freire¹

1.Universidade Federal da Paraíba, João Pessoa (PB), Brazil.

*Corresponding author: gabrielaalmeidamed@gmail.com

BACKGROUND

Pruritus in systemic sclerosis (SSc) is present in about 40% of patients, with a significant impact on quality of life (QOL) and uncertain pathophysiology. It is associated with worsening of the skin and gastrointestinal involvement and is most commonly seen on the head, back and extremities. Despite the high frequency, its management remains uncertain and based on the use of symptomatic.

CASE REPORT

Woman, 61-year-old, diagnosed with systemic lupus erythematosus (SLE) 20 years ago, polyarthritis, serositis, alopecia, oral ulcers and nuclear speckled 1:640 ANA (SLICC criteria, 2012), in irregular follow-up and use of azathioprine 2 mg/kg/day. It evolved 5 years ago with generalized pruritus, unintentional weight loss and Raynaud's phenomenon (RP), with significant impact on QOL and progressive worsening for 6 months. Admitted to university hospital for investigation. On admission, she presented generalized pruritus without skin lesions (grade 9/10), telangiectasias in the face and mucosa and sclerodactyly. Complementary exams showed: mammography, upper digestive endoscopy, colonoscopy, thyroid ultrasonography (USG), transvaginal pelvic USG and abdominal computed tomography (CT) without alterations; serology for hepatitis B and C, syphilis and HIV negative; chest CT with nonspecific interstitial lung disease (NSIP), presence of ground glass in approximately 30% of the parenchyma, spirometry not available; anti-RNP positive at low titers (1:40), anti-Ro, anti-La, anti-Jo-1, anti-Scl-70 negative. Given the presence of sclerodactyly, RP, telangiectasias and interstitial lung disease, she met ACR/EULAR 2013 criteria for SSc. Treatment was initiated with hydroxychloroquine 400 mg/day, cyclophosphamide (CYC) 1 g/month for pulmonary involvement, nifedipine 20 mg/day and, for the treatment of pruritus, loratadine 10 mg/day, dexchlorpheniramine 4 mg/day, prednisone 15 mg/day and amitriptyline 50 mg/day. After starting CYC, the itching improved significantly (score 2/10), with progressive weaning from antihistamines and prednisone.

CONCLUSION

The management of pruritus in SSc remains a challenge in clinical practice. Clinical trials that seek to find more effective therapeutic alternatives for the treatment of pruritus should be encouraged, given the high impact on the QOL of these patients. The reported case shows a significant pruritus response with the use of CYC, which may corroborate the hypothesis of an autoimmune component in the pathophysiology of this symptom.

KEYWORDS

Pruritus, Autoimmunity, Systemic sclerosis.