Papulosquamous Diseases

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Mei Di, Dong-Fang Wan, Su-Qin Xu, Yan Lu, Jian-Ming Shen, Wen-Yuan Zhu, Cheng Tan, Ru-Zhi Zhang, Fu-Quan Long, Hui Zhang, Jian-Min Chang, Lei Wang, Yang Cao,
Cheng-Rang Li, and Jie Yan

Abstract

Papulosquamous diseases that will be shown in this chapter are palmoplantar psoriasis, inverse psoriasis, pityriasis lichenoides et varioliformis acuta, purpuric pityriasis rosea, generalized papular pityriasis rosea, pigmentary pityriasis rosea, keratosis follicularis squamosa (Dohi), hypertrophic lichen planus, lichen planopilaris, lichen planus pigmentosus-inversus, linear lichen planus pigmentosus, nail lichen planus, coincidence of lichen planus and vitiligo, acute generalized lichen planus, dermatosis papulosa nigra, lichen sclerosus, erythromelanosis follicularis faciei et colli, lichen aureus, generalized lichen nitidus, and pityriasis rotunda.

M. Di
Department of Dermatology and Venereology, Beijing Anzhen Hospital, The Capital University of Medical Sciences, Beijing, China

D.-F. Wan
Department of Dermatology, The Third Anyang Municipal People's Hospital, Anyang, China

S.-Q. Xu
Department of Dermatology, The Forth Hospital of Hebei Medical University, Shijiazhuang, China

Y. Lu, M.D. · W.-Y. Zhu, M.D. · L. Wang · C.-R. Li, M.D.
Department of Dermatology, The First Affiliated Hospital of Nanjing Medical University, Nanjing, China

J.-M. Shen, M.D.
Department of Dermatology, The Second Affiliated Hospital of Tianjin Medical University, Tianjin, China

C. Tan, M.D.
Department of Dermatology, Affiliated Hospital of Nanjing University of Chinese Medicine, Nanjing, China
e-mail: tancheng@yeah.net

R.-Z. Zhang, M.D. (✉)
Department of Dermatology, The Third Affiliated Hospital of Suzhou University, Changzhou, China

F.-Q. Long
Department of Dermatology, The Second Affiliated Hospital of Kun Ming Medical University, Kunming, China

H. Zhang
Department of Dermatology, Xinhua Hospital Affiliated to Shanghai Jiao Tong University School of Medicine, Shanghai, China

J.-M. Chang, M.D.
Department of Dermatology, Beijing Hospital, Beijing, China

Y. Cao
Department of Dermatovenereology, The Jin Ling Hospital, Nanjing, China

J. Yan
Department of Dermatology, Peking Union Medical College Hospital, Chinese Academy of Medical Sciences and Peking Union Medical College, Beijing, China

W.-Y. Zhu et al. (eds.), *Atlas of Skin Disorders*, https://doi.org/10.1007/978-981-10-8037-1_9

9.1 Palmoplantar Psoriasis [1]

- Palmoplantar psoriasis (PP) refers to a localized psoriasis variant and accounts for 3–4% of all psoriasis cases.
- The classical lesions display some thick scaly, hyperkeratotic plaques, which cause a significant psychological impact on the sufferer and hampers his/her daily movements.
- Diagnosis is not always straightforward considering the frequent clinical overlap with chronic eczema. It is worth noting that syphilis can be observed in the palmoplantar area mimicking psoriasis, and therefore simple screening and verification tests should be performed in suspected patients.
- Palmoplantar psoriasis is a treatment challenge. Topical treatment is always preferred as the first-line therapy, including emollients, keratolytics, tazarotene, calcipotriol, PUVA, and UVB. Systemic therapy is needed when the topical treatments fail or when the disease becomes more severe.

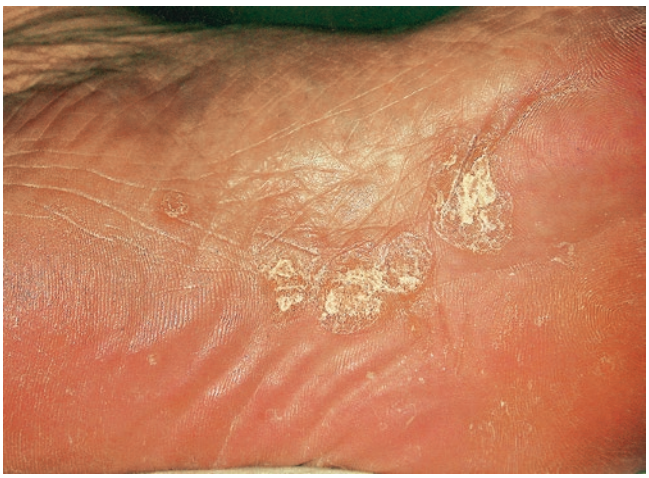


Fig. 9-1-1 Demarcated, hyperkeratotic plaques covered with scales on the left sole

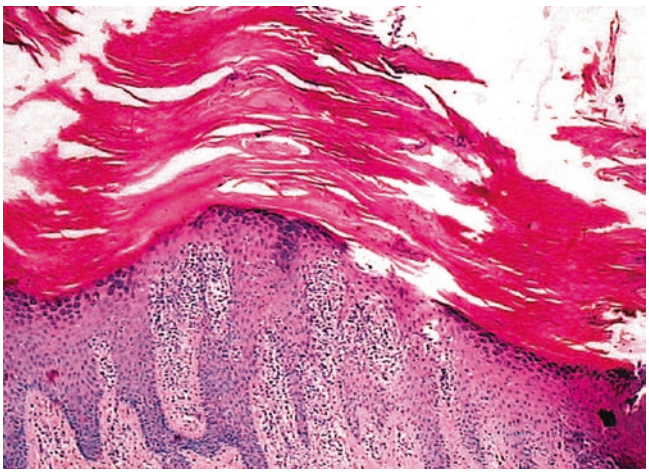


Fig. 9-1-2 Parakeratosis, the rete ridges show regular elongation with thickening in the lower portion, the papillae are elongated and the capillaries dilated, and lymphocytes infiltrated around the vessels in the upper dermis (HE stain, ×40)

9.2 Inverse Psoriasis [2]

- Inverse psoriasis (IP), also referred to as intertriginous psoriasis, is not considered a separate entity but rather a special site of involvement of plaque psoriasis, making up approximately 3–7% of patients with psoriasis.
- This condition is characterized by the development of erythematous, shiny, non-scaly plaques at intertriginous areas such as the buttocks, gluteal cleft, and axillae. Although the lesions lack the typical scaling of plaque psoriasis, minimal scaling can be seen in the more keratinized regions.
- Inverse psoriasis is treatment resistant. The preferred therapies of IP are local glucocorticosteroids and vitamin D analogues. It also requires a modified therapeutic method because it is usually less responsive to standard therapeutic regimens.

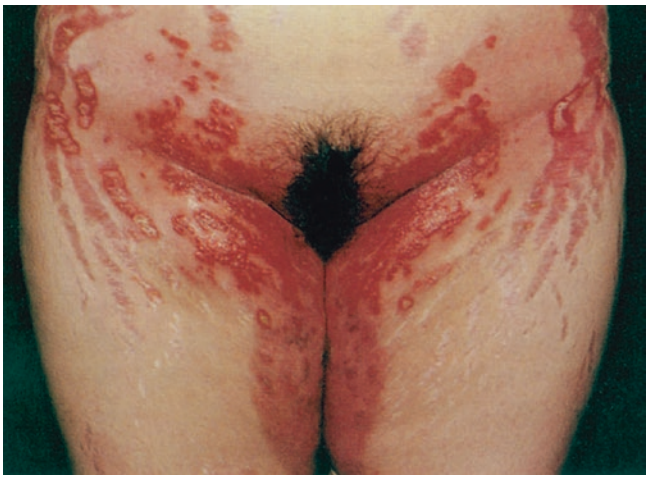


Fig. 9-2-1 Circumscribed large erythematous patches in inguinal region

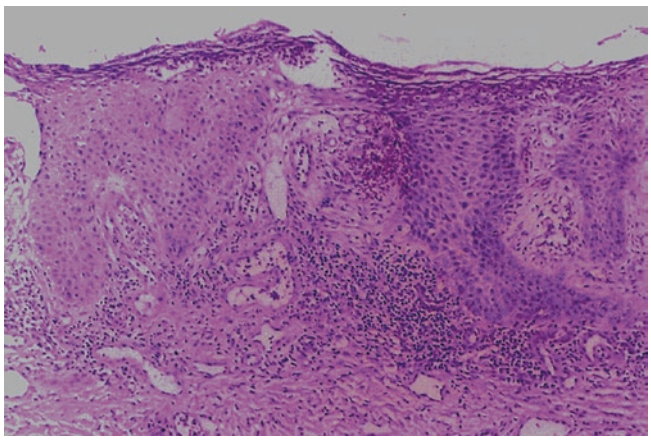


Fig. 9-2-2 Hyperkeratosis, parakeratosis, acanthosis, and elongated rete ridges; a Munro microabscess present in the stratum corneum (HE stain, ×100)

9.3 Pityriasis Lichenoides Et Varioliformis Acuta [3]

- Pityriasis lichenoides et varioliformis acuta (PLEVA) is primarily associated with lymphoproliferative reactions and may be triggered by extrinsic agents, including microbial pathogens, certain drugs, and vaccinations.
- PLEVA frequently appears on the limbs, torso, and flexural regions, which might emerge as diffuse patterns or generalized patterns. The eruption is polymorphous because lesions exist in all phases of development, which can last from a few weeks to months or years with burning and pruritus.
- PLEVA features erythematous macules that quickly evolve into papules with a fine micaceous scale. The papule usually has a central punctum that becomes vesiculopustular, goes through hemorrhagic necrosis, and then becomes ulcerated, covered with reddish brown crusts.
- PLEVA tends to be self-limited in its course, except the febrile ulcerative variant, which can be fatal. Therapy is targeted mainly at relieving itching.

