Psoriasis

5

Kanchana Mallawaarachchi

5.1 Plaque Psoriasis

Epidemiology

The worldwide point prevalence of psoriasis among adult population is 2–3% (Burden et al. 2016). The prevalence of psoriasis among Sri Lankan population is over 0.4%, and 8.3% have reported a positive family history (Gunawardena et al. 1978). There are two peaks of age of onset of psoriasis. The first peak occurs around 16–22 years of age (Type 1 psoriasis), while the second one is between 57–62 years of age (Type 2 psoriasis). Both men and women are equally affected.

Pathophysiology

Genetics. There is strong evidence to suggest the genetic influence in the pathogenesis of psoriasis. Lifetime risk for psoriasis if both parents are affected is as high as 65% (Burden et al. 2016).

Environmental Factors. A number of environmental factors are associated with inducing and exacerbating psoriasis in genetically susceptible individuals.

Infections. Streptococcal tonsillitis can trigger psoriasis, particularly guttate psoriasis. HIV may exacerbate existing psoriasis.

Medications. Lithium, antimalarials, interferon-α and TNF-α could exacerbate psoriasis. In addition, β-blockers, nonsteroidal anti-inflammatory drugs and angiotensin-converting enzyme inhibitors are also mentioned in small case series and case reports.

Alcohol Misuse and Cigarette Smoking. There is well-established evidence that excessive alcohol intake is associated with psoriasis. Alcohol increases the risk of severe psoriasis, anxiety and depression and cardiovascular morbidity in patients with psoriasis. Similarly, Cigarette smoking increases the risk of developing psoriasis. These individuals develop more severe disease and are also at a higher risk of developing psoriatic arthritis.

Psychological Distress. Psychological distress may cause flair up of their disease. Also, moderate to severe psoriasis results in increased level of depression and anxiety.

Sunshine. Sunshine is beneficial for most patients with psoriasis. However, 5–20% may report exacerbation of their disease with increased sun exposure.

Trauma. Psoriasis demonstrate Koebner phenomenon in which lesions occur in sites of old scars and cutaneous trauma.

Clinical Features

Pruritus, although not as severe as that in eczema, is the dominant symptom in most patients. Scaling can be extensive especially in scalp psoriasis. Burning pain may occur in certain types of

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psoriasis such as pustular psoriasis, erythrodermic psoriasis and flexural psoriasis.

Psoriasis belongs to the group of papulosquamous diseases, and plaque psoriasis is the commonest presentation. Plaques are usually erythematous on the fair skin (Figs. 5.1, 5.2, 5.3). However, erythema may not be so prominent on the coloured skin where the plaques become more hyperpigmented in appearance (Figs. 5.4, 5.5). Plaques are well demarcated from the surrounding skin and usually covered with an adherent silvery scale. The number of the plaques, which is distributed symmetrically mostly on the extensor surfaces of the skin, can be varied. The size ranges from one to several centimetres in diameter. The shape of the plaque, which is usually oval or round, may vary depending on the clinical subtype (Figs. 5.6, 5.7). Annular lesions develop due to gradual clearance of the centre of the plaque, and linear lesions may develop due to Koebner phenomenon (Fig. 5.8). The plaque usually resolves leaving a hypopigmented or hyperpigmented patch.

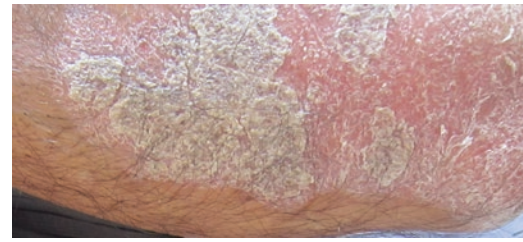

Fig. 5.1 Erythematous well-demarcated scaly plaque is characteristic of psoriasis (photographed by Dr. Kanchana Mallawaarachchi)

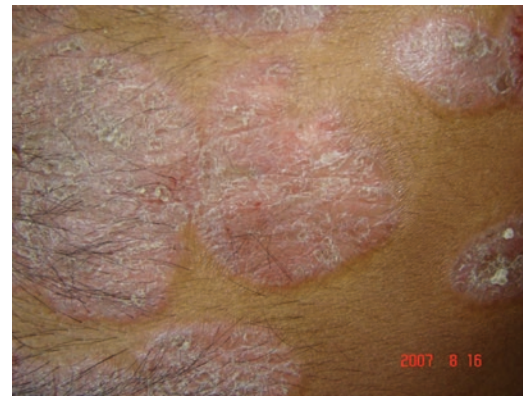

Fig. 5.2 Erythematous plaques with mild silvery scales (photographed by Dr. Ranthilaka R. Ranawaka)

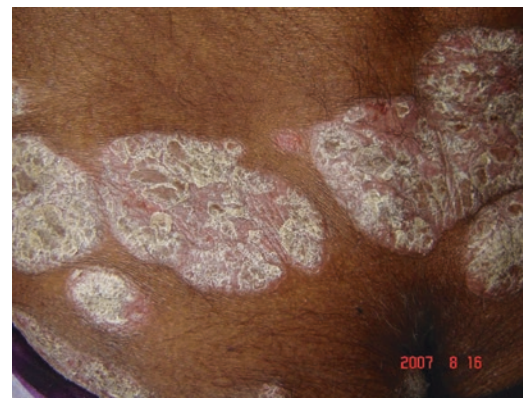

Fig. 5.3 Psoriatic plaques with thick silvery scales (photographed by Dr. Ranthilaka R. Ranawaka)

5.1.1 Plaque Psoriasis Affecting Specific Sites

5.1.1.1 Scalp Psoriasis

Scalp psoriasis is better felt than seen. Multiple scaly erythematous plaques develop within the hair-bearing area. The plaques may cross the hair line and extend a short distance beyond. Hair loss is not a usual feature in chronic plaque psoriasis of the scalp (Figs. 5.9, 5.10, 5.11 and 5.12).

5.1.1.2 Flexural Psoriasis

Flexural psoriasis, which is also known as inverse psoriasis, usually present with psoriasis elsewhere on the skin. However, it may pose a diagnostic difficulty when presents in isolation. Typical sites involved are the axillae, groin area, napkin area in infants, gluteal cleft, umbilicus

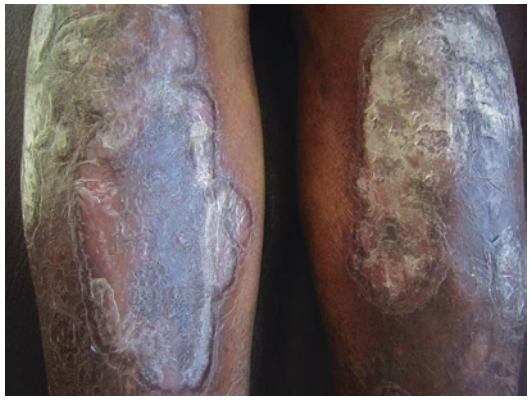

Fig. 5.4 Silvery coloured scales adherent on a rather hyperpigmented plaque (photographed by Dr. Kanchana Mallawaarachchi)

