Lichen Planus, Nitidus, and Striatus

22

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Contents

22.1 Lichen Planus

22.1.1 Epidemiology

Lichen planus (LP) is an inflammatory disease of unknown cause affecting the skin, scalp, nails, and mucous membranes. LP primarily affects middle-aged adults (ages 30–60) and is estimated to affect 0.5–1.0 % of the population worldwide [1]. Childhood LP typically accounts for 5 % or less of cases, and some studies report a higher incidence in patients with skin of color, particularly South Asians and African-Americans [2, 3].

22.1.2 Etiology

LP is characterized by a T-cell-mediated immunological reaction affecting the dermis and epidermis, leading to keratinocyte apoptosis [4]. While the role of specific trigger factors is controversial, LP has been linked to hepatitis C infection [5].

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22.2 Clinical Features

22.2.1 Distribution and Arrangement

The lesions of LP have a predilection for the flexor wrists, trunk, medial thighs, shins, scalp, oral mucosa, and genital mucosa (Figs. 22.1, 22.2, 22.3, 22.4, 22.5, 22.6, 22.7, and 22.8). LP may also affect the palms and soles. Involvement of the face is rare. The Koebner phenomenon is a common occurrence in patients with LP (Fig. 22.9). Many clinical variants of LP have been reported, including actinic (Fig. 22.10), linear, annular (Fig. 22.11), hypertrophic, and ulcerative LP. Some variants are reported to occur more often in certain patient populations. Actinic LP is characterized by lesions in the sun-exposed areas of the face, arms, hands, and neck. It primarily affects the Middle Eastern population with usual onset in the spring and summer [6]. Linear LP, in which lesions are limited to one band or streak, has been reported in up to 10 % of cases of LP in Japan compared to less than 1 % of patients elsewhere [7]. Lichen planopilaris is a disease of the scalp which is characterized by scaly papules at the base of affected hairs leading to permanent alopecia (Figs. 22.12 and 22.13).

