Rheumatologic Diseases

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Abstract

Rheumatologic diseases have a diversity of cutaneous and systemic manifestations. This chapter presents with annular erythematous type of subacute cutaneous lupus erythematosus, bullous systemic lupus erythematosus, lupus erythematosus tumidus, neonatal lupus erythematosus, antiphospholipid syndrome, nodular scleroderma, atrophoderma of Pasini and Pierini, acrodermatitis chronica atrophicans, linear atrophoderma of Moulin, rheumatoid neutrophilic dermatitis, and graft-versus-host disease.

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10.1 Annular Erythematous Type of Subacute Cutaneous Lupus Erythematosus [1]

- Subacute cutaneous lupus erythematosus (SCLE) comprises approximately 10% of LE and exhibits either non-scarring papulosquamous (two-thirds) or annular polycyclic (one-third) lesions, with positive circulating SSA/anti-Ro antibodies.
- Certain genetic backgrounds favor development, with ultraviolet light and drugs, as potentially the most important triggers. Histopathological examination reveals interface dermatitis with vacuolar degeneration of basal keratinocytes and dermal mucinosis.
- The treatments are variable, including topical sunscreens, tacrolimus, pimecrolimus, moderate potency steroids, oral antimalarials, and glucocorticoids.

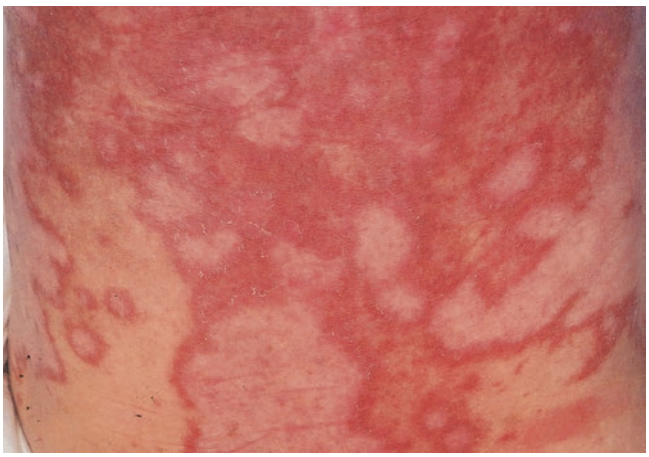


Fig. 10-1-1 Annular and polycyclic erythema with thin scales and elevated edges on the back

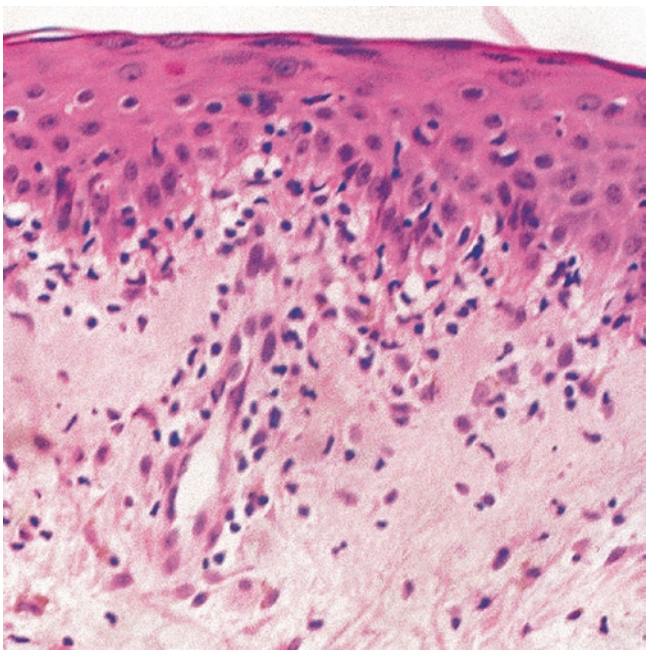


Fig. 10-1-2 The epidermis is atrophic. There is liquefaction degeneration of the basal cells and lymphocytic and eosinophilic perivascular infiltrations in the upper dermis (HE stain, ×400)

10.2 Bullous Systemic Lupus Erythematosus [2]

- Bullous systemic lupus erythematosus (BSLE) is an acquired autoimmune dermatosis that has a preference for patients with SLE. It typically presents with multiple, tense, and clear fluid-filled vesicles and bullae overlying erythematous plaques.
- Although the relationship between bullous eruptions and symptom flares in SLE has not been determined, BSLE may lead to a risk for developing lupus nephritis and may be related to a worse prognosis and refractory disease.
- The autoimmune characteristics of bullous lupus erythematosus are manifested as the presence of circulating anti-VII collagen antibodies. Histopathological examination reveals subepidermal blisters with a neutrophil-predominant infiltrate in the upper dermis.
- According to the findings during treatment, corticosteroids are ineffective, while dapsone generally improves the condition of the skin.

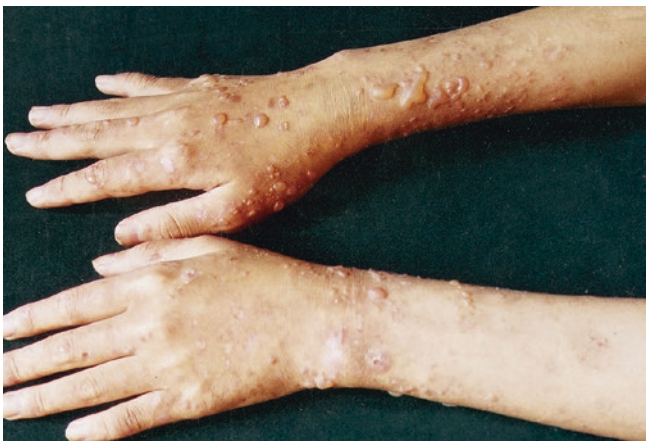


Fig. 10-2-1 Numerous blisters on the forearms and dorsum of both hands

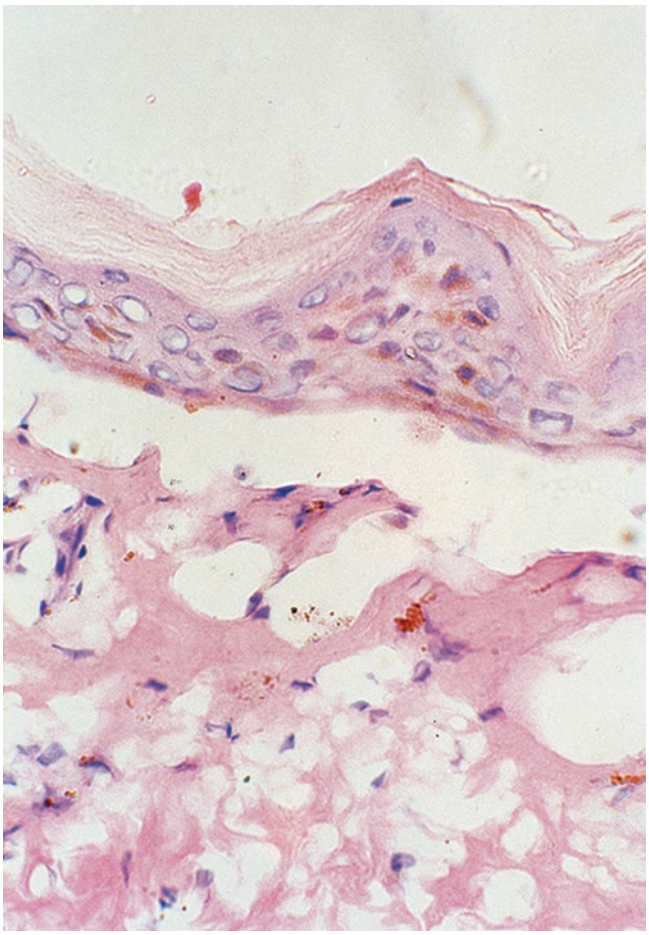


Fig. 10-2-2 A subepidermal bulla and liquefaction degeneration of basal cells (HE stain, ×400)

