Widespread induration of the skin

A 63-year-old male farmer presented with thickening and whitish discoloration of skin of the arms, forearms, upper chest and back of six months duration. Complaints started with itching over the right arm, which over a period of time became indurated. Skin tightness soon progressed to involve both upper limbs, chest and the back. At the same time the patient also gave history of multiple asymptomatic hypopigmented papules which coalesced to form atrophic plaques over the upper chest, forearms and back. There was no history of fever, joint pain, insect bites, circumcision, drug intake prior to the onset of lesions. There was no history of similar skin tightness over the face, hands or any other systemic complaints. Cutaneous examination revealed widespread induration of the skin of the upper back, chest, arms and both forearms. The skin of the right forearm appeared shiny because of induration [Figures 1-5]. The lower limbs and face were not affected.

Multiple hypopigmented atrophic papules and plaques were seen on the back, chest and arms. Hypopigmented plaques with prominent follicular plugging were seen on the right arm. Palms, soles, nails and genitalia showed no abnormality. Systemic examination was normal. All the rou tine hematological and biochemical parameters were normal. The antinuclear antibody (ANA) profile showed a weak positive reaction to Sm and SS-A antibodies. Findings of the skin biopsy done from the indurated lesion on the right arm. A biopsy performed from the shiny lesion on the right forearm showed flattened epidermis with homogenization of the dermal collagen and a patchy perivascular infiltrate in the dermis.

WHAT IS YOUR DIAGNOSIS?

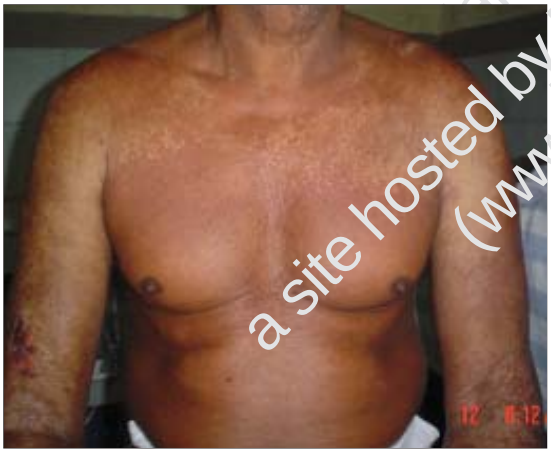

Figure 1: Multiple hypopigmented macules on the chest along with an ulcer on the right forearm overlying the hypopigmented plaque

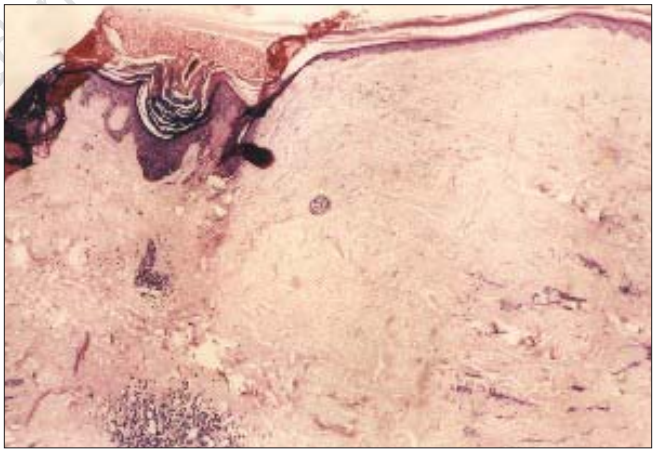

Figure 2: Thinning of the epidermis with follicular plugging, scanty dermal infiltrate with homogenisation of collagen in the dermis (x50)

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Diagnosis: Lichen sclerosus et atrophicus coexisting with morphea