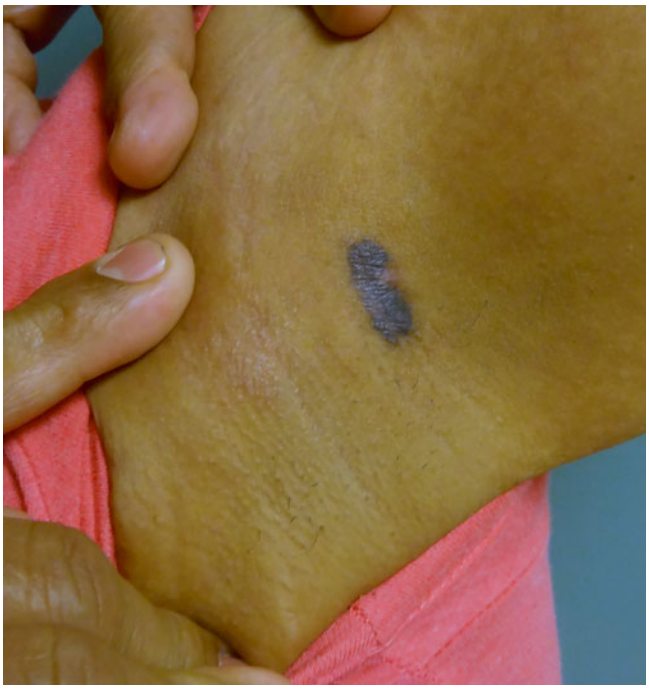

Fig. 22.1 Lichen planus of axilla in a Hispanic female

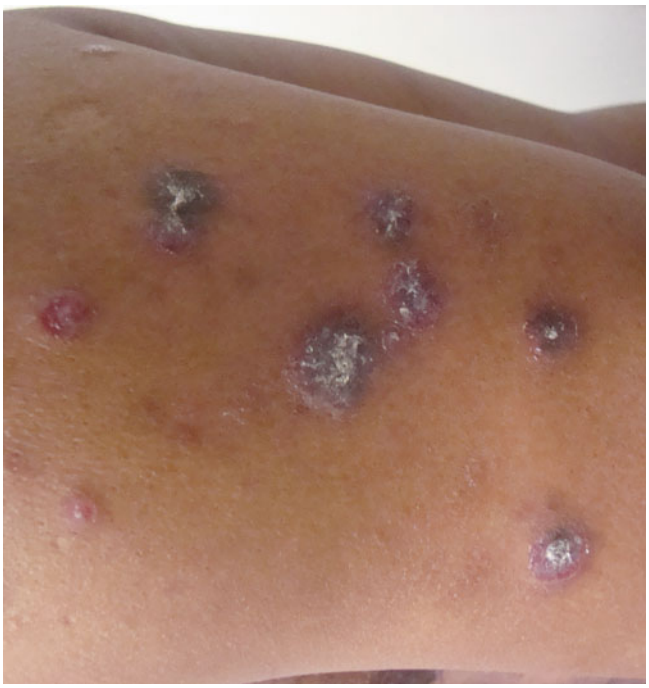

Fig. 22.3 Lichen planus of upper arm in an AA male

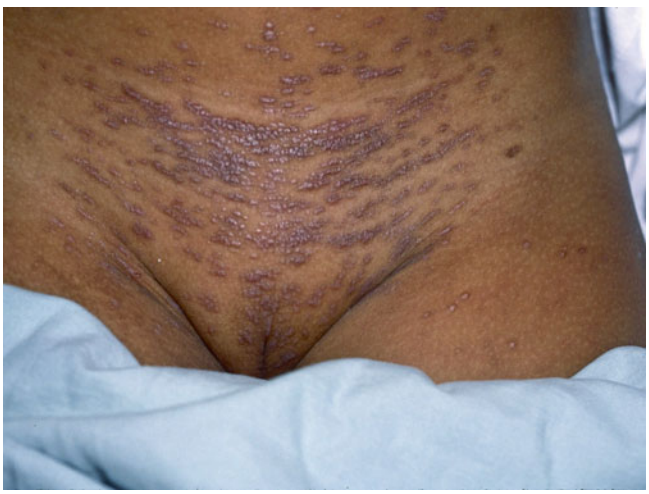

Fig. 22.2 Lichen planus of lower trunk and genital region in an AA female (Courtesy of Dr. Chauncey McHargue)