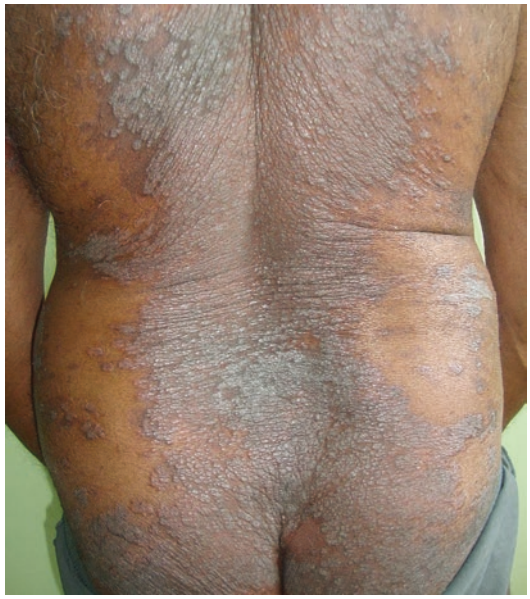

Fig. 5.5 Psoriatic plaques can be hyperpigmented and erythema is not apparent in dark skin. Therefore, PASI (Psoriasis Area Severity Index) score may be inaccurate in pigmented skin (photographed by Dr. Ranthilaka R. Ranawaka)

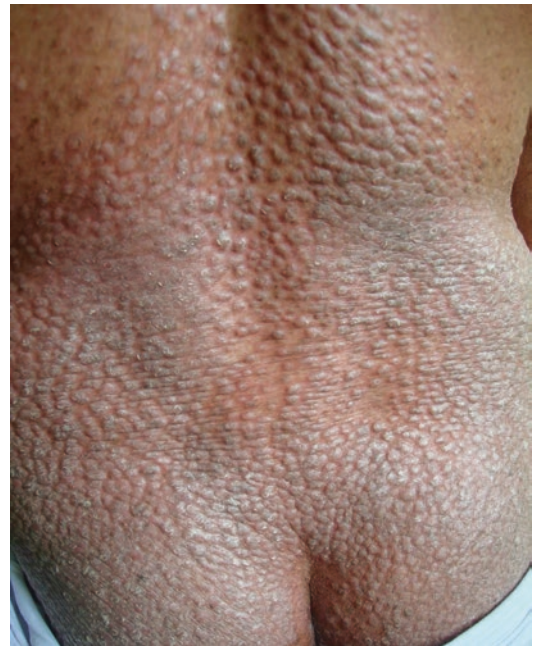

Fig. 5.6 Follicular psoriasis (psoriasis around hair follicle openings) (photographed by Dr. Ranthilaka R. Ranawaka)

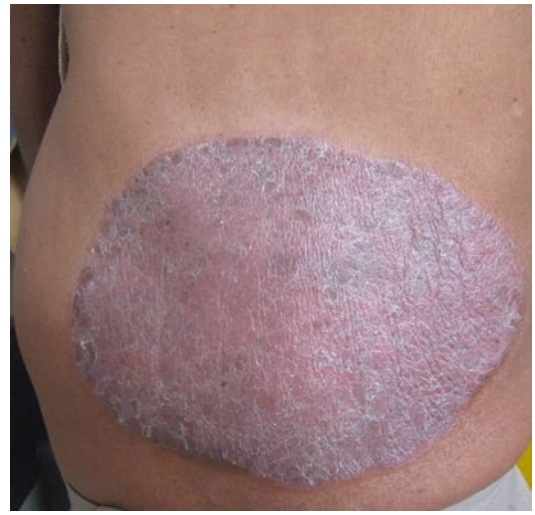

Fig. 5.7 A large psoriatic plaque on the trunk (photographed by Dr. Kanchana Mallawaarachchi)

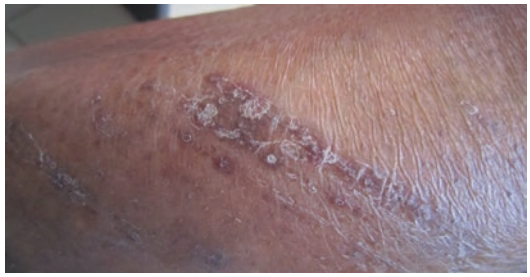

Fig. 5.8 Koebner phenomenon (photographed by Dr. Kanchana Mallawaarachchi)

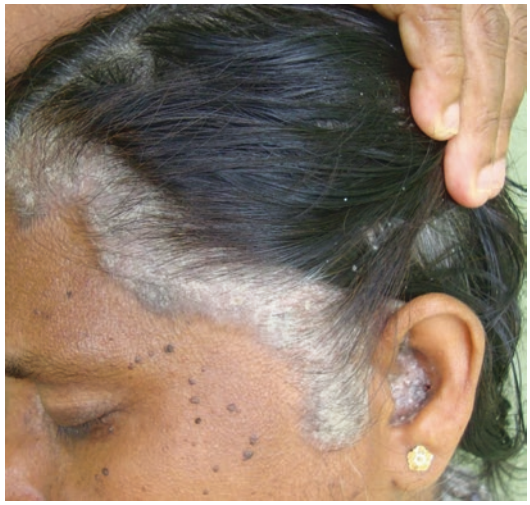

Fig. 5.9 Scalp psoriasis. Note well-demarcated thick scaly plaques extending a short distance beyond the hairline (photographed by Dr. Ranthilaka R. Ranawaka)

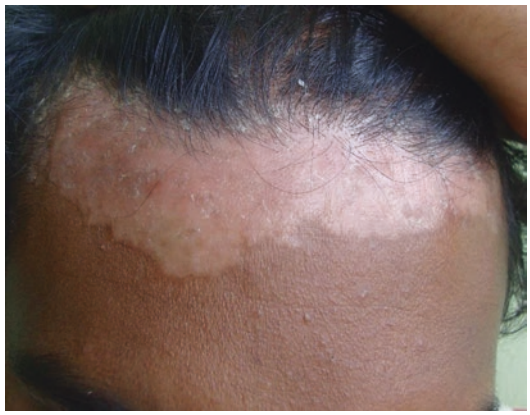

Fig. 5.10 Scalp psoriasis. Erythematous well-demarcated lesion extending beyond the hairline. Note scales are mild in this patient (photographed by Dr. Ranthilaka R. Ranawaka)

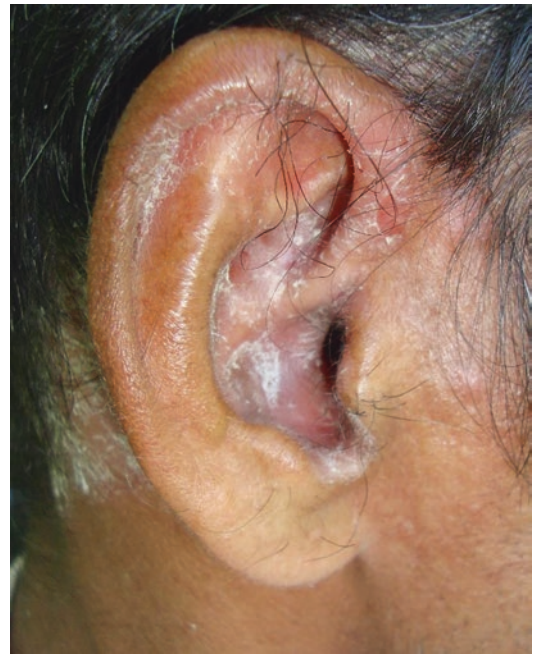

Fig. 5.11 A patient with scalp psoriasis having external ear involvement (photographed by Dr. Ranthilaka R. Ranawaka)

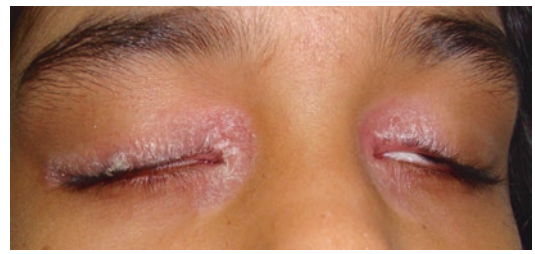

Fig. 5.12 Psoriasis around the eyes (photographed by Dr. Ranthilaka R. Ranawaka)

and submammary folds. Plaques are well defined and erythematous but may lack scaling. Maceration, fissuring and secondary infections may be seen due to their location within the skin folds (Figs. 5.13, 5.14, 5.15 and 5.16).

