Chapter 5
Itch

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Case Vignette

55-year-old male patient with a disease duration of 5 years, a diffuse cutaneous form of SSc (mRSS 20), including GI involvement and stable lung fibrosis, is presenting with dryness of skin and an increased sensation of itch, involving mostly the extremities and parts oft he trunk. Itch is so severe that he gets awakened at night. Use of ointment from a drug store provided no relief. His previous medical history is remarkable for allergic rhino-conjunctivitis due to grass pollen. Furthermore, his only son suffers from atopic eczema.

Background

Itch (pruritus) is a very common problem in the general population with a 12-month incidence of up to 7% [1, 2]. In patients with SSc, itch may be present in up to 60% of cases and a correlation with disease severity has been described

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[3–6]. In addition, it has also been shown that the presence of itch correlates independently with the severity of skin hardening and greater GI involvement and remained predominantly stable in the course of the disease [5, 7].

In general, Itch may be directly associated with skin diseases or can be secondary due to internal organ involvement .. The exact causative mechanisms for itch in SSc still have to be elucidated. It is tempting to speculate that several pathophysiological events contribute to the clinical symptom of itch in SSc patients. Inflammatory processes in the dermis, the subsequent loss of skin appendages by the fibrotic process in combination with the loss of the ability to synthesize appropriate amounts of "endogenous lipids and emollients" contributes to skin xerosis are probably major factors [7]. Furthermore, entrapping of sensory nerve fibers can also contribute to the development of pruritus.

Due to comorbidities, itch is often not taken sufficiently into consideration with its effect on well being of the patient, although it may significantly affect quality of life, disturb sleeping and contribute to mood disorders and depression [7]. Therefore adequate symptomatic therapy of itch is an important component of multimodal therapy in SSc patients.

Differential Diagnosis

Differential diagnosis of pruritus encompasses a wide array of dermatological and non-dermatological diseases (see Fig. 5.1). Medical history (including onset, duration, location, quality and severity of itch) and skin examination are the basis of an initial assessment [8].

In chronic pruritus (i.e., persisting longer than 6 weeks) a complete dermatological examination is mandatory to exclude comorbidities and underlying systemic dieseases. It also has to be considered that a number of drugs may induce or maintain chronic pruritus [8].

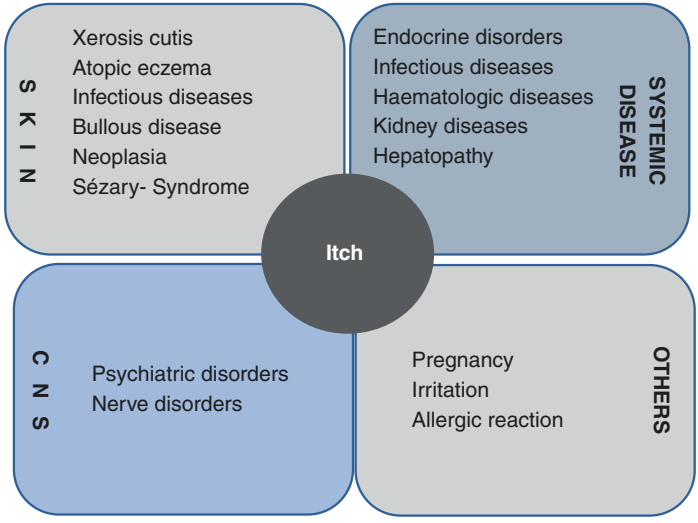

FIGURE 5.1 Differential diagnosis of SSc associated Itch

Therapy

The patient in this vignette suffers from a diffuse cutaneous SSc, but also has a history of atopic disease, indicating an increased risk to have dry skin (xerosis cutis) and develop atopic eczema.

As a first step, the patient should be informed about general pruritus-relieving measures as keeping room temperatures stable and applying moisturizers to improve skin barrier and reduce itching [2]. Although many patients report that hot or cold showers reduce itch, no scientific studies have been performed to confirm this observation and repeated showering leads to drying up of the skin. Avoidance of frequent bathing and showers is strongly recommended.

Topical therapy will be the mainstay of initial treatment. Usually, the patients apply emollients that have a high-water content favoring resorption, but having no effect on the skin texture and reducing itch sensation. Hence, an emollient containing urea (3–10%), lactic acid (1–5%, increasing hydra-

tion) and a higher percentage of glycerol (up to 20%) should be recommended. Topical application of glucocorticoids can reduce itching for some time. Also calcineurininhibitors can be used. Capsaicin (0.025% in a lipophilic vehicle), is an alkaloid contained in chili peppers interacting with sensory neurons, which is recommended for localized pruritus [2].

Antihistamines are widely used as a treatment for chronic itch in systemic diseases. However, conventional doses of antihistamines, either sedating first generation or non-sedating second generation antihistamines have not yet proven effective in controlled studies.

Gabapentin and pregabalin are antiepileptic and anxiolytic drugs which are also used in neuropathic pain and chronic severe pruritus. In severe cases, these drugs may therefore be considered as treatment options [2, 9].

UV-based therapy is well established for treating pruritus. UV modalities comprise UVB (290–320 nm) and UVA (320–400 nm). Usually, UV therapy is combined with topical treatment. A number of uncontrolled studies indicate a beneficial effect of UVA1 and photochemotherapy (Psoralen + UVA) on fibrosis in SSc leading to softening of the skin [10, 11] (Morita et al., 1995, 2000). In addition, UV therapy is also reducing the inflammatory response. Therefore, in severe pruritus these options should be considered. Systemic therapy of the underlying disease by mycophenolate mofetil, rituximab or cyclophosphamide in severely affected patients with dsSSc leads to a reduction of the inflammation and can influence the extent of itching.

In conclusion, management of itch in SSc patients comprises proper diagnosis and exclusion of other factors [12]. Treatment consists of topical treatment modalities accompanied by systemic and UV-therapy. In protracted, recalcitrant itch an interdisciplinary approach involving a dermatologist is mandatory.

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