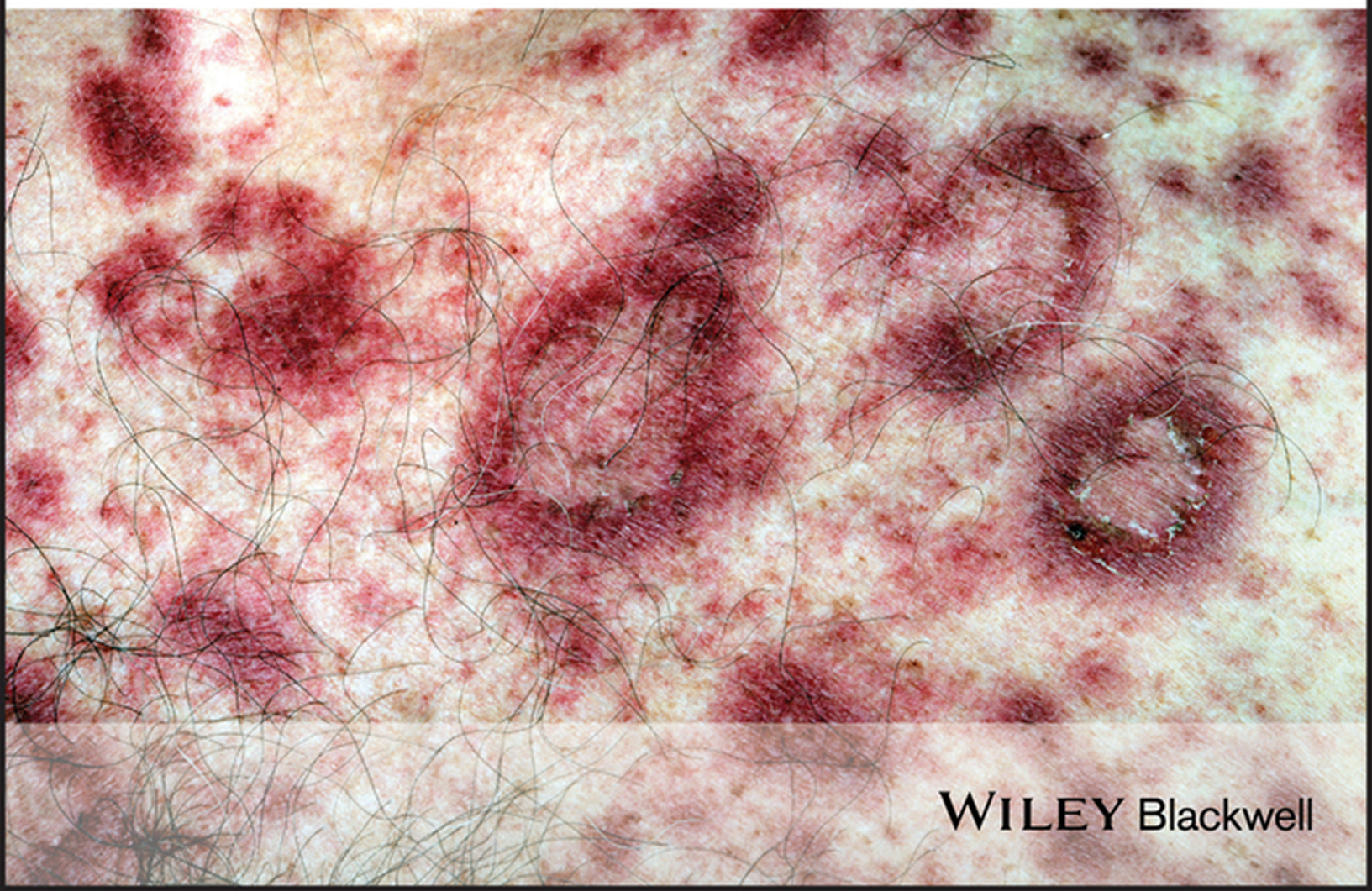
ABC of Dermatology

SEVENTH EDITION

Edited by Rachael Morris-Jones



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Seventh Edition

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Preface

The 7th edition of the *ABC of Dermatology* incorporates all the latest scientific advances in genetics, pathophysiology, and management strategies whilst at the same time remaining a practical clinical approach to dermatology. There are also additional new chapters to reflect current learning needs around cosmetic dermatology procedures and genital dermatology. The emphasis as always is to produce a valuable resource for any medical and nurse practitioner who is diagnosing and managing skin disease.

As well as a wholly practical approach to clinical dermatology, the 7th edition gives insights into the latest thinking around the pathophysiological processes that explain the characteristic features of skin disease and the current approach to the management of skin disease, including the newer biological agents for treating inflammatory disease and tumours.

The fascination of dermatology lies partly in the visual nature of discipline but also in one's ability to diagnose systemic disease through examination of the skin surface. Manifestations of underlying disease can form specific patterns in the skin which in some instances are pathognomonic. However, where there is any diagnostic doubt a simple skin biopsy for histopathology/immunohistochemistry and/or culture is fantastically helpful in most cases.

For those working in resource-poor settings there may be little access to modern investigations for skin disease patients and therefore the clinical diagnosis will be the benchmark on which skin disease is managed. To this end the 7th edition is full of clinical photographs eliciting the appearances of skin disease in a multitude of different pigmented skin tones and ethnic groups. Descriptions of skin management include simple and relatively cheap interventions as well as sophisticated cutting-edge immunotherapies.

