

# MEDICAL PRACTICE

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## *Clinical Progress*

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### Psoriasis—Clinical Features

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Few dermatoses are as easily recognized and as morphologically distinctive as classical psoriasis. Every clinical student quickly learns to recognize the sharply defined salmon-red plaques, capped by silvery scales, occurring typically on the extensor aspects of the limbs. Much psoriasis is indeed classical but few other than dermatologists appreciate the number of morphological variants with which this disease may present. Misdiagnosis of “atypical” psoriasis is common and leads to prognostic and therapeutic errors. The clinical features of psoriasis may be influenced by a variety of endogenous and environmental factors. This article will describe briefly the less well recognized variants of psoriasis, some common and relatively minor, others uncommon yet devastating in their impact. Attention will also be drawn to the ways in which therapy may modify the clinical picture.

#### Impact of Age

Classical psoriasis, with large discoid lesions on the trunk and limbs, is seen chiefly in the years of sexual maturity. At the extremes of life, both in young and old, psoriasis may look very different.

#### NAPKIN PSORIASIS

Traditionally, psoriasis has been rightly taught to be rare before the age of 5 years, but the paediatrician and the general practitioner know that eruptions evolving from the napkin area in infants may be very psoriasiform in appearance. The creases are affected and the erythematous plaques have well-defined edges on the thighs and abdomen. The axillae, neck, and scalp

may also be involved.<sup>1</sup> Warin coined the term “napkin psoriasis” for such eruptions, which in the short term have an excellent prognosis and which respond well to topical corticosteroids. The relationship of this eruption to true psoriasis is disputed, candidosis and seborrhoeic dermatitis having claims to its lineage. Until long-term follow up studies have been undertaken its nature will be debated, but its occurrence in psoriatic families and occasional reappearance as ordinary psoriasis later lend support to the contention that it occurs in infants with the psoriatic predisposition.<sup>2</sup>

#### GUTTATE PSORIASIS

This exanthem is typically seen in children aged 5 to 12 years, often as the first manifestation of the disease. A sore throat is followed after ten to twenty days by an eruption, mainly on the trunk, of pink guttate maculopapules. Infectious disease may be suspected but after a few days the papules become more typically psoriatic and the absence of malaise or any enanthem makes the diagnosis clear. Guttate psoriasis often clears within three or four months without much treatment but may evolve into chronic discoid psoriasis.

#### FLEXURAL PSORIASIS

In the elderly, psoriasis has a tendency to be present in the axillae, submammary, or anogenital folds, though not usually exclusively. Flexural psoriasis is usually seen in postmenopausal women and older men, though it may occur at any age. The intertriginous site modifies the appearance of the psoriasis, which lacks scales and presents as a smooth glazed well-defined erythematous plaque. Similar psoriasis is found on the glans penis of the uncircumcized.

#### Localized Patterns

The clinical spectrum of psoriasis is enormous, both in space and time. At one extreme is the trivial and the transient; at the

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other, chronic universal or near-universal disease. Persistent localized psoriasis will not evade recognition if simple rules of differential diagnosis are remembered.

*Scalp.*—Psoriasis often presents in and may be confined to the scalp, especially at its margins. It is the likely diagnosis in the patient complaining constantly of severe dandruff. Unlike dandruff, psoriasis in the scalp can usually be felt as well as seen and is patchy as elsewhere. It rarely causes alopecia.

*Flexures.*—Psoriasis may present as pruritus ani and be confined to the perianal skin. Solitary perivulval and pubic plaques are not rare.

*Penis.*—Psoriasis is a common cause of persistent erythematous macules or plaques on the glans penis. Whether scale is present or not depends on whether the member is circumcized.

*Nails.*—Psoriasis may effect the nail matrix or the nail bed.<sup>3</sup> Psoriatic involvement of the matrix at the nail folds produces pitting or ridging of the emerging nail plate. Involvement of the bed produces onycholysis and subungual thickening. There may be no overt involvement of the skin of the nail folds. Pitting of the nail plate and onycholysis are usually seen in the fingers and subungual thickening in the toes.

The diagnosis of all of these localized patterns is unlikely to be missed if the whole body surface is inspected.

### Other Morphological Variants

#### PUSTULAR PSORIASIS

The pathology of psoriasis is characterized by accelerated and abnormal formation of the epidermis, capillary hypertrophy in the papillary dermis, and the immigration of polymorphonuclear leucocytes into the epidermis to produce microscopic (sterile) microabscesses. If the last named feature is exaggerated relative to the others, localized leucocytic aggregations of macroscopic proportions develop, apparent clinically as subcorneal pustules. Pustular psoriasis is uncommon. It is usually chronic and confined to the palms and soles. Usually seen in middle life, it is characterized by well-defined areas of erythema and variable scaling, in which erupt flat yellow pustules which slowly desiccate to leave small brown stains in the desquamating corneum. The thenar and hypothenar eminences, the heels, and the insteps are particularly affected. There may or may not be ordinary psoriasis elsewhere.

