The skin in psoriasis: assessment and challenges

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Key words: plaque psoriasis, generalised pustular psoriasis, palmoplantar pustulosis, PASI, DLQI, BSA, PGA, comorbidity ABSTRACT

The coexistence of psoriasis arthritis (PsA) and psoriasis vulgaris in about 20% of patients with psoriasis leads to a need for rheumatologic-dermatologic team work. We summarise the role of dermatologists in assessment of the skin in psoriasis.

Chronic plaque psoriasis must be differentiated from other subtypes such as generalised pustular psoriasis (GPP) or palmoplantar pustulosis (PPP). Therapeutic management is based on the evaluation of the disease severity. Quantitative scoring of skin severity includes calculation of the Psoriasis Area and Severity Index (PASI), body surface area (BSA) as well as the Dermatology Life Quality Index (DLQI). These scoring systems do not replace the traditional dermatologic medical history and physical examination of the patient. The skin should be examined for additional skin diseases; moreover, patients should be monitored for comorbidity, most importantly PsA and cardiovascular comorbidity.

Introduction

Psoriasis is a chronic inflammatory skin disease affecting about 2% of the Caucasian population (1). About 20% of the patients have psoriatic arthritis (PsA) (2, 3). Many patients with psoriasis are not aware of their PsA; and the prevalence of undiagnosed PsA is still high as has been shown in a recent systematic meta-analysis. Accordingly, up to 15.5% of patients with psoriasis had undiagnosed PsA (4). Dermatologists usually are the doctors consulted in cases of new-onset psoriasis (5). In collaboration with rheumatologists they should screen their patients with psoriasis for PsA, as PsA is a progressive disease, and a subgroup of patients develops progressive damage and loss of function in the first few years of the disease (4). On the other hand, in about 6 to 18% PsA may precede skin lesions (3, 4). Then it usually is the rheumatologist, who first diagnoses the skin disease. Hence, both specialties play an important role in early disease detection and determining the course regarding further treatments of PsA as well as psoriasis.

This paper refers to the tasks of dermatologists in assessment of the skin in a patient with psoriasis and/or PsA. Clinically challenging aspects will be addressed to strengthen the collaboration of dermatology, rheumatology, as well as general medicine, to care for patients with psoriasis. First of all the diagnosis of psoriasis must be identified. The differential diagnoses includes eczema or mycosis fungoides, and the distinct type of skin psoriasis should be defined, *i.e.* psoriasis vulgaris (PV) manifesting as chronic plaque psoriasis has to be distinguished from generalised pustular psoriasis (GPP), palmoplantar pustulosis (PPP), or acrodermatitis continua suppurativa (6, 7). We focus on PV and summarise the specific clinical tools which are commonly used for the assessment of disease severity, and describe some challenges that may occur. Assessment of the skin of patients with PV or PsA is a component of a comdermatological examination: plete thoroughly performed it takes into account important individual aspects of the skin status such as the number of melanocytic nevi or the tendency to skin dryness and atopy (Table I). Importantly, patients have to be monitored for skin tumours or precancerous lesions such as basal cell carcinoma, squamous cell carcinoma or actinic keratoses, respectively, taking into account the often increased cumulative risk of carcinogenic sun exposure, UV light treatment and/or immunosuppressive therapies (8).

Key features of skin psoriasis

Psoriasis vulgaris (PV) is diagnosed by the characteristic psoriatic plaques consisting from salmon red sharply bordered macules covered with silvery

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Table I. Dermatological assessment of patients with psoriasis.

Definition of psoriasis type/s	Concomitant skin disorders or history of these				
Plaque psoriasis (Psoriasis vulgaris)	Infections				
Palmoplantar pustulosis (PPP)	Tumours				
Acrodermatitis continua suppurativa	Eczemas				
Generalised pustular psoriasis (GPP)	Lichen planus				
± Psoriasis arthritis (PsA)	Vitiligo				
Predilection sites of the skin	Alopecia areata				
Scalp (retroauricular)	Urticaria				
Extensor sites	Dermatitis herpetiformis Duhring				
Nails	Cutaneous lupus erythematodes				
Flexural / genitals (gluteal cleft)	Scars (after tumour excision)				
General aspects of the skin	Comorbidity / cardiovascular risk factors				
Pigmentation type	Body Mass Index (BMI)				
Number of nevi	Hyperlipidaemia				
Skin dryness	Hypertension				
Mucous membrane (tonsils)	Other diseases or history of these*				
Teeth	Rheumatologic / orthopaedic				
Conjunctivae	Gastrointestinal / hepatic / renal				
	Neurological / psychiatric				
	Cancer / haemolymphatic				
	Allergies				