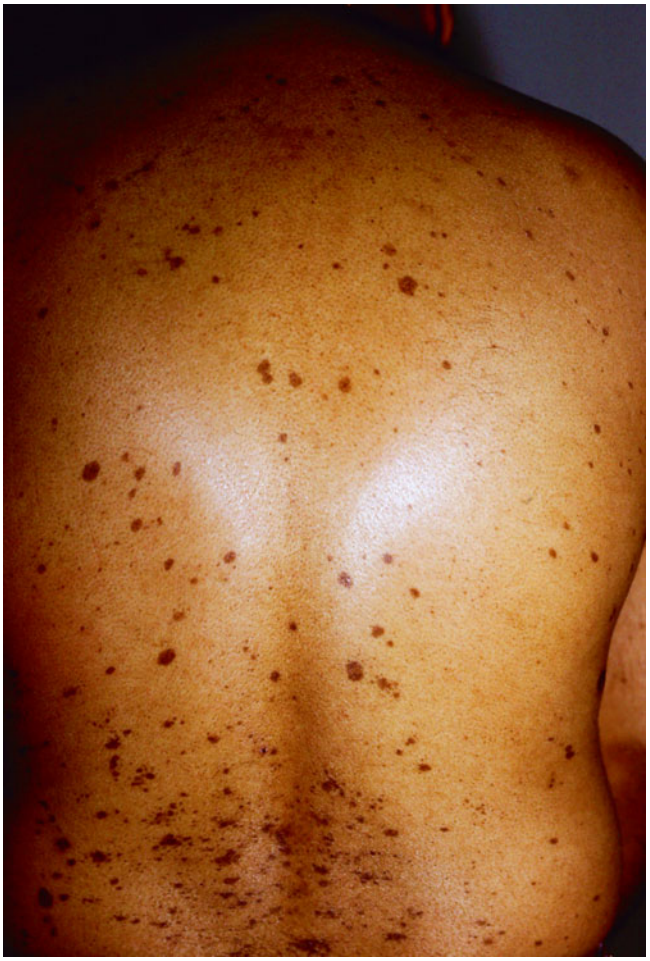

Fig. 22.4 Lichen planus of the back in a Hispanic male

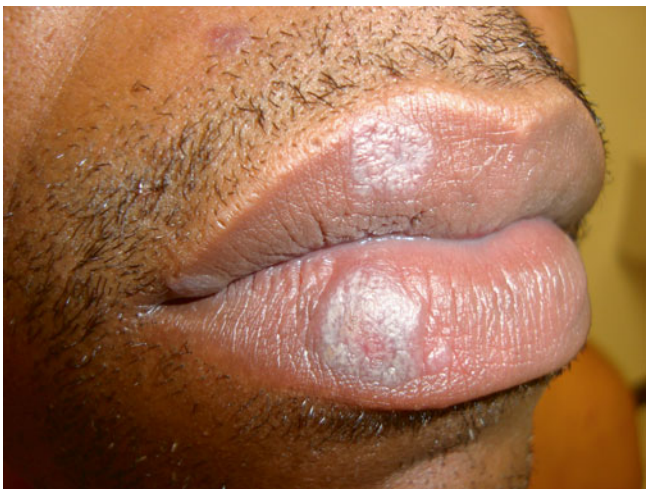

Fig. 22.5 Lichen planus of lips in an AA male

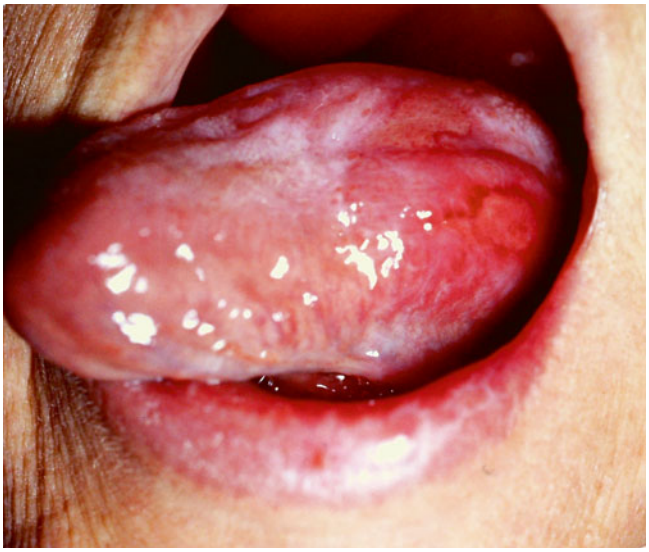

Fig. 22.6 Lichen planus of the tongue in a Hispanic female

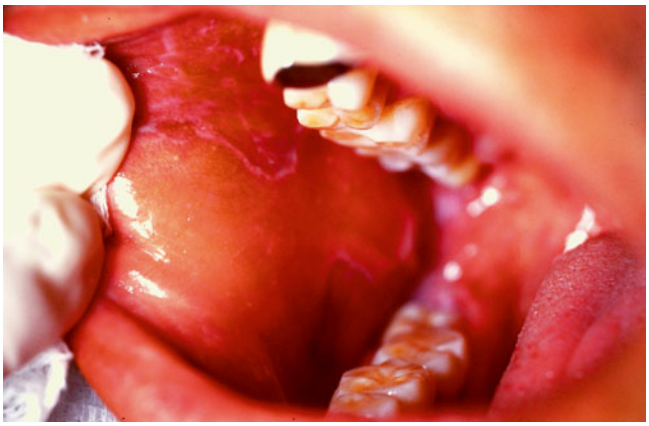

Fig. 22.7 Lichen planus of buccal mucosa in a Hispanic male

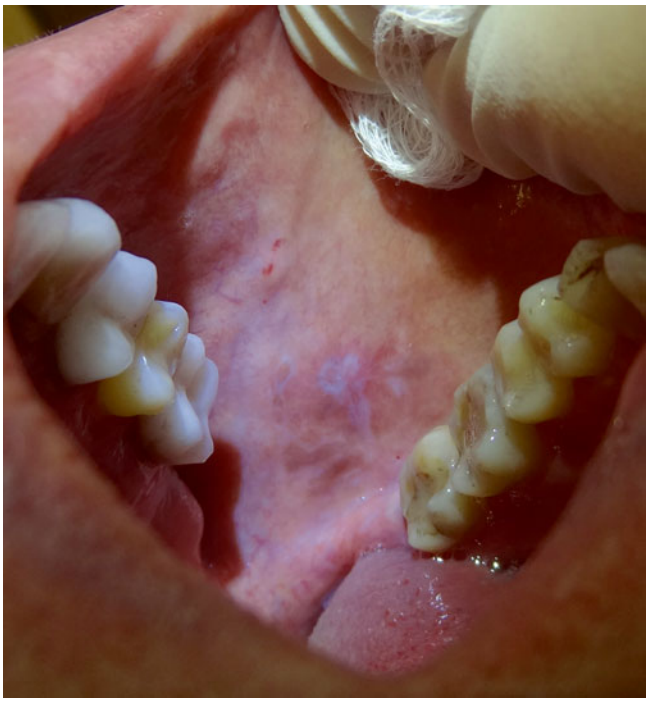

Fig. 22.8 Lichen planus of buccal mucosa in a Hispanic female

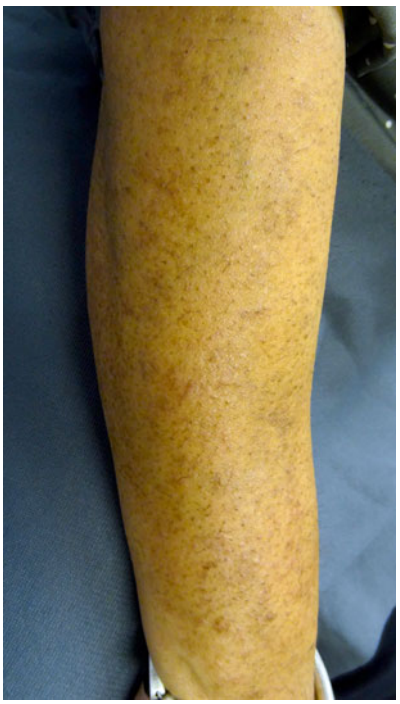

Fig. 22.10 Actinic lichen planus of the forearm in a South Asian female

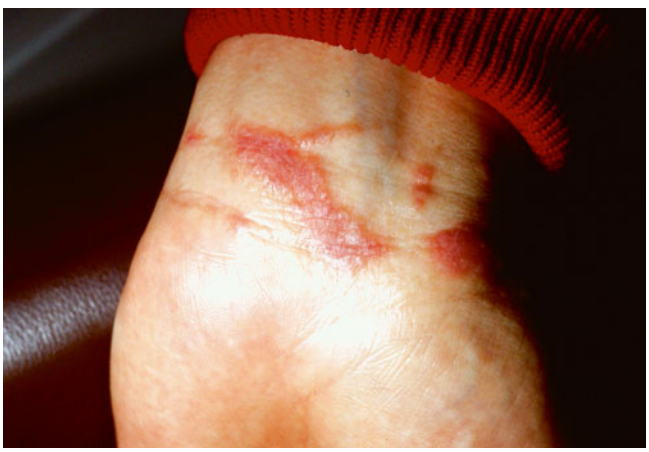

Fig. 22.9 Lichen planus of wrist in a Hispanic female

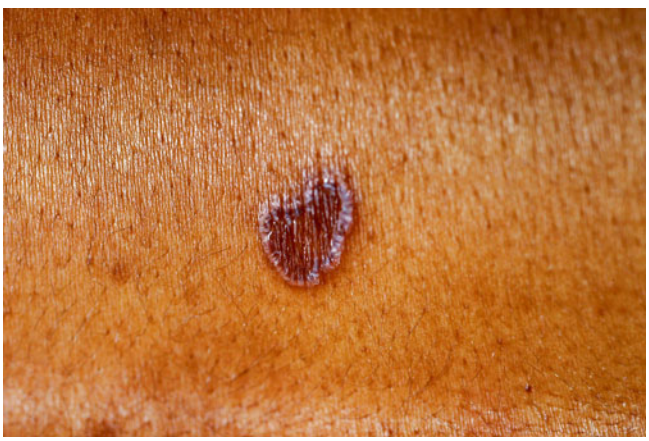

Fig. 22.11 Annular lichen planus of the arm in an AA male

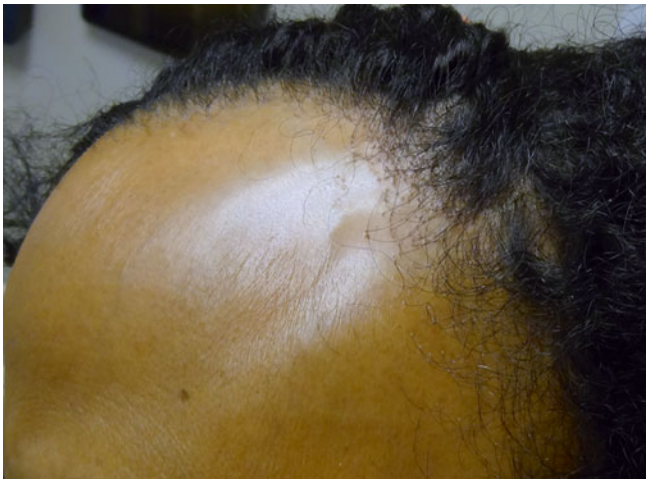