10.3 Lupus Erythematosus Tumidus [3, 4]

- Lupus erythematosus tumidus (LET) is an uncommon photosensitive dermatosis. However, there are no changes on the epidermal surface and scarring on resolution in most cases. The succulent, edematous, and non-scarring plaques preferentially occur on sun-exposed regions.
- Histologically, perivascular and periadnexal infiltration of lymphocytes and interstitial deposition of mucin are observed.
- Usually, LET is a benign disease. Photo-protective measures and antimalarials are often effective. In addition, corticosteroids, tacrolimus ointment, and methotrexate may play a role.

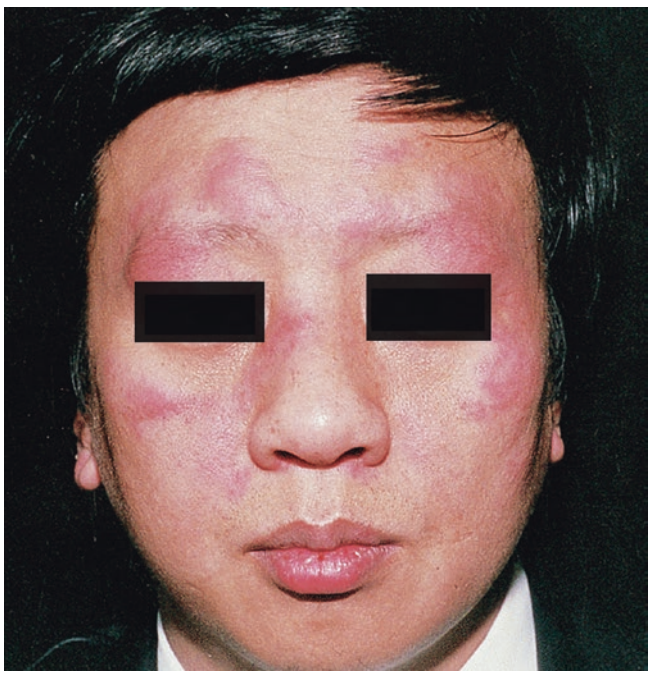


Fig. 10-3-1 Edematous erythema on the face (Reproduced with the permission from [3, 4])

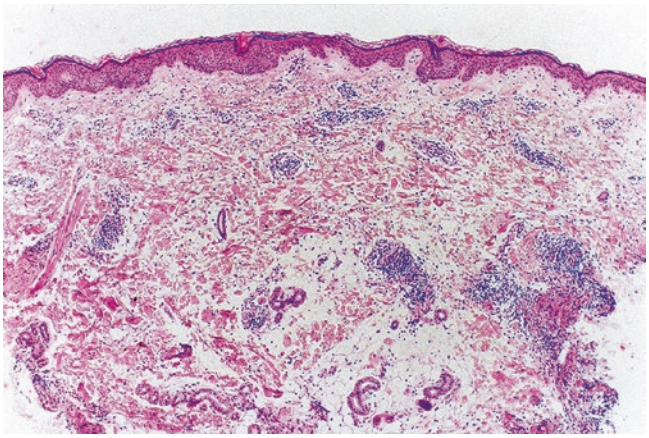


Fig. 10-3-2 Lymphohistiocytic infiltration in superficial and deep perivasculatures and around the appendages of the skin (HE stain, ×40)

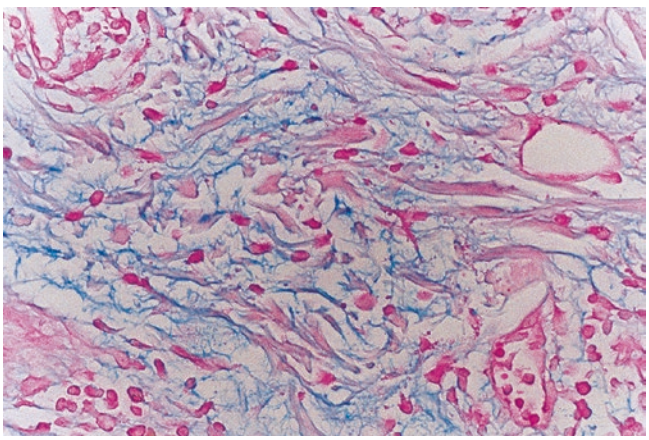


Fig. 10-3-3 Alcian blue stain showed abundant mucin deposition in collagen bundles of dermis (Alcian blue stain, ×100)

10.4 Neonatal Lupus Erythematosus [5]

- Neonatal lupus erythematosus (NLE) is a relatively uncommon autoimmune disease. The typical clinical features are cutaneous lesions, hematological or hepatic abnormalities, and congenital heart block.
- Generally, NLE is related to the transplacental passage of maternal IgG against Ro/SSA, La/SSB, and U1-RNP after 3 months of pregnancy.
- Typical lesions comprise erythematous, centrally atrophic plaques that are annular or polycyclic and preferentially affect the face and scalp. They usually begin in the first weeks of life and improve within 4–6 months.
- The diagnosis depends on the typical clinical features and the occurrence of autoantibodies in maternal or infant serum.