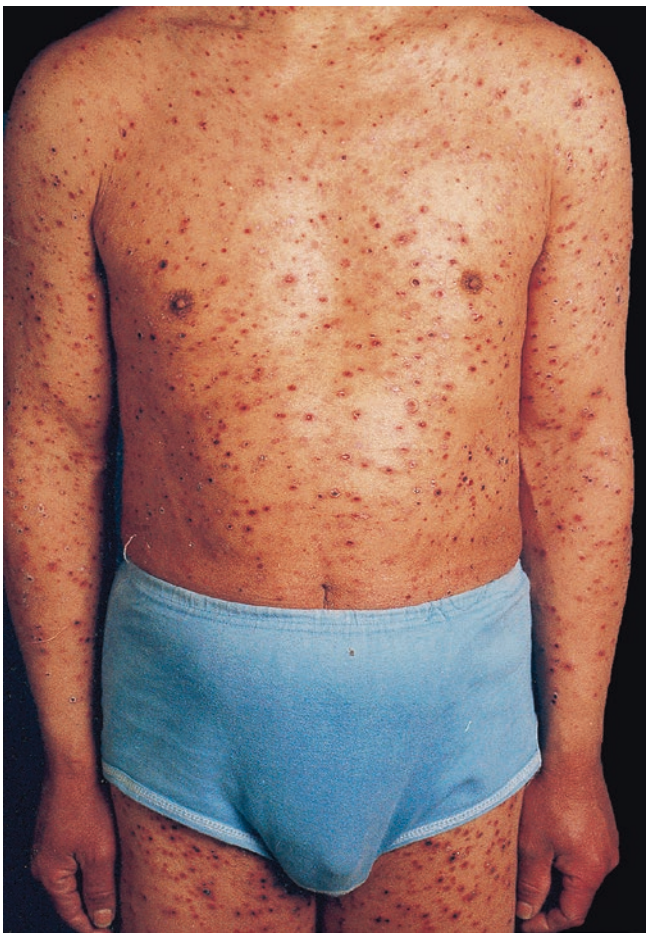


Fig. 9-3-1 A large number of papules and vesicles covered with scales and dark crests on the trunk and extremities

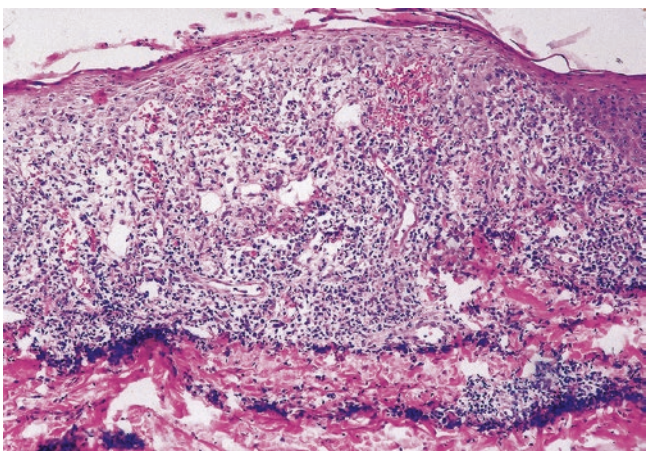


Fig. 9-3-2 Predominantly lymphocytic infiltrate invades the epidermis. The necrotic keratinocytes and liquefaction degeneration of basal cells appear in the epidermis (HE stain, ×100)

9.4 Purpuric Pityriasis Rosea [4]

- Purpuric pityriasis rosea (PPR), an uncommon type of pityriasis rosea (PR), is characterized by round to oval purpuric macules and papules over the body and proximal limbs. A herald patch might be observed. The lesions are usually oriented along skin fragmentation lines ("Christmas tree" pattern).
- The histological examination of PPR reveals patchy parakeratosis, spongiosis, dermal edema, and extravasation of erythrocytes without evidence of capillaritis or vasculitis.
- The course and prognosis of PPR are similar to classical PR, which is self-limiting. The treatment only requires emollients and oral antihistamines. To control pruritus and hasten the resolution of lesions, low-potency topical steroids may be added.

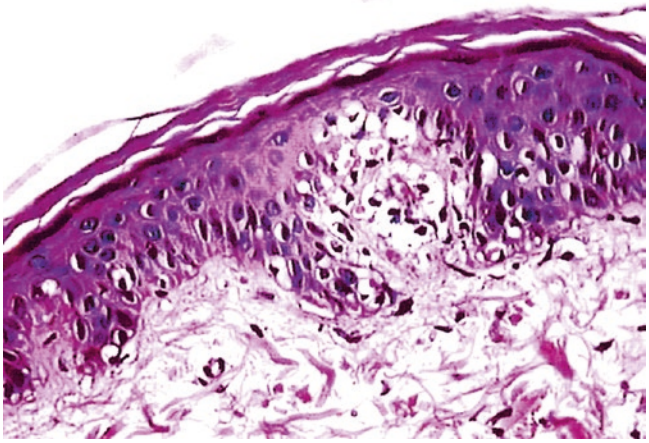
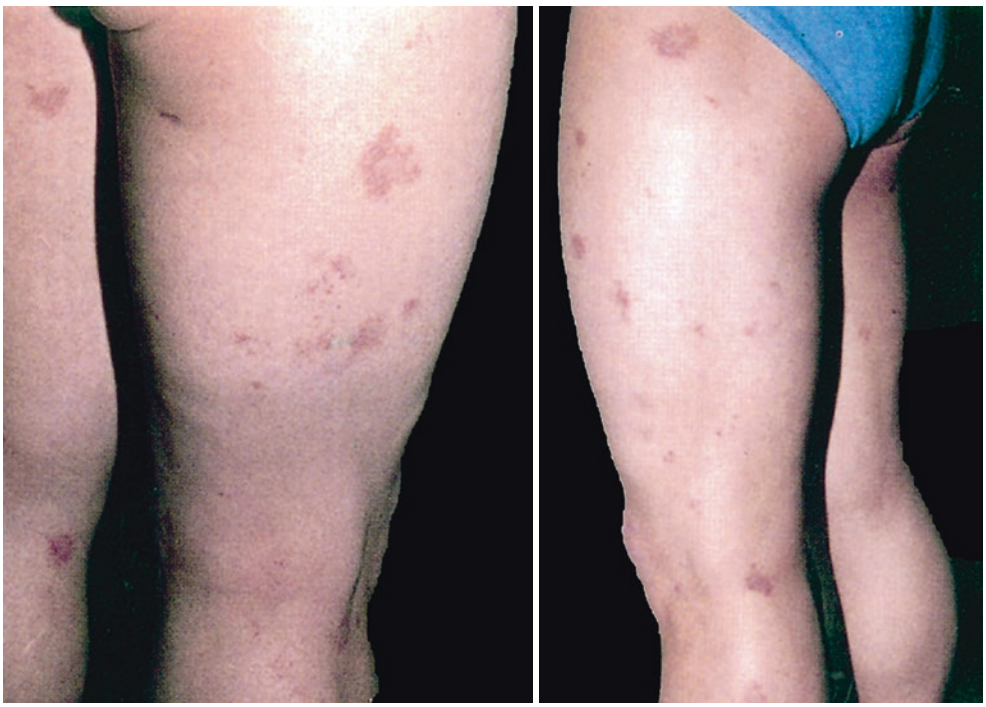


Fig. 9-4-3 Intracellular and intercellular edema, hydropic degeneration of basal cells in the epidermis, edema, and dilated vessels with extravasation of erythrocytes in the papillary dermis (HE stain, ×400)



Figs. 9-4-1, 9-4-2 Petechiae and ecchymoses covered with finely crinkled scaling on the lower limbs

9.5 Generalized Papular Pityriasis Rosea [5]

- Atypical forms of pityriasis rosea (PR) are not uncommon, accounting for up to 20% of all cases and comprising five types: relapsing, persistent, pediatric, pregnancy, and PR-like eruption.
- In terms of morphology, papular, vesicular, and purpuric forms are more common variants of PR.
- The papular form occurs in a minority of patients but is common in children and pregnant women, emerging as various papules (1–2 mm). The lesions may be observed together with classic PR lesions.
- The popular PR is easily confused with a wide variety of similarly appearing cutaneous disorders, posing a challenge for an accurate and timely diagnosis.

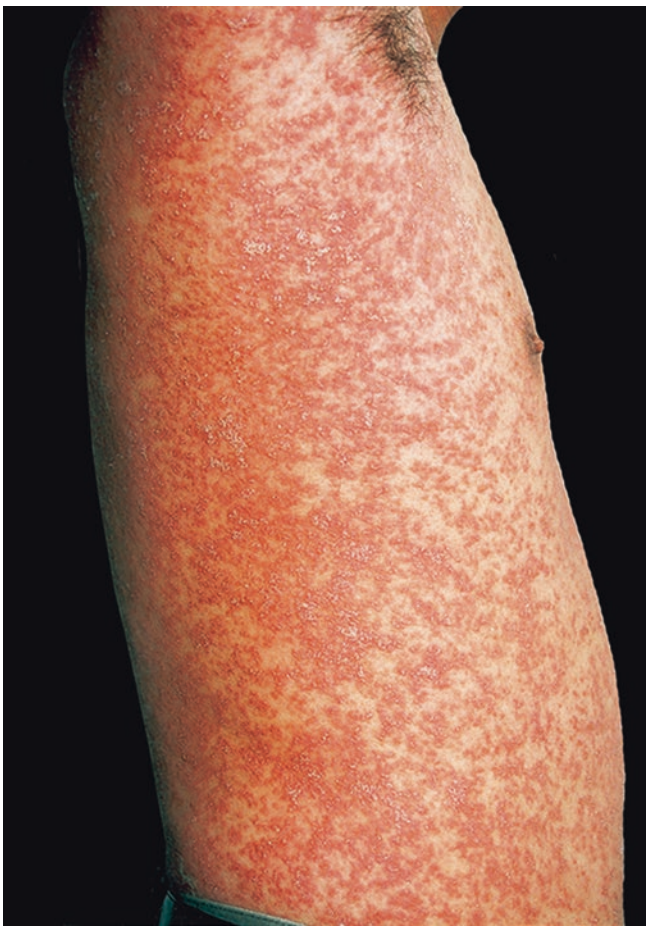


Fig. 9-5-1 Dense red scaly papules on the right flank and waist, part of them confluent with macules

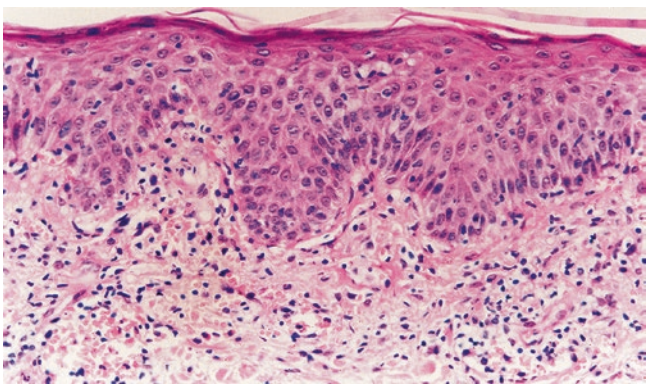


Fig. 9-5-2 Parakeratosis and slight acanthosis, spongiosis, intracellular edema, dilated capillaries, erythrocyte extravasation, and perivascular lymphocyte infiltrate were present (HE stain, ×200)

9.6 Pigmented Pityriasis Rosea [6]

- The incidence rate of atypical pityriasis rosea is 10–15%. According to their morphology, atypical pityriasis rosea can be grouped under the following varieties: purpuric, vesicular, urticaria, generalized popular, lichenoid, erythrodermic, and EM-like forms.
- Many light or dark brown ovular macules without scales occur on the trunk and minimally sun-exposed surfaces. These are arranged along the long axis and run parallel to the lines of cleavage. Skin lesions may be asymptomatic and continue for a long time.