Fig. 22.12 Lichen planopilaris of scalp in an AA female

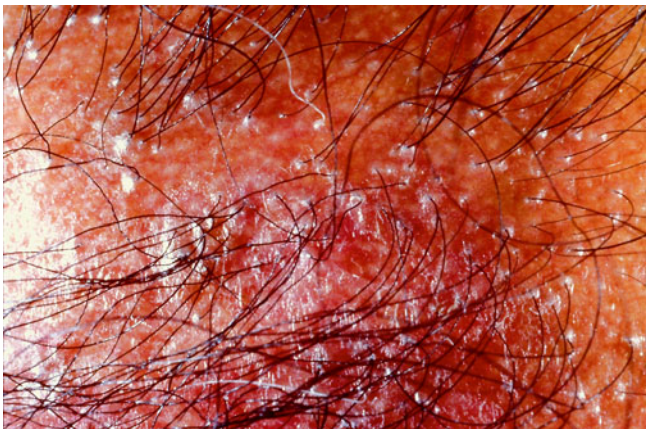

Fig. 22.13 Lichen planopilaris of scalp in a Hispanic female

22.2.2 Morphology

The primary lesions of LP are polygonal, violaceous, flat-topped papules that may coalesce into plaques. Pruritus is a prominent feature of the disease. The lesions are often covered by scant, adherent scales or a reticular network of fine white lines (Wickham striae). Postinflammatory hyperpigmentation is a common finding as the lesions clear, particularly in people with skin of color [6]. Oral lesions may be reticulate, annular, erythematous, or ulcerative and are often painful. Genital lesions are more common in men, often presenting on the glans penis, sometimes with an annular pattern. Women may have linear, white striae on the vulva and vagina. LP often affects the nails, typically with simultaneous involvement of several nails. Longitudinal ridging and splitting are common findings. Other observed changes include thinning of the nail plate, pterygium, trachyonychia, onycholysis, and subungual hyperkeratosis [1].

22.3 Natural History and Prognosis

The natural history of LP is highly variable. The majority of LP cases with skin lesions will resolve within 1–2 years. Oral disease is often chronic and resistant to treatment [4]. Recurrences of LP are common [6].

22.4 Histopathologic Features

LP is characterized by a dense, band-like lymphocytic infiltrate in the superficial dermis. Degenerative keratinocytes (Civatte or colloid bodies) are also present in the superficial dermis [4]. A "sawtooth" pattern of epidermal hyperplasia may also be seen.