On a global scale the number of people with access to the internet via computers or mobile devices is increasing at a rapid pace. This enables them access to a multitude of resources including those related to the diagnosis and management of human disease. Many patients are increasingly using the internet to attempt to self-diagnose ('Doctor Google') their own skin conditions, identify any known underlying causes and gain some insights into the possible treatment strategies. An informed patient can be hugely beneficial to everyone involved in the provision of healthcare. However, at times this can lead to patients becoming overly anxious or misinformed. There is an increasing use of teledermatology in many parts of the world where populations are a long distance from a skin specialist, where images of the patient's skin complaint are taken and sent to an expert for a 'virtual opinion'. Mobile phone consultations with a remote doctor are also being seen as a way to meet the increasing demand for GP consultations. However, remote dermatology can be tricky as it may be difficult to examine the patient thoroughly and there is no way of feeling the skin texture and induration of rashes/lesions. Remote dermatology can be immensely helpful, but ultimately the gold standard for accurate diagnosis and management of skin disease remains seeing patients in person, preferably by a practitioner with knowledge of skin disease.

We trust the 7th edition of the ABC of Dermatology will not only introduce the reader to a fascinating clinical discipline but will also help them to diagnose and manage skin disease in whichever part of the world they are working.

Rachael Morris-Jones

Acknowledgements

I would very much like to sincerely thank all my co-contributors, whose expertise in specialist areas of dermatology has been invaluable in ensuring that this Seventh Edition is right up to date and written by experts in their field.

Dr Fiona Lewis has written a chapter on genital dermatology, which is a very valuable new edition to the *ABC of Dermatology*; genital disease can be a challenging area even for seasoned dermatologists, so her practical approach to the diagnosis/management of genital disease is hugely welcomed. Cosmetic procedures are increasing globally, with many more women and men undertaking rejuvenation procedures. This area of medicine remains a bit of a mystery to those of us who have only benefited from conventional medical training, so the addition of a chapter dedicated to cosmetic dermatology is fantastically helpful for all of us. Dr Emma Craythorne (alongside her skin cancer surgery work) has expertise in treating scars and performing cosmetic procedures and has written this new chapter on cosmetic procedures to give us an insight into what can be done and what the outcomes and pitfalls might be. Even if we don't practice cosmetic dermatology ourselves, patients may ask for our advice and we may also see patients with complications following procedures, so well informed is hopefully well prepared.

Tissue viability clinical nurse specialist Bernadette Byrne works alongside plastic surgeons, and they have contributed to her wound chapter in this Seventh Edition, so we can understand more about biological dressings, which are being increasingly used in challenging wounds. Bernadette has an impressive depth of knowledge as well as decades of experience managing literally thousands of complex wounds in patients from the outpatient setting to the intensive care unit. Her clinical practical approach is an invaluable guide to wound management in any setting.

Dr Saqib Bashir has taken over writing the chapter on lasers and photodynamic therapy and intense pulsed light from Alun Evans. Saqib has a huge wealth of expertise in using multiple different lasers and light devices. He has great skill in prudently treating patients with a variety of skin tones, and he brings this expertise and his high standards of care to the chapter.

Dr Aisling Ryan is a dedicated dermatology consultant with a wealth of expertise in medical dermatology, specifically in the rapidly expanding field of biological therapies. She has taken over writing the Formulary chapter from Karen Watson. Aisling helps keep us up to date with the new molecular targets for biological therapy, indications, outcomes and adverse events. In the future most of us will all be looking after patients who may be suitable for or already taking biological therapies, so this area is hugely important for all of us to keep us abreast of cutting-edge medicine.

Dr John Ferguson has taken over the Skin and Photosensitivity chapter from me. He has developed an area of expertise in photobiology and had just joined the specialist clinic at St John's Institute of Dermatology. He has hugely enhanced this chapter with his detailed knowledge of how UV light can affect the skin.

A large proportion of the illustrations in the Seventh Edition of the *ABC of Dermatology* comes from King's College Hospital, London, UK. I am indebted to the medical photography department at King's for their very professional, high-quality clinical images, without which this book would be of little use. Many of the images in the hair and scalp, genital and cosmetic chapters have been provided by the St John's Institute of Dermatology, St Thomas' Hospital, London, UK. Dr Stephen Morris-Jones, consultant in Infectious Diseases, University College Hospital, London, UK, provided some of the cutaneous infection images and we have retained some of Dr Barbara Leppard's photographs in the tropical dermatology chapter that she took whilst working in Africa. Bernadette Byrne and the Plastics Team at King's College Hospital, London, UK, have provided the wound chapter photos in addition to some photographs retained from previous editions which come from the Victoria Hospital, Kirkcaldy and Queen Margaret Hospital, Dunfermline, Fife, the Royal Infirmary, Edinburgh and from Paul Buxton's own collection. Dr Jon Salisbury, a consultant histopathologist at King's College Hospital, London, UK, provided all the histopathology images to demonstrate cutaneous disease at the cellular level and Dr Edward Davies, consultant immunologist at King's College Hospital, London, UK, provided the direct immunofluorescence images of the skin in immunobullous disease.