#### RUPIOID PSORIASIS AND KERATODERMA BLENORRHAGICA

In this variant gross parakeratotic scale is heaped over a pustular lesion. It is usually seen in the palms and soles but may occur elsewhere. It is morphologically and histologically identical with the so-called keratoderma blenorrhagica seen in some men with Reiter's disease. In my view keratoderma blenorrhagica and psoriasis are one and the same. The psoriatic lesions seen in Reiter's syndrome are usually atypical—pustular, rupioid, penile, or perionychial lesions being particularly prominent. These lesions tend to settle with the rest of the syndrome but may evolve into chronic banal psoriasis. Possibly keratoderma blenorrhagica occurs only in men who have inherited the psoriatic diathesis.

#### ACRAL PSORIASIS (ACRODERMATITIS CONTINUA)

In this puzzling pattern the disease begins at one or more finger tips or around the nail fold simulating paronychia, often after trivial local infections or trauma. More fingers or toes are progressively affected, with much discomfort and disability. The disease involves the nail bed and fold and leads to gross nail dystrophy or shedding. It may spread to the palm or sole. Rarely, intractable and eventually fatal generalized pustular psoriasis may ensue, particularly in old people.

### Aetiology and Pathogenesis

The only undisputed factor in the aetiology is the genetic one. This assertion is based on the facts of familial aggregation (in the absence of any aggregation in spouses), on twin studies, and on individual pedigrees and kindreds.<sup>4</sup> Familial aggregation has been established in several large surveys in N.W. Europe. Twin data may reflect selective reporting but the concordance rate is much higher in published identical as compared with non-identical twin pairs. As with other common diseases, the mode of inheritance is difficult to establish. In a minority of families the disease appears to be inherited as a simple Mendelian dominant. Attempts at complete genetic analysis of pooled epidemiological data have foundered on the difficulty that the psoriatic diathesis may not be manifested until late in life, if at all. Unfortunately, no certain method of recognizing the latent psoriatic has yet been devised. The weight of evidence favours dominant inheritance with incomplete penetrance, but multiple factors may be concerned.

#### PREVALENCE

At any given time the disease is found in 1.5 to 2.0% of adults in North West Europe.<sup>5 6</sup> There are major racial variations, the disease being much less common in the yellow-brown races and very uncommon in the negro.<sup>7</sup> If Mendelian inheritance is assumed a predisposition frequency can be calculated if the incidences in the population and families of probands are known.<sup>5</sup> This has been found to be about 11%, assuming dominant inheritance.

#### TRIGGERING FACTORS

In a subject with the psoriatic predisposition a number of postnatal and environmental factors may trigger or perpetuate the disease.

#### Trauma

Possibly the napkin acts in this way. Psoriasis may be provoked by surgical incisions, sutures, scratches, etc. (Koebner phenomenon), especially if the disease is in an eruptive phase. The Koebner phenomenon has recently attracted experimental observation, but its mechanism is still not fully understood.<sup>8</sup>

#### Infection

Upper respiratory infection, especially streptococcal, has already been mentioned as a cause of guttate psoriasis of childhood. Remote infection may also precipitate palmoplantar or the generalized types of pustular psoriasis. Rarely childhood psoriasis seems to be perpetuated by chronic upper respiratory infection and cannot be controlled until the infection is eliminated—for instance, by tonsillectomy.

#### Sexual Epochs

Endocrine crises seem to influence psoriasis. There are peaks of incidence at the menarche and menopause.<sup>9</sup> Psoriasis usually improves during pregnancy only to relapse post partum. Very rarely generalized pustular psoriasis is precipitated by pregnancy.

#### Light

Many psoriatics are better in summer, and the disease often clears dramatically during a holiday in the sun. Conversely, psoriasis may be precipitated in exposed skin by exposure to the sun.

### Drugs

Chloroquine sometimes aggravates psoriasis. Corticosteroids have a two-edged effect on psoriasis. While producing improvement (sometimes dramatic) in the short term, they tend to render the disease unstable in the long term and may perpetuate erythrodermic psoriasis. Their withdrawal is usually promptly followed by relapse and rarely generalized pustular psoriasis may be precipitated.<sup>10</sup>

Any drug eruption in a psoriatic may provoke widespread psoriasis in its wake.

### Hypocalcaemia

Accidental parathyroidectomy has rarely precipitated fulminating generalized pustular psoriasis (sometimes called impetigo herpetiformis); hypocalcaemia has also provoked ordinary psoriasis.<sup>11</sup> Low serum calcium levels may be seen during the course of fulminant psoriasis; several factors seen to contribute including hypoalbuminaemia, hypervolaemia, and transient malabsorption.<sup>12</sup>

### Emotional Factors

Emotional factors are difficult to measure and their importance has been exaggerated in the past. Nevertheless, there is little doubt that major emotional stress can sometimes contribute to the exacerbation and chronicity of this disease.