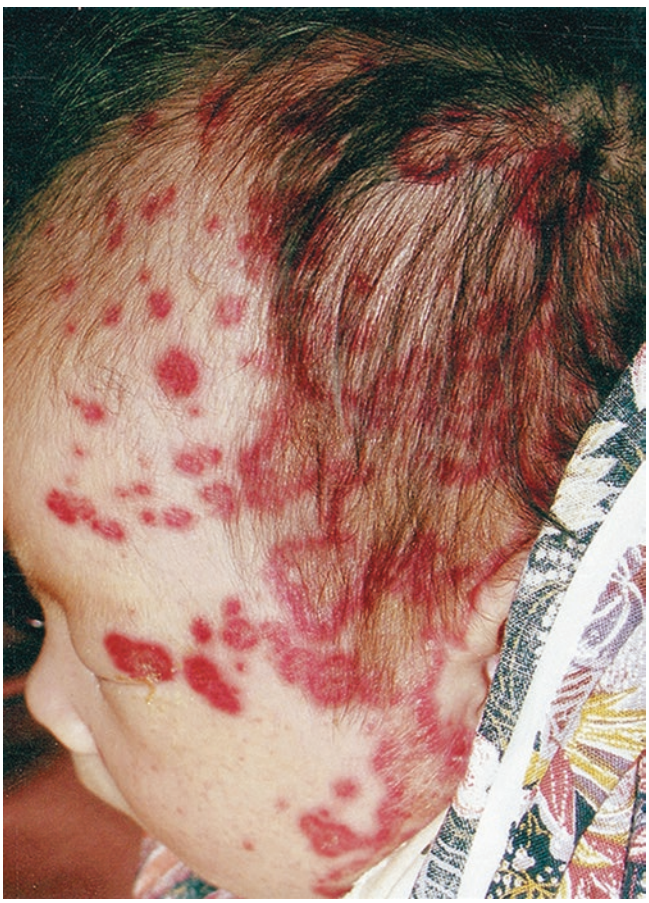


Fig. 10-4-1 Annular violaceous macules on the head and face

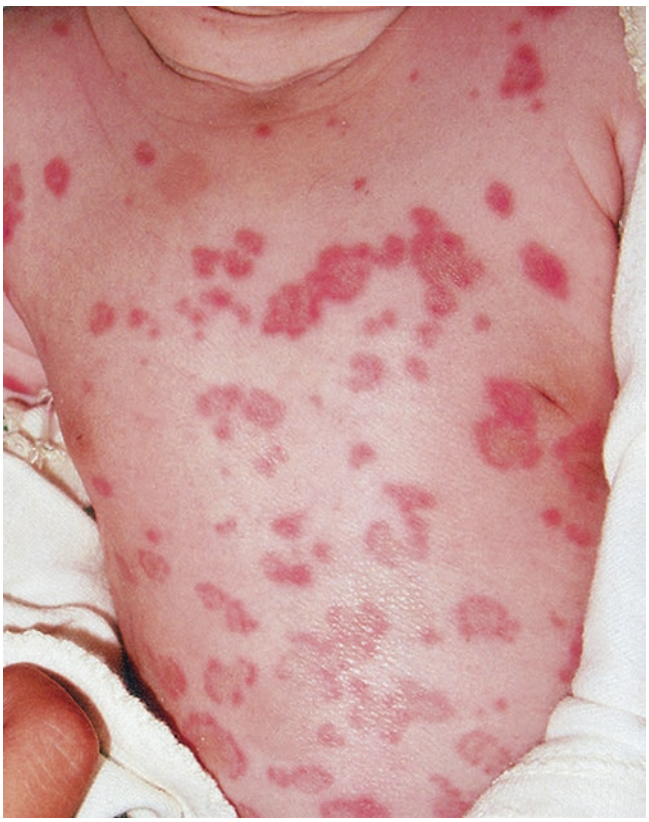


Fig. 10-4-2 Annular slight red macules on the chest and abdomen

10.5 Antiphospholipid Syndrome [6]

- Antiphospholipid syndrome, also called antiphospholipid antibody syndrome (APS or APLS), is an autoimmune disease presenting with characteristic antiphospholipid antibodies. The blood of the patient often shows a hypercoagulable state.
- APS may result in the formation of blood clots (thrombosis) in arteries and veins as well as pregnancy-related complications, for instance, miscarriage, stillbirth, preterm delivery, and severe preeclampsia.
- To make an accurate diagnosis, typical clinical manifestations and the lupus anticoagulant or anti-β_2-glycoprotein-I are essential.
- Anticoagulant may decrease the risk of further episodes of thrombosis and improve the prognosis of pregnancy of patients with APS.

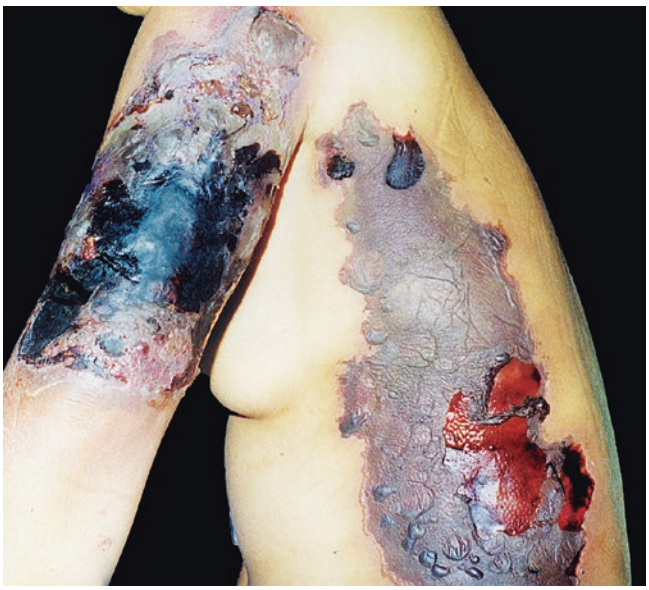


Fig. 10-5-1 Infiltrate erythema, dark crusts, and erosions on the extensor of the left upper arm and flank

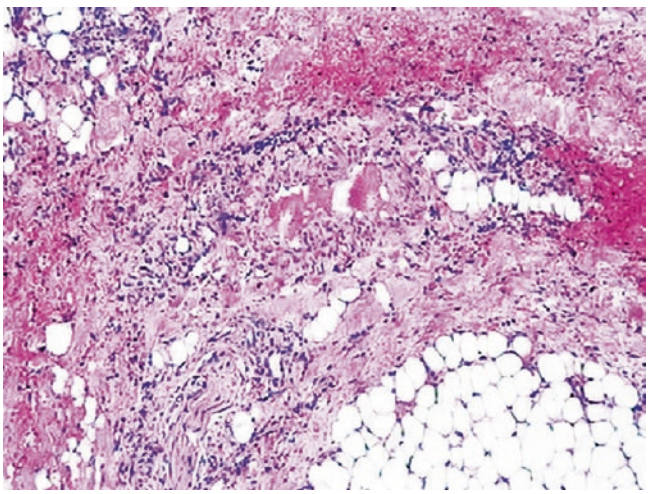


Fig. 10-5-2 Inflammatory infiltrate around small perivascular in subcutaneous and embolism in the small vascular (HE stain, ×100)

10.6 Nodular Scleroderma [7]

- Nodular scleroderma (NS) is a very unusual variant of scleroderma and preferentially occurs in middle-aged females.
- Typical manifestations comprise firm nodules or plaques resembling keloid, distributed predominantly on the proximal extremities.
- Steroids, topical calcipotriene, cyclosporine, D-penicillamine, methotrexate, photochemotherapy, and excision may improve symptoms of patients with NS.