I owe a huge debt of gratitude to all my Dermatology colleagues at King's College Hospital who diagnosed and managed many of the patients you will see in the illustrations in this Seventh Edition. I would specifically like to thank Dr Daniel Creamer, Dr Sarah Walsh, Dr Saqib Bashir and Prof Roderick Hay and Dr Tanya Basu.

I am especially indebted to all the patients for consenting to include their clinical images in the *ABC of Dermatology* to help us to demonstrate the features presenting in a multitude of skin/nail and hair disorders far better than any written description would do.

Dr Rachael Morris-Jones

CHAPTER 1

Introduction

Rachael Morris-Jones

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OVERVIEW

- The clinical features of skin lesions are related to the underlying pathological processes.
- Skin conditions broadly fall into three clinical groups: (i) those with a well-defined appearance and distribution; (ii) those with a characteristic pattern but with a variety of underlying clinical conditions; (iii) those with a variable presentation and no constant association with underlying conditions.
- Skin lesions may be the presenting feature of serious systemic disease, and a significant proportion of skin conditions threaten the health, well-being, and even the life of the patient.
- Clinical descriptive terms such as macule, papule, nodule, plaque, induration, atrophy, bulla, and erythema relate to what is observed at the skin surface and reflect the pathological processes underlying the affected skin.
- The significance of morphology and distribution of skin lesions in different clinical conditions are discussed.

Introduction

The aim of this book is to provide an insight for the non-dermatologist into the pathological processes, diagnosis, and management of skin conditions. Dermatology is a broad specialty, with over 2000 different skin diseases, the most common of which are introduced here. Pattern recognition is key to successful history-taking and examination of the skin by experts, usually without the need for complex investigations. However, for those with less dermatological experience, working from first principles can go a long way in determining the diagnosis and management of patients with less severe skin disease. Although dermatology is a clinically oriented subject, an understanding of the cellular changes underlying the skin disease can give helpful insights into the pathological processes. This understanding aids the interpretation of clinical signs and overall management of cutaneous disease. Skin biopsies can be a useful adjuvant to reaching a diagnosis; however, clinico-pathological correlation is essential in order that interpretation of the clinical and pathological patterns is put into the context of the patient.

Interpretation of clinical signs on the skin in the context of underlying pathological processes is a theme running through the chapters. This helps the reader develop a deeper understanding of the subject and should form some guiding principles that can be used as tools to help assess almost any skin eruption.

Clinically, cutaneous disorders fall into three main groups.

1. Those that generally present with a characteristic distribution and morphology that leads to a specific diagnosis – such as chronic plaque psoriasis (Figure 1.1), basal cell carcinoma, and atopic dermatitis.
2. A characteristic pattern of skin lesions with variable underlying causes – such as erythema nodosum (Figure 1.2) and erythema multiforme.
3. Skin rashes that can be variable in their presentation and/or underlying causes – such as lichen planus and urticaria.

A holistic approach in dermatology is essential as cutaneous eruptions may be the first indicator of an underlying internal disease. Patients may, for example, first present with a photosensitive rash on the face, but deeper probing may reveal symptoms of joint

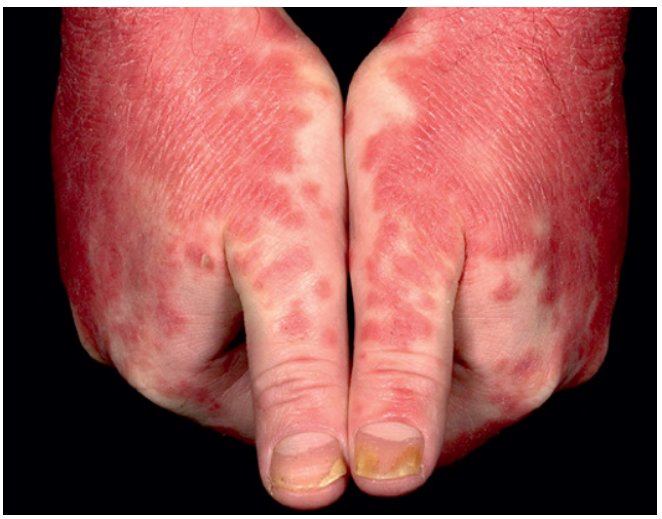

Figure 1.1 Psoriasis with nail changes.

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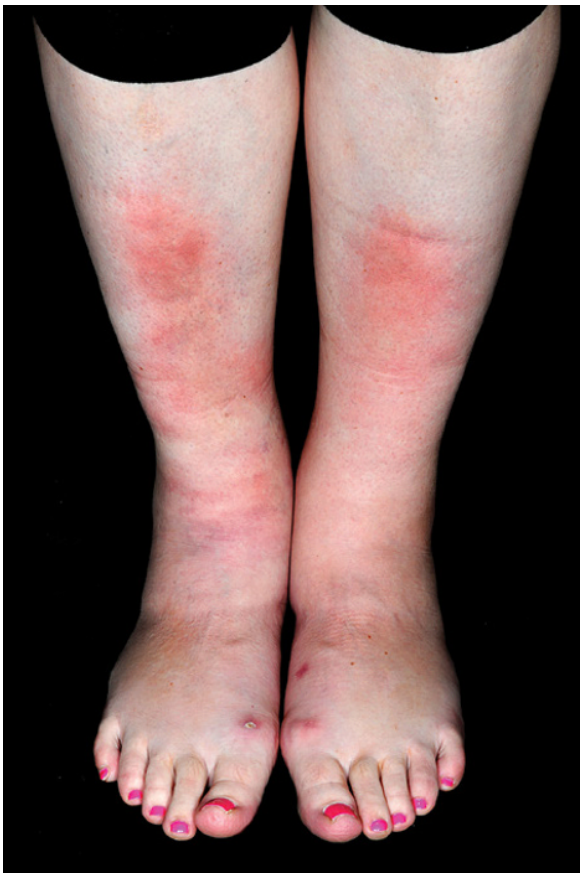