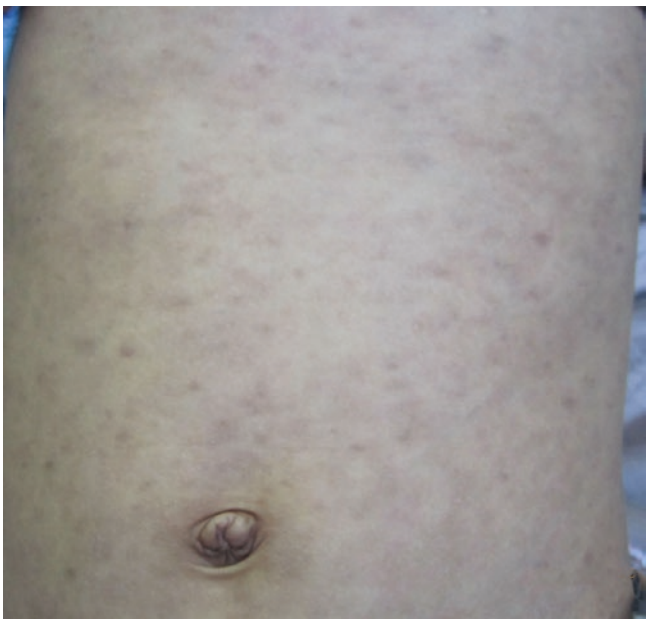


Fig. 9-6-1 Many oval or circinate pigmented patches were arranged along the long axis of the ribs

9.7 Keratosis Follicularis Squamosa (Dohi) [7, 8]

- Keratosis follicularis squamosa (KFS) is characterized by 3–10-mm-diameter asymptomatic scaly patches with follicular plugs on the trunk.
- Bacterial infection, hormonal disorders, and heredity have been considered as methods of pathogenesis of KFS.
- Five cases of KFS have been reported. The skin lesions of KFS were caused by a tight brassiere, belt, and swimsuit, respectively.

Fig. 9-7a-1 The brownish scales with black follicular horny plugs in the center were limited to the area beneath the cloth brassiere

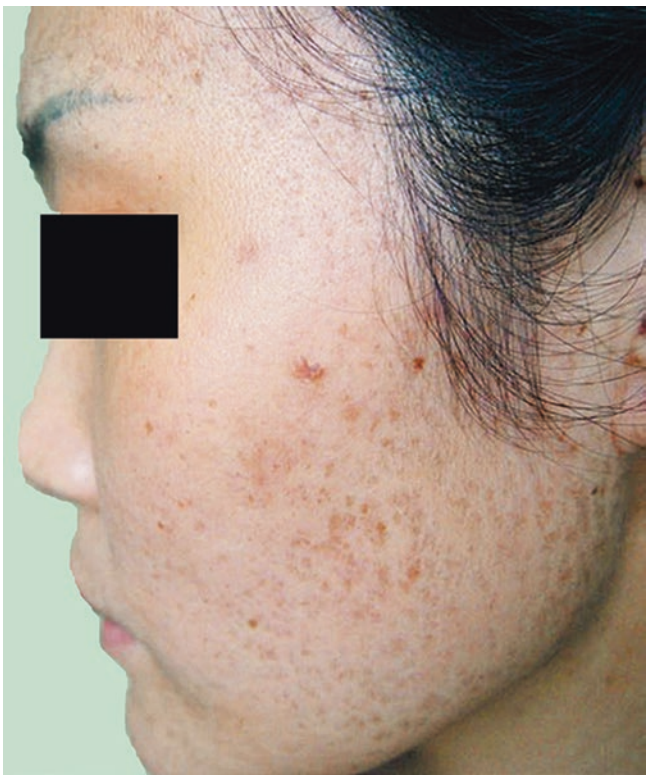


Fig. 9-7b-1 Atrophic acne scars like lesions, with a tiny brownish follicular plug in the center

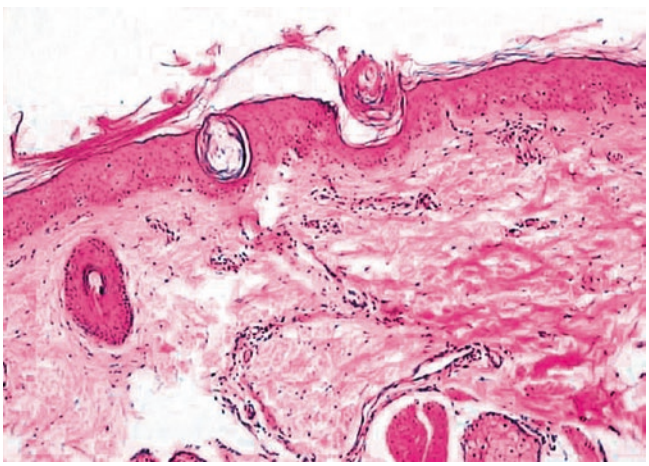


Fig. 9-7b-2 Dilated follicles with keratotic plugs and orthohyperkeratosis of the epidermis. Lateral margins of the scales were slightly detached from the granular layer (HE stain, ×40)

9.8 Hypertrophic Lichen Planus [9, 10]

- Hypertrophic lichen planus (HLP) is a distinct variant of cutaneous LP that features prominent proliferation in the epidermis and severe itching. It prevails on the lower limbs, particularly the tibia and ankle joints.
- Histological examination of HLP suggests excessive hyperplasia of the epidermis, acanthosis, a thickening stratum granulosum, and compact or lamellated hyperkeratosis, with pseudoepitheliomatous observations and liquefaction of the basal cells and a band infiltration of inflammatory cells in the higher dermis.
- The favored treatment for cutaneous LP is high-potency topical corticosteroids. Local triamcinolone acetonide is a satisfactory treatment for HLA. Acitretin (Soriatane) is used for more severe cases that do not respond to topical treatment.

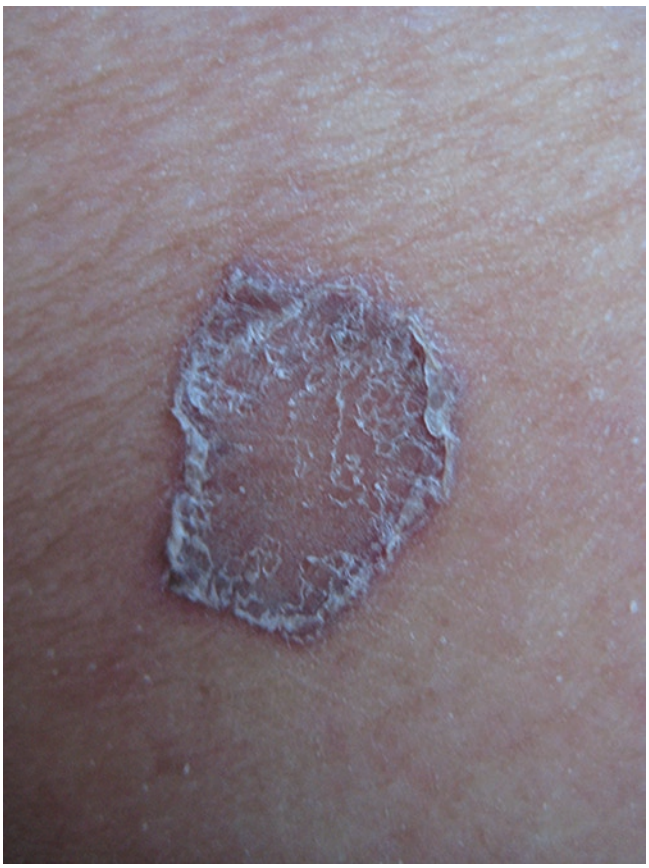


Fig. 9-8-1 A large verrucous plaque with a few scales had an elevated border and center pigmentation

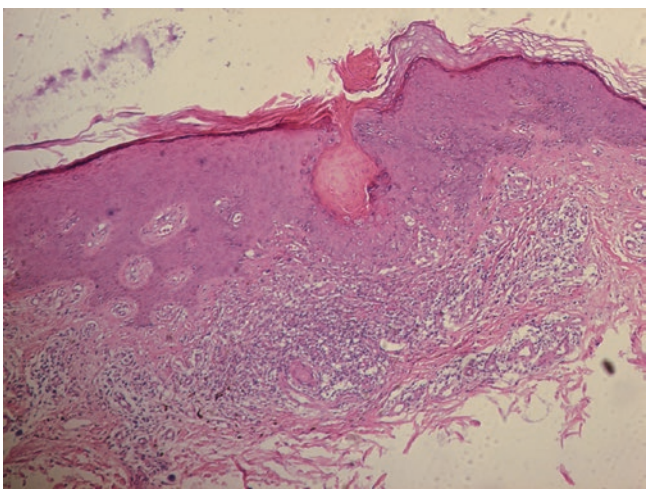


Fig. 9-8-2 Hyperkeratosis and epidermal hyperplasia, liquefaction of the basal cells, and a band-like infiltration of inflammatory cells in the upper dermis

9.9 Lichen Planopilaris [11]

- Lichen planopilaris (LPP), also known as follicular lichen or follicular lichen planus, is a common form of primary cicatricial alopecia that is seen in 80% of middle-aged women.
- The clinical presentation of LPP varies, with early forms presenting slight hair loss. Dermatoscopy and scalp biopsy are helpful for a diagnosis of LP.
- Its treatment is controversial and often unusual. The main goal of therapy is to prevent the progression of cicatricial alopecia.