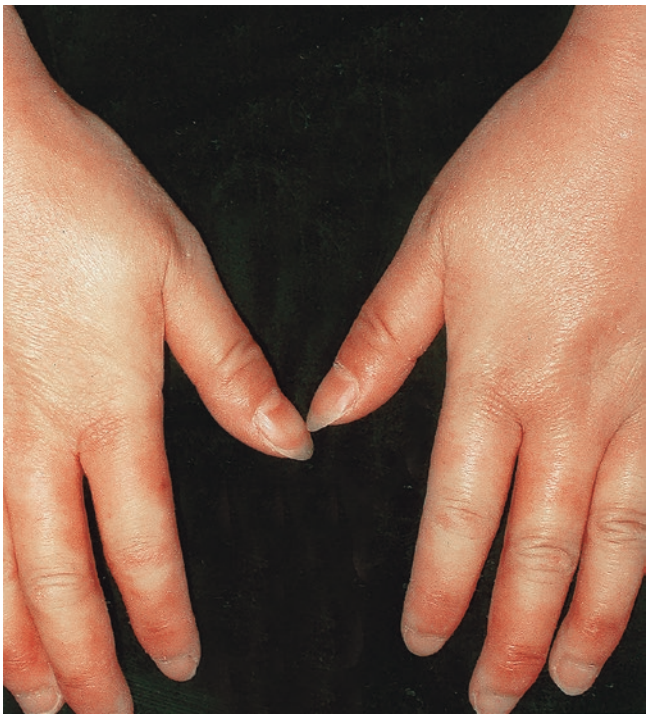


Fig. 10-6-1 Slight-red maculae on the fingers and dorsum of both hands

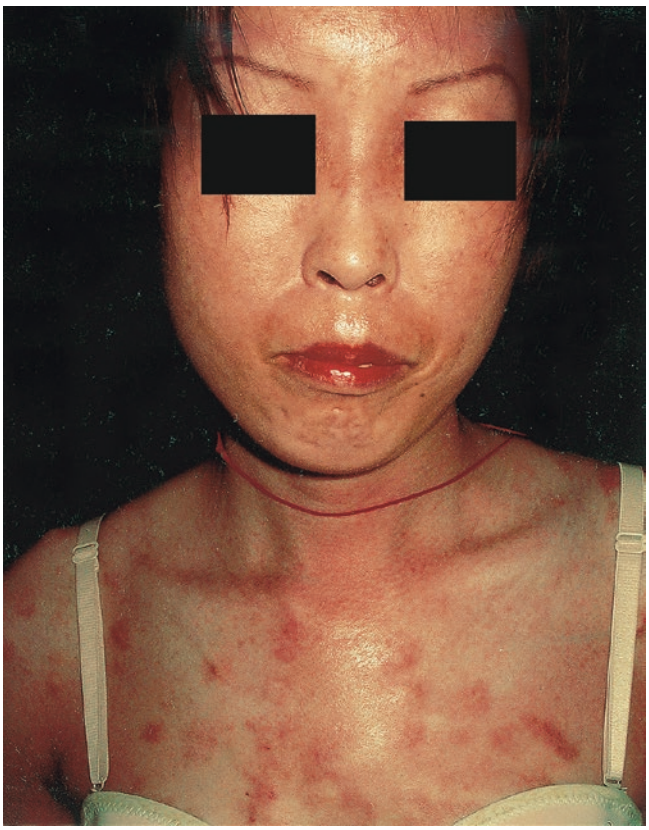


Fig. 10-6-2 Skin texture decreased on the forehead and plaques on the chest (Reproduced with the permission from [7])

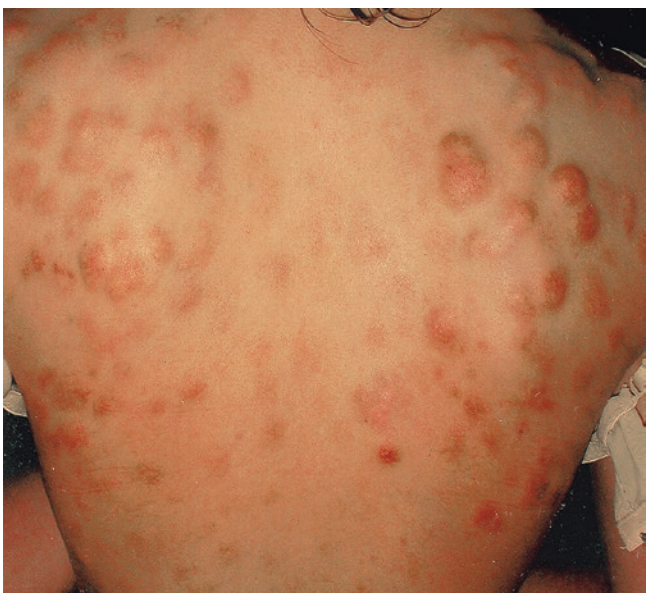


Fig. 10-6-3 Several slight red nodules on the shoulder and back

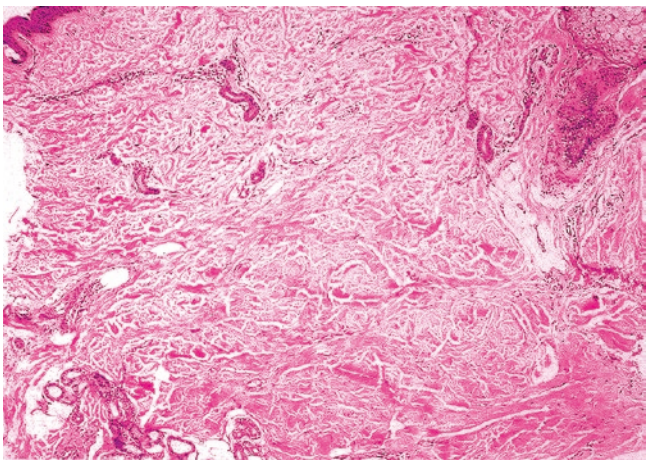


Fig. 10-6-4 Collagen fibers increased and packed in the dermis, a few blood vessels decreased and hair follicle atrophy (HE stain, ×40)

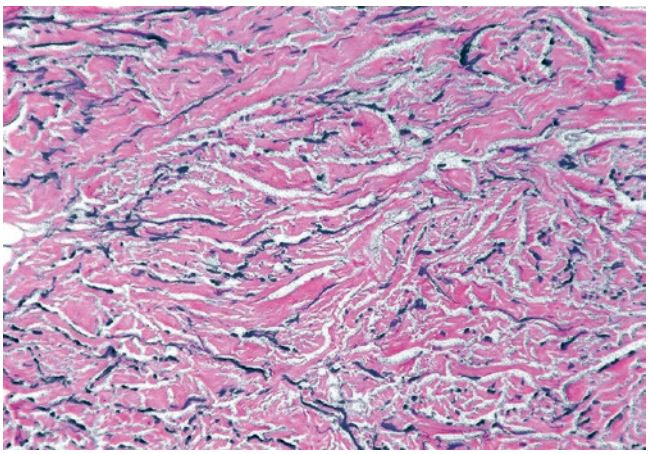


Fig. 10-6-5 Elastic fibers increased in the lower dermis (combined stain, ×100)

10.7 Atrophoderma of Pasini and Pierini [8, 9]

- Atrophoderma of Pasini and Pierini (APP) is an infrequent dermatologic condition. Its main feature is an asymptomatic, violaceous brownish discolored patch, generally showing one or more sharply demarcated depressed lesions. Clinically, the lesions most commonly occur in the lumbosacral region.
- Genetic factors, neurogenic causes, immunological factors and abnormal metabolism of dermatan sulfate may play roles in the pathogenesis of AP.
- Most patients with APP show a bilateral symmetric distribution. Occasionally, a segmental zosteriform distribution may be observed in a small subset of patients. APP could be associated with crossed total hemiatrophy, which implies facial atrophy and contralateral atrophy of the trunk and extremities.
- Considering the possibility of an underlying borrelial infection, it has been suggested that early cases of APP be treated with a course of appropriate oral antibiotics. Because the condition is asymptomatic and limited to the skin, most patients do not have to accept any treatment.

