CASE 24

A 49-year-old woman from Samutprakarn

Chief complaint : multiple of white papules for two months

Present illness : She presented with multiple pruritus papules symmetrically distributed on ankles and abdomen. The new lesions gradually developed.The patient stated that few papules appeared three years ago. She denied any systemic symptoms.

Past history : Migraine for 4 years
osteoarthritis for 5 months

Family history: Nil

Physical examination

General : normal consciousness , not pale no jaundice

Heart, Lung and abdomen: WNL

Dermatological examination

Groups of shiny white pinhead papules at lateral side of both elbows , ankles and abdomen . (Fig.24.1, 24.2)

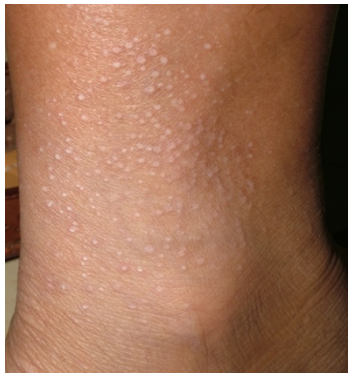
Fig. 24.1

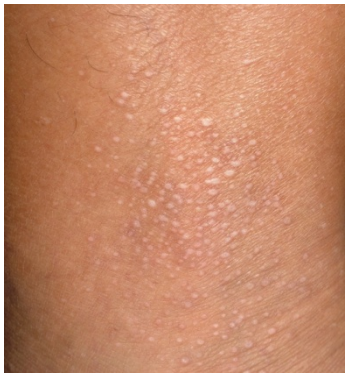
Fig. 24.2

Histopathology : (S08-12733) (Fig. 24.3, 24.4)

- Orthokeratosis and hypergranulosis

- Homogenized collagen and dilated blood vessels in the thickened papillary dermis

- Superficial infiltrate of lymphocytes beneath papillary dermis

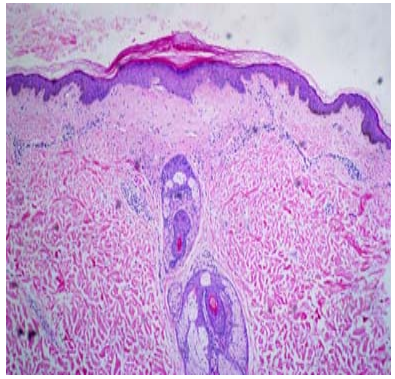
Fig. 24.3

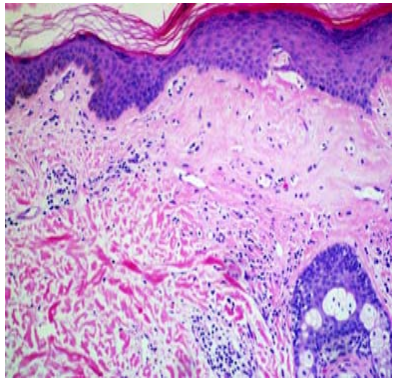
Fig. 24.4

Diagnosis : Lichen sclerosus et atrophicus

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Discussion:

Lichen sclerosus et atrophicus (lichen sclerosus , Kraurosis vulvae , Balantinitis xerotica obliterans (lichen planus of the penis))

Lichen sclerosus is a clinically distinct inflammatory disease primarily of the superficial dermis that leads to white scar-like atrophy. In non-genital skin, lichen sclerosus may itch and be cosmetically disturbing. Genital lichen sclerosus causes both dryness and severe, persistent pruritus, and it often leads to progressive atrophy and functional impairment. Phimosis and scarring of the vaginal introitus are the most frequent complications.

As in most inflammatory diseases, genetic predisposition contributes to the development of lichen sclerosus. In addition, association with the MHC class II antigen HLA-DQ7.[1]

Francois Henri Hallopeau's report on a 'lichen plan atrophique' in 1887 is considered to be the first description of lichen sclerosus.[2]

Lichen sclerosus is relatively uncommon, more common in women than in men.[3,4] It is a disease that can affect both the extragenital skin and the anogenital region. Lichen sclerosus of the oral cavity, the palms or soles is rare. They can lead to superficial scar-like atrophy or erosions. Extragenital lichen sclerosus normally does not cause symptoms except for dryness and associated pruritus. Most patients present with slightly sclerotic, white scar-like lesions that are guttate, aggregated or coalescent into a shiny, livid to ivory, relatively soft scar with a wrinkled surface. In more advanced stages, telangiectasias or follicular plugging can be seen. The flattened interface of the epidermis and dermis results in fragility of the dermal–epidermal junction; therefore, lichen sclerosus is occasionally complicated by the occurrence of bullae that tend to become hemorrhagic.

Lichen sclerosus has a specific histologic pattern. In the beginning, superficial dermal edema dominates, associated with a band-like lymphocytic infiltrate beneath that zone. The epidermis is thinned, with orthohyperkeratosis and vacuolar degeneration of the basal layer. Hyperkeratosis is especially pronounced at follicular openings and may lead to plugging. The vacuolar degeneration at the dermal–epidermal interface and the flattening of the rete ridges predisposes to the development of blisters, which may become hemorrhagic. The most important changes are found in the superficial dermis, where the pale staining reflects homogenized dermal collagen and extreme edema in early stages. Loss of elastic fibers is typical for lichen sclerosus and is not found in morphea. Clefting and hemorrhage within homogenized papillary dermis is often seen.

Treatment

Corticoseroid, clobetasol propionate 0.05% cream was applied for 12-24 weeks.[5] Clinical improvement was confirmed by histopathology. Safety and efficiency of clobetasol in the treatment of genital lichen sclerosus were documented for all age groups and in both sexes.[6,7] However, topical glucocorticoids do not cure the disease and relapses may occur.

Calcineurin inhibitors, the macrolide immunosuppressants pimecrolimus (1% cream) and tacrolimus (0.1% ointment) have been used as topical treatments for vulvar lichen sclerosus.[8]

Vitamin D derivatives , In vitro, calcipotriol inhibits the proliferation of cultured fibroblasts and topical calcipotriene 0.005% ointment has been used for the treatment of morphea.[9]

Vitamin A derivatives, reports have described improvement of both vulvar lichen sclerosus and morphea. Vitamin A derivatives, etretinate or acitretin at doses of 10-50 mg/day is effective in localized scleroderma or lichen sclerosus. Retinoids can inhibit TGF-β, one of the key cytokines that promote collagen synthesis by fibroblasts.

Hormone, topical testosterone or progesterone preparations were frequently used in the treatment of genital lichen sclerosus.

References :

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