B. Acquired keratoses

a. Inflammatory keratoses

1. Psoriasis (Fig. 15.21)

Outline

- It most frequently occurs in young and middle-aged men and women. Erythema and papules are accompanied by thick silvery-white scales. Inflammation in the epidermis and accelerated turnover of epidermal cells are found.
- It is classified by clinical features into five types: psoriasis vulgaris, guttate psoriasis, pustular psoriasis, psoriatic erythroderma and psoriatic arthritis.
- Auspitz phenomenon and Köbner phenomenon are the characteristic features.
- The pathological findings are thickening of the epidermis, hyperkeratosis associated with parakeratosis, vasodilation in the papillary dermis, and neutrophilic infiltration directly under the horny cell layer (Munro’s microabscess).
- The main treatments are topical vitamin D₃ ointments, topical steroids, PUVA therapies and narrow-band UVB. Immunosuppressants (methotrexate and cyclosporin) and retinoids are also useful, and biological therapies are also used for severe cases.

Classification

Psoriasis occurs in 1% to 2% of Caucasians and about 0.1% of ethnic Japanese. Men outnumber women by 2 to 1. Psoriasis is classified by symptoms into five types: psoriasis vulgaris (mainly keratotic erythema accompanied by scaling), guttate psoriasis (scattered small lesions with a diameter of 1 cm or less), pustular psoriasis (mainly pustular eruptions), psoriatic erythroderma and psoriatic arthritis (Table 15.5). Some cases change from one type to another. Psoriasis vulgaris accounts for an overwhelming majority of all cases.

Clinical features

The onset of psoriasis is mostly in adolescence through middle age. Remissions and exacerbations recur through life. Some cases have complete remission. Each type is described below.

Pathogenesis

The essential pathogenesis is unknown. The turnover from basal cell to horny cell to exfoliation, which normally takes 28 days, takes only 4 to 7 days, because of enhanced proliferation of epidermal cells. Psoriasis is strongly familial, especially in
Caucasians. For this reason, multiple genes are certainly involved. When one identical twin has psoriasis, the other is reported to have a 65% chance of also having the disorder. HLA-Cw6 is associated with the occurrence. Recent study suggests that dysfunction of T cells is the primary event and that dyskeratosis of the epidermis occurs secondarily. Additional inductive factors include irritants, injury, sunlight, infection (hemolytic streptococcus, in particular), and drugs (e.g., lithium, β blockers, calcium antagonists).

### Pathology

Inflammation occurs, most severely in the upper epidermal layer (Fig. 15.22). As the epidermal turnover is abnormally enhanced, epidermal cells forming the horny layer retain their nuclei (parakeratosis). Hyperkeratosis is present. Munro’s microabscesses caused by infiltration of neutrophils are found directly below the horny cell layer. Because epidermal cells move to the horny cell layer before they produce keratohyaline granules, the granular layer thins and disappears, resulting in thickening of the suprabasal cell layer. Because the epidermal rete ridges become club-shaped and extend toward the dermis, the dermis protrudes directly below the horny cell layer in some areas. In pustular psoriasis, multiple neutrophils infiltrate into the upper suprabasal cell layer, and epidermal cells are destroyed to form spongiosis called Kogoj’s spongiform pustule (Fig. 15.23).

### Laboratory findings

Köbner phenomenon and Auspitz phenomenon are present. Inflammatory findings such as elevated erythrocyte sedimentation rate, leukocytosis and hyperproteinemia may be caused in pustular psoriasis and psoriatic erythroderma. In psoriatic arthritis, rheumatoid factors are usually negative.
Psoriasis is diagnosed by the characteristic clinical findings; however, a biopsy may be conducted for differential diagnosis. In pustular psoriasis, pustules are sterile.

**Diseases to be differentiated from psoriasis**

See Table 15.6.

**Treatment**

The main treatments are topical active forms of vitamin D₃ ointments and steroids, PUVA therapies, and narrowband UVB therapies. In severe cases, retinoids or immunosuppressants (e.g., methotrexate, cyclosporine A) are orally administered. Oral steroids tend not to be used because they may induce pustular psoriasis. The combination of tar with UV light (Goeckerman regimen) is no longer widely used due to its oncogenicity. Biological therapy has come to be used.