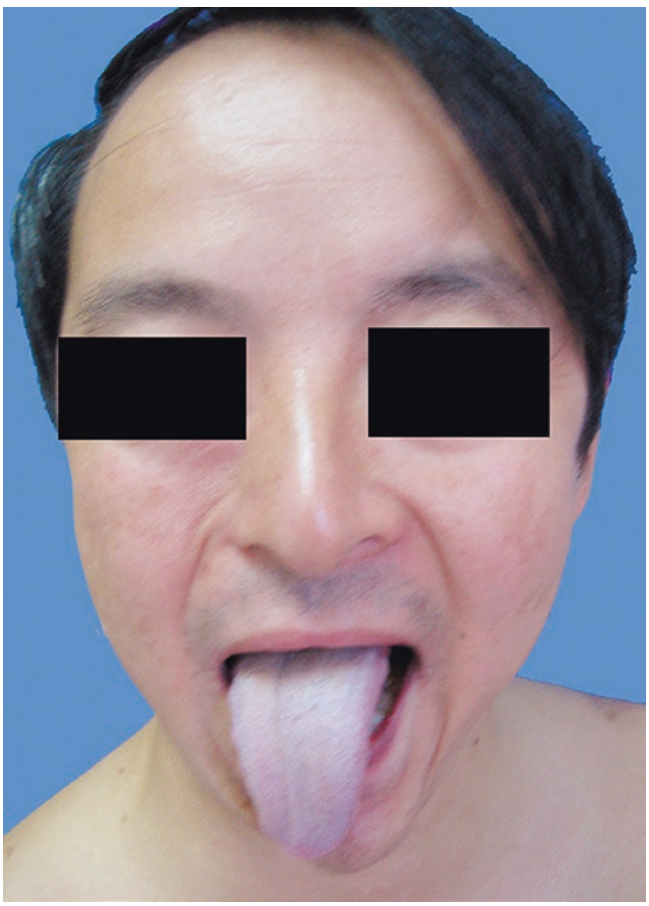


Fig. 10-7-1 Significant atrophy in right side of the tongue (Reproduced with the permission from [8, 9])

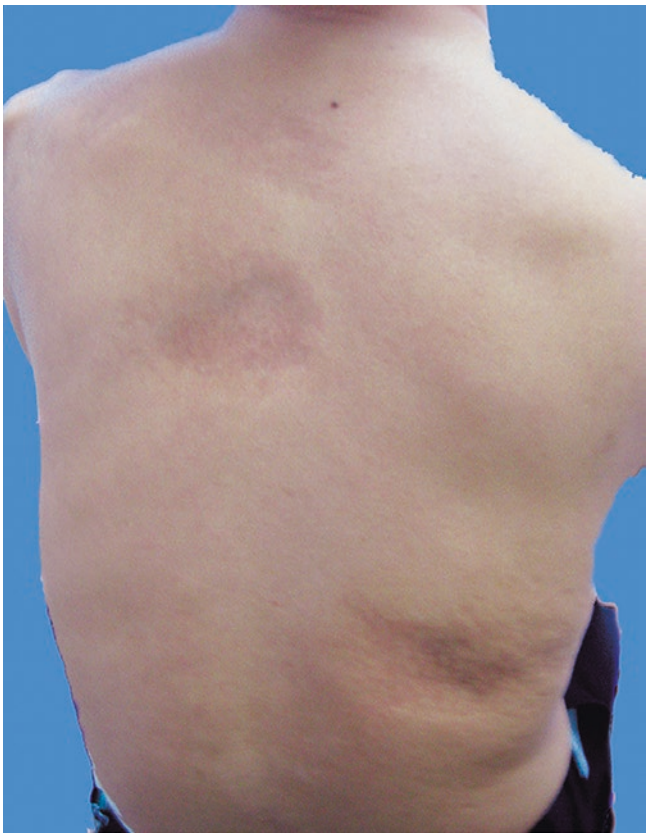


Fig. 10-7-2 A few brownish patches appeared on his back and lumbar part

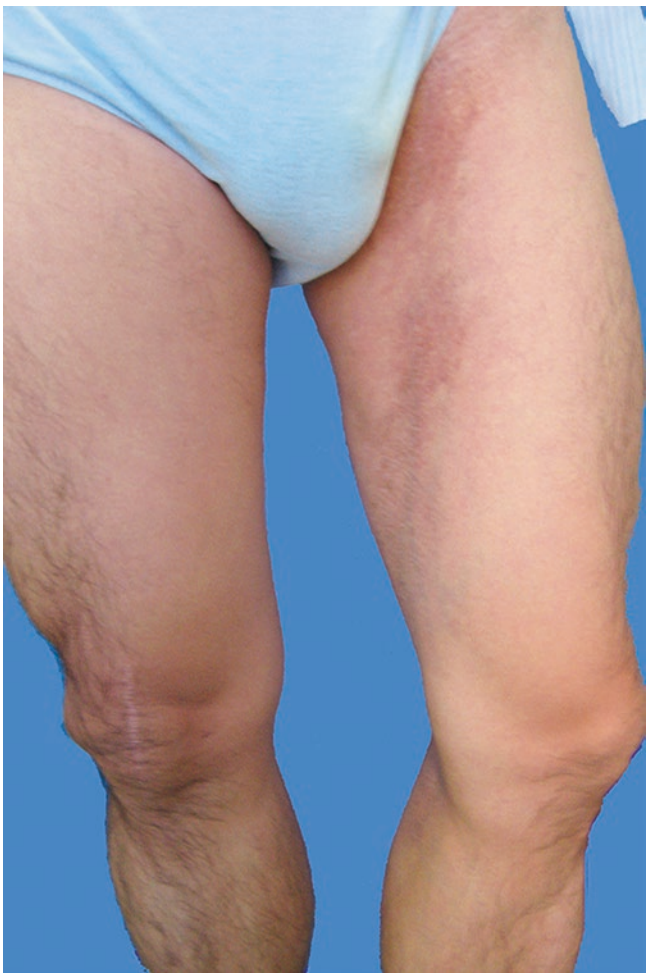


Fig. 10-7-3 A few brownish patches appeared on medial region of the left thigh. In the centers of these patches, variciform superficial veins could be seen clearly without of redness or induration