Figure 1.2 Erythema nodosum in pregnancy.

pains etc. leading to the diagnosis of systemic lupus erythematosus. Similarly, a patient with underlying coeliac disease may first present with blistering on the elbows (dermatitis herpetiformis). It is therefore important not only to take a thorough history (Box 1.1) of the skin complaint but in addition to ask about any other symptoms the patient may have and examine the entire patient carefully.

Box 1.1 **Dermatology history-taking**

- Where – site of initial lesion(s) and subsequent distribution
- How long – continuous or intermittent?
- Trend – better or worse?
- Previous episodes – timing? Similar/dissimilar? Other skin conditions?
- Who else – Family members/work colleagues/school friends affected?
- Symptoms – Itching, burning, scaling, or blisters? Any medication or other illnesses?
- Treatment – prescription or over-the-counter? Frequency/time course/compliance?

The significance of skin disease

Seventy per cent of the people in developing countries suffer skin disease at some point in their lives, but of these, 3 billion people in 127 countries do not have access to even basic skin services. In developed countries the prevalence of skin disease is also high; up to 15% of general practice consultations in the United Kingdom are concerned with skin complaints. Many patients never seek medical advice and will use the internet to self- diagnose and self-treat using over-the-counter preparations.

The skin is the largest organ of the body; it provides an essential living biological barrier and is the aspect of ourselves that we present to the outside world. It is therefore not surprising that there is great interest in 'skin care' and 'skin problems', with an associated ever-expanding cosmetics industry and so-called cosmeceuticals. At the other end of the spectrum, impairment of the normal functions of the skin can lead to acute and chronic illness with considerable disability and sometimes the need for hospital treatment.

Malignant change can occur in any cell in the skin, resulting in a wide variety of different tumours, the majority of which are benign. Recognition of typical benign tumours saves the patient unnecessary investigations and the anxiety involved in waiting to see a specialist or waiting for biopsy results. Malignant skin cancers are usually only locally invasive, but distant metastases can occur. It is important therefore to recognise the early features of lesions such as melanoma (Figure 1.3) and squamous cell carcinoma before they disseminate.

Underlying systemic disease can be heralded by changes on the skin surface, the significance of which can be easily missed by the unprepared mind. So, in addition to concentrating on the skin changes, the overall health and demeanour of the patient should be assessed. Close inspection of the whole skin, nails, and mucous membranes should be the basis of routine skin examination. The general physical condition of the patient should also be determined as indicated.

Most skin diseases, however, do not signify any systemic disease and are often considered 'harmless' in medical terms. However, due to the very visual nature of skin disorders, they can cause a great deal of psychological distress, social isolation, and occupational difficulties, which should not be underestimated. A validated measure of how much skin disease affects patients' lives can be made using the Dermatology Life Quality Index (DLQI). A holistic approach to the patient both physically and psychologically is therefore highly desirable.

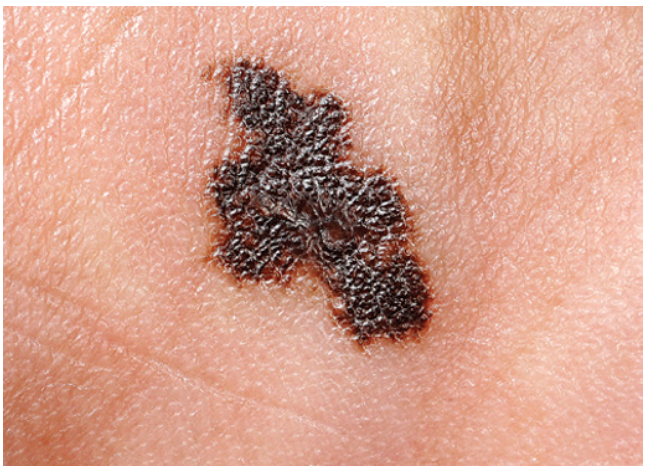
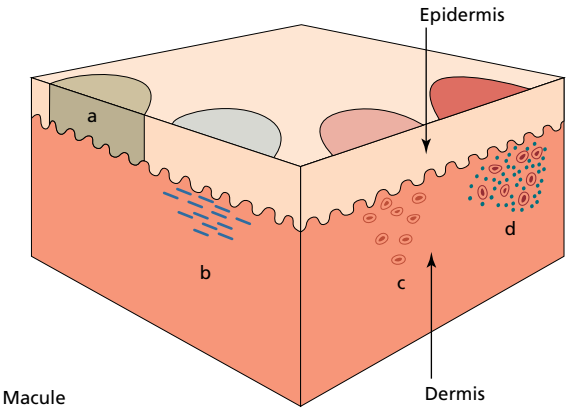


Figure 1.3 Superficial spreading melanoma.

