
Clinical Spectrum and Severity of Psoriasis

Matthew Meier · Pranav B. Sheth

Department of Dermatology, University of Cincinnati College of Medicine, Cincinnati, Ohio, USA

Abstract

Psoriasis is a chronic inflammatory skin disease. Associated comorbidities or risks may include psoriatic arthritis, obesity, depression, smoking, diabetes, hyperlipidemia, an increased risk of cardiovascular disease with myocardial infarction, or an increased risk of lymphoma. The clinical presentation of psoriasis can range from the more common red scaling elevated plaques on the elbows, knees, or scalp to the less common superficial pustules scattered on the palms or soles, or in rare cases widespread pustules on the body. More specifically, the clinical spectrum of psoriasis includes the plaque, guttate, small plaque, inverse, erythrodermic, and pustular variants. The determinants of the clinical severity of psoriasis, the risk of comorbidities, and the quality of life of a psoriatic patient are influenced by multiple factors. At the minimum, these include variations in the quality and type of psoriasis, the quantity of skin involved, and the distribution of skin lesions (including special areas such as the scalp, nails, face, intertriginous regions, and palmoplantar surfaces). Objective measures used to quantify the severity of psoriasis, including the body surface area involved, Physician's Global Assessment, Psoriasis Area and Severity Index, and quality of life measures, are all assessments that can be useful in guiding approaches to management and therapeutics. In this paper, we review the clinical spectrum of psoriasis, the differential diagnoses, measures and determinants of severity, and the recommendations on when to refer a patient to a specialist in psoriasis. We also briefly review the comorbidities, and note the importance of referring the psoriatic patient to the internist/general practitioner for evaluation and management for these comorbidities.

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Psoriasis is a chronic inflammatory disorder of the skin that can affect a person at any age. It can present in various patterns and forms. The most common morphologic presentation of psoriasis is that of the plaque type, with the second form being the pustular type. Its course is variable and unpredictable. It may be episodic with short or long periods of reported complete clearance, or be unrelenting and persistent with waxing and waning of activity influenced by identifiable or unidentifiable triggers and alleviators. Initially it may be indolent and virtually unrecognizable as 'psoriasis'

by the patient or physician, only to present itself in a more classic presentation during times of emotional, physical, or medical stress.

Throughout history, psoriasis has been understood and misunderstood as a disease solely of the skin. Its consequences on the social, psychological, physical, and spiritual fabric of the individual and those close to him/her have been increasingly recognized. However, by the end of the 20th century, the psoriasis model had evolved to become a disorder of the skin and joints. Accepted as the consequence of an immune system gone awry, psoriasis has become a model of a skin disease with 'systemic inflammation'. Much work is now being done to understand the association of comorbidities with psoriasis and their impact on the patient and society.

With the advances in technology and medical research, the current pathogenetic model for psoriatic disease includes a combination of genetic predisposition, immunologic dysfunction, and keratinocyte factors that lead to the formation of psoriasis. Along with these, the roles of the peripheral and central nervous systems, vascular system, adaptive and innate immune systems, environmental factors, and infectious agents contribute to the formation of psoriasis. In this paper, we will review the clinical spectrum, the differential diagnosis, and the severity of psoriasis as they relate to quality of life, therapeutic options, and signs that indicate when further evaluation by a dermatologist or a specialist in psoriatic disease is warranted.

Epidemiology

It is estimated that approximately 2% of the US population is affected by psoriasis. Similar prevalence values have been obtained in Europe, with the exception of slightly higher values seen in Norway and the Faeroe Islands. Racial differences do exist. In a recent US population-based survey, the prevalence in Caucasians was estimated to be 2.5%, while in African-Americans this was 1.3%. Males and females are affected equally. Psoriasis can present at any age, but most commonly presents in bimodal peaks between the ages of 15 and 30, and after 40 years of age [1, 2].

Genetics

The inheritance of psoriasis is quite complex, and does not fit into a simple recessive or dominant Mendelian inheritance pattern. Other factors, including environmental ones, may be involved. It is known that having a first-degree relative with psoriasis confers an increased risk of disease [3]. Psoriasis is a complex genetic disease, accounting for the difficulty in identifying susceptibility genes. What is known is that there are multiple susceptibility regions that have been identified, which contain genetic polymorphisms that confer an increased risk of developing psoriasis. The best

characterized is PSORS1, in which HLA-Cw6 was recently identified as the definitive allele which confers the elevated risk [4, 5].