5.1.1.3 Nail Psoriasis

The prevalence of nail involvement among patients with psoriasis ranges from 50% to 80% (Bardazzi et al. 2019). This is higher among those with psoriatic arthritis and may represent poor prognosis. Nail psoriasis affect both nail matrix

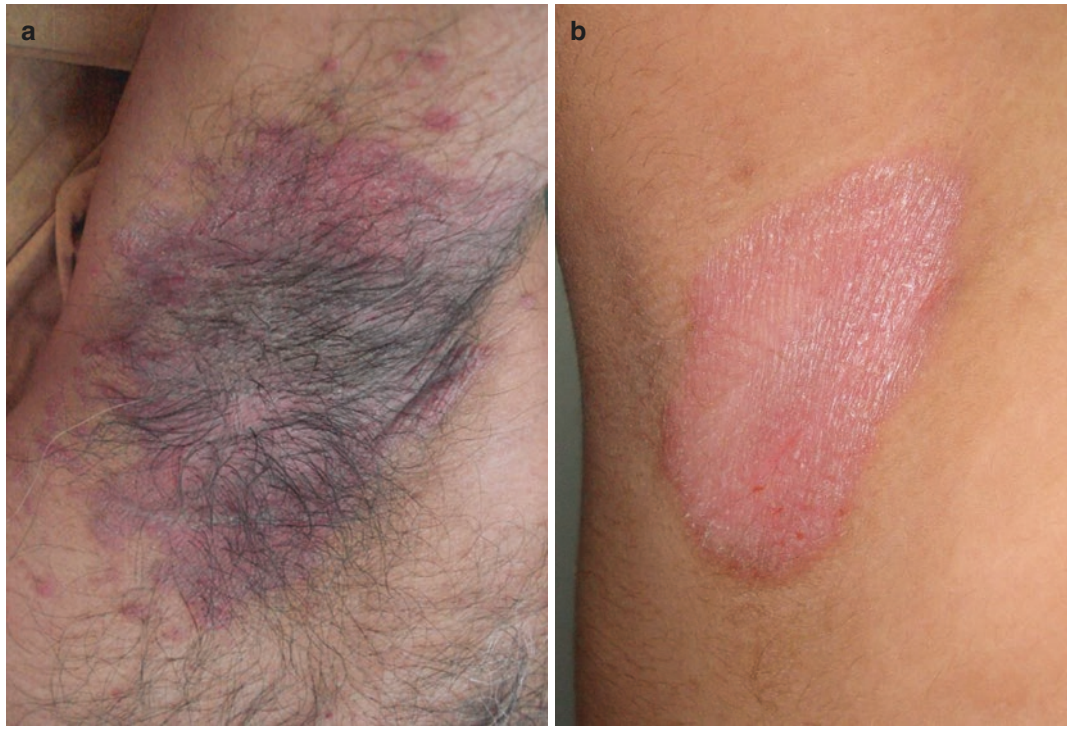

Fig. 5.13 (**a**, **b**) Flexural psoriasis in the axillae. Note plaques are thin and scaling is minimal, but the edges are well defined and retain its characteristic colour (photographed by Dr. Kanchana Mallawaarachchi and Dr. Ranthilaka R. Ranawaka)

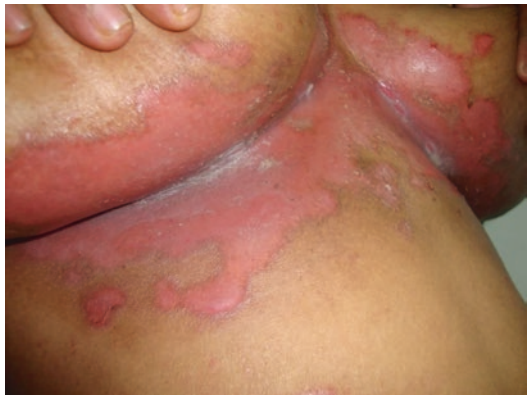

Fig. 5.14 Submammary flexural psoriasis. Flexural plaques are thin, and scaling is absent, but edges are well defined and retain its characteristic erythematous colour. Note the fissuring of the folds and secondary candida infection which leads to itching and burning sensation (photographed by Dr. Ranthilaka R. Ranawaka)

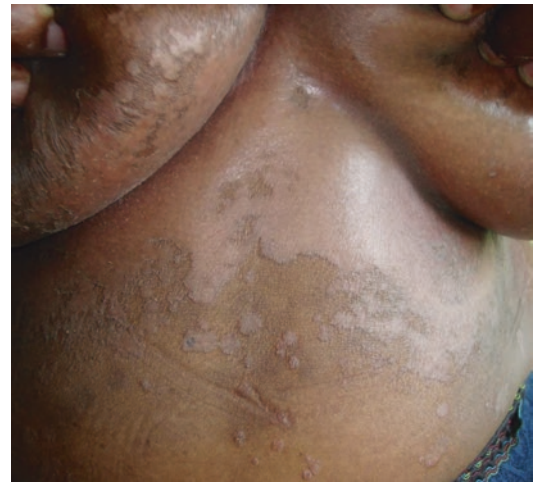

Fig. 5.15 Submammary flexural psoriasis. Plaques are well defined, and the surface has a glazed hue, but the erythema may be minimal in dark skin as in this patient (photographed by Dr. Ranthilaka R. Ranawaka)

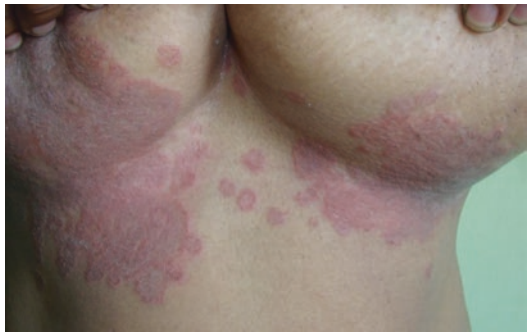

Fig. 5.16 Submammary tinea infection. Differentiating from tinea infection may be difficult if patient has co-existing psoriasis. Well-defined erythematous lesions. But note the characteristic raised inflammatory edge in tinea infection (photographed by Dr. Ranthilaka R. Ranawaka)

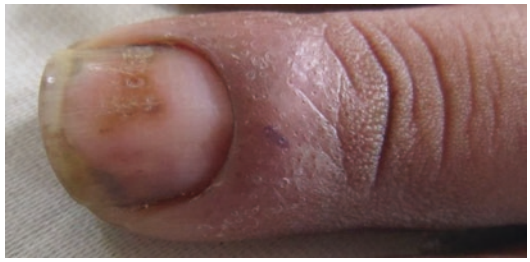

Fig. 5.17 Oil drop sign and coarse pitting in nail psoriasis (photographed by Dr. Kanchana Mallawaarachchi)

and nail bed. Nail matrix involvement is represented by coarse pitting, ridging and grooves of the nail plate, whereas nail bed involvement is characterised by 'oil drop sign', subungual hyperkeratosis and onycholysis (Figs. 5.17, 5.18, 5.19 and 5.20).