Descriptive terms of clinical inspection

All specialties have their own common terms, and familiarity with a few of those used in dermatology aids with diagnostic skills as they relate to the underlying pathology. The most important are defined below.

- *Macule* (Figure 1.4). Derived from the Latin for a stain, the term *macule* is used to describe changes in colour (Figure 1.5) without any elevation above the surface of the surrounding skin. There may be an increase in pigments such as melanin, giving a black or brown colour. Loss of melanin leads to a white macule. Vascular dilatation and inflammation produce erythema. A macule with a diameter greater than 2 cm is called a *patch*.
- *Papules and nodules* (Figure 1.6). A *papule* is a circumscribed, raised lesion, of epidermal or dermal origin, 0.5–1.0 cm in diameter (Figure 1.7). A *nodule* (Figure 1.8) is similar to a papule but greater than 1.0 cm in diameter. A vascular papule or nodule is known as a *haemangioma*.
- *A plaque* (Figure 1.9) is a circumscribed, superficial, elevated plateau area 1.0–2.0 cm in diameter (Figure 1.10).



Macule
a) Melanin pigment *in* epidermis
b) Melanin pigment *below* epidermis
c) Erythema due to dilated dermal blood vessels
d) Inflammation in dermis

Figure 1.4 Section through skin.

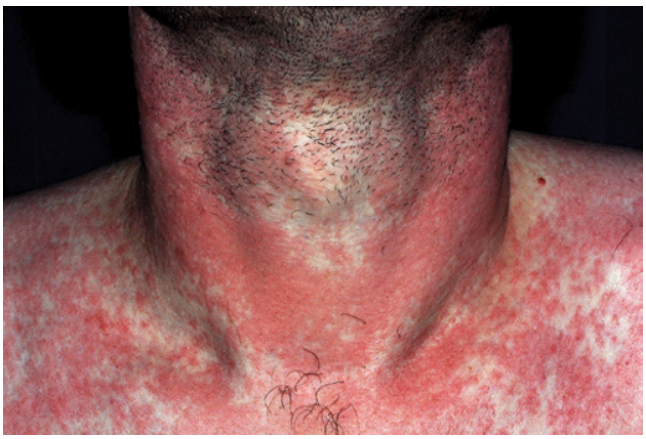


Figure 1.5 Erythema due to a drug reaction.

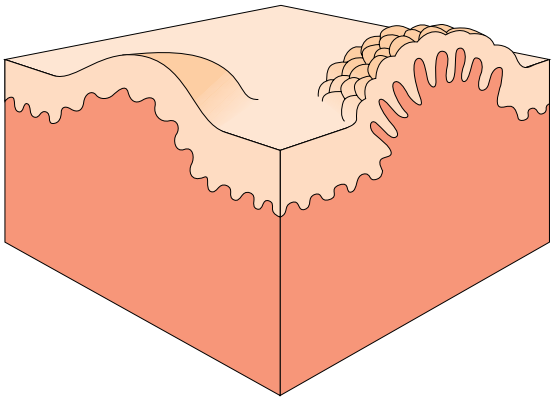


Figure 1.6 Section through skin with a papule.

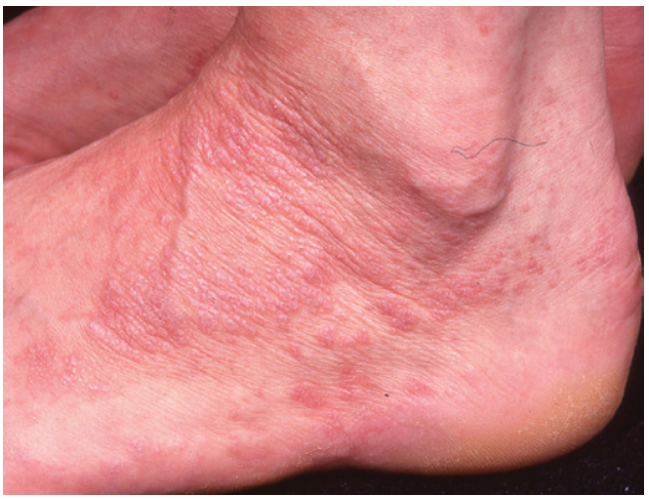


Figure 1.7 Papules in lichen planus.

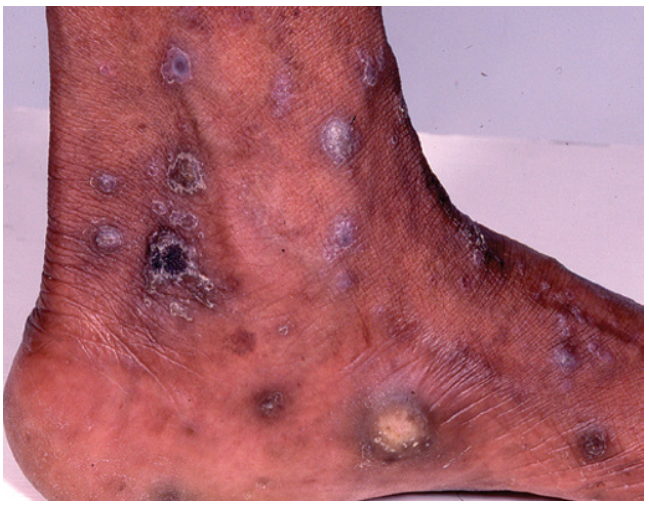


Figure 1.8 Nodules in hypertrophic lichen planus.