*incl. screening for psychological distress, fatigue, smoking and alcohol consumption.

scales. Knees, elbows, scalp and umbilicus are commonly affected (Fig. 1). Importantly, the diagnosis of inverse psoriasis, in which only the flexural folds are affected, should not be missed. It presents with erythematous sharply demarcated areas, typically without silvery scaling. Patients may not address symptoms of inverse psoriasis. Hence, psoriasis cannot be excluded if the gluteal cleft, groins, and retro-auricular areas have not been examined. Involvement of the lips is possible; involvement of the mucous membrane would be extremely unusual (1, 9). However, considering potential differential diagnoses such as lichen planus or adverse reactions of systemic therapies (Fig. 2) inspection of the mouth should be performed in all patients with psoriasis. Nail psoriasis is extensively described elsewhere in this supplement. In short, 15-50% of patients with psoriasis have nail changes. This figure increases to 85% in patients with psoriatic arthritis (10). Nail pitting, oil spots and onycholysis are highly diagnostic (11).

Distinct subtypes of psoriasis

Plaque psoriasis should be differentiated from other psoriasis forms, which are clinically distinct and have a different genetic background (1, 9, 12). GPP is now regarded an autoinflammatory skin diseases (DIRA/DITRA, etc.) (13). It has a different, more rapid disease



Fig. 1. Clinical examples of distinct forms of psoriasis and special localisations of psoriasis vulgaris: severe chronic plaque psoriasis (PASI 21.6) (\mathbf{a}), palmoplantar pustulosis in a female patient with Sapho syndrome (\mathbf{b}), generalised pustular psoriasis in a patient with IL36 receptor mutations (\mathbf{c}), acrodermatitis continua suppurativa in a patient also suffering from psoriasis vulgaris (\mathbf{d}), psoriasis capitis as most common location of psoriasis vulgaris (\mathbf{e}), severe nail psoriasis (\mathbf{f}), inverse psoriasis first misdiagnosed as mycosis (\mathbf{g}), isolated palmar psoriasis vulgaris (\mathbf{h}), and psoriasis of the external ear canal and scalp (\mathbf{i}).



Fig. 2. Clinical examples of concomitant skin diseases in patients with psoriasis: squamous cell carcinoma in a patient, who regularly used sun bathing as self-therapy (\mathbf{a}) , basal cell carcinoma origination from a scare in a patient suffering from psoriasis arthritis treated with methotrexate (\mathbf{b}) , mild lichen planus of the gingiva in a patient under biologics (\mathbf{c}) , multiple actinic keratoses in a patient with plaque psoriasis, who had received a high cumulative dosage of UVB light therapy (\mathbf{d}) , palmar pustulosis (\mathbf{e}) and mild exanthema (\mathbf{f}) in a female with nail psoriasis and psoriasis arthritis, who had received a TNF-alpha blocker.

course characterised by flares, continuous development of pustules, often with fever (1, 6, 13). PPP is a very chronic disease of the feet and/or hands characterised by persisting sterile pustules with or without hyperkeratotic dermatitis (1, 14).

Several reports document that biologic therapies with TNF-antagonists may cause *de novo* occurrence or exacerbation of this form of psoriasis (15). Acrodermatitis continua suppurativa is defined by pustular eruptions, initially affecting the tip of the fingers and nails, often affecting the bony structures of the distal phalanxes (1, 6) (Fig. 1). Coexistence of different types of psoriasis may occur. PsA can be associated with each of them, but frequency and type of the arthritic component might differ, *e.g.* PPP may be more often associated with SAPHO syndromic sternoclavicular and sternomanubrial tenderness and pain (16).

Table II. Examples of relevant clinical severity scores for plaque psoriasis and their items.