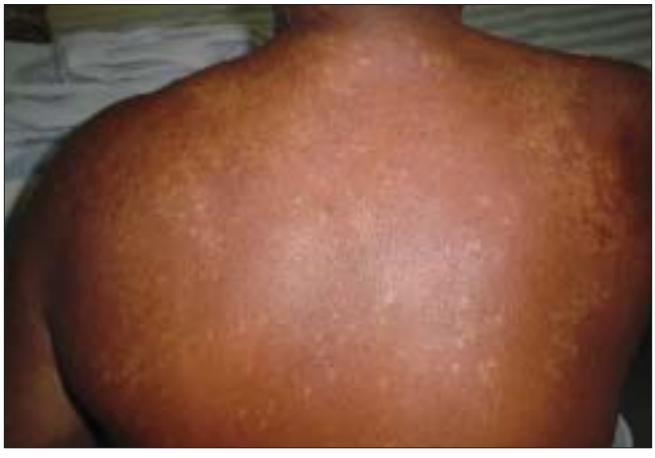

Figure 3: Hypopigmented macules on the back

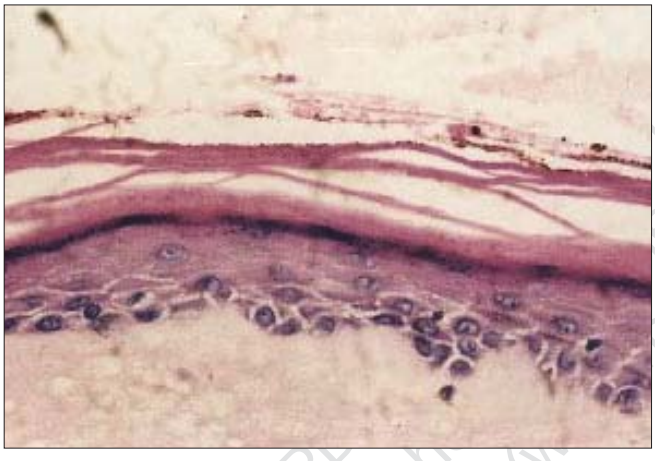

Figure 4: Basal cell degeneration (400x)

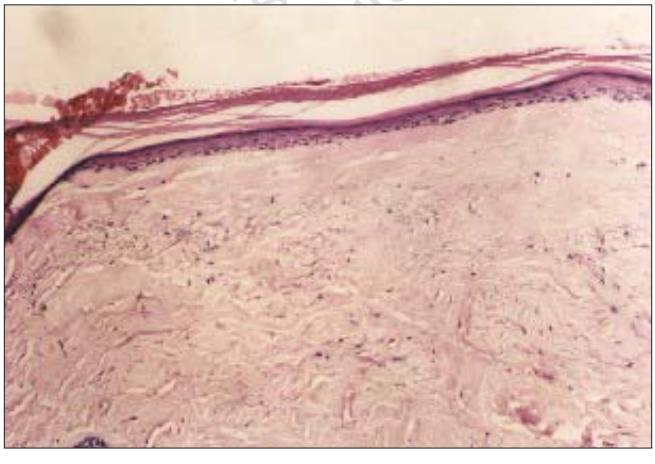

Figure 5: Homogenisation of collagen bundles in the dermis (100x)

DISCUSSION

Lichen sclerosus et atrophicus (LSEA) is an inflammatory disease of unknown cause and incompletely characterized pathogenesis. Hallopeau is usually credited with the first clinical description of what was to be later called LSEA. It can affect individuals of all age groups with reported onset from six months of age to late adulthood.[1] The majority of patients affected are women with a female to male ratio of 10:1. More cases of genital LSEA are reported than nongenital LSEA. In females, prepubertal, perimenopausal and postmenopausal vulvar and perineal involvement are most common. The symptoms include pruritus, burning pain, dyspareunia, dysuria, vaginal discharge, labial stenosis leading to "keyhole" or "figure of 8" appearance.[2] In males most cases reported are on the glans penis and prepuce with sudden onset of phimosis in a previously retractable foreskin, adhesions to the glans, decreased sensation on the glans, painful erection, meatal stenosis[3] etc. Extragenital LSEA occurs commonly on the neck and shoulders and is generally asymptomatic in both sexes. The eruption typically begins as white polygonal papules that coalesce to plaques. The appearance of being "splashed with whitewash" led to the early appellation of "white spot scleroderma". Characteristic are comedo-like plugs on the surface of the plaque that correspond to the appendageal ostia. The plugs eventually disappear with time leaving a smooth porcelain white plaque.[4] Other presentations of LSEA include bullous,[5] vesicular, hyperkeratotic,[6] vitiligoid variants[7] where pigmentary changes exist without papules and plaques, palmoplantar,[8] scarring alopecia[9] etc. Ten to fifteen per cent of LSEA cases occur in children, the majority of which involve the female genitalia. At least half the cases of genital LSEA in girls will involute by menarche. The earlier the onset of LSEA, the more likely is the resolution.[10] Premalignant and frankly malignant changes can occur in patients with LSEA. Squamous cell carcinoma (SCC) may develop subsequently in genital LSEA in male and female adults and in female children. However, SCC never occurs in extragenital lesions. Lichen sclerosus et atrophicus exhibits the isomorphic or "Koebner" phenomenon which along with trauma and infection has been thought to contribute to genital LSEA. The concept that friction from clothing may cause lichen sclerosus lesions on the shoulder and neck has also been proposed. Lichen sclerosus et atrophicus may appear in postsurgical scars and sunburns also.[11] Extragenital LSEA associated with morphea has been mentioned in the literature.[12] There are many reports in the literature linking Borrelia burgdorferi to LSEA which may play a role in its pathogenesis. There is no confirmed effective

treatment for the extragenital lesions. Calcipotriol may be effective and low-dose UVA 1 has been reported to be of benefit. For genital lesions potent topical corticosteroids give both symptomatic relief and prevent scarring. Estrogen or testosterone-containing creams, ACTH injections, etretinate, potassium p aminobenzoate, liquid nitrogen cryoprobe, carbon dioxide laser have been tried in isolated cases.[13] Our patient was started on oral minipulse therapy with six tablets of betamethasone phosphate twice weekly. At the end of six months, the patient reported significant improvement in the cutaneous induration.

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