- *Vesicles and bullae* (Figure 1.11) are raised lesions that contain clear fluid (blisters) (Figure 1.12). A bulla is a vesicle larger than 0.5 cm. They may be superficial within the epidermis or situated in the dermis below it. The more superficial the vesicles/bullae, the more likely they are to break open.

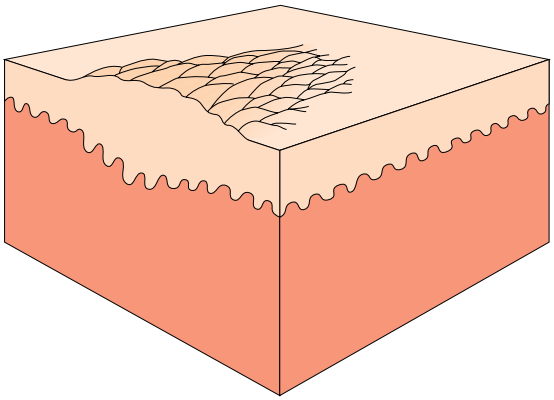


Figure 1.9 Section through skin with plaque.

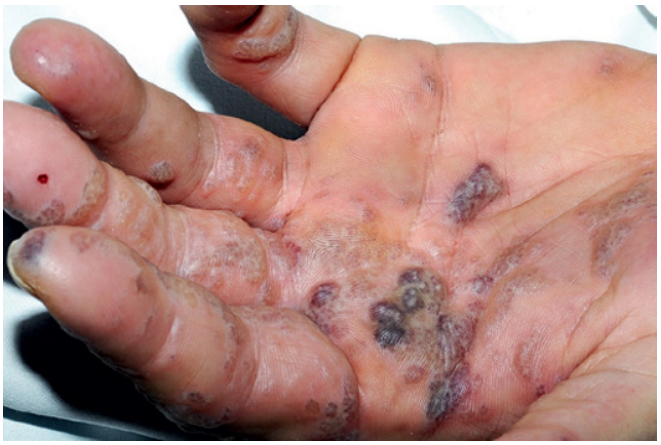


Figure 1.11 Bullae on the palm from multidermatomal shingles.

- *Lichenification* is a hard thickening of the skin with accentuated skin markings (Figure 1.13). It commonly results from chronic inflammation and rubbing of the skin.
- *Discoid lesions*. These are 'coin-shaped' lesions (Figure 1.14).
- *Pustules*. The term *pustule* is applied to lesions containing purulent material – which may be due to infection – or sterile pustules (inflammatory polymorphs) (Figure 1.15) that are seen in pustular psoriasis and pustular drug reactions.
- *Atrophy* refers to loss of tissue, which may affect the epidermis, dermis, or subcutaneous fat. Thinning of the epidermis is characterised by loss of normal skin markings; there may be fine wrinkles, loss of pigment and a translucent appearance (Figure 1.16). In addition, sclerosis of the underlying connective tissue, telangiectasia or evidence of diminished blood supply may be present.
- *Ulceration* results from the loss of the whole thickness of the epidermis and upper dermis (Figure 1.17). Healing results in a scar.
- *Erosion*. An erosion is a superficial loss of epidermis that generally heals without scarring (Figure 1.18).
- *Excoriation* is the partial or complete loss of epidermis as a result of scratching (Figure 1.19).
- *Crusted*. Dry serous fluid forming a crust (underlying epidermis or dermis is usually disrupted) (Figure 1.20).
- *Fissuring*. Fissures are slits through the whole thickness of the skin.
- *Desquamation* is the peeling of superficial scales, often following acute inflammation (Figure 1.21).
- *Annular* lesions are ring-shaped (Figure 1.22).
- *Reticulate*. The term *reticulate* means 'net-like'. It is most commonly seen when the pattern of subcutaneous blood vessels becomes visible (Figure 1.23).

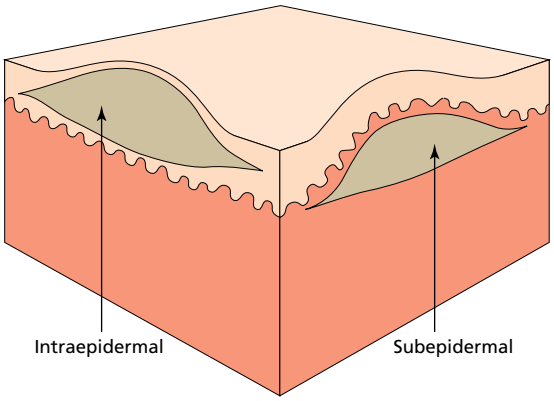


Figure 1.12 Section through skin showing sites of vesicle and bulla.