	Erythema	Desqua- mation	Infiltration	BSA	Psychosocial impact	History of the illness and treatment	Calculated by
PASI	+	+	+	+	-	-	Physician
BSA	-	-	-	+	-	-	Physician
PGA	+	+	+	-	-	-	Physician
LS/PGA	+	+	+	+			Physician
SPI	+	+	+	+	+	+	Physician
saSPI	+	+	+	+	+	+	Patient
proSPI	+	+	+	+	+	+	Physician
SAPASI	+	+	+	+	-	-	Patient

PASI: Psoriasis Area and Severity Index; BSA: Body Surface Area; PGA: Physicians Global Assessment; LS: Laatice System Physician's Global Assessment; SPI: Salford Psoriasis Index Simplified Psoriasis Index; saSPI: self-assessment Simplified Psoriasis Index; proSPI: professional Simplified Psoriasis Index; SAPASI: Self Administered Psoriasis Area Severity Index. [adapted from Puzenat *et al.* 2012 (24)].

Plaque psoriasis: assessment of disease severity

In psoriasis initiation of therapy and monitoring of the therapeutic effectiveness are based largely on evaluation of the disease severity. Therefore, scoring of skin severity is a necessity in psoriasis care. More than 44 different scoring systems were used in 171 randomised clinical trials of psoriasis therapies between 1997 and 2000 (17). Common tools to score psoriasis include determination of the area involved in relation to the whole body surface (Body Surface Area, BSA) (18, 19), the Physician Global Assessment (19) and the Psoriasis Area and Severity Index (PASI), which was constructed by Frederiksson and Pettersson (19, 20) in order to assess the severity of PV. The PASI score includes a number of welldefined dermatological parameters, e.g. skin redness and infiltration corresponding to the inflammatory component of the disease, and allows for scoring of skin area involvement (see below).

The Salford Psoriasis Index (SPI) is derived from combining a converted figure of the PASI, a second score indicating psychosocial disability, and a third score based on historical information (21). Chularojanamontri et al. recently published a modified version of the SPI renamed to Simplified Psoriasis Index (22). However, no single instrument captures all dimensions of psoriasis severity (23). Puzena et al. selected six relevant clinical severity scores (PASI, BSA, PGA, LS-PGA, SPI and SAPASI) (Table II) and compared their methodological validations and quality (24). They conclude that the PASI is the most thoroughly validated score and can be recommended for quantitative evaluation of clinical severity of psoriasis. This conclusion is in agreement with the recommendations of current consensus guidelines for the management of psoriasis (8, 25).

The Psoriasis Area and Severity Index (PASI):

everyday clinical practice

For calculation of the PASI four main body areas are assessed: the head (h), the trunk (t), the upper extremities (u) and the lower extremities (1), corresponding to 10, 20, 30 and 40% of the total body area, respectively (Fig. 3). The area of psoriatic involvement of these four main areas (Ah, At, Au and Al) is given a numerical value: 0 = noinvolvement; 1 = <10%; 2 = 10<30%; 3 = 30 < 50; 4 = 50 < 70%; 5 = 70 < 90%,and 6 = 90-100%. To evaluate the severity of the psoriatic lesions three target symptoms, namely erythema (E), infiltration (I), and desquamation (D) are assessed according to a scale 0-4, where 0 means a complete lack of cutaneous involvement and 4 represents the severest possible involvement. The severity rating for the three main target symptoms is multiplied with the numerical value of the areas involved and with the various percentages of the four body areas. These values are then added to obtain the PASI. The formula can be written as follows (20):

 $\begin{array}{l} PASI = 0.1 \ x \ Ah \ x \ (Eh + Ih + Dh) + 0.3 \ x \\ At \ x \ (Et + It + Dt) + 0.2 \ x \ Au \ x \ (Eu + Iu + Du) \\ + 0.4 \ x \ Al \ x \ (El + Il + Dl) \end{array}$

The index varies in steps of 0.1 units from 0.0 to 72.0 (20); and there are multiple online tools for PASI training and computing (Table III). As such, the