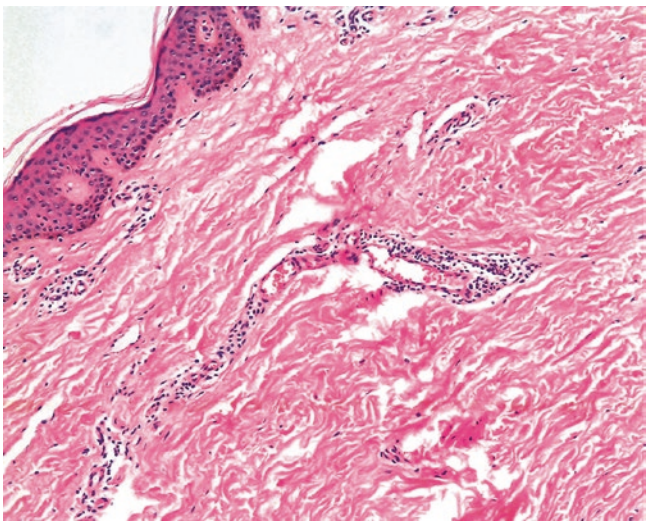
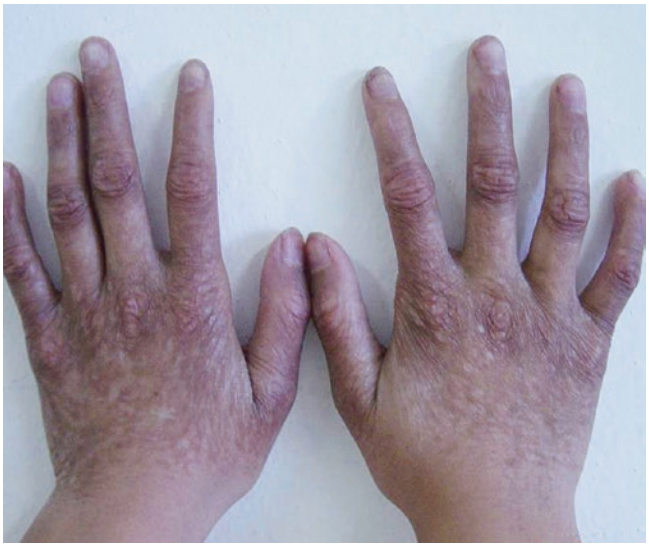
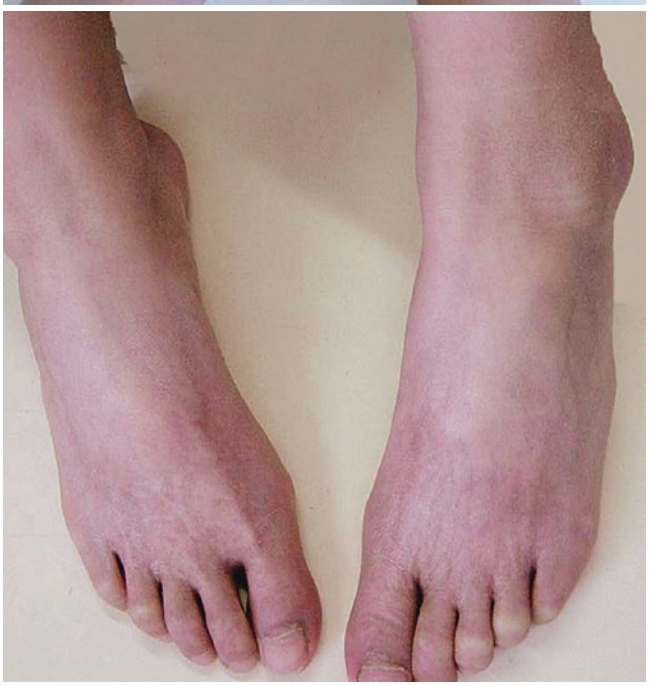


Fig. 10-7-4 Normal epidermis with increased pigment granules in the basal layer, perivascular mononuclear infiltration of the atrophic dermis, and subcutaneous adipose tissue moving up (HE stain, ×100)

10.8 Acrodermatitis Chronica Atrophicans [10]

- Acrodermatitis chronica atrophicans (ACA) is an uncommon late manifestation of tick-borne *Borrelia burgdorferi* infection, with inflammatory and atrophic lesions on acral skin.
- A correct diagnosis may depend on the characteristic clinical manifestations, histologic findings, and serum IgG antibody against borrelial antigens.
- The course of disease includes two stages: the inflammatory stage with bluish red discoloration and cutaneous swelling, and the atrophic phase.
- Doxycycline and penicillin may be effective for the acute case.





Figs. 10-8-1, 10-8-2 Brown plaques with keratotic and slightly atrophic appearance distributed on the back of the hands (1), fingers, and feet (2), especially on the joints

10.9 Linear Atrophoderma of Moulin [11]

- Linear atrophoderma of Moulin (LAM) is an acquired cutaneous disorder. Children and adolescents are mainly involved.
- Clinically, it is characterized by atrophic pigmentation spots along the unilateral Blaschko line of the body without special uncomfortable symptoms, without a preceding inflammation, and with subsequent induration or scleroderma.
- Histopathology, hyperpigmentation of the basal epidermis and a normal dermis with unaltered connective tissue and elastic fibers can be observed.
- There is no normative treatment for LAM, the progress of which is slow and can be self-cured.

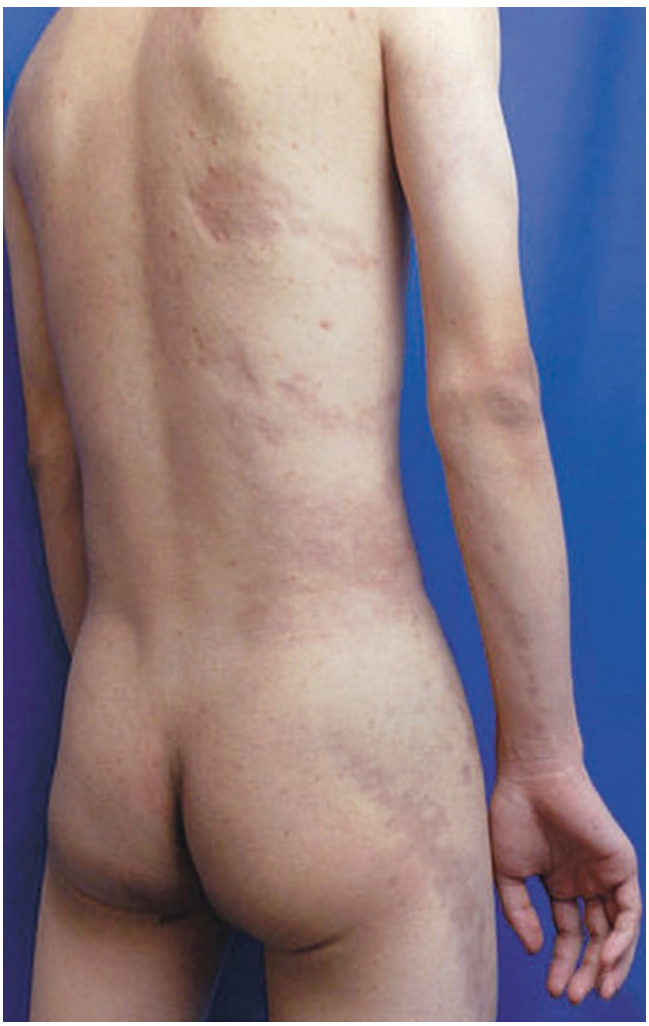


Fig. 10-9-1 Dark-brown atrophic patches along Blaschko's lines on the right backside, right forearm, and buttock