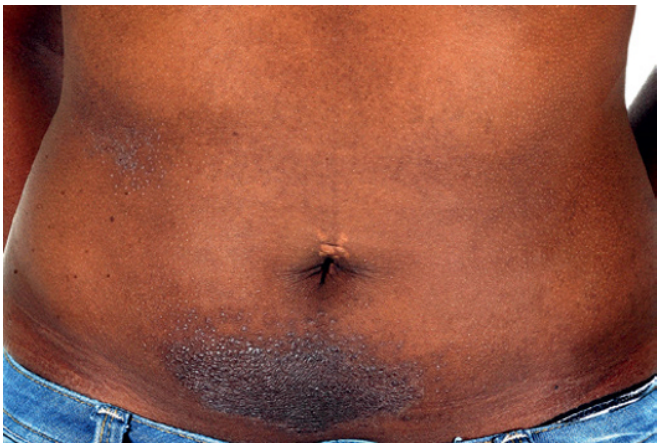


Figure 1.13 Lichenification due to chronic eczema in nickel allergy.

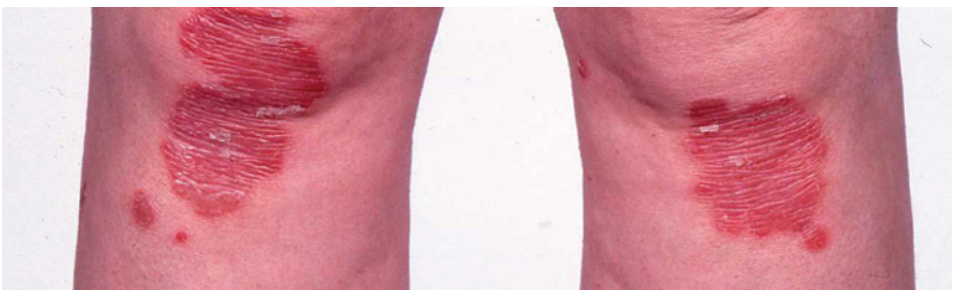


Figure 1.10 Psoriasis plaques on the knees.

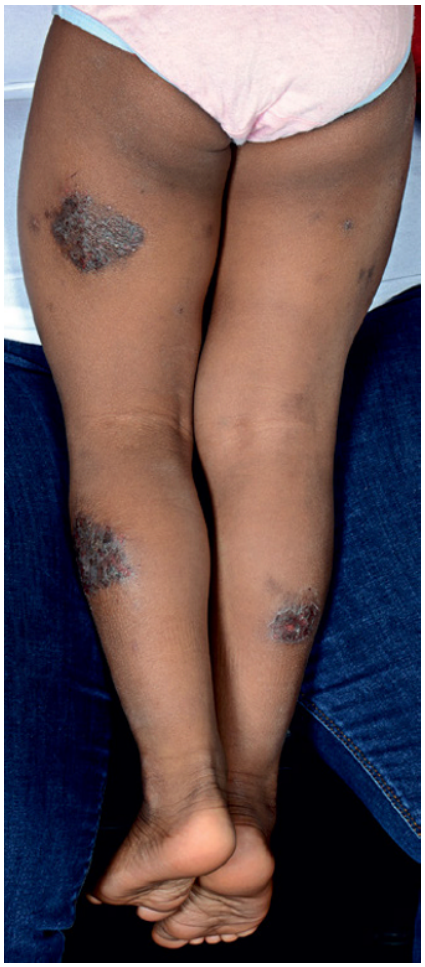


Figure 1.14 Discoid lesions in discoid eczema.

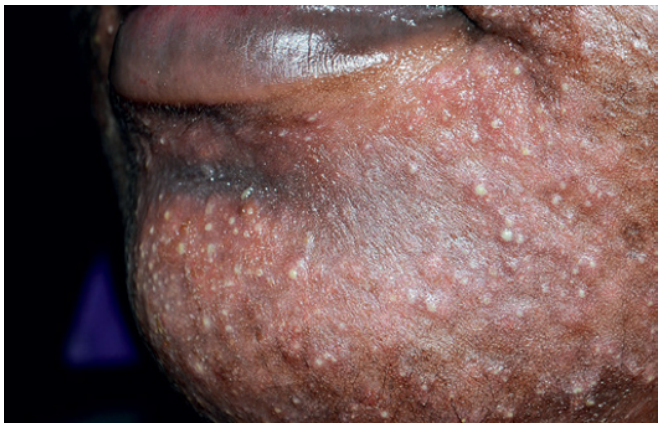


Figure 1.15 Inflammatory pustules secondary to contact dermatitis with Argon oil.

Clinical approach to the diagnosis of rashes

A skin rash generally poses more problems in diagnosis than a single, well-defined skin lesion such as a wart or tumour. As in all branches of medicine, a reasonable diagnosis is more likely to be reached by thinking firstly in terms of broad diagnostic categories rather than specific conditions.

