Psoriasis

Psoriasis may be divided into psoriasis vulgaris, generalized pustular psoriasis, and localized pustular psoriasis.

*Psoriasis Vulgaris*    *Clinical Features*
Psoriasis vulgaris is a common chronic inflammatory skin disorder that affects approximately 1.5% to 2%
associated with acute group A ~-hemolytic streptococcal infections. Involvement of the nails is common...
Spongiform pustules of Kagoj in Psoriasis =
Psoriatic arthritis characteristically involves the terminal interphalangeal joints, but frequently the large joints...
This cutaneous eruption is characterized by the presence of variable numbers of sterile pustules appearing in a spongiform pattern, most frequently in plaques. Several exacerbations may occur, and lesions of ordinary psoriasis may be seen in the intervals between them.

The four variants of generalized pustular psoriasis show considerable resemblance and overlapping in their clinical picture and also have a similar histologic appearance. They differ mainly in the mode of onset and the distribution of the lesions. Frequently, all four diseases show oral pustules, particularly on the tongue.

Acute generalized pustular psoriasis of von Zumbusch is generally diagnosed when the eruption occurs in patients with preexisting psoriasis, either of the plaque type or of the erythrodermic type. Frequently, the eruption occurs after systemic steroid therapy withdrawal. The exanthematous type of generalized pustular psoriasis refers to a group of patients with later onset of psoriasis, atypical distribution of the lesions, and a rapid and apparently spontaneous pustular eruption.
Generalized pustular psoriasis of pregnancy is a rare pustular eruption that appears during the last trimester of pregnancy. It starts with flexural lesions of ... may occur repeatedly during successive pregnancies. Some authors have considered it to be the same disease as impetigo herpetiformis, but others claim that they stand as separate entities.

In some instances of subacute annular pustular psoriasis or gyrate lesions show a clinical resemblance ...
Very rarely, children develop generalized pustular psoriasis, also known as infantile and juvenile pustular psoriasis. This disease has a benign course with frequent spontaneous remissions.

**Localized Pustular Psoriasis**

**Clinical Features**
There are three types of localized pustular psoriasis: (a) "psoriasis with pustules", in which only one or a few areas of skin develop pustules; (b) "psoriasis with pustules and plaques", in which pustules develop on pre-existing plaques; and (c) "psoriasis with pustules and plaques with pustules", in which pustules develop on plaques that subsequently become pustules themselves.
Acrodermatitis continua of Hallopeau is the term used when the pustular eruption involves the distal portions of the hands and feet. In the localized type of Acrodermatitis continua of Hallopeau, only the fingers or toes are affected. Atrophy of the skin and permanent nail loss may occur on the fingers and toes.

Pustulosis palmaris et plantaris is a chronic, relapsing disorder occurring on the palms, soles, or both. Crops of small, deep-seated pustules are seen within areas of erythema and scaling. In the earliest stage, the lesions are often covered with a yellowish crust, which may become detached, leaving a raw surface. An acute variant called "pustular bacterid" describes a rare eruption of large and sterile pustules on hands and feet.
Psoriasis and Acquired Immunodeficiency Syndrome
The association between psoriasis and human immunodeficiency virus (HIV) infection is commonly seen. The prevalence of virus infection in patients with psoriasis may be increased (108). Clinically, psoriasis may have a more severe course with sudden exacerbations and may be refractory to treatment (109). Extensive erythrodermic psoriasis may occur, and a trend of low peripheral T-cell CD4+ (helper-inducer) counts and a more severe clinical course have been noted.
Psoriasis Vulgaris

Histopathology
The histologic picture of psoriasis vulgaris varies considerably with the stage of the lesion and usually is diagnostic only in early, scaling papules and near the margin of advancing plaques.

The earliest pinhead-sized macules or smooth-surfaced papules show subtle histologic changes with a preponderance of parakeratosis, spongiosis, and hyperkeratosis.
exocytosis of neutrophils, they may aggregate in the uppermost portion of the spinous layer to form small
In the fully developed lesions of psoriasis, as best seen at the margin of enlarging plaques, the histologic picture is characterized by the following features:

(a) Spongiform pustules of Kagoj
(b) Epidermal hyperplasia
(c) Keratinocyte vacuolization
(d) Hypergranulosis
(e) Munro microabscesses
(f) Vacuolar degeneration of the epidermis
(g) Elongation and edema of the dermal papillae
(h) Dilated and tortuous capillaries
Of the listed features, only the spongiform pustules of Kogoj and Munro microabscesses are truly diagnostic of psoriasis. These pustules are characterized by the presence of eosinophils and neutrophils within the lesions. The pustules are usually located in the epidermis and are often accompanied by a surrounding inflammatory infiltrate. The presence of these pustules is a key feature in the diagnosis of psoriasis, as they can rarely be made with certainty on a histologic basis. The changes in active psoriasis are discussed in detail later.