5.1.1.4 Seborrheic Psoriasis

Seborrheic psoriasis, which is also known as sebopsoriasis, demonstrates well-demarcated erythematous plaques over seborrheic areas of the skin. Seborrheic psoriasis characteristically follows the distribution pattern of seborrheic dermatitis and usually occurs in patients with plaque psoriasis elsewhere on the skin. When occurs in isolation, it may be difficult to differentiate from seborrheic dermatitis.

5.1.1.5 Genital Psoriasis

Genital psoriasis is usually associated with classical plaque psoriasis or more frequently with flexural

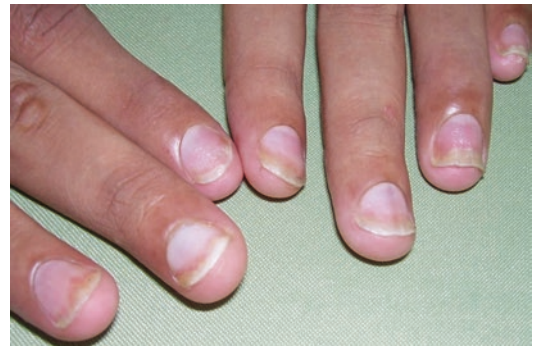

Fig. 5.18 Coarse pitting of the nail plate and distal onycholysis (photographed by Dr. Kanchana Mallawaarachchi)

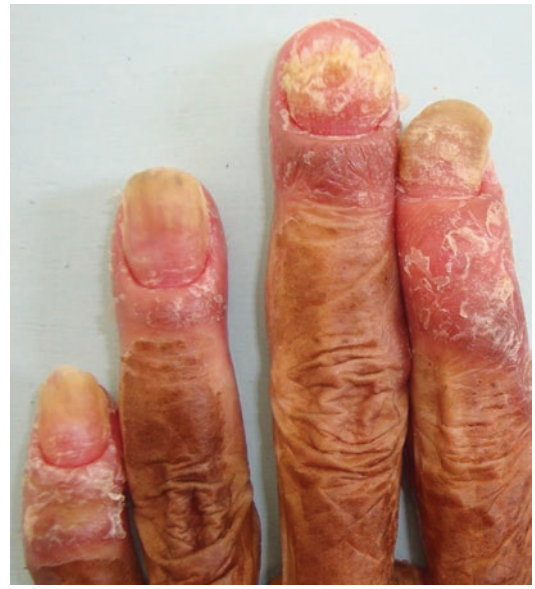

Fig. 5.19 Subungual hyperkeratosis in nail psoriasis (photographed by Dr. Ranthilaka R. Ranawaka)

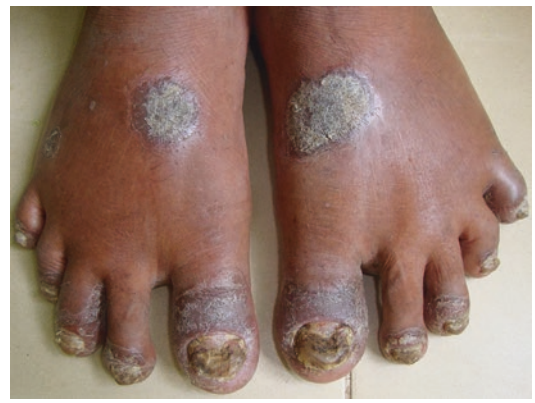

Fig. 5.20 Subungual hyperkeratosis and total onychodystrophy in nail psoriasis (photographed by Dr. Ranthilaka R. Ranawaka)

Fig. 5.21 Genital psoriasis. Symmetrical, erythematous, non-scaly, well-demarcated thin plaques of psoriasis affecting the labia majora and adjacent skin (photographed by Dr. Ranthilaka R. Ranawaka)

psoriasis. When genital lesions are the only manifestation of psoriasis, a diagnostic biopsy may be required. It may be symptomatic with pruritis being the dominant symptom in women and causes significant degree of sexual dysfunction in both sexes. In men, glans penis is most frequently affected, while scrotal skin and the penile shaft may also be involved. Plaques are usually well-defined and erythematous, but scaling is not seen in uncircumscribed men. Symmetrical, erythematous plaques with minimum scaling over labia majora is the usual presentation in women (Fig. 5.21).

5.1.1.6 Non-pustular Palmoplantar Psoriasis

Psoriasis presents with typical characteristics on palms and soles. Sharply defined margins and absence of vesicles may be helpful in distinguishing palmoplantar psoriasis from eczema. On the dorsum hands, the knuckles frequently get affected. Nail changes usually occur concurrently (Figs. 5.22, 5.23, 5.24 and 5.25).

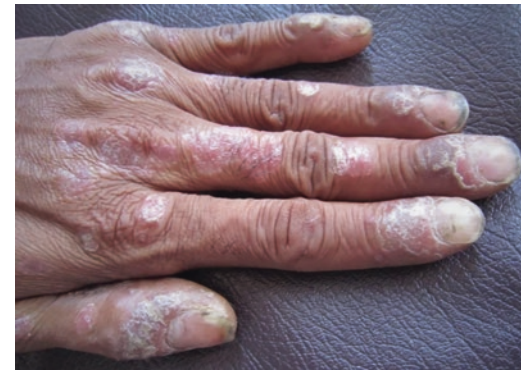

Fig. 5.22 Non-pustular palmoplantar psoriasis. On the dorsum hands, the knuckles frequently get affected (photographed by Dr. Kanchana Mallawaarachchi)

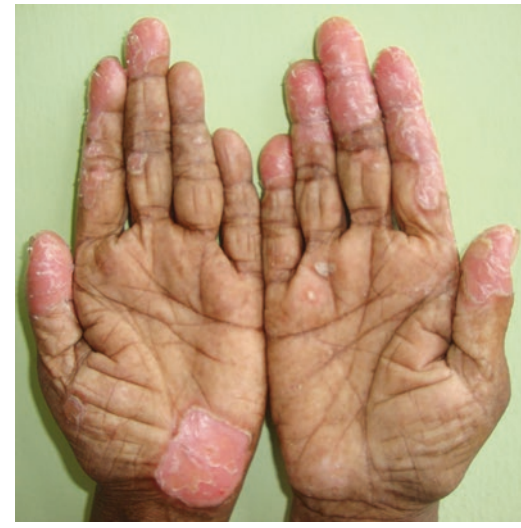

Fig. 5.23 Non-pustular palmoplantar psoriasis. Sharply defined margins and absence of vesicles are helpful in distinguishing palmoplantar psoriasis form eczema (photographed by Dr. Ranthilaka R. Ranawaka)