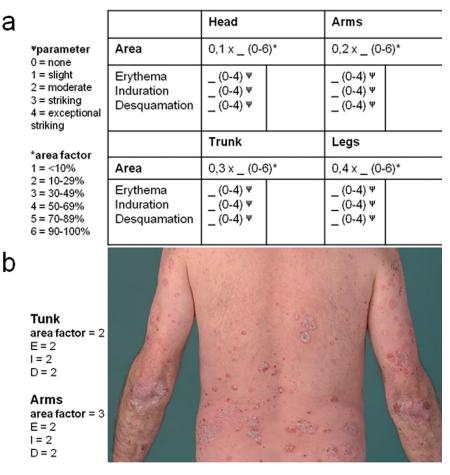


Fig. 3. Assessment of disease severity in psoriasis vulgaris: PASI scheme and calculation; the neck is assessed together with the heat; buttocks are assessed with the legs (a). Example of a patient with plaque psoriasis (arms and trunk); the total sum of the PASI of this patient was 15.3 (b).

index can be used in everyday clinical practice to manage patients with plaque psoriasis, in particular, if a systemic treatment is considered (23, 25). However, the PASI is not applicable for GPP or PPP and does not specifically consider the severity of nail involvement (11, 26).

Scoring of nail psoriasis

Nail psoriasis has a substantial impact on patients' quality of life (11, 26). Several scoring systems have been proposed to assess nail psoriasis severity, *e.g.* the NAPSI (Nail Psoriasis Severity Index) has often been used in clinical studies (27) (see elsewhere in this supplement). For therapy monitoring of nail psoriasis we recommend regular photo-documentation and evaluation of the dynamic Physician Global Assessment (dynamic PGA) of the nails, *e.g.* as a 5-point ordinal rating ranging from "clear" to "very severe" (11, 19).

General aspects of skin assessment

General aspects of skin examination often influence the treatment options and strategies, e.g. skin dryness may increase pruritic symptoms of psoriasis, so that systemic therapy should be combined with regular moisturising. For first-line treatment of plaque psoriasis, a high number of melanocytic nevi may be regarded a contraindication for UV treatment. Moreover, skin tumours or a history of these may represent a relative contraindication for immunosuppressive therapies. Hence, dermatological monitoring is necessary before and during treatment (1, 8). Patients with PV or PsA under treatment with biologic agents should be observed for the development of other immune-mediated skin diseases, e.g. lichen planus, vitiligo, alopecia areata, and of course certain drug eruptions should be seen by the dermatologist (Table I, Fig. 2).

Dermatologic evaluation includes assessment of health-related quality of life and comorbidity

The assessment of psoriasis severity should take into account its burden on health-related quality of life (HR-QoL) (23). As mentioned above, the PASI is insufficient to assess functional disability secondary to specific localisations of skin lesions, e.g. on the face, hands or nails. Moreover, patients with psoriasis often suffer from pruritus, cutaneous pain, burning sensations, bleeding, and/ or social-life impairment (23, 25). In clinical practice evaluation of all these symptoms might be perceived as cumbersome; however, validated scales assessing the burden of plaque psoriasis on HR-QoL are the Dermatology Life Quality Index (DLOI) (28), the Short-Form 36 (SF-36) and the Skindex 29 and Skindex 17 (23) that may be completed by the patients in 3–15 minutes. The DLQI consists of 10 questions concerning symptoms and feelings, daily activities, leisure, work, and school, personal relationships and treatment. All questions relate "to the last week", and the score ranges from 0 (no impairment of life quality) to 30 (maximum impairment). The tool has been translated into at least 21 different languages. There is a children's version of the DLQI (29), the Children's Dermatology Life Quality Index (CDLQI), and a text and cartoon version of this has been described (30) (Table III).

A definition of the different scores of the DLQI and their impact on patients' life allows a reliable grading of the impact on quality of life (31). By using this definition in psoriasis, a DLQI <5 would indicate only mild impact on an individual patients' quality of life (23, 25, 32).

Comorbidity

From the dermatological perspective, PsA is the most important comorbidity; and of course its diagnosis and treatment should not be delayed. The initial dermatological assessment of patients with psoriasis or PsA should include a concise query, whether there is a history of specific gastrointestinal, hepatic, renal, endocrinologic, neurologic, psychiatric, orthopaedic or other rheumaTable III. Internet resources for the assessment and management of skin psoriasis*.