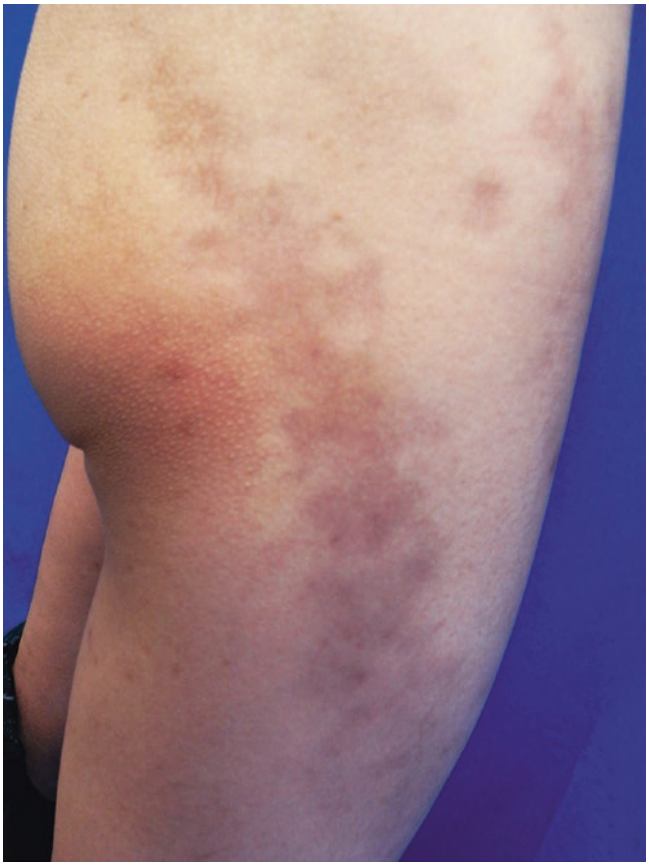


Fig. 10-9-2 Hyperpigmented atrophic patches without angiotelectasis on the right buttock

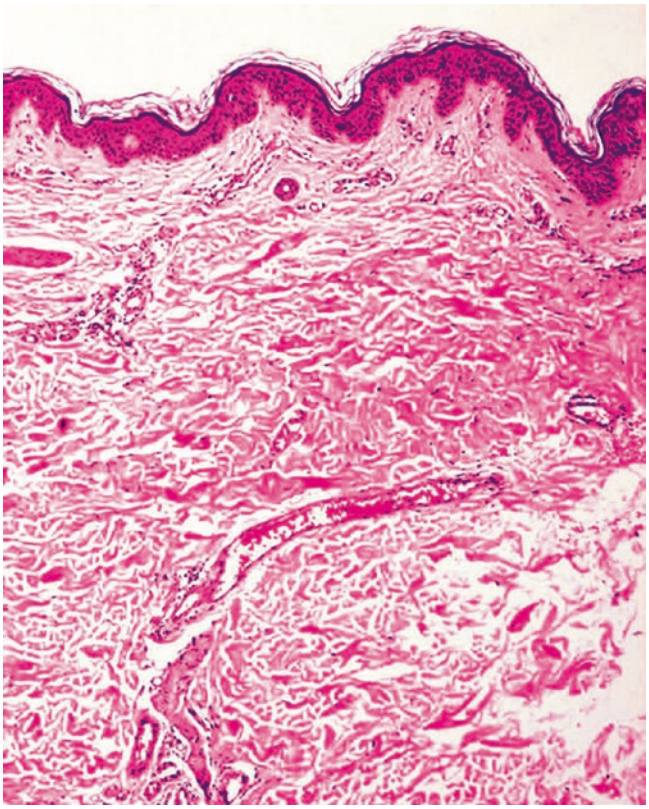


Fig. 10-9-3 Normal epidermis with increased melanin in the basal layer; telangiectasia and perivascular mononuclear cell infiltration in the upper dermis (HE stain, ×40)

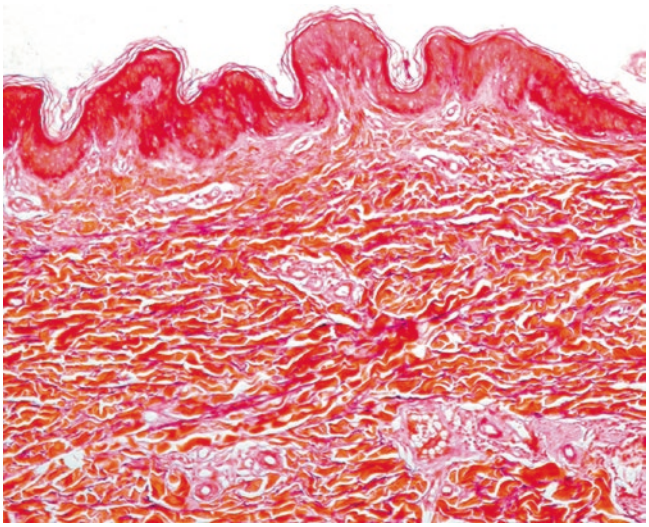


Fig. 10-9-4 Reduced elastic fibers without fragmentation in the upper dermis (Elastic fiber stain, ×100)

10.10 Rheumatoid Neutrophilic Dermatitis [12, 13]

- Rheumatoid neutrophilic dermatitis (RND) is a unique skin complication of severe seropositive RA.
- Clinically, it shows symmetrically distributed nodular erythema or plaque, mainly occurring on the joints and exterior surfaces of the extremities.
- Pathological changes in RND comprise heavy dermal infiltration of neutrophils without vasculitis. Microabscesses are infrequently observed in the dermal papillae.
- Although RND generally heals without scarring, residual atrophic scars have been documented.
- Other skin manifestations of RA include rheumatoid nodules, interstitial granulomatous dermatitis, vasculitis, pyoderma gangrenosum, urticarial, vitiligo, and neutrophilic lobular panniculitis.

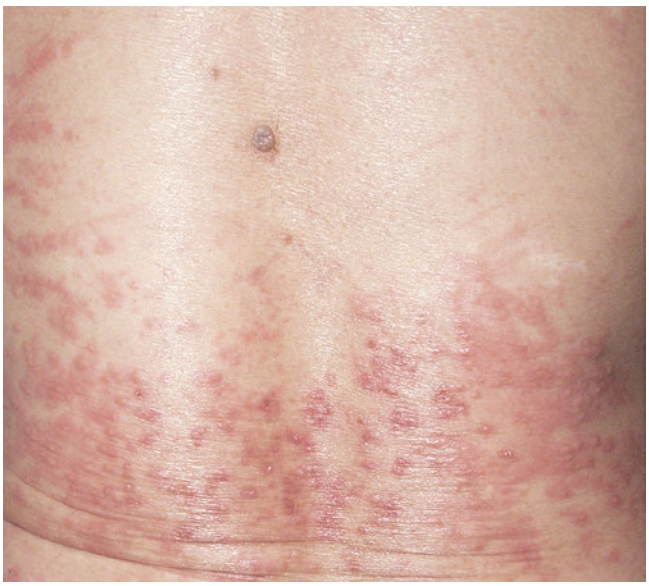


Fig. 10-10-1 On the waist and abdomen, there were densely distributed red- or skin-colored papules and nodules, ranging from 2 to 4 mm in diameter. Erosions, crusts, and ulceration were not discovered