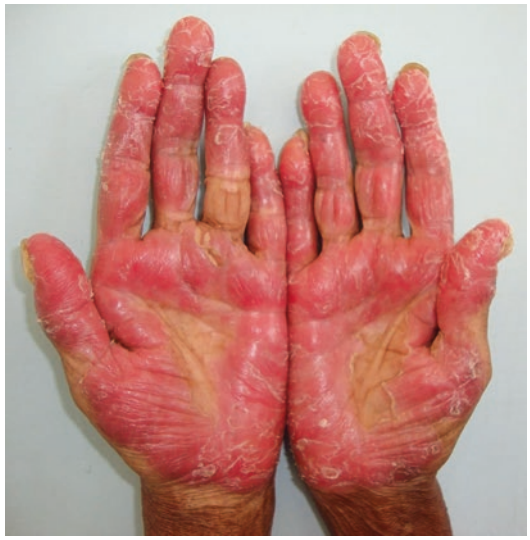

Fig. 5.24 Non-pustular palmoplantar psoriasis. Sharply defined margins and absence of vesicles are helpful diagnostic features (photographed by Dr. Ranthilaka R. Ranawaka)

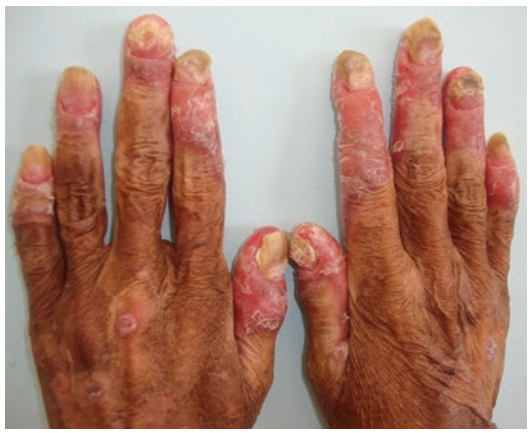

Fig. 5.25 Non-pustular palmoplantar psoriasis. Nail changes usually occur concurrently (photographed by Dr. Ranthilaka R. Ranawaka)

5.1.2 Clinical Variants of Plaque Psoriasis

5.1.2.1 Guttate Psoriasis

Guttate psoriasis, which is commoner among children and young adults than among adult population, usually presents as a sudden onset skin eruption where crops of small lesions appear on the body. The prevalence of guttate psoriasis is variable, and a Japanese population study found it to be 3.2% of all cases of psoriasis (Ito et al. 2018). The prevalence of guttate psoriasis among psoriatic patients in Malaysia is 2.9% (Affandi et al. 2018). Guttate psoriasis frequently follows an episode of pharyngitis due to Group A streptococci.

The size of an individual lesion ranges from few millimetres to 1 centimetre, and they are distributed mostly on the trunk and the proximal limbs. The scaly lesions are round or oval in shape. Lesions usually resolve within 3 months, but few individuals progress into develop chronic plaque psoriasis later in life (Figs. 5.26 and 5.27).

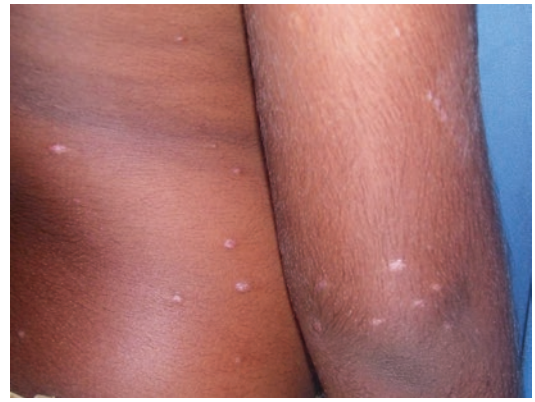

Fig. 5.26 Guttate psoriasis, distribution on the trunk and proximal limbs (photographed by Dr. Kanchana Mallawaarachchi)

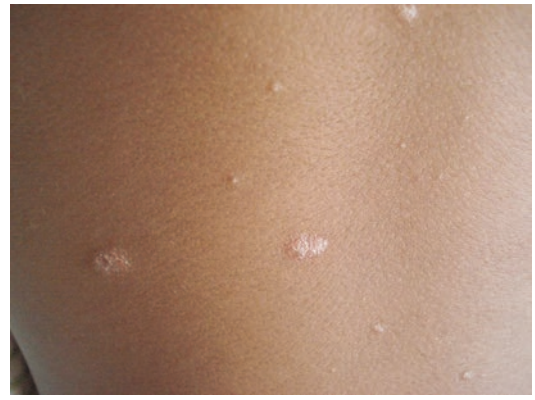

Fig. 5.27 Oval-shaped scaly plaques of guttate psoriasis (photographed by Dr. Kanchana Mallawaarachchi)

5.1.2.2 Erythrodermic Psoriasis

Erythrodermic psoriasis occurs when more than 90% of the skin is covered by psoriasis. It may result from the extension and spread of stable psoriatic plaques or more acutely due to unstable psoriasis which develop and spread within a short period of time. When happen due to stable plaque psoriasis, the process is chronic, and psoriasis lesions retain typical characteristics of psoriasis. The patient is systemically well. Erythrodermic psoriasis due to unstable psoriasis results in a systemically ill patient with fever and significant amount of itching. It is difficult to identify areas of uninvolved skin, and typical features of psoriasis are often lost. This flare-up may precipitate due to various factors such as irritating topical applications like coal tar and salicylic acid, sudden withdrawal of oral steroids, methotrexate and ciclosporin, alcohol abuse and systemic disease. Erythrodermic psoriasis can lead to skin failure and its complications (Fig. 5.28).

5.1.2.3 Linear and Segmental Psoriasis

Linear and segmental psoriatic lesions are rare. They are usually unilateral or Blaschko linear. Being a polygenic disease, psoriasis may present with either isolated linear lesions or with segmental lesions superimposed sequentially or concurrently in a patient with chronic plaque psoriasis. The isolated linear lesions occur due to postzygotic new mutation of an otherwise healthy embryo, while superimposed segmental psoriasis is due to postzygotic loss of wild-type allele in a heterozygotic embryo (Happle 2007) (Figs. 5.29 and 5.30).

5.1.3 Histopathology of Plaque Psoriasis

The epidermis shows parakeratosis, focal orthokeratosis and absence of the granular layer. Collections of neutrophils in the stratum corneum known as Munro microabscesses and those in the spinous layer known as spongiform pustules of Kogoj are seen. Rete ridges are elongated and branched along with suprapapillary thinning. Prominent blood vessels with neutrophilic and mononuclear cell infiltrates are seen in the papillary dermis.