22.5 Diagnosis and Differential Diagnosis

Diagnosis is often made clinically by the classic morphologic characteristics of the lesions, but a punch biopsy may be helpful to confirm the diagnosis in atypical cases. The differential diagnosis of LP includes lichenoid drug reaction, pityriasis rosea, guttate psoriasis, lichen nitidus, and lichenoid syphilid [1, 6]. The differential diagnosis of oral lesions includes leukoplakia, lupus erythematosus, candidiasis, squamous cell carcinoma, and autoimmune bullous diseases.

22.6 Treatment

For limited skin or oral lesions, high-potency topical steroids are appropriate first-line therapies. Widespread lesions may respond to oral corticosteroids. Other therapies to consider include retinoids (topical or systemic), phototherapy (UVB, UVA1, PUVA), topical calcineurin inhibitors, antifungal agents (for oral LP), and low molecular weight heparin [8, 9].

22.7 Lichen Nitidus

22.7.1 Introduction

Lichen nitidus (LN) is a chronic inflammatory condition of the skin that primarily affects children and young adults. The cause of LN is unknown although some have reported cases of LN concurrent with Crohn's disease. Others have reported an association between LN and atopic dermatitis. Familial cases of LN are rare [3].

22.7.2 Clinical Features

Lesions of LN are often localized to a few areas, including the penis, lower abdomen, upper extremities, and chest (Figs. 22.14, 22.15, and 22.16). In some cases LN assumes a widespread distribution and the lesions may coalesce into plaques with fine scale. The Koebner phenomenon is common (Figs. 22.17 and 22.18) [6].

The lesions are discrete, flat-topped, uniform papules, no larger than 1–2 mm. The color varies from yellow/brown to dark red in contrast to the violaceous lesions of LP. Unlike LP, LN is usually asymptomatic. LN occasionally affects the palms and soles with multiple hyperkeratotic papules that may coalesce into plaques.

Variants of LN have been reported in African-Americans and dark-skinned patients from the Middle East and Indian subcontinent. This disease is called summertime LN actinicus, actinic LN, or actinic lichenoid eruption. LN actinicus affects both children and adults. Lesions are identical in appearance and histology to LN but with a distribution in sun-exposed areas of the dorsal hands, forearm, and posterior neck [3]. Facial presentation of LN actinicus in three African-American girls has also been reported [10].

22.7.3 Histologic Features

LN is characterized by a focal, circumscribed infiltrate of lymphocytes and histiocytes. The epidermis often grows around this infiltrate, giving a "ball and claw" appearance. The inflammation is often granulomatous with multinucleate giant cells and epithelioid histiocytes [3].

22.7.4 Natural History and Treatment

The course of LN is slowly progressive with spontaneous remission after several years. Treatment is unnecessary in most cases. Patients who desired treatment can be treated with topical corticosteroids, dinitrochlorobenzene, PUVA, astemizole, or acitretin [3].