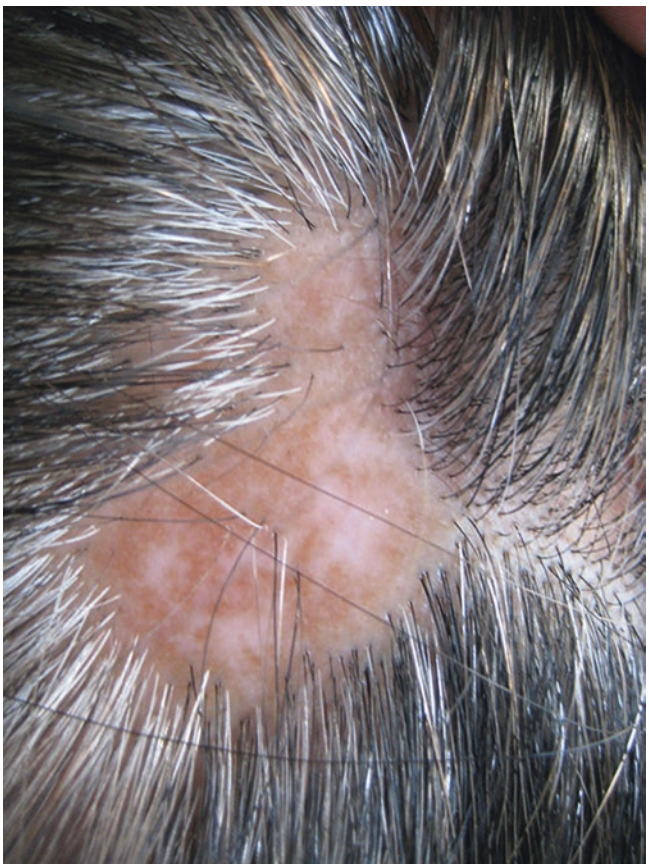


Fig. 9-9-1 The ivory white irregular patches of pseudopelade on the vertex

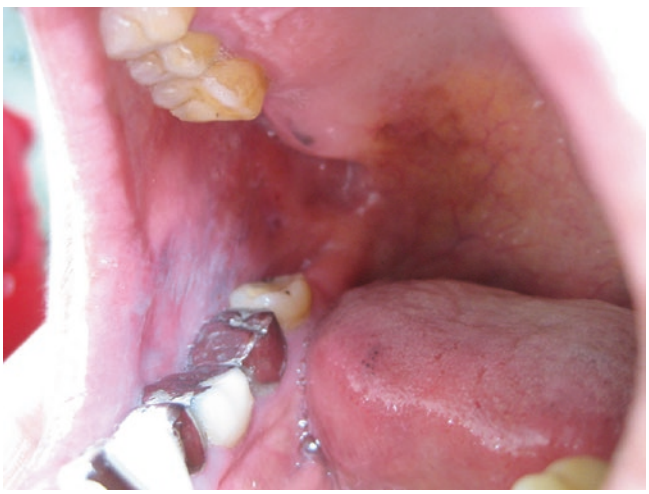


Fig. 9-9-2 The lacy white lesions were on the buccal mucosa

9.10 Lichen Planus Pigmentosus-Inversus [12]

- Lichen planus pigmentosus-inversus (LPPI) features hyperpigmented macules, predominantly over the intertriginous and flexural skin folds.
- Rarely, the lesions in LPPI coincide with Langer's lines of cleavage. External mechanical stimulus to intertriginous areas is a reasonable explanation for its occurrence.

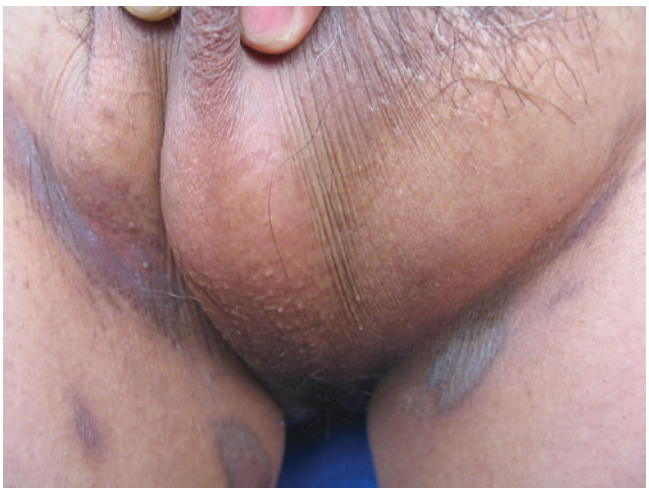


Fig. 9-10-1 Brownish to grayish macules distributed along Langer's lines in the bilateral gluteal areas

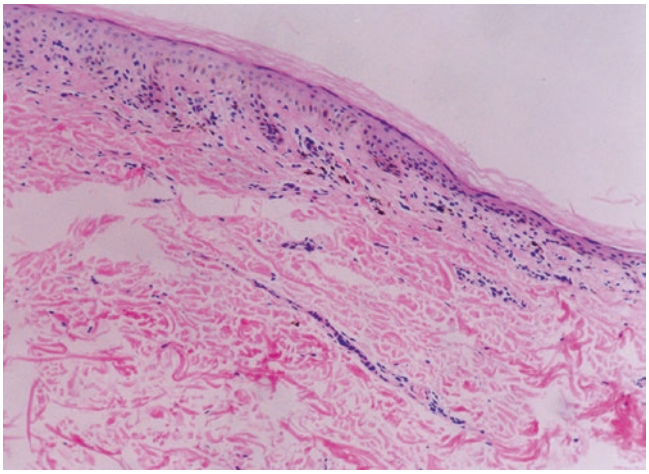


Fig. 9-10-2 Hyperkeratosis, epidermal atrophy, and lymphocytic infiltration in the dermis (HE stain, ×100)

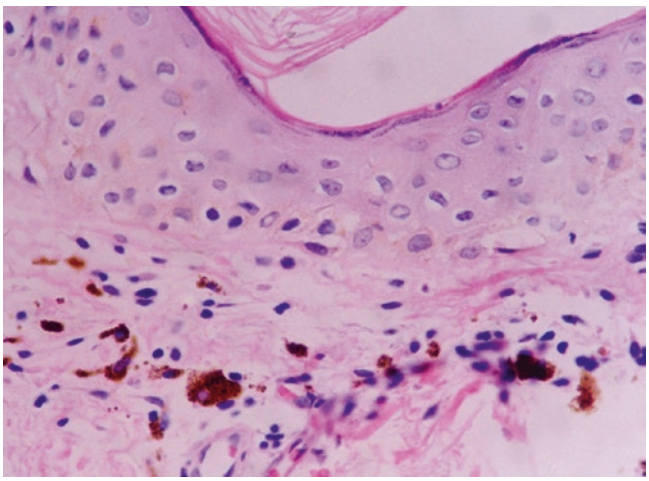


Fig. 9-10-3 Liquefaction degeneration of basal layer cells; melanophages can be noted in the dermis (HE stain, ×400)

9.11 Linear Lichen Planus Pigmentosus [13]

- Lichen planus pigmentosus (LPP), a subtype of lichen planus, is a relatively infrequent cutaneous disorder that runs along Blaschko's lines.
- The representative presentations of LPP are spotted or retiform hyperpigmented, with nigger-brown macules or papules. Lesions of LPP preferentially emerge in regions that are usually exposed to the sun and in flexural folds.
- The differential diagnosis of LPP involves linear dermatosis such as lichen striatus, postinflammatory hyperpigmentation, linear and ashy dermatosis, and whorled nevoid hypermelanosis. At present, hydroquinone may be a pivotal kind of topical agent.

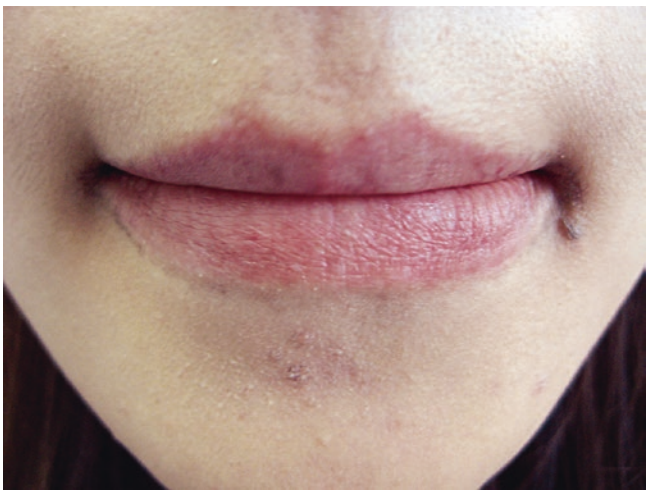


Fig. 9-11-1 There was a well-defined dark brown streak paralleling near the lower lip line

Fig. 9-11-2 A few dark porphyreous speckles clustered on the skin near to the nail fold of right fingers except the thumb

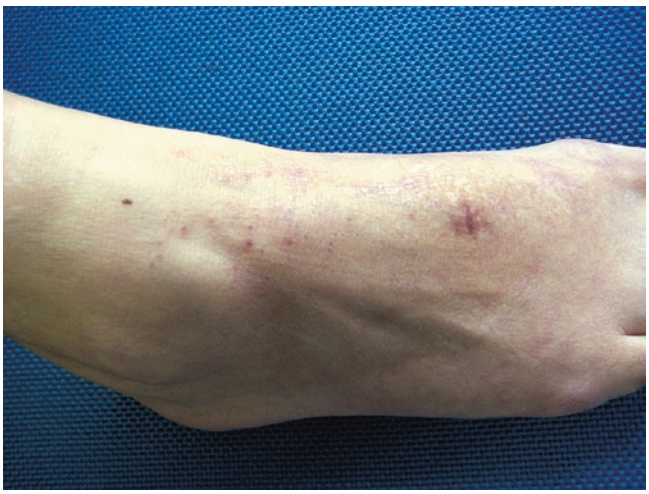


Fig. 9-11-3 On the dorsum of the right foot, many flat-topped, shiny, purple, and polygonal papules are presented, showing a distribution of zosteriform pattern