PASI

http://www.pasitraining.com/calculator/step_1.php http://pasi.corti.li
http://www.dermnetnz.org/scaly/pasi.html
DLQI
http://www.cardiff.ac.uk/dermatology/quality-of-life
http://www.pasitraining.com/dlqi
Dermatological guidelines for the management of patients with psoriasis
http://www.ncbi.nlm.nih.gov/pubmedhealth
http://www.awmf.org/leitlinien

*last accessed 6-9-2015.

tologic diseases, neoplasm or allergy (Table I). From epidemiologic studies it has been well established that severe psoriasis is associated significantly with a moderate increase of risk for diabetes and obesity (5, 33). A recent population-based Swedish register study showed that mild and severe psoriasis are associated with increased mortality rates as patients with severe psoriasis die on average 2.6 years younger than age-, sex-, and residency-matched control subjects. The increases in all-cause mortality observed were largely attributed acutely to increased cardiovascular mortality (34). For this reason, new dermatological guidelines do not only refer to the skin, but also recommend to determine the Body Mass Index (BMI) (upper limit: 30 kg/m²) and/or waist circumference (upper limit: 94 cm in men, 80 cm in women) in patients with moderate to severe psoriasis (23).

Combination of skin assessment tools and therapy algorithm

A recent consensus program for the treatment of plaque psoriasis defined a number of important items related to psoriasis assessment and therapy (8, 25). Current guidelines distinguish between "mild" and "moderate to severe" psoriasis, but, as discussed above, a single assessment tool for disease severity is not sufficient to reflect all clinical situation. For plaque psoriasis the following definitions have been consented (25, 32):

Definition of "mild" plaque psoriasis BSA \leq 10 and PASI \leq 10 and DLQI \leq 10

Definition of "moderate-to-severe" plaque psoriasis

BSA >10 and PASI >10 and DLQI >10 The presence of the following disease manifestations may have a substantial impact on the dynamic or static Physician Global Assessment (PGA) (19), which can alter the classification of mild disease to moderate-to-severe disease (23):

- involvement of visible areas
- involvement of major parts of the scalp
- involvement of genitals
- involvement of palms and/or soles
- onycholysis or onychodystrophy of at least two fingernails
- pruritus leading to scratching
- presence of single recalcitrant plaques

Definition of treatment effectiveness The reduction in PASI of \geq 75% (Δ PASI ≥75) has been considered to indicate treatment success after an antipsoriatic treatment has been initiated (8, 25). Clinical studies on the effectiveness of systemic therapy in plaque psoriasis may note that a certain proportion of patients experienced a 75% reduction in their PASI scores over a 3-months treatment period and report this as a percentage of people achieving "PASI 75". With the development of new and highly effective biologic agents, treatment goals are being newly defined as Δ PASI 90 or even Δ PASI 100 (35).

In patients who have a Δ PASI of \geq 50% but <75%, the impact of the disease on quality of life, which can be estimated from the DLQI, may be of value to decide either to continue or modify a treatment regimen. Therefore, it is advisable to assess the DLQI in patients with skin psoriasis before initiation of a systemic treatment and during the follow-up visits. For nail psoriasis, the DLQI may be used in combination with the dynamic PGA of the nails (11).

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Conclusion

Assessment of the skin in patients with psoriasis is based on quantitative scoring tools. The Psoriasis Area and Severity Index (PASI) is the best validated score for defining disease severity in psoriasis vulgaris (23). New treatment goals are defined as Δ PASI 90 (90%) clearing of skin lesion) or Δ PASI 100 (disease free skin). According to the current dermatological guidelines, calculation of PASI should be combined with the Body Surface Area (BSA), as well as the Dermatology Life Quality Index (DLQI) (25). The static or dynamic Physician's Global Assessment (PGA) should be estimated and may be highly important if special disease manifestations exist, e.g. nails are involved (11). However, these scoring systems do not replace the dermatologic clinical examination and medical history of the patient, i.e. assessment of PASI, DLQI or PGA alone is not sufficient. The skin should be examined for additional skin diseases, which may occur in patients with psoriasis. Moreover, patients should be monitored for comorbidity beyond the skin, most importantly PsA (4) and cardiovascular comorbidity (5). Since the coexistence of PsA and skin psoriasis is about 20-30%, an early diagnosis of a progressive PsA is needed. Management of the patient with psoriasis clearly is the crystallising point of a rheumatologic-dermatologic team work.

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