Pathogenesis

Psoriasis involves a complex interplay between various cells of the immune system and skin, including dermal dendritic cells, T cells, neutrophils, and keratinocytes. CD8+ T cells populate the epidermis, while macrophages, CD4+ T cells, and dermal dendritic cells reside in the superficial dermis. A multitude of cytokines, chemokines, and cell surface receptors are involved in a web of molecular pathways leading to clinical disease. Psoriasis is considered to be an immune-mediated disease characterized by a predominantly Th1-type cytokine profile in lesional skin with elevated levels of interferon- γ , TNF- α , IL-12, and IL-18, among others. More recently, the Th17 pathway has proven to be critical for maintenance of the chronic inflammatory process. At the center of this pathway is the CD4+ T cell, whose maintenance is supported by IL-23 secreted by antigen-presenting cells (dermal dendritic cells). These Th17 CD4+ T cells secrete IL-17 and IL-22, contributing to the enhancement and maintenance of inflammation and epidermal proliferation [6, 7].

Morphologic Subtypes

In its most classic morphologic presentation, psoriasis is characterized by red scaling elevated plaques. These correlate to the inflammation, vascular dilatation, and altered epidermal proliferation and differentiation (regular hyperplasia and hyperparakeratosis) seen histologically. Variations of this plaque morphology include red scaling patches (more often seen in the scalp, inverse, and erythrodermic forms of psoriasis) and the red scaling papules seen in early or flaring, guttate, or follicular psoriasis.

The second morphologic presentation is one of superficial pustules, characterized by intra-epidermal neutrophil accumulation with only mild epidermal hyperplasia on histology. Variations of this morphology include discrete and/or confluent superficial yellow-white pustules, either on a smooth erythematous edematous base or overlying normal-appearing skin (as seen in pustular psoriasis of von Zumbusch) or superficial discrete dirty yellow-brown pustules often found on the hands or feet (as seen in palmoplantar pustulosis).

Although there are 2 main morphologies, there are multiple subtypes of psoriasis that have been described based upon a combination of morphology, distribution, and pattern. These subtypes often occur alone. However, there may be an overlap or transition from one subtype to another due to various triggers or evolution of the disease. The subtypes are described in the following sections.



Fig. 1,2. Plaque psoriasis. Back and buttocks (1) and lower extremities (2) of two patients with plaque psoriasis, showing the variations in erythema, scale, and symmetry in this type of psoriasis. **Fig. 3.** Scalp psoriasis. Extension of plaques onto the forehead in a patient with diffuse involvement of the scalp associated with focal hair thinning.

Plaque Psoriasis

This is the most common and well-recognized form of psoriasis. It is characterized by well-defined raised erythematous papules and plaques with silvery coarse scale. There can be great variation in the intensity of erythema (ranging from light pink to bright red to deep purplish red), elevation of the lesion (flat to very thick and elevated), and amount of scale (scattered light diffuse white scale overlying the lesion to thick micaceous hyperkeratotic scale in which the color and thickness of the lesion are not easily discernable; fig. 1, 2). The distribution is typically symmetric, and sites of predilection include the extensor surfaces of the extremities, particularly the elbows and knees, sacrum, scalp, nape of the neck, and to a lesser extent the remainder of the trunk, genitalia, face, and ears [8] (fig. 3). Individuals may present with hyperkeratotic scaling disease localized to the scalp only, making it difficult to discriminate from severe seborrheic dermatitis or tinea capitis. This presentation has been called tinea amiantacea, and requires evaluation to distinguish between these 3 entities. Plaque psoriasis localized to the palms and soles can have a significant impact on the patient's quality of life and function (fig. 4). At times, ill-defined or partially treated plaques localized only to the palms and soles can be difficult to distinguish from chronic hand dermatitis as both conditions can have erythema, scaling, fissuring, pain, itching, and nail changes. Biopsies are often equivocal and may suggest a psoriasiform dermatitis.

Additional features of psoriatic plaques include the Auspitz sign and Woronoff's ring. Auspitz sign is the presence of pinpoint bleeding at the base of a plaque after scale is forcibly removed. Its presence can sometimes be helpful, but it is not present in all cases and can also be seen in other disorders [9]. Woronoff's ring refers to the presence of a white ring around erythematous plaques undergoing topical treatment or phototherapy [10]. Interestingly, some patients develop lesions at sites of trauma, including those from sunburn. This phenomenon is known as the isomorphic or Köbner phenomenon (fig. 5).

Fig. 4. Palmar plaque psoriasis and psoriatic arthritis. This 10-year-old female has restricted use of her hands due to tightness, fissuring, itching, and pain. Note the flexion contracture of the 5th digit from psoriatic arthritis.

Fig. 5. Köbner phenomenon. A patient with a history of scalp psoriasis who developed lesions on the upper back 3 weeks following a sunburn to the area.

Fig. 6. Inverse psoriasis. Thin plaques localized to inframammary regions in a patient with psoriasis that also involved the axillary, inguinal, abdominal, and gluteal folds.

Fig. 7. Guttate psoriasis. Acute generalized scaly papules in a patient with asymptomatic streptococcus colonization of the pharynx.