### Complications

#### SECONDARY INFECTION

Unlike eczematous dermatitis, psoriasis is rarely clinically infected, whether by bacteria, dermatophytes, or yeasts. Psoriatic scale can harbour and disseminate pathogenic staphylococci occasionally with disastrous results. An instance has been reported where a psoriatic anaesthetist was responsible for several postoperative infections and deaths.<sup>13</sup> Personnel with psoriasis working in surgical theatres, obstetric, or neonatal units should be bacteriologically screened regularly.

#### GENERALIZED EXFOLIATIVE (ERYTHRODERMIC) PSORIASIS

Psoriasis may spread until the skin is universally involved, spontaneously or because of injudicious topical therapy, over-exposure to ultraviolet light, chloroquine administration, or withdrawal of systemic steroid therapy. The distinct and well-defined plaques of discoid psoriasis become submerged in a generalized inflammation in which the skin is uniformly erythematous and oedematous with varying degrees of exfoliation. If persistent, superficial lymphadenopathy may follow and the hair may be lost. Itching and discomfort may be intense and the whole picture in the acute case is complicated by malaise and an upset in the regulation of heat loss. Several factors may contribute to the latter. Obstruction of the sweat pores causes hypohidrosis, but the increase in cutaneous blood flow and temperature raises radiant heat losses, so that the patient may be hyperthermic or hypothermic according to the ambient conditions.

The increased metabolic activity of the skin and the rise in blood flow to sustain this make demands on the heart, leading to a hyperdynamic circulatory state and rarely to high output cardiac failure. Prolonged exfoliation with its consequent loss of the fibrous protein, keratin, leads to protein depletion—one

of the factors contributing to hypoalbuminaemia. Temporary malabsorption may further complicate the metabolic situation. These and other metabolic consequences (which are not peculiar to erythrodermic psoriasis but may follow erythroderma of any cause) have been well characterized and described by Shuster and his school over the last decade.<sup>14</sup>

#### GENERALIZED PUSTULAR PSORIASIS

In its most acute form generalized pustular psoriasis is a dramatic intervention in the course of psoriasis.<sup>15</sup> The onset may be sudden, heralded by apprehension and nausea. Waves of widespread or universal erythema, accompanied by malaise, occur, during which the skin is both painful and tender. Within a few hours the sheets of erythema are studded with masses of pinhead and larger intraepidermal pustules. The pustules may become confluent to form yellowish-green lakes of pus beneath the stratum corneum. Fever and leucocytosis are present. Waves of scarlatiniform peeling follow, removing the desiccating pustules. This series of events may occur just once, or may recur every few days for weeks. It may supervene on any previous pattern of psoriasis or may rarely occur *de novo*.<sup>10</sup>

### Psoriatic Arthritis

Psoriasis and an inflammatory polyarthritis go together more often than chance alone would indicate.<sup>6, 6</sup> In addition, there are clinical and serological reasons for separating psoriatic arthritis from rheumatoid and other arthropathies. Most patients never develop rheumatoid serum factors, either overt<sup>16</sup> or masked<sup>17</sup>—i.e., psoriatic arthritis is a seronegative arthropathy.

Four major clinical patterns are usually recognized:<sup>18</sup>

*“Distal” Arthritis.*—In this the disease starts either in one or more terminal interphalangeal joints in the fingers or in the interphalangeal joints of the toes. It may produce “sausage”-like deformities of digits. Eventually, it often affects more proximal joints, large or small.

*Arthritis Mutilans.*—This is characterized by severe peripheral joint destruction with striking osteolysis with or without bony ankylosis. Spondylitis is common in these patients, and the sacroiliac joints are usually obliterated.

*“Indistinguishable” Arthritis.*—This resembles rheumatoid but tends to be more asymmetrical.

*Spondylitis.*—Spondylitis resembling ankylosing spondylitis may be seen in the absence of peripheral arthritis.

Rarely in any of these four patterns striking paravertebral ossification is seen.<sup>19</sup>

Nail dystrophy is twice as common in psoriatic arthritis as in uncomplicated psoriasis. Erythrodermic and generalized pustular forms are more often associated with arthritis. In general the psoriasis which accompanies chronic psoriatic arthritis tends to be more severe, persistent, and resistant to treatment than uncomplicated psoriasis.

The cause of psoriatic arthritis and the nature of the association of psoriasis and arthritis are not understood but a genetic association seems possible. Atypical polyarthritis has been sporadically reported in psoriatic families, and recently psoriatic arthritis has been found to be aggregated in the first-degree relatives of probands with the disease 50 times more often than expected.<sup>18</sup>

The prognosis of psoriatic arthritis is usually favourable if corticosteroids are avoided, but rapidly evolving arthritis mutilans may leave in its wake severe hand damage with loss of function.