**Types of psoriasis**

1) **Psoriasis vulgaris**

Rose pink papules appear and extend to coalesce gradually into sharply circumscribed erythematous plaques with thick silvery scales on the surface (Figs. 15.24-1 to 15.24-4). The eruptions are usually asymptomatic; however, itching is present in some cases. Areas that are subjected to external stimulation, such as the elbows, patellae, scalp and buttocks are most commonly involved. The disorder may also occur in the intertriginous areas of obese people.

2) **Guttate psoriasis**

Multiple keratotic erythema of up to 1 cm in diameter occurs on the trunk and proximal sides of the extremities with a relatively acute course (Figs. 15.25-1 and 15.25-2). Individual eruptions are the same as those of psoriasis vulgaris. It is often seen in children. Streptococcal infection or drugs can be the causative factors.

**Laboratory findings on psoriasis: Wax-fragment phenomenon, Auspitz phenomenon**

When psoriatic eruptions are scraped with a fingernail, white scales that resemble wax flakes are seen. With the peeling off of the scales, petechia is easily caused; this is called Auspitz phenomenon. It is caused by the intrusion of dermal papillae directly into the horny cell layer. Stimulation such as injury may induce eruptions at normal sites of skin in patients with psoriasis (Köbner phenomenon).
3) Pustular psoriasis

Pustules are the main clinical feature. The disorder is subdivided into a generalized type and a localized type (Table 15.7). In the generalized type, fever, systemic fatigue and bodily chills accompany erythema on which multiple sterile pustules occur and coalesce. The pustules rupture spontaneously to form erosions. Exudative fluid may cause hypoproteinemia, leading to marked systemic aggravation in some cases. It may occur in the course of psoriasis vulgaris, or it may develop suddenly without any history of psoriasis (Fig. 15.26).

4) Psoriatic erythroderma

Psoriatic skin lesions appear all over the body and become erythroderma. Proteins are consumed in large amounts in the lesions. The horny cell layer forms incompletely, bringing hypoproteinemia, dehydration and electrolyte abnormality.

5) Psoriatic arthritis

Arthritis symptoms may accompany psoriasis. The majority of cases are the peripheral type, which affects distal interphalangeal (DIP) joints. There is a type in which vertebra and sacroilitis are involved. Arthritis proceeds without psoriatic skin lesions in many cases. There is association with the HLA-Cw6 gene.

Table 15.6 Differential diagnosis of psoriasis.

<table>
<thead>
<tr>
<th>Disease</th>
<th>Differential points</th>
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<tbody>
<tr>
<td>Seborrheic dermatitis</td>
<td>The clinical findings resemble those of psoriasis, but the affected sites are relatively localized at seborrheic areas.</td>
</tr>
<tr>
<td>Chronic eczema</td>
<td>Various, localized skin lesions including erythema, scales, papules, and blisters. Intense itching. The lesions are less clearly margined than in psoriasis.</td>
</tr>
<tr>
<td>Parapsoriasis</td>
<td>Pigmentation and atrophy are often present. Histopathological differentiation may be necessary.</td>
</tr>
<tr>
<td>Pityriasis rosea (Gibert)</td>
<td>Psoriasis-like lesion appears after manifestation of the first eruption, and disappears in 1 or 2 months.</td>
</tr>
<tr>
<td>Mycosis fungoides</td>
<td>Clinical findings may resemble those of psoriasis. Histopathological infiltration of atypical lymphocytes to the epidermis (Pautrier's microabscess).</td>
</tr>
<tr>
<td>Syphilitic psoriasis</td>
<td>Psoriasis-like eruptions on the palms and soles. History-taking and serologic test for syphilis are important.</td>
</tr>
<tr>
<td>Drug eruption</td>
<td>History-taking on drugs and tolerance test are conducted.</td>
</tr>
<tr>
<td>Ankylosing spondylitis</td>
<td>Psoriasis-like eruption in some cases; differentiation from psoriatic arthritis is important.</td>
</tr>
</tbody>
</table>

Clinical images are available in hardcopy only.