The rete ridges show considerable elongation and extend downward to a uniform level, resulting in regular acanthosis.
show thickening ("clubbing") in their lower portion. Not infrequently, adjacent rete ridges seem to coalesce...
The suprapapillary epidermis appears relatively thin in comparison with the markedly elongated rete ridges.
In some instances the cornified layer consists entirely of confluent parakeratosis forming a platelike scale...

Munro microabscesses are located within the parakeratotic areas of the cornified layer. They consist of...
The dermal papillae, in accordance with the elongation and basal thickening of the rete ridges, are elongated and club shaped. Lymphocytes are present in the papillae, except in early lesions, in which neutrophils are also present in the upper portion of the papillae.
An entirely typical histologic picture as described earlier is not always found, even if the biopsy specimens
indicate focal parakeratosis, or, occasionally, alternating layers of orthokeratosis and parakeratosis. The last-named pattern
indicates a fluctuation in the activity of the psoriasis.

The bleeding points that may be produced by gentle scraping of the skin (Au spitz sign) correspond to the
Guttate or eruptive psoriasis shows the histologic features of an early or active lesion of psoriasis, where there is more pronounced inflammatory infiltrate, with neutrophils or lymphocytes, underlying the parakeratotic scale and the cornified layer overlying the mounds of parakeratosis with neutrophils, which, in turn, may appear loosely arranged.

The histologic picture of erythrodermic psoriasis in some instances shows enough of the characteristics of psoriasis to allow this diagnosis. Frequently, however, the histologic appearance is indistinguishable from that of a chronic eczematous dermatitis.

Generalized Pustular Psoriasis
Histopathology
Whereas in ordinary psoriasis the spongiform pustule of Kogoj is a very small micropustule and is seen only in early stages. As these pustules move up into the cornified layer, they become pyknotic and assume the appearance of a large Munro abscess.
In addition to the large spongiform pustules, the epidermal changes in generalized pustular psoriasis are
In the healing stage, the lesions of all types of generalized pustular psoriasis may present the same histologic appearance as ordinary psoriasis.

*Localized Pustular Psoriasis*
Histopathology

In the variants of localized pustular psoriasis "psoriasis with pustules" and localized annular pustular psoriasis, the histologic picture is the same as that described for generalized pustular psoriasis.

In localized acrodermatitis continua of Hallopeau, the nail bed is mainly affected, showing marked epithelial hyperplasia and hyperkeratosis with mounds of parakeratosis with neutrophils. The nail matrix is only occasionally involved.
In pustulosis palmaris et plantaris there is a fully developed large intraepidermal unilocular pustule. It is elevated...
Very early lesions may show spongiosis and exocytosis of lymphocytes in the lower epidermis overlying...
Histopathology

The histologic picture in most cases is similar to that of psoriasis. In others, the histologic sections may show some plasma cells. As in other dermatitides related to AIDS, eosinophils may be present in the inflammatory infiltrate.
Pathogenesis of Psoriasis Vulgaris

Although the cause of psoriasis is unknown, there is increasing evidence of a complex interaction among...
Electron Microscopy
The earliest recognizable morphologic events in psoriasis have been investigated in lesions that cleared...
Spongiform pustules of Kagoj in Psoriasis
Ultrastructural studies of the spongiform pustule of Kogoj, one of the most characteristic histologic structures in psoriasis, show distinctive features. The spongiform pustule is characterized by a spongelike network composed of degenerated and flattened keratinocytes. The ultrastructure of the capillary loops in the dermal papillae shows them to be different from normal capillary loops, resulting from the deposition of amorphous substances and accumulation of collagen fibrils in the basement membrane zone.
Epidermal Cell Cycle Kinetics
The rate of epidermal cell replication is markedly accelerated in active lesions of psoriasis, as shown by
Early calculations made it appear likely that in psoriatic lesions there was a great acceleration of the transit time of the cell layer, from approximately 53 days in normal epidermis to only 7 days in the epidermis of active psoriatic lesions.