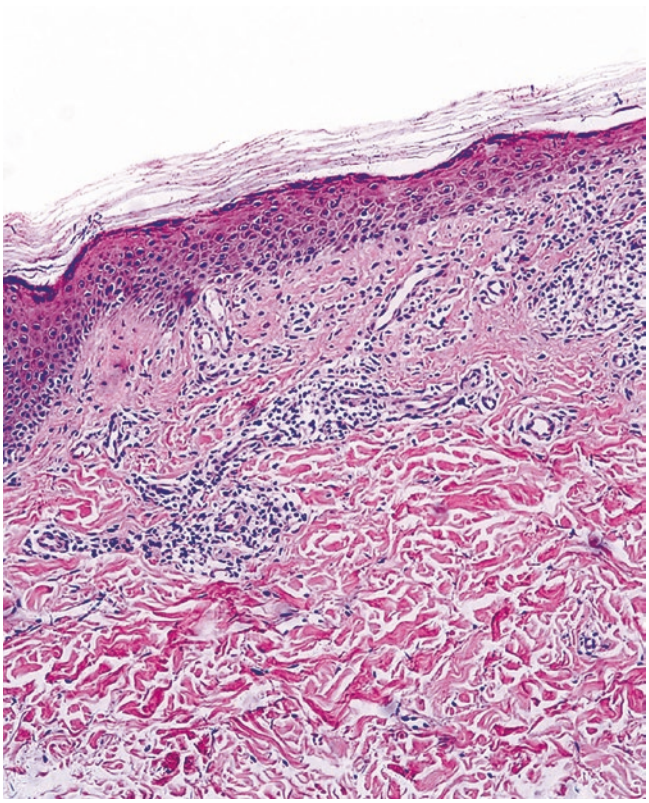


Fig. 9-11-4 Hyperkeratosis, parakeratosis, and atrophy of the epidermis with liquescent degeneration of the basal cells. A perivascular lymphohistiocytic infiltration and pigmentary incontinence in the dermis were also noted (HE stain, ×200)

9.12 Nail Lichen Planus [14]

- The incidence rate of nail lichen planus (NLP) is approximately 10% in patients with lichen planus (LP). NLP features destruction of the nail plate, particularly the fingernails.
- NLP is usually located in the matrix, presenting with nail plate thinning, longitudinal ridging, and fissuring, with occasional onycholysis with or without subungual hyperkeratosis.
- Early diagnosis is vital because of its aggressive behavior. Histopathology should be carried out in suspected cases, while dermatoscopy has proven to be useful for its diagnosis.
- It is hard to treat this disorder, which has a high rate of recurrence. Alitretinoin is a satisfactory treatment option.

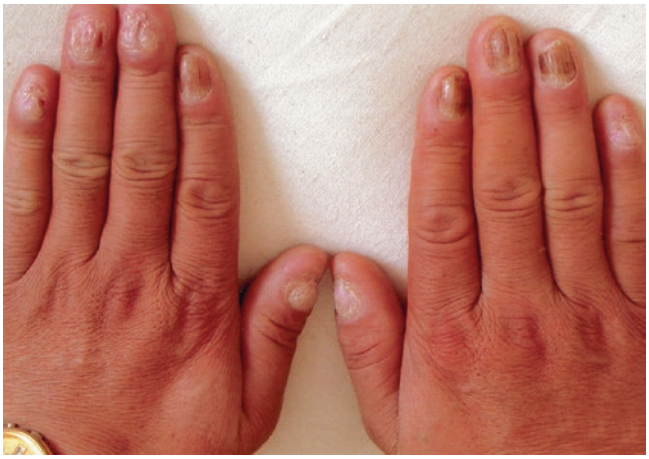


Fig. 9-12-1 Irregular longitudinal grooving and ridging of the nail plate, thinning of the nail plate, pterygium formation, and shedding of the nail plate with atrophy of the nail bed

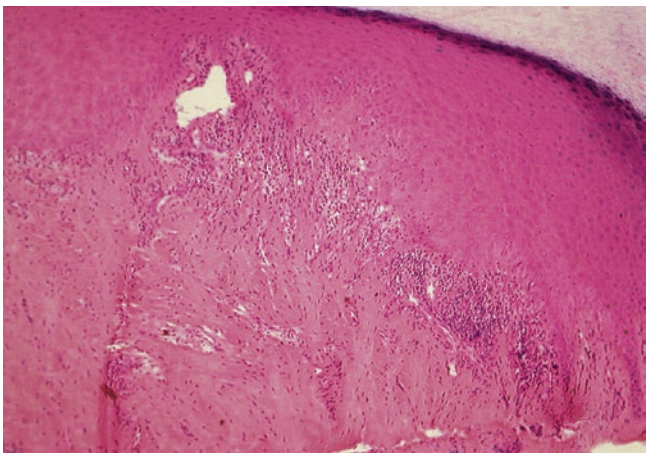


Fig. 9-12-2 Acanthosis, liquefaction degeneration of basal layer cells, and lymphocyte band-like infiltrate in the upper dermis (HE stain, ×100)

9.13 Coincidence of Lichen Planus and Vitiligo [15]

- Lichen planus and vitiligo are relatively frequent cutaneous diseases. Given a relevant prevalence of 0.5~1% of vitiligo and lichen planus, the coexistence of these disorders in one patient is predictable.
- However, the appearance of both conditions in one patient is not just an occasional phenomenon based on the probability of an autoimmune background or usual pathogenesis.
- Immunity has a direct effect on the etiology of lichen planus and vitiligo. The coincidence of these two skin disorders has been reported solely or concomitant with other autoimmune disorders.

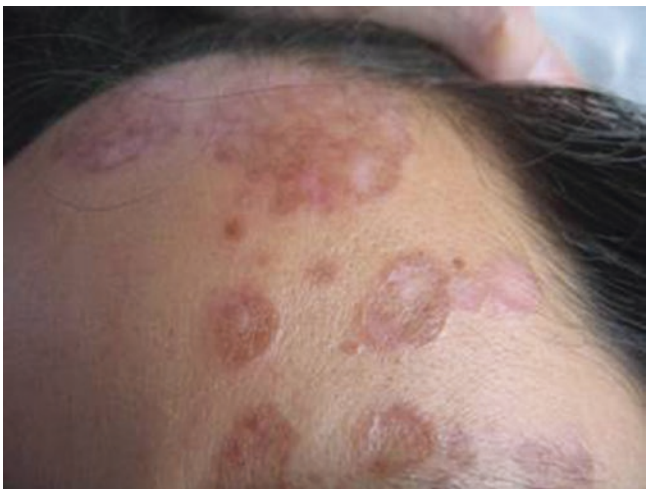


Fig. 9-13-1 Madder red or brown, round or polygonal, 0.3~3.0 cm in diameter maculae or patches, with an elevated borderline on the left side of the frontal region

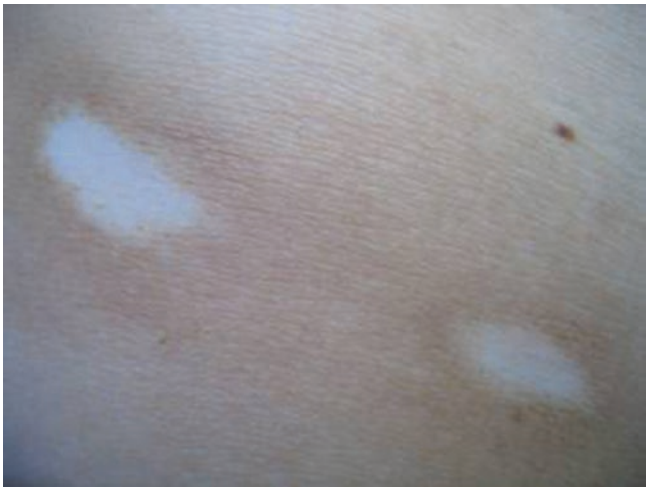


Fig. 9-13-2 Irregular depigmented patches on the right side of the waist (2 cm × 2 cm ~ 2 cm × 5 cm)

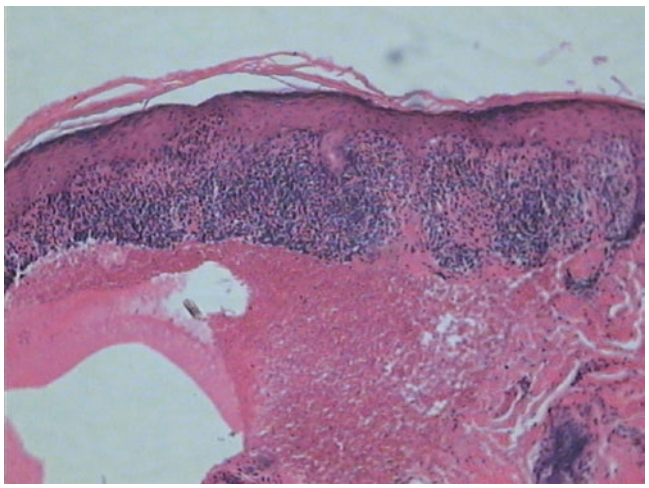


Fig. 9-13-3 Hyperkeratosis, focal hypergranulosis, and irregular acanthosis in the epidermis (HE stain, ×40)

9.14 Acute Generalized Lichen Planus [16]

- Acute generalized lichen planus presents as generalized, erythematous papules and hyperpigmented macules with a flat to. The shape of the lesions is round, polygonal, or umbilicated. Patients mostly present severe pruritus.
- This condition may pose a therapeutic challenge for dermatologists if the condition persists or flares after topical or systemic corticosteroid therapy. Another choice of therapy for this disease might be acitretin.