5.1.4 Management of Plaque Psoriasis

Psoriasis poses various problems in the management which can be unique to the individual patient. Therefore, the management strategy

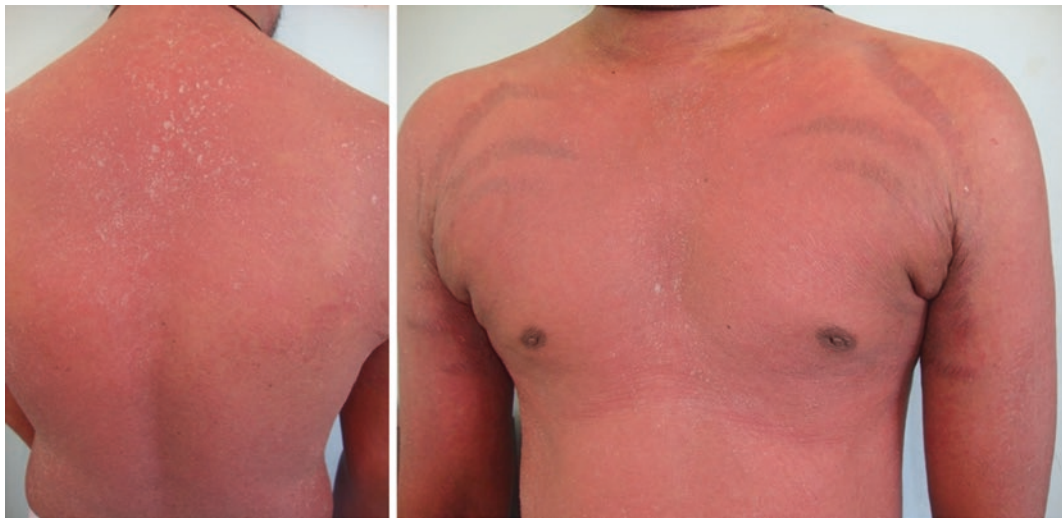

Fig. 5.28 Striae and erythrodermic psoriasis in a 32-year-old man who had been on long-term unsupervised oral prednisone for his widespread psoriasis (photographed by Dr. Ranthilaka R. Ranawaka)

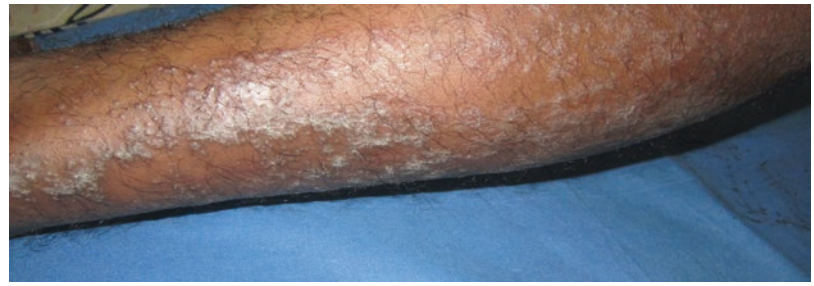

Fig. 5.29 Isolated linear lesions (photographed by Dr. Kanchana Mallawaarachchi)

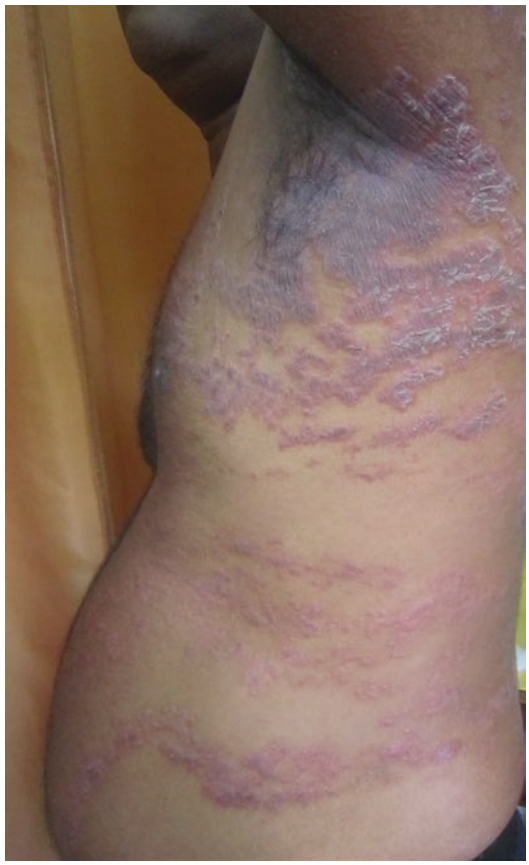

Fig. 5.30 Classic psoriatic plaques with superimposed Blaschko linear lesions (photographed by Dr. Kanchana Mallawaarachchi)

needs to be individualised, and attention must be paid to patients' general physical, social and psychological wellbeing. Associated comorbidities need to be assessed, and advice from the multidisciplinary team should be sought.

Topical treatments are the mainstay of treatment in patients with limited plaque psoriasis. They are still useful in treating patients with severe psoriasis, when used in association with systemic agents. Topical corticosteroids are used as the main topical agent in the management of psoriasis. It can be used to treat any subtype including unstable and erythrodermic psoriasis. Potent and very potent topical steroids are used in treating the scalp, palms and soles, while milder topical steroids are used when treating the face and the flexural skin. Topical steroids can be used in combination with other keratolytic agents such as coal tar and salicylic acid. Vitamin D analogues, calcitriol, calcipotriol and tacalcitol, are effective topical agents. The combination of topical steroid and calcipotriol is a very effective topical treatment for psoriasis. Dithranol, which has some antimitotic properties, may still use in the management of psoriasis.

Narrowband ultraviolet B therapy is used as a first-line treatment in moderate to severe psoriasis and is known to offer superior efficacy and low toxicity than PUVA. Systemic therapy is offered for patients with severe psoriasis and for those with psoriasis causing a significant impact on their lives. However, this should be a combined decision of the patient and the treating clinician. Methotrexate, ciclosporin, acitretin and biologics are the most frequently used systemic agents. Constant monitoring and close supervision are needed in using systemic agents in the management of psoriasis as toxicity of these drugs can be life-threatening (Fig. 5.31). Following guidelines in using these medicines is helpful in achieving maximum possible disease control and avoiding hazardous side effects.

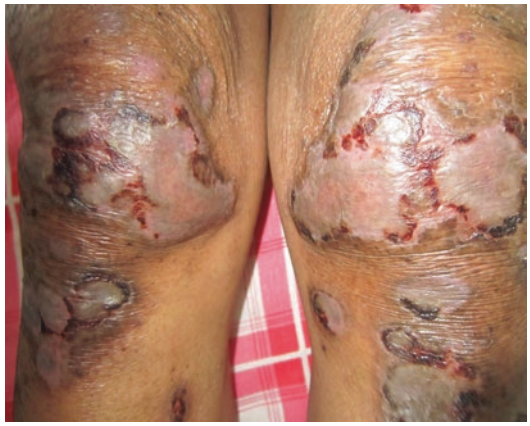

Fig. 5.31 Ulceration of psoriatic plaques in a patient with methotrexate toxicity (photographed by Dr. Kanchana Mallawaarachchi)

5.2 Pustular Psoriasis

Pustular psoriasis is now considered as a separate group under the psoriasis spectrum of diseases due to differences in the phenotype, genetics, pathogenesis and response to the treatment when compared with psoriasis vulgaris. The European Rare and Severe Psoriasis Expert Network (ERASPEN) classify pustular psoriasis into generalised pustular psoriasis, acrodermatitis continua of Hallopeau and palmoplantar pustulosis (Navarini et al. 2017). However, pustular psoriasis should be differentiated from the pustules that appear within the plaques of unstable plaque psoriasis.