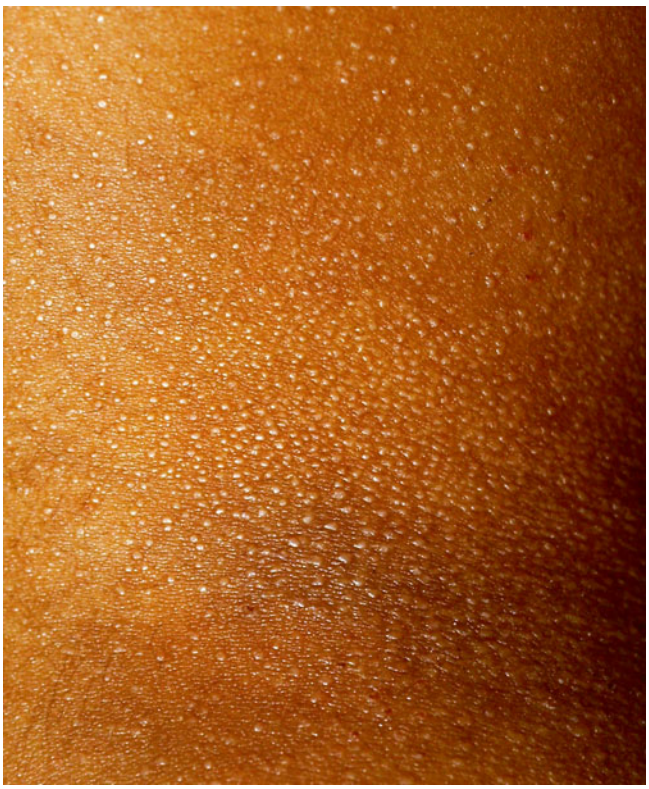

Fig. 22.14 Lichen nitidus of trunk in a Hispanic girl

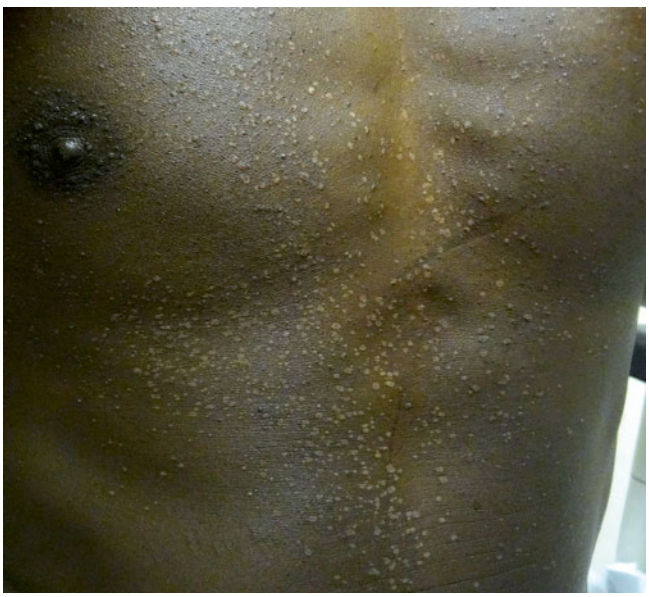

Fig. 22.15 Lichen nitidus of trunk in an African male

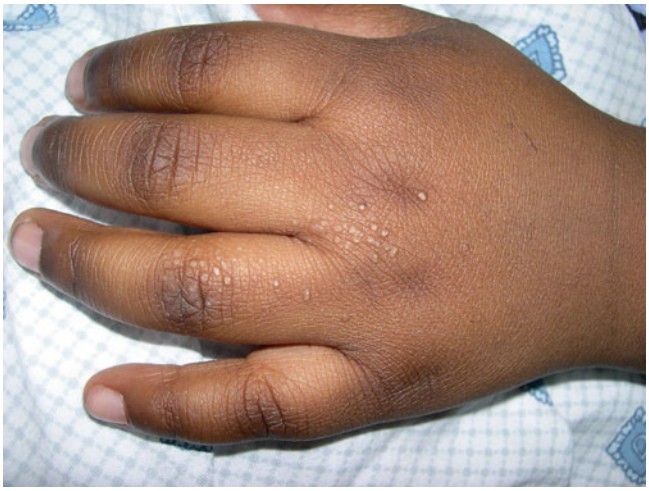

Fig. 22.16 Lichen nitidus of hand in an AA male (Courtesy of Dr Tor Shwayder)

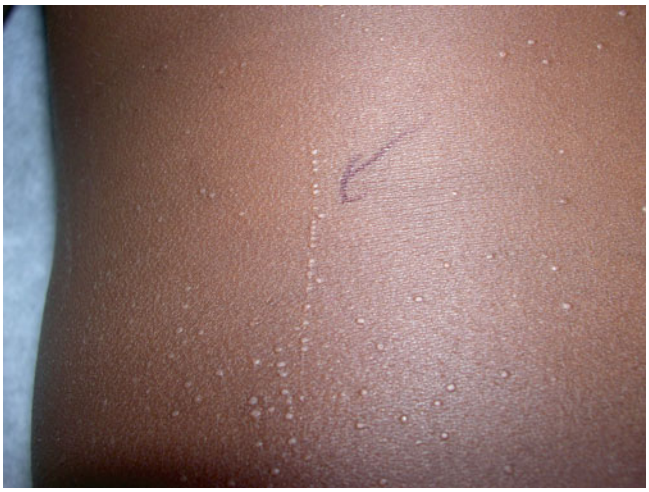

Fig. 22.17 Lichen nitidus in an AA male with Koebner phenomenon (Courtesy of Dr Tor Shwayder)

