5.2 Lichen sclerosus

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Definition

Lichen sclerosus (LS) is an inflammatory scarring dermatosis that has a predilection for the genital skin in both sexes [1].

Etiology and Pathogenesis

For the development of LS, immunological alterations seem to be of crucial importance: Pathogenetically relevant autoantibodies were identified which are directed against the extracellular matrix protein-1 (ECM-1) [2]. These autoantibodies have been detected in both sexes [1]. Patients with LS have an increased risk to develop other autoimmune conditions such as thyroid disease, alopecia areata, vitiligo or pernicious anaemia [1,3]. LS may be observed at any age, although two peaks of incidence occur, in the prepubertal and postmenopausal periods. The female-to-male ratio can be up to 10:1 [4].

Clinical features

The most common symptoms include sclerosing plaques and variable atrophy and fissures of the skin, combined with perifocal erythema. LS may cause anatomical changes if left untreated. Patients may complain of pruritus, soreness, dyspareunia and painful defecation [3].

Practical Guidance

All patients should be advised to avoid irritant and fragranced products [1].
III Special Treatment Indications

Therapy

Topical application of potent corticosteroids such as clobetasol propionate (CP) 0.05% ointment is considered the first-line treatment for all females (regimen for 3 months) and men (regimen for 1–3 months) with (ano)genital LS [1].

Following a number of case series demonstrating the clinical effectiveness of TCI in patients with LS [5–7], several studies investigated the safety and efficacy of tacrolimus ointment for treating genital and extragenital LS.

Topical treatment with tacrolimus: Published evidence in LS

Phase-II and double-blind studies

A multicentre phase II trial carried out to assess the safety and efficacy of topical tacrolimus in 84 patients with long-standing, active LS included a follow-up of 18 months (Hengge et al.). From the evaluable patients (n = 70), 54 patients (77%) reached an objective response with tacrolimus ointment 0.1% applied twice daily to all external lesions. Complete response (primary endpoint) was achieved in 16% and 43% of patients by weeks 16 and 24 of treatment, respectively (Fig. 8a–b). Treatment led to a significant reduction of the total lesional area (p < 0.01; Fig. 9a–b) and to a significant decline in the total symptom score (p < 0.005). There were no severe adverse events reported and, aside from the initial burning and pain, tacrolimus ointment was well tolerated. Disease recurred in 3 (9%) patients during the follow-up period [3].

Funaro et al. evaluated the safety and efficacy of tacrolimus ointment 0.1% compared with CP ointment 0.05% in 55 patients with vulvar LS. Patients were randomized and applied the respective treatment nightly for up to 3 months. A significant reduction in signs and symptoms was seen in both treatment groups, but the authors found that CP was significantly more effective than tacrolimus [8].

Ebert et al. showed that tacrolimus ointment 0.1% applied immediately after surgery of fully established LS is a tolerable and most probably safe adjuvant treatment option: All 20 boys (mean age: 9.7 years) with fully developed LS completed the topical treatment without any relevant side effects. Topical tacrolimus treatment led to disease control for a median duration of > 1 year in all patients [9].
Fig. 8  
(a) LS in a 10-year girl before treatment with topical tacrolimus.  
(b) After 3 weeks of treatment with tacrolimus ointment 0.1%.

Fig. 9  
(a) LS in an adult woman before treatment with topical tacrolimus.  
(b) After 3 weeks of treatment with tacrolimus ointment 0.1%.
A number of smaller clinical or retrospective studies evaluated the efficacy of topical tacrolimus in female [10–13], male [14] or mixed LS-populations [15]. In addition, many case reports have been published describing the use of tacrolimus ointment in LS, including different regions affected by LS [5–7, 16–32].

References