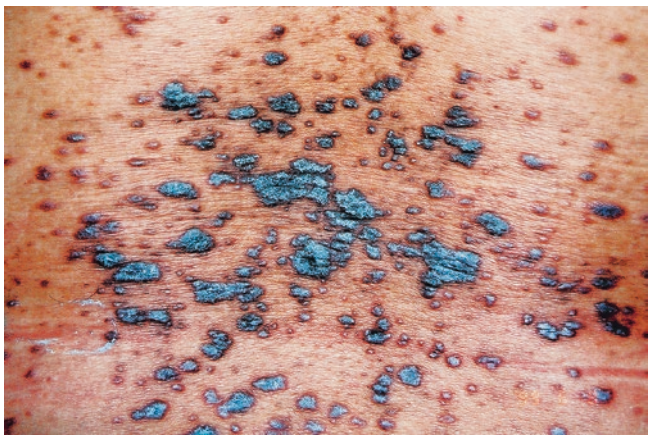


Fig. 9-14-1 Thick, polygonal, violaceous, hyperkeratotic papules on the waist

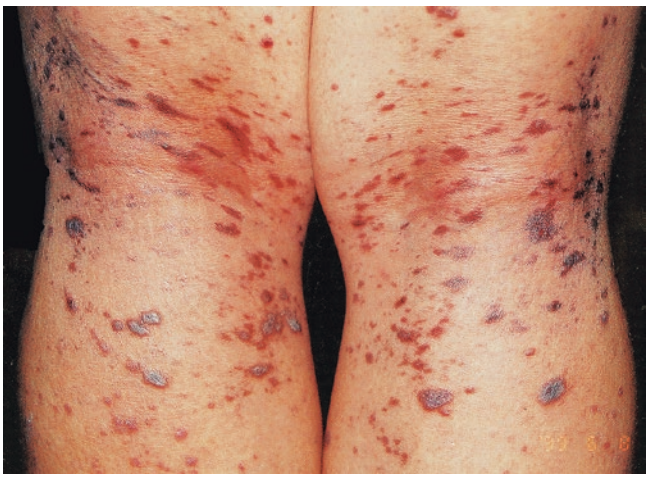


Fig. 9-14-2 Dense violaceous papules on the flexor surfaces of both lower limbs

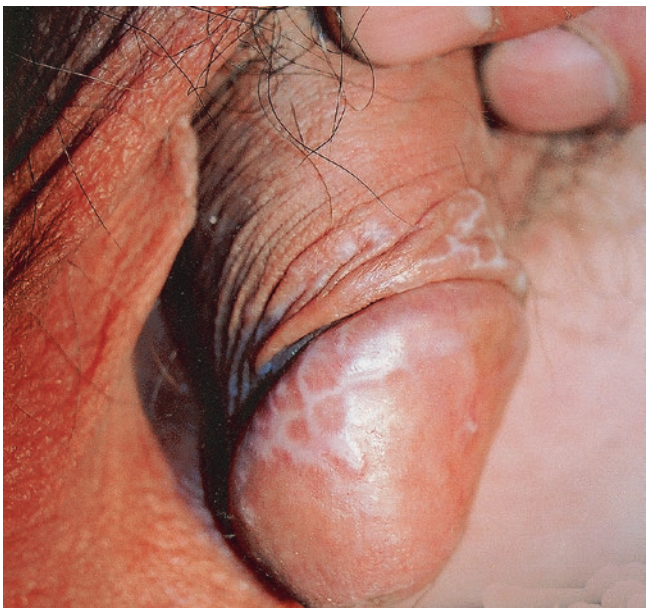


Fig. 9-14-3 Dendritic whitish maculae on the glans penis and foreskin

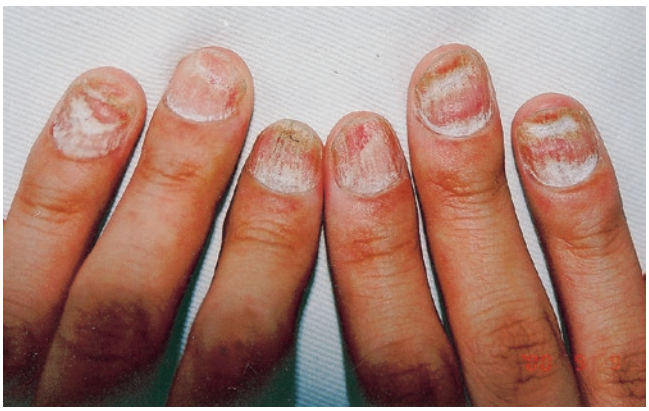


Fig. 9-14-4 Roughened nails with white scales on the fingers

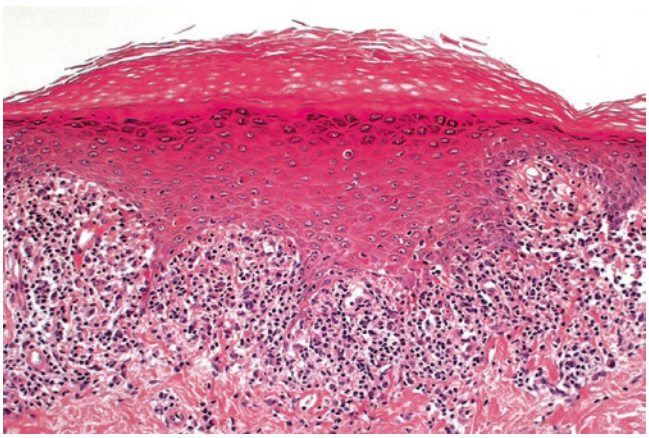


Fig. 9-14-5 Hyperkeratosis, hypergranulosis, acanthosis, basal cell liquefaction degeneration, and a band-like lymphocyte infiltrate in the upper dermis (HE stain, ×100)

9.15 Dermatosis Papulosa Nigra [17]

- Dermatosis papulosa nigra (DPN) is considered to be a benign cutaneous disorder. The manifestation of DPN presents as multiple, small (1–5 mm), excessive pigmentation macules and papules. Lesions frequently occur in the malar regions of the face, neck, forehead, and trunk.
- It usually begins in adolescence and is far more common in females than males, with a ratio of two to one. The histological findings of DPN are the same as those of seborrheic keratosis (acanthotic type). Horn pseudocysts may sometimes occur.
- The mainstay treatments for DPN cover curettage, surgical excision, cryotherapy, laser procedure, and electrodesiccation.

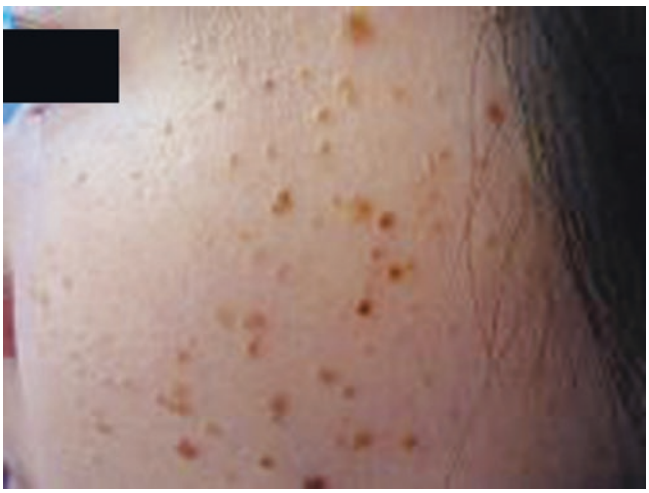


Fig. 9-15-1 Diffuse symmetric distribution of flat, round papules with papillomatous tops in sizes of 1–5 mm in diameter

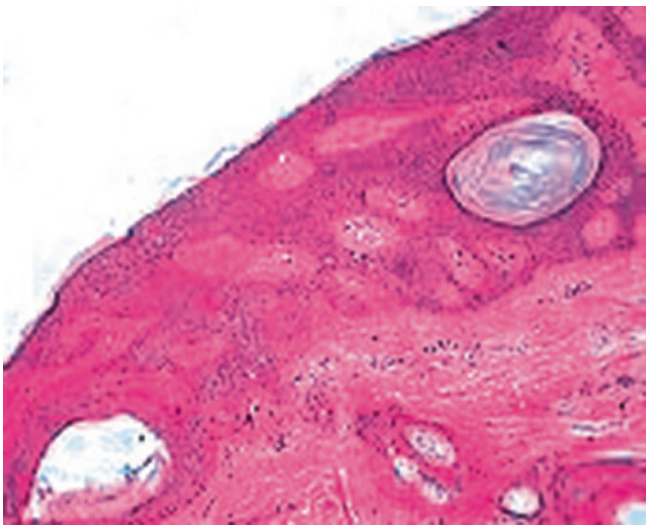


Fig. 9-15-2 Hyperkeratosis, mild acanthosis, and normal granular layer; elongation of rete ridge forming net-like structure; two keratin cysts in the spinous layer and increased melanin granules in the basal layer; a few of lymphocytes around the small vessels in the super dermis (HE stain, ×40)

9.16 Lichen Sclerosus [18]

- The terms lichen sclerosus et atrophicus, kraurosis vulvae, and balanitis xerotica obliterans are replaced by the single term lichen sclerosus, which can present from childhood to old age and occurs most commonly in females.
- In females, white, polygonal, flat-topped papules, plaques, or atrophic patches with severe itching may be seen in the vulvar and perianal areas. In males, the white atrophic macules may present on the glans penis, penile shaft, perianal area, and scrotum.
- Phimosis and paraphimosis are common complications of lichen sclerosus. Extragenital lesions preferentially occur on the upper back, chest, and breasts. Between 44% and 55% of cases of penile SCC are associated with lichen sclerosis.
- The histopathology shows atrophy of the epidermal layer with hydropic degeneration of basal cells and edema, accompanied by homogenization of the collagen in the dermis.