Inverse Psoriasis

Inverse psoriasis is characterized mainly by its distribution: it is localized predominantly to intertriginous regions including the axillae, inframammary regions, gluteal cleft, genitals, abdominal folds and inguinal folds. These lesions also differ in morphology from that of typical plaque psoriasis lesions in that they are well-defined shiny erythematous patches or thin plaques without significant scale [6, 11] (fig. 6). The presentation may initially be confused for bacterial (often accentuated in the crease), candidal (appearance of superficial pustules and 1- to 2-mm superficial round erosions often helpful for diagnosis), or fungal intertrigo (less involvement of the crease, peripheral desquamative-type scale and possible fungal involvement elsewhere, such as the feet, gluteal cheeks, or toenails). Scrapings or cultures are sometimes needed to discriminate between these entities.

Guttate Psoriasis

This form of psoriasis is characterized by an acute generalized eruption of smaller round to oval-shaped well-defined erythematous scaly papules and plaques up to 1 cm in size [12] (fig. 7). This is considered to be more of an eruptive form of psoriasis and is often associated with infection, especially streptococcal pharyngitis [13]. More common in children and young adults, guttate psoriasis may initially respond well to antibiotics, heliotherapy, or phototherapy and go into remission, only to recur with reinfection.

Patients with guttate psoriasis may have a higher risk of developing plaque psoriasis later in life. On the other hand, for some patients, guttate psoriasis may be the initial manifestation of chronic psoriasis, triggered by infection or stress, only to evolve into the plaque-type over time. Guttate psoriasis can also be easily confused with the papular type of acutely flaring plaque psoriasis. The nature of the prior course, triggering factors, and intensity of erythema and scale (less in guttate) may help in differentiating these. Guttate psoriasis may also be confused with pityriasis rosea (PR), but differs in the nature of the scale (psoriatic scale involves the entire lesion and is more coarse, rather than the finer localized trailing ring pattern of the scale in PR), pattern (psoriasis does not present in a Christmas tree pattern and is less likely to appear in linear arrangements in the axilla and neck), and course (guttate psoriasis often lasts longer than 8 weeks unless appropriately treated, while PR often resolves within 6–8 weeks without treatment).

Although guttate psoriasis is highly associated with streptococcal infections, there is little evidence-based data, as documented in a Cochrane library review, to support treatment of these patients with antibiotics [14]. In the authors' experience, patients with an acute papular psoriatic eruption with a throat culture positive for streptococcus or markedly elevated antistreptolysin titers have a moderate chance of improving and sometimes clearing their psoriasis with oral antibiotics.

Erythrodermic Psoriasis

When involvement of psoriasis becomes so diffuse that it involves more than 90% of the total body surface area, a patient is considered to be erythrodermic. With confluence of the lesions and diffuse involvement, the lesions may be thinner or truly flat, and it may be difficult to identify lesions of classic plaque psoriasis. The skin may be warm to the touch due to increased perfusion, and the nature of erythema may vary depending on the patient's skin color and acuity of disease (brighter red with acuity or flaring). The scaling is also of a different quality; it is more fine, flaky, and desquamative rather than the typical silvery coarse thick scale of plaque psoriasis (fig. 8, 9). Of note, facial involvement may lead to ectropion.

Although not specific to erythrodermic psoriasis, as it can be seen in patients with erythroderma of any cause, associated clinical findings may include lymphadenopathy, fever or hypothermia, tachycardia, and peripheral edema. Associated laboratory findings can include an elevated erythrocyte sedimentation rate, hypoalbuminemia, leukocytosis or leukopenia, anemia, and elevations of lactate dehydrogenase, liver transaminases, uric acid, and calcium [15]. Precipitants for erythrodermic psoriasis include the tapering or discontinuation of systemic medications (such as corticosteroids, cyclosporine, or methotrexate), phototherapy-related toxicity, irritants (such as tar), and systemic illness or infection. Significant morbidity may occur due to dehydration from extensive fluid and electrolyte disturbances, protein losses, high-output cardiac failure, and infection.

Psoriasis is only one of many causes of erythroderma. Cutaneous T cell lymphoma – typically in the setting of Sezary syndrome, atopic dermatitis, drug reactions, other papulosquamous skin diseases (such as pityriasis rubra pilaris), as well as connective tissue diseases such as lupus or dermatomyositis among others – can also present as exfoliative erythroderma. Presence of classic cutaneous, scalp, or inverse lesions, nail findings, or personal or family history of psoriasis may provide clues to the etiology of erythroderma [16]. Skin biopsies are often unhelpful in confirming erythroderma secondary to psoriasis, but may be helpful in ruling out cutaneous T cell lymphoma or connective tissue disease.

Small Plaque Psoriasis

This form of psoriasis is similar in morphology to guttate psoriasis with discrete papules and plaques, with lesions as large as 3 cm in size. However, small plaque psoriasis represents a chronic form of psoriasis rather than an acute eruptive process [12]. This variant may not have the pattern of accentuation on the extensor extremities, scalp, elbows, and knees as in classic psoriasis, and may have a more randomly distributed, scattered, and diffuse pattern.