5.2.1 Generalised Pustular Psoriasis

Generalised pustular psoriasis (GPP) is characterised by widespread eruption of sterile pustules on non-acral sites of the body. It may or may not be associated with systemic inflammation and usually has more than one episode or persists for more than 3 months. Some patients may have plaque psoriasis before or after the episode of GPP, but most patients have pustulosis as the sole presentation. It is a rare condition with peak age of onset between 40 and 60 years of age (Burden et al. 2016).

Precipitating factors: Certain environmental factors may precipitate or exacerbate GPP. These include systemic infections, certain topical and systemic medications, sudden withdrawal of systemic corticosteroids, pregnancy and hypocalcaemia.

Histology: Histology reveals significant papillary oedema, spongiosis, and infiltration of neutrophils with spongiform pustule formation of the sub-corneal tissue. In addition, histopathological features of psoriasis such as elongated rete ridges, parakeratosis and acanthosis may be seen.

Clinical presentation: Patients usually present with abrupt onset waves of pustules on inflamed erythematous skin. Sensation of burning of the skin and tenderness are signs that the patient is progressing into an unstable form. Fever and malaise are usually associated with the eruption, and inflammatory markers are usually elevated. If the patient has pre-existing plaque psoriasis, they become severely inflamed, erythematous and studded with pustules. At the same time, pustules develop in uninvolved skin and the flexures. A similar condition that occurs during pregnancy is known as impetigo herpetiformis, and a subacute annular form is also seen (Figs. 5.32, 5.33, 5.34, 5.35 and 5.36).

Treatments: Most cases need systemic therapy, and acitretin is usually the first line (Burden et al. 2016). High-dose ciclosporin and methotrexate are also effective, while biologics are considered as second-line treatment. Prednisolone may be used especially during the pregnancy, but withdrawal should always be covered by another systemic agent to prevent major relapse. Few case reports of hydroxyurea being effective are also found in the literature (Fonseka et al. 2008).

5.2.2 Palmoplantar Pustulosis

Palmoplantar pustulosis (PPP) is a chronic persistent condition which tends to be resistant to most of the treatment. PPP is the commonest of three pustular psoriasis subtypes and typically occurs in isolation (Navarini et al. 2017). The association between plaque psoriasis and PPP

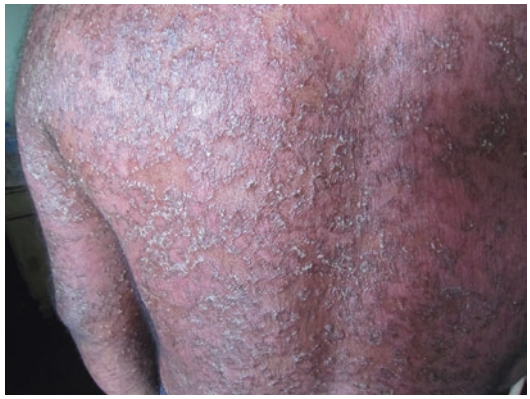

Fig. 5.32 Lakes of pustules in GPP (photographed by Dr. Kanchana Mallawaarachchi)

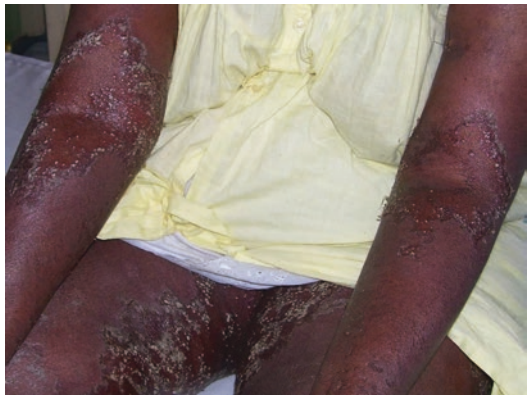

Fig. 5.33 Flexural involvement in GPP (photographed by Dr. Kanchana Mallawaarachchi)

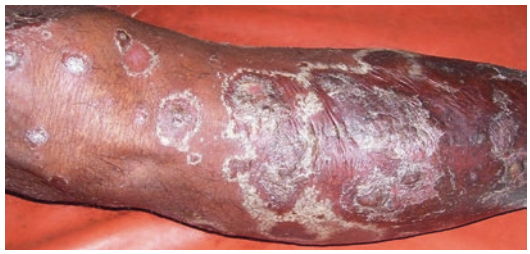

Fig. 5.34 Pustules developing within the existing plaques (photographed by Dr. Kanchana Mallawaarachchi)

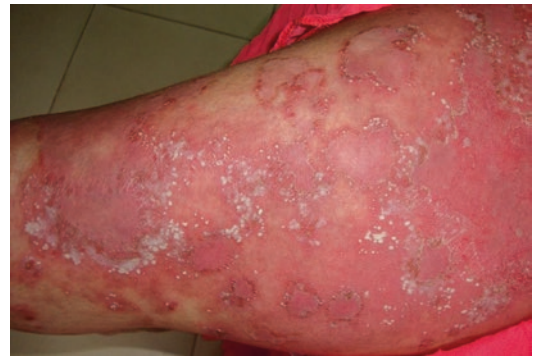

Fig. 5.35 Abrupt onset waves of pustules on inflamed erythematous skin (photographed by Dr. Ranthilaka R. Ranawaka)

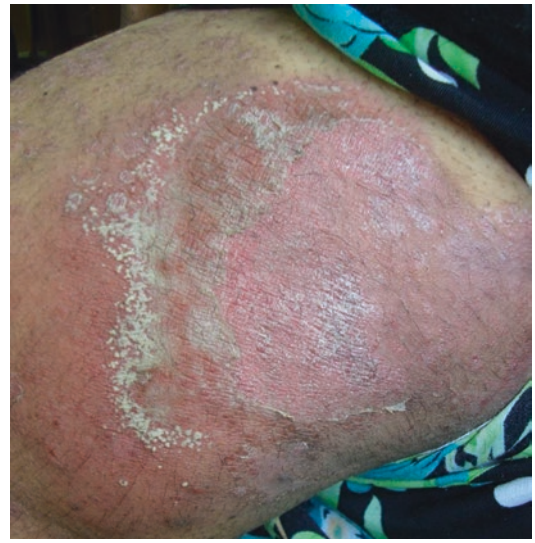

Fig. 5.36 Generalised pustular psoriasis showing waves of pustules on inflamed erythematous skin (photographed by Dr. Ranthilaka R. Ranawaka)

can vary between 18% and 24% (Burden et al. 2016). However, differences in the morphology, genetics and response to treatments have led clinicians to consider PPP as a distinct entity. The global prevalence is around 0.01–0.05%, while a Japanese study reveals a slightly higher prevalence of 0.12% (Kubota et al. 2015). The peak age of onset is between 30 and 50 years, and it is more common in women.

Clinical presentation: PPP usually starts with one or few well-demarcated erythematous plaques studded with pustules. Scaling and fissuring can also appear later. It progresses to become symmetrical involvement of the hand and/or feet. Thenar eminence is affected more

than other areas of the hand, while instep, medial and lateral borders and back of the heel are frequently affected areas of the foot. Plaques on the dorsal aspects and digits are less frequent (Figs. 5.37 and 5.38).

Differential diagnosis: Tinea and eczema are the usual differential diagnosis. Tinea is mostly asymmetrical and unilateral, and toe webs are usually involved. Eczema with secondary infection can closely mimic PPP, but it is usually painful, and contact dermatitis of the feet does not affect the instep.

