Pruritus in Autoimmune Connective Tissue Diseases

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KEYWORDS

• Pruritus • Itch • Connective tissue • Lupus • Dermatomyositis • Systemic sclerosis • Sjögren

KEY POINTS

- Autoimmune connective tissue diseases (ACTDs) have prominent skin manifestations and commonly present with pruritus.
- Triggers of pruritus in ACTDs are diverse from disease related to medication side effects and malignancy development.
- It's important to work-up pruritus in ACTDs to treat patients appropriately according to the underlying cause independent of disease activity.

INTRODUCTION

The autoimmune connective tissue diseases (ACTDs) are a broad range of diseases (eg, the most common being lupus, dermatomyositis [DM], systemic sclerosis [SSc], and Sjögren syndrome), often with multisystem involvement. Their unifying feature is that a person's own immune system is dysregulated. Although these diseases are commonly primarily managed by rheumatologists, they often have prominent skin manifestations. The primary symptom of skin findings is either pruritus, or burning. Just as the diseases themselves are diverse, however, so are the causes of these symptoms. This article addresses the most common triggers for pruritus in these patients, the appropriate work-up, and the evidence behind different treatment regimens when pruritus occurs, whether dependent on or independent of disease activity.

DERMATOMYOSITIS

Itch and Cutaneous Dermatomyositis

Pruritus is a prominent feature in DM and has a significant impact on patients' quality of life.[1] A pruritus questionnaire of 70 DM subjects showed that subjects had a mean score of 44.6 on the 100-mm visual analog scale (VAS) in response to effect of itching on daily life.[2] A prospective study[3] conducted to compare clinical characteristics of DM, its relationship to malignancy, and treatment between 2 tertiary medical centers in the United States and Singapore reported that itch was the most common initial symptom among both populations, representing 63% and 80% of patients in the United States and Singapore, respectively.

Although photosensitivity in DM is similar to that of SSc lupus erythematosus (SLE),[4] pruritus evaluated by a VAS and a 0 to 10 scale in DM and in cutaneous lupus erythematosus (CLE) populations found that DM produces more pruritus than CLE ($P<.0001$).[5] This is possibly consistent with the observation that DM patients are more diffusely erythematous than SLE patients even when disease is relatively quiescent. Photoprotection is important and DM patients must be counseled to use broad-spectrum sunscreens that incorporate UV-A and UV-B blocking elements, to reapply

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sunscreen frequently, to remain out of direct sunlight as much as possible (especially between the hours of 10:00 AM and 2:00 PM), and to use physical barriers, such as broad-brimmed hats and long sleeves. The use of topical and SSc immunosuppressants can be helpful, although patients with DM are more likely to have an adverse cutaneous reaction to antimalarial therapy than SLE patients.[6]

Itch and Comorbidity Considerations in Dermatomyositis

The contribution of pharmacotherapy either as itch as a medication side effect or the medications effects on internal organs should be considered in the itchy DM patient (**Fig. 1**). Although DM is significantly associated with malignancy, with up to 30% of patients with DM having an associated malignancy,[7] hemoproliferative forms are extremely rare in DM.

LUPUS

Itch and Cutaneous Systemic Sclerosis Lupus Erythematosus

The term, *lupus*, can refer to disease limited to the skin, such as chronic CLE, also known as discoid lupus erythematosus (DLE); to subacute cutaneous lupus (SCLE), a different skin-only form; or to the SSc multisystem form SLE. Even in SLE, DLE and SCLE may occur. Itch can occur, however, with or without cutaneous involvement.

In a review of 91 SLE patients meeting American College of Rheumatology diagnostic criteria, 81% of SLE patients had clinical photosensitivity. Although some of these were SLE-specific photosensitivity, others had conditions, such as polymorphous light eruptions and solar urticaria.[4] These latter reactions were found more common in SLE than in the general population and it is unclear if the photosensitive triggering of different conditions is a comorbidity or if SLE photosensitivity may itself be more polymorphous in its presentation than previously thought. This same study found, however, that the more severe photosensitive cutaneous findings were associated with more severe disease flares, regardless of the form, possibly supporting the latter contention. Pruritus may be an early warning sign, because questionnaire data suggest that both SLE and DM patients experience skin pruritus on average 9.5 years before diagnosis, a finding not found in the same study in SSc patients.[8] Although some investigators suggest that photosensitivity varies by ethnic origin, with darker skin showing less photosensitivity,[9] others have not found this the case.[4] Recommendations for photoprotection for SLE patients are similar to those for DM patients.

Immunosuppression, topical with corticosteroids, calcineurin inhibitors, or calcipotriene, can assist in decreasing inflammation and thereby the symptom of pruritus, and some patients need SSc suppression with oral corticosteroids or steroid-sparing agents, such as methotrexate, mycopheolate mofetil, azathioprine, and thalidomide, among other options. Minimally

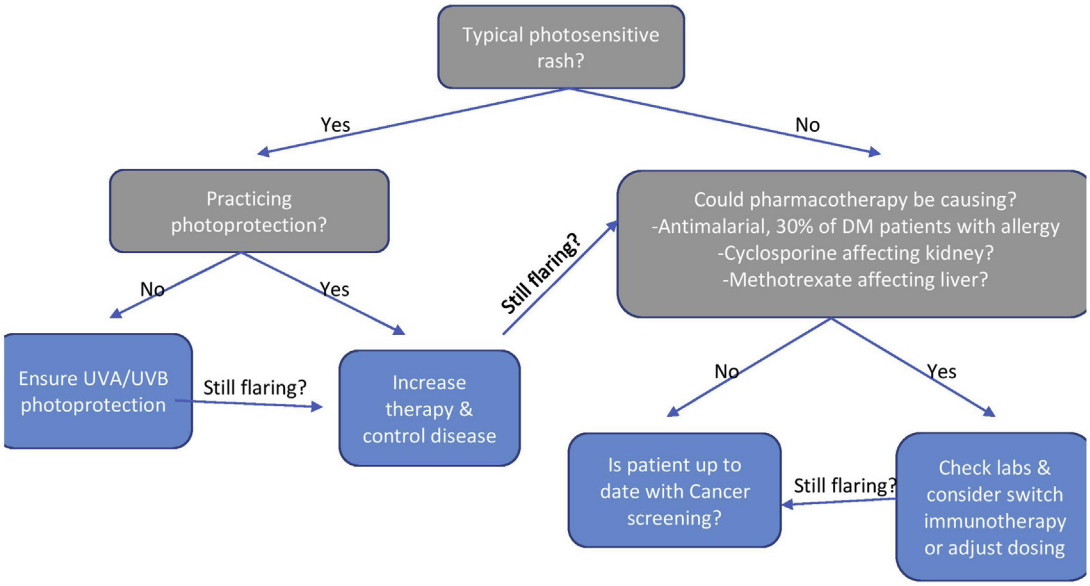

Fig. 1. Pruritus work-up and management in DM patients.