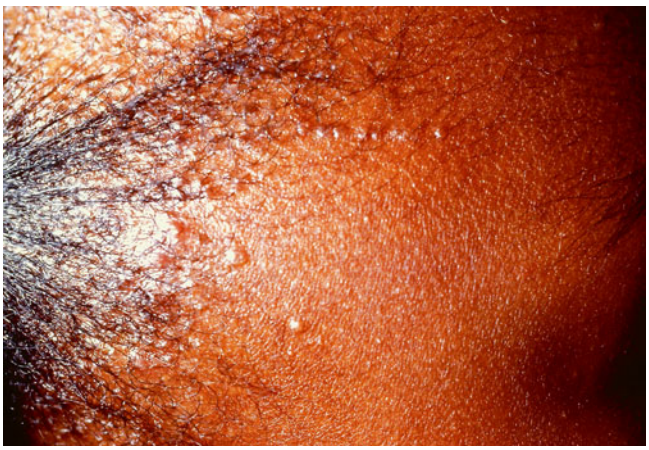

Fig. 22.18 Lichen nitidus of the temple in an African-American girl

22.8 Lichen Striatus

22.8.1 Introduction

Lichen striatus is an asymptomatic, self-limited eruption of small, scaly, erythematous papules seen primarily in children. The cause of lichen striatus is unknown [3].

22.8.2 Clinical Features

The 1–3 mm papules characteristic of lichen striatus coalesce to form a band which progresses down the extremity (more common) or around the trunk (less common) and characteristically follows the lines of Blaschko (Fig. 22.19). Hypopigmentation is prominent in persons with skin of color. Nail lesions are rare and typically restricted to a single nail [3].

22.8.3 Brief Description of Histopathologic Features

The histologic appearance of lichen striatus is variable. There is a dense lymphocytic infiltrate of the dermis which may be perivascular or band-like. Often there is a dense infiltrate around the eccrine sweat glands and ducts as well, which may help distinguish lichen striatus from LP [3].

22.8.4 Natural History and Treatment

On average lichen striatus spontaneously resolves within 1 year, but it may persist for up to 4 years. Postinflammatory hypopigmentation may last for months or several years as

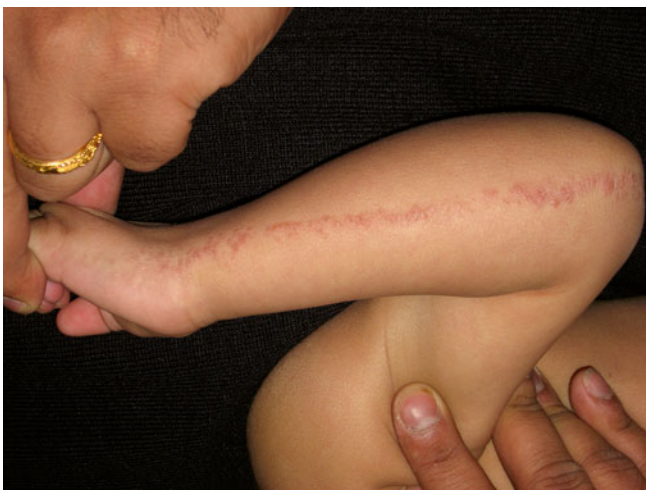

Fig. 22.19 Lichen striatus in a South Asian girl (Courtesy of Dr Tor Shwayder)

well. Recurrences are uncommon. The diagnosis is usually straightforward, but the differential diagnoses may include linear lichen planus, linear psoriasis, and inflammatory linear verrucous epidermal nevus. Treatment is usually not necessary although corticosteroids may be used to help the appearance of the lesions [3].

References

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