Further investigations have found that (a) the germinative cell cycle is shortened from 311 to 36 hours, indicating that the keratinocytes, (b) there is a doubling of the proliferative cell population in psoriasis from 27,000 to 52,000 cells/mm$^2$ of epidermal surface area, and (c)
100% of the germinative cells of the epidermis enter the growth fraction instead of only 60% for normal subjects. This leads to a 10-fold rather than an 8-fold acceleration.

The source of the cycling cells in the suprabasal layers of the epidermis is not well defined. They could be...
Recent studies suggested that psoriatic epidermis shows aberrant expression of apoptosis-related molecules.
Keratinocyte Differentiation
Keratinocytes undergo the process of differentiation as they migrate upward through the epidermis from
Spongiform pustules of Kagoj in Psoriasis
Immunopathology
Immunologic factors play a very important role in the pathogenesis of psoriasis. Psoriasis is now regarded...
CD4+ T cells produce a variety of cytokines, including interleukin-2 (IL-2), tumor necrosis factor-a (TN Fa}, and y-interferon (yIFN), which is also produced by CD8+ T lymphocytes.
Keratinocytes stimulated by TNFs may produce IL-1B, which is a potent T-lymphocyte and neutrophil chemoattractant present in increased amounts in psoriatic epidermis. This cytokine may be involved in the formation of Munro microabscesses.
Spongiform pustules of Kagoj in Psoriasis
IFN is believed to play an important role in the initiation of psoriatic lesions as demonstrated by the induction of pinpoint lesions of psoriasis at sites of IFN injection in previously uninvolved skin.
yIFN induces the expression of the ICAM-1 in keratinocytes and endothelial cells. This molecule mediates...
not to be responsive to the growth inhibition effects of yIFN, leading to their hyperproliferative state in the disease.
Increased expression of p53 and downregulation of Bcl-2, consistent with the dynamics of psoriasis, have...
Pathogenesis of Localized Pustular Psoriasis
A relationship of pustulosis palmaris et plantaris with psoriasis is not generally accepted, although two facts favor a relationship: (1) a leukotactic factor identical to that noted in psoriasis has been found in pustulosis palmaris et plantaris.
Pathogenesis of Psoriasis and AIDS
There is evidence of the role of both CD8+ and CD4+ T lymphocytes and yIFN in the pathogenesis of psoriasis.
Paradoxically, as T-helper cell counts decline, it appears that psoriatic lesions exacerbate until a preterminal stage, it was shown that ylFN serum levels were much higher in HIV-positive psoriatic patients than HIV-negative subjects.
The immunodysregulation resulting from HIV infection may trigger psoriasis in those genetically predisposed.
Differential Diagnosis
Two histologic features are of great value in the diagnosis of psoriasis vulgaris: (a) mounds of parakeratosis with elongation of the rete ridges and parakeratosis, can be found also in chronic eczematous dermatitis, such as atopic dermatitis, nummular dermatitis, or allergic contact dermatitis, which then may appear to be "psoriasiform." However, the elongation of rete ridges is uneven. Although mild spongiosis may be seen in treated lesions of psoriasis and in those with superimposed allergic contact dermatitis secondary to topical treatments.

Lichen simplex chronicus is considered in the differential diagnosis of fully developed psoriatic plaques. In contrast to psoriasis, it shows a more uniform acanthosis, and fibrosis of the papillary dermis with collagen bundles aligned perpendicular to the skin surface.

Seborrheic dermatitis may be very difficult to distinguish from psoriasis vulgaris, especially if overlap occurs. Accentuated spongiosis, accentuated acanthosis at the follicular ostia, and more irregular acanthosis are histologic features suggestive of seborrheic dermatitis.

Pityriasis rubra pilaris shares some histologic features with psoriasis, namely, acanthosis and parakeratosis. However, it could be differentiated from psoriasis because the lesions of pityriasis rubra pilaris show a more uniform acanthosis with formation of rete ridges, and a more uniform inflammatory infiltrate. Although the Kogoj spongiform pustule is highly diagnostic of the psoriasis group of diseases,
including Reiter’s disease, histologically typical spongiform pustules may occur also in pustular dermatophytosis, bacterial impetigo, pustular drug eruptions, and candidiasis, particularly if pustules are clinically present. Periodic acid-Schiff (PAS) and Gram stains are useful for identifying microorganisms. Histologically, spongiform pustules are characterized by the presence of vacuolated keratinocytes and a neutrophilic infiltrate, whereas Munro microabscesses are smaller and more circumscribed. In particular...
Because of the clinical and, particularly, the histologic resemblance of the tongue lesions in pustular psoriasis.