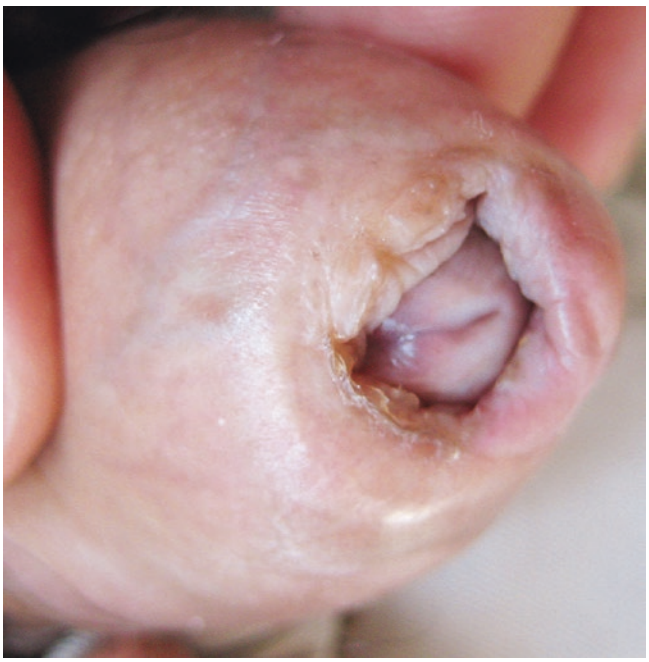


Fig. 9-16a-1 White and sclerosus plaques involved the penile shaft and the glans penis

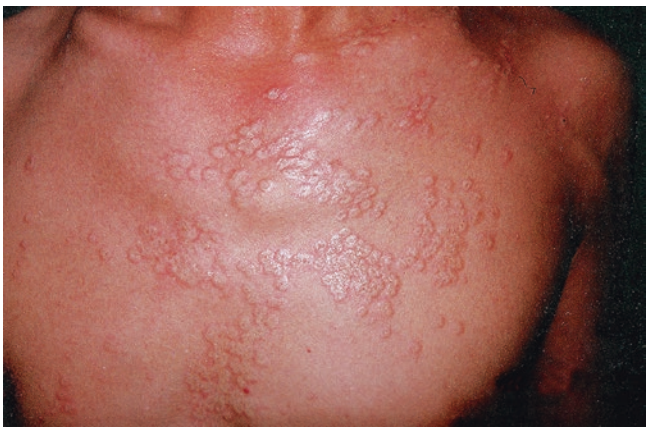


Fig. 9-16b-1 White flat-topped, discoid papules or plaques covered with horny plugs and surrounded by an erythematous halo on the chest

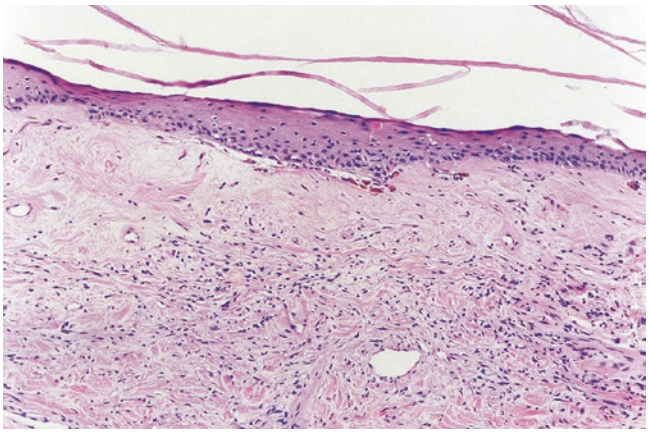


Fig. 9-16b-2 Hyperkeratosis with follicular plugging, atrophy of the stratum malpighii with hydropic degeneration of basal cells, pronounced edema and homogenization of the collagen in the upper dermis, and an inflammatory infiltrate in the middermis (HE stain, ×20)

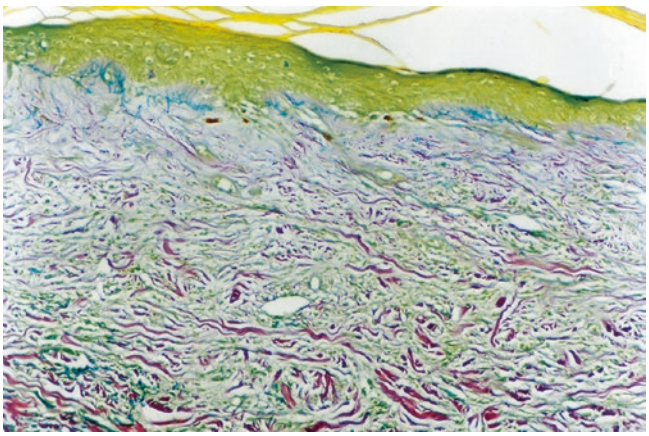


Fig. 9-16b-3 The diminution of elastic fibers sparse in the upper dermis (Verhoeff stain, ×20)

9.17 Erythromelanosis Follicularis Faciei Et Colli [19]

- Erythromelanosis follicularis faciei et colli (EFFC) is considered to be a relatively infrequent cutaneous disorder. Clinically, erythema, follicular papules, and light brown pigmentation are easily observed. The lesions preferentially present on the preauricular and maxillary region.
- EFFC occurs mostly in men during adolescence and occasionally in women. Keratosis pilaris occurring on the arms and shoulders is frequently found.
- No treatment has proven satisfactory for EFFC. Various options have been attempted, but the lesions recur after discontinuing treatment.

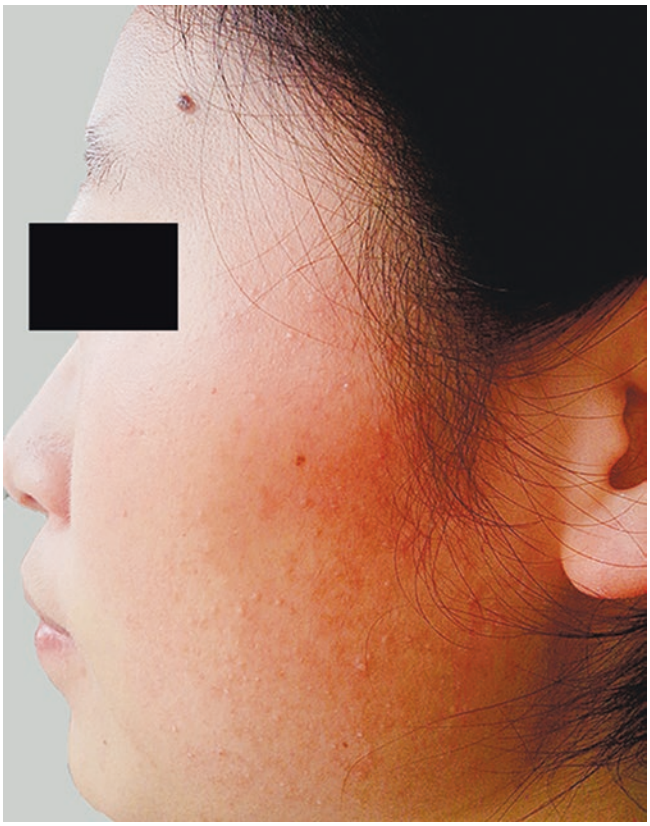


Fig. 9-17-1 Follicular papules and reddish-brown spots or patches on the cheeks and temples

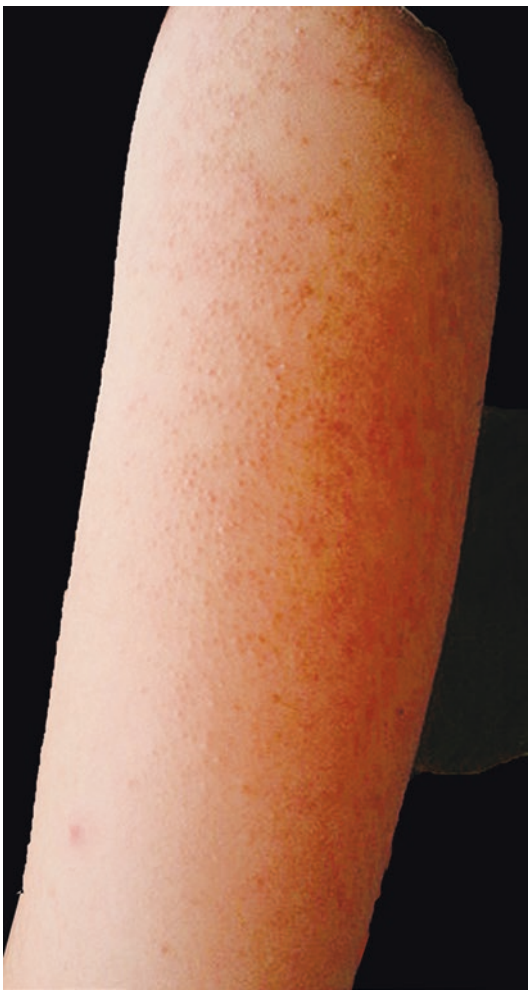


Fig. 9-17-2 Follicular keratosis papules on the extensor aspect of the arms. Her mother had similar condition

9.18 Lichen Aureus [20, 21]

- Lichen aureus (also called "lichen purpuricus") is an uncommon subtype of pigmented purpuric dermatosis with a chronic and benign course.
- Clinically, lichen aureus usually shows asymptomatic, unilateral, and solitary plaques, which are more frequently located on the lower extremities. Skin lesions vary in color, ranging from dark brown to copper and a golden hue.
- The histopathological examination of lichen aureus suggests infiltration of lymphocytes and histiocytes and extravasation of erythrocytes and hemosiderin, with little or no epidermal alteration.
- Treatment is difficult. The therapeutic arsenal includes topical corticosteroids, calcineurin inhibitors, and PUVA.