Treatments: Super potent topical steroids are the first-line treatment. Acitretin, ciclosporin and oral PUVA are also effective. Methotrexate, alitretinoin and biologics are used in resistant cases.

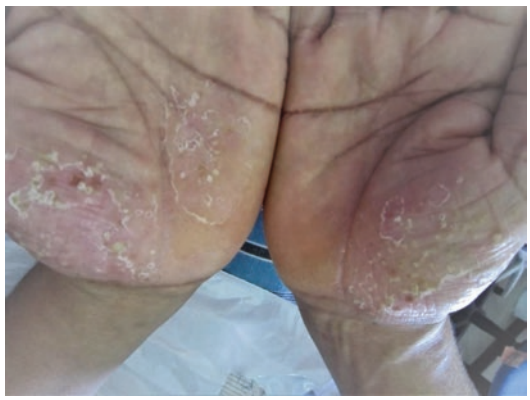

Fig. 5.37 Palmoplantar pustulosis (photographed by Dr. Kanchana Mallawaarachchi)

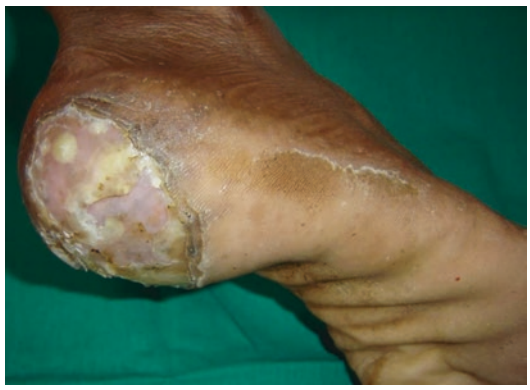

Fig. 5.38 Palmoplantar pustulosis (photographed by Dr. Ranthilaka R. Ranawaka)

5.2.3 Acrodermatitis Continua of Hallopeau

Acrodermatitis continua of Hallopeau (ACH) is a rare form of localised pustular psoriasis. It characteristically affects fingertips and nails destructively and tends to be resistant to treatment and chronic. It is common in middle-aged females (Benjegerdes et al. 2016).

Clinical features: ACH initially affects the finger. The changes usually start at the fingertip which demonstrates erythematous scaly eruption with pustules. Gradual involvement of nail folds and nail bed results in nail destruction. ACH gradually progresses proximally on the finger and eventually involves other fingers and toes. Complete destruction of the nail plate, bony involvement and tapering and wasting of the distal phalanx are seen in the advanced disease (Figs. 5.39 and 5.40).

Treatments: ACH can be refractory to treatment. Potent topical steroids, tacrolimus and calcipotriol are used as topical applications. Acitretin, ciclosporin and biologics are used in the management of refractory cases.

5.3 Psoriatic Arthritis

Psoriatic arthritis is a seronegative arthritis which is included under the spondyloarthropathy spectrum (Coates and Helliwell 2017). It affects nearly 40% of patients with moderate to severe psoriasis (Burden et al. 2016). Psoriatic arthritis comes under the scope of rheumatologists, but dermatologists who manage psoriatic patients on daily basis should be able to identify symptoms and signs as early as possible and refer patients to relevant specialities accordingly. The CASPAR (Classification Criteria for Psoriatic Arthritis) criteria guide clinicians in diagnosing psoriatic arthritis (Taylor et al. 2006). However, a lengthy discussion on psoriatic arthritis is beyond the scope of this book (Figs. 5.41 and 5.42).

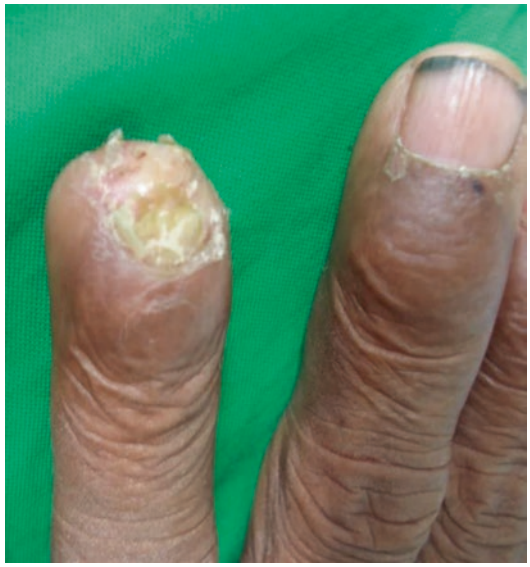

Fig. 5.39 Acrodermatitis continua of Hallopeau (photographed by Dr. Chathurarya Siriwardena, Consultant Dermatologist, General Hospital Nuwaraeliya, Sri Lanka)

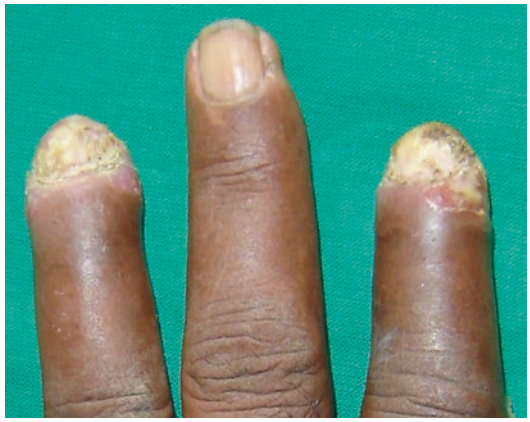

Fig. 5.40 Acrodermatitis continua of Hallopeau (photographed by Dr. Ranthilaka R. Ranawaka)

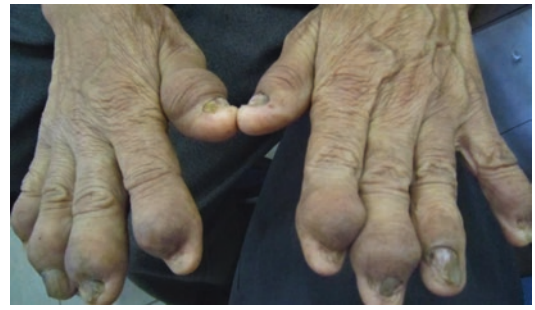

Fig. 5.41 Deforming psoriatic arthritis with nail changes (photographed by Dr. Kanchana Mallawaarachchi)

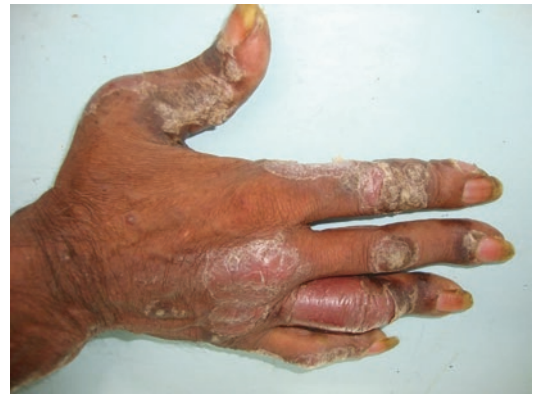

Fig. 5.42 Psoriatic arthritis. Dactylitis (sausage fingers) and swollen, tender joints (photographed by Dr. Ranthilaka R. Ranawaka)

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