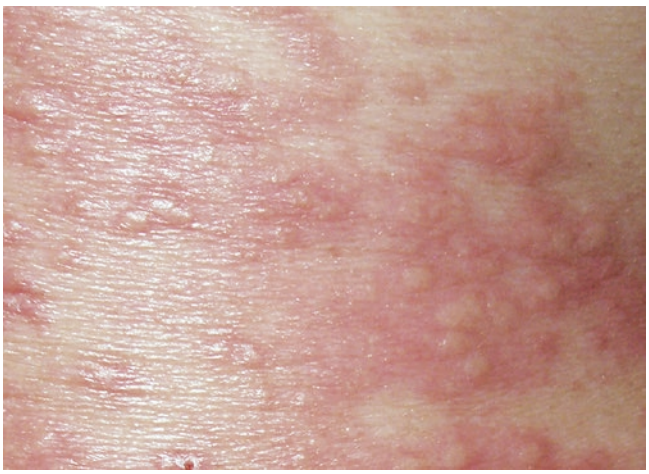


Fig. 10-10-2 Urticarial erythema or pseudo-vesicles upon amplification of the lesions

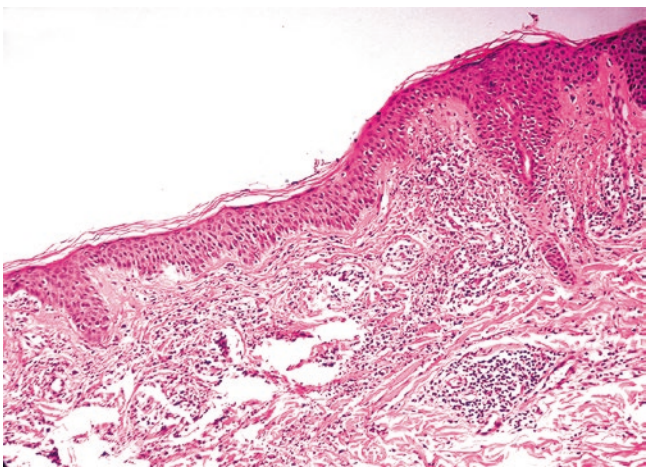


Fig. 10-10-3 Edema in the dermis, with neutrophilic cell infiltration in a band-like pattern (HE stain, ×100)

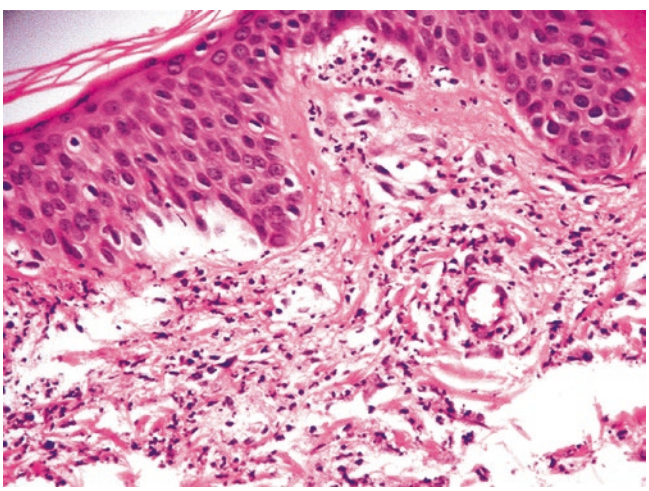
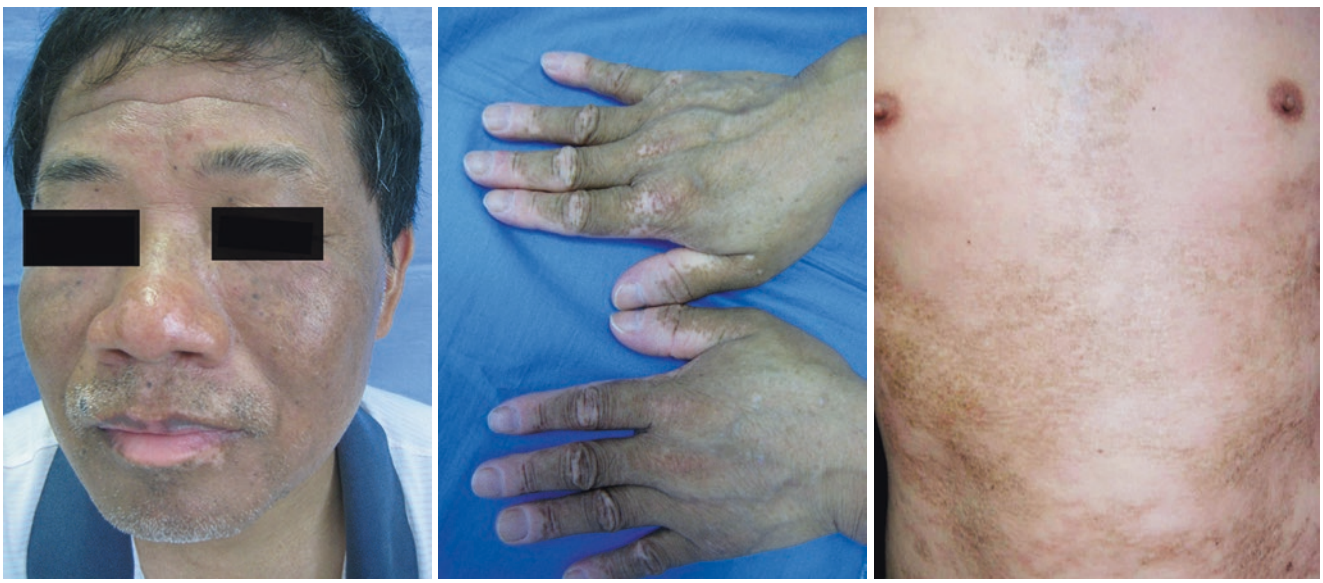


Fig. 10-10-4 Skin biopsy revealed leukocytoclasis without fibrinoid necrosis, thrombosis, or extravasation of erythrocytes There is collagen degeneration in foci, as well as neutrophilic microabscess (HE stain, ×400)

10.11 Graft-Versus-Host Disease [14]

- The cutaneous eruptions of graft-versus-host disease (GVHD) occur between the fourth and the fifth weeks after transplantation.
- GVHD may be divided into acute and chronic. Morbilli-like lesions, erythroderma, and conditions mimicking toxic epidermal necrolysis are mainly observed in acute patients. In chronic cases, lichen planus-like lesions, multiple sclerosis, hypo- or hyperpigmentation, atrophy, and alopecia are observed.
- In 30–40% of patients, chronic GVHD develops 3–5 months after grafting. The vast majority of GVHD patients present cutaneous involvement.



Figs. 10-11-1, 10-11-2, 10-11-3 Chronic GVHD with diffuse dyschromia and hyperpigmentation on the face (1), hands (2), and trunk (3) (Reproduced with the permission from [14])

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