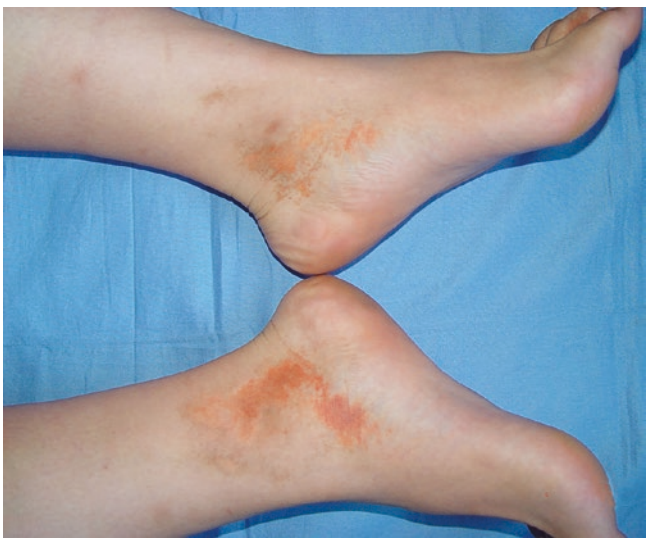


Fig. 9-18-1 Golden pigmented maculae presented as "cayenne pepper" on the dorsum of both feet. The skin lesions are confluent on the inner malleolus

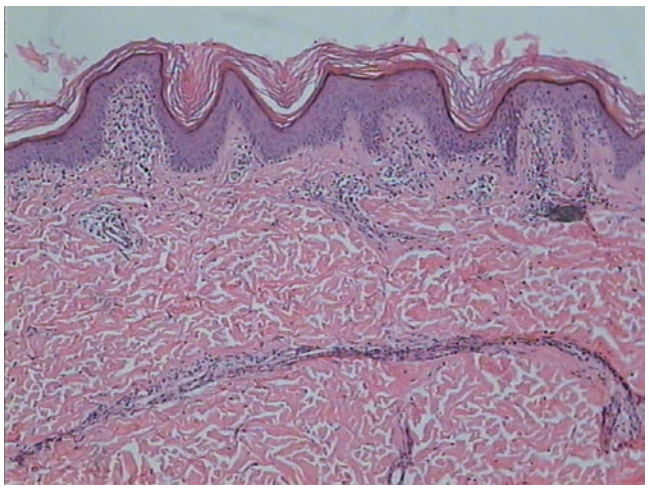


Fig. 9-18-2 A band-like infiltration of mononuclear and histiocytic cells in the upper dermis (HE stain, ×100)

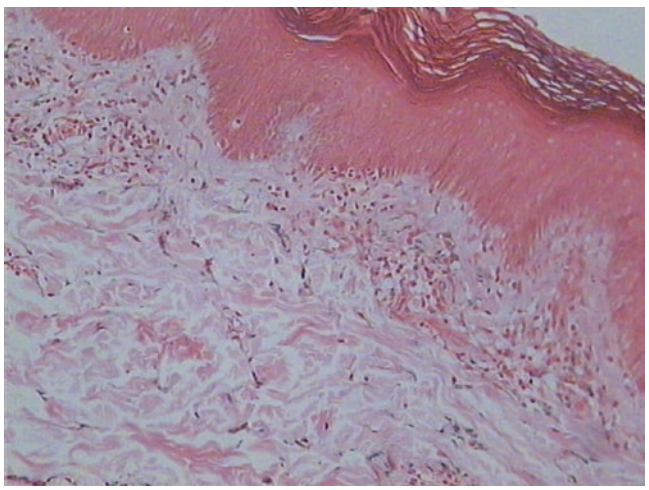


Fig. 9-18-3 Perls staining showed the presence of massive hemosiderin in the upper dermis (HE stain, ×200)

9.19 Generalized Lichen Nitidus [22, 23]

- Lichen nitidus (LN) is a rare chronic inflammatory dermatosis. Small (1–2 mm) and skin-colored or erythematous papules may be observed in many cases. Children and young adult are among the most vulnerable. Commonly, it occurs on the penis, abdomen, genital region, and extremities.
- LN can be segmented into seven types, including vesicular, hemorrhagic, follicular, linear, actinic, and generalized variants. Generalized LN is extremely unusual and can be associated with Down syndrome and multiple endocrine neoplasia (MEN).
- Local steroids, antagonists, antituberculous agents, narrowband UVB phototherapy, acitretin, and low-dose cyclosporine may also play a role.

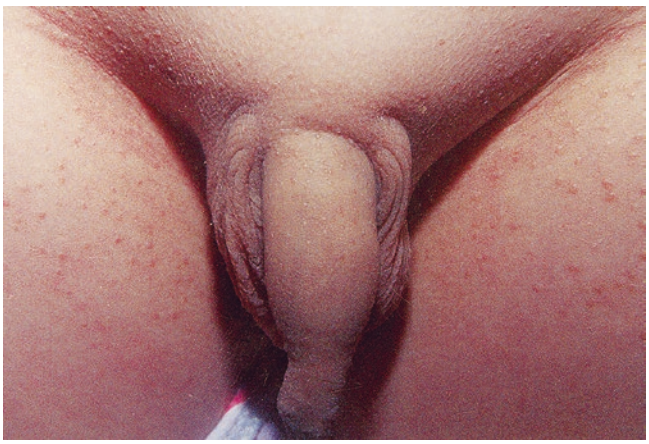


Fig. 9-19-1 Numerous flat-topped, skin-colored, round or polygonal papules without coalescence on the penis and foreskin

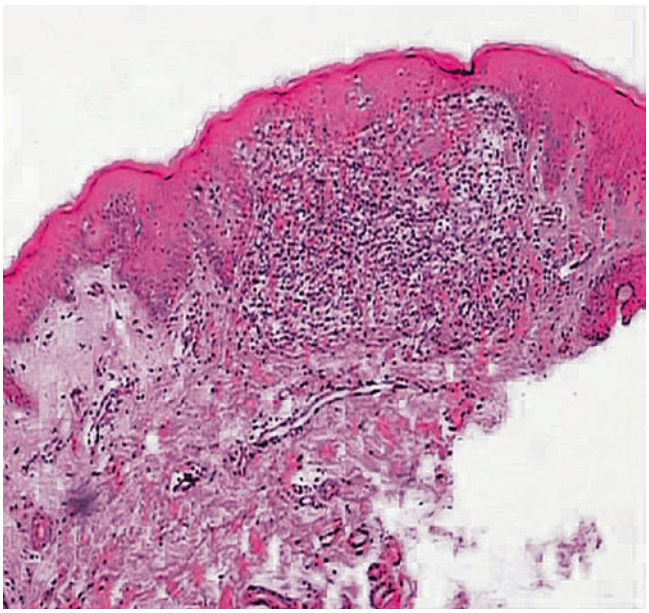


Fig. 9-19-2 Circumscribed nest of cells in the dermis papillary; the infiltrate consisted of lymphocytes, epithelioid cells, histiocytes, and a few multinucleated giant cells (HE stain, ×100)

9.20 Pityriasis Rotunda [24]

- Pityriasis rotunda (PR), also called "pityriasis circinata" and "tinea circinata," is a relatively rare disorder. Typical features are scaly, circular, well-demarcated, hypo- or hyperpigmented fine plaques over the trunk and extremities.
- PR may be idiopathic or related to infections or malignancy. The number may range from 1 to rarely greater than 100, with a typical diameter of 2–3 cm that may, in some cases, exceed 20 cm.
- Histopathological examination of PR shows hyperkeratosis, an absent granular layer, a pigmented basal layer, pigmentary incontinence, and a perivascular lymphocytic infiltrate.
- Conventional topical therapies have shown no benefit. Lactic acid lotion and oral vitamins may improve the lesions. In some cases, successful treatment of the underlying disease leads to clearance of the lesions.

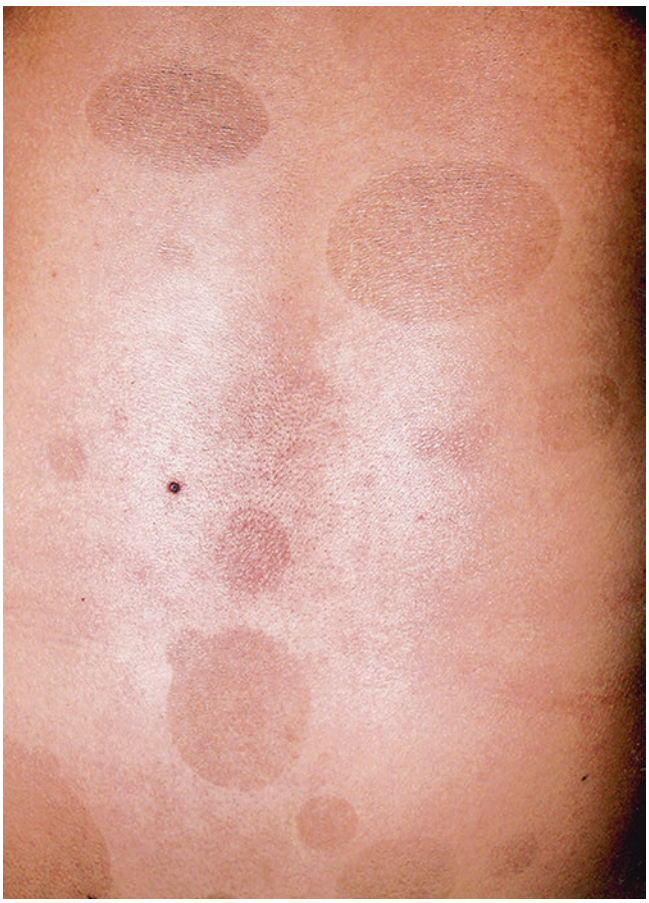


Fig. 9-20-1 Multiple, strikingly circular, or oval scaly patches on the trunk

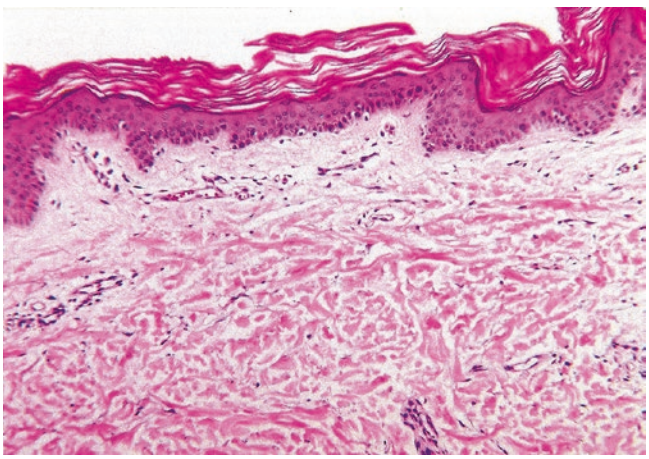


Fig. 9-20-2 Hyperkeratosis and infiltrate of a few lymphocytes and histiocytes around blood vessels in the dermis (HE stain, ×100)

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