Answers to Dermato-venereological Quiz on page 41

1. Physical examination revealed a soft porcelain-white plaque with slight epidermal atrophy. There were several similar porcelain-white macules surrounding the main plaque lesion. No other lesions were found in whole body skin examination. The differential diagnoses are: lichen sclerosis, vitiligo, morphea, tinea versicolor, lichen planus.
2. The most likely diagnosis is extra-genital lichen sclerosis (LS). Skin biopsy was performed and confirmed the diagnosis of extra-genital LS. The histopathology showed epidermal atrophy with loss of rete ridges and vacuolar changes of basal keratinocytes. The superficial dermis showed fibrosis with hyalinization. Abundant lymphocytic infiltrates were noted in deep dermis.
   Typical histopathology features of LS may include:
   - Epidermal hyperkeratosis with follicular plugging
   - Epidermal atrophy with flattening of rete ridges
   - Vacuolization of the basal layer of the epidermis
   - Marked edema in the superficial dermis (early lesions)
   - Homogenized collagen in the upper dermis (established lesions)
   - Lymphohistiocytic infiltrate underlying the zone of homogenized collagen
3. Topical steroid and phototherapy are the first line treatments most would use. Systemic treatments are required for extensive disease not control with first line therapy. Typically potent to super-potent topical steroid is used as daily application for two to three months. If response is satisfactory, topical steroid can be tapered to maintenance therapy two days per week and continue for three months. Topical steroid can be tapered off if disease remained in control. Phototherapy in the form of UVA1, NBUVB and PUVA are all reported to be effective in case series. Topical calcineurin inhibitors may be used but are less effective in extra-genital LS. Methotrexate alone or in combination with systemic steroid may be used for extensive disease or those who failed first line therapy.
4. Extra-genital LS occurs in about 10-15% of patients with LS. Extra-genital LS is rare in children. Concomitant genital LS may be present. The prognosis is difficult to characterise due to the rarity of the disease and paucity of reports in children. The disease usually follows a chronic course over several years with possibility of exacerbation and quiescence. Squamous cell carcinoma is not associated or rare with extra-genital LS.[1]

   The patient was treated with 0.1% mometasone ointment daily and clinical improvement was noted after seven months and remained static at one year with tapering of topical steroid (Figure 2).

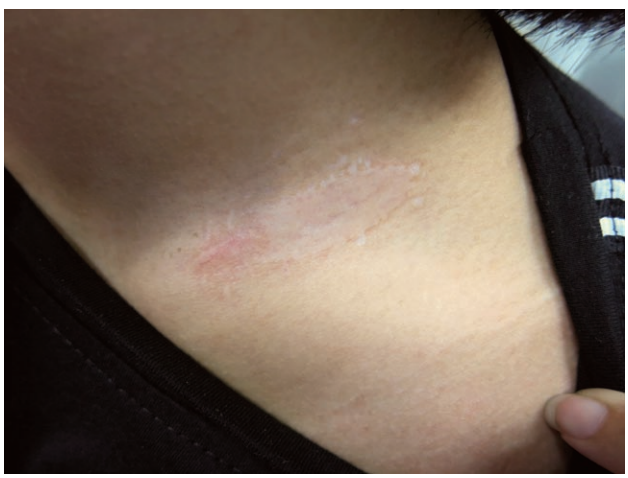

Figure 2.

Reference

1. Lewis FM, Tatnall FM, Velangi SS, Bunker CB, Kumar A, Brackenbury F, et al. British Association of Dermatologists guidelines for the management of lichen sclerosus, 2018. Br J Dermatol 2018;178:839-53.