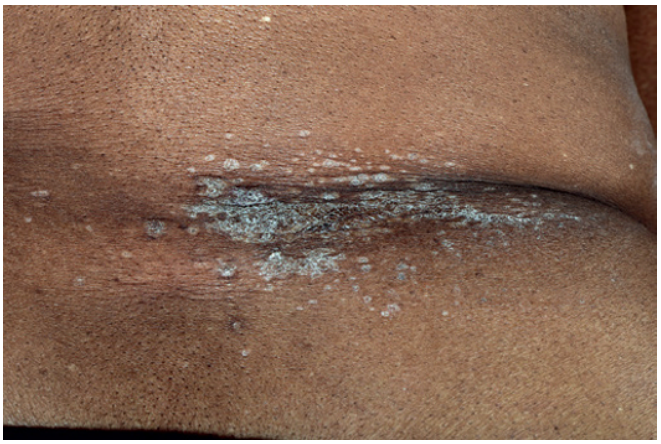


Figure 1.16 Epidermal atrophy in extra-genital lichen sclerosus.

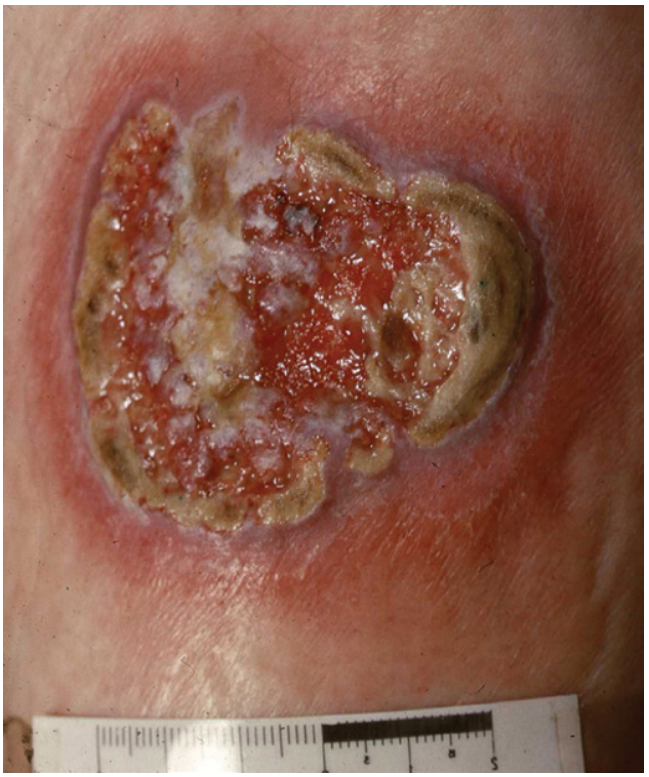


Figure 1.17 Ulceration in pyoderma gangrenosum.

There may be a history of recurrent episodes such as occurs in atopic eczema due to the patient's constitutional tendency. In the case of contact dermatitis, regular exposure to a causative agent leads to recurrences that fit from the history with exposure times. Endogenous conditions such as psoriasis can appear in adults who have had no previous episodes. If several members of the same family are affected by a skin rash simultaneously then a contagious condition, such as scabies, should be considered. A common condition with a familial tendency, such as atopic eczema, may affect several family members at different times.

A simplistic approach to rashes is to classify them as being from the 'inside' or 'outside'. Examples of 'inside' or endogenous rashes

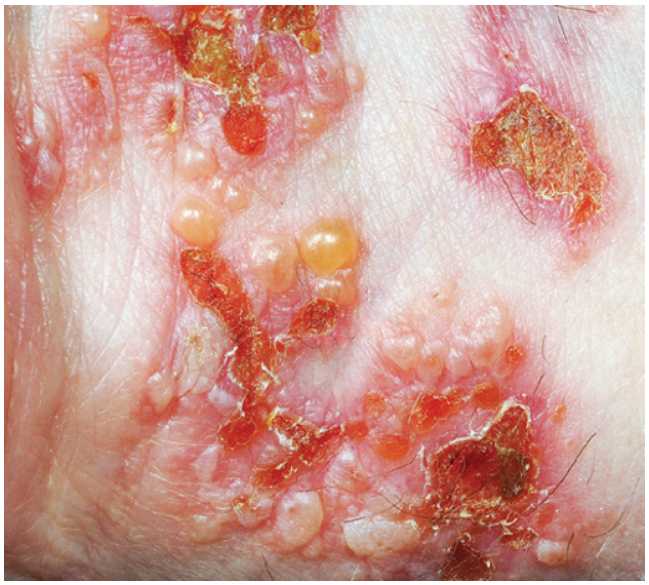


Figure 1.18 Erosions (loss of epidermis) in paraneoplastic bullous pemphigoid.

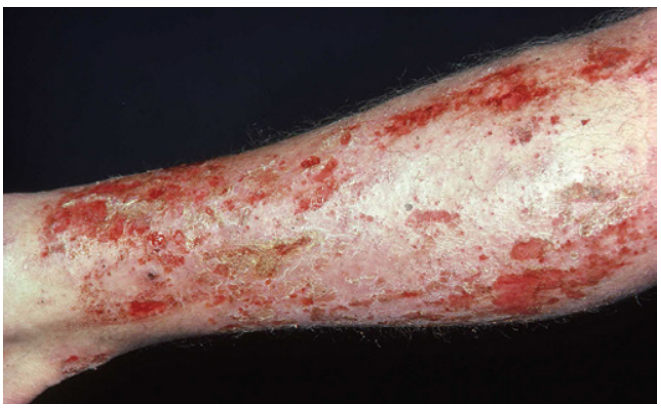


Figure 1.19 Excoriation of epidermis in atopic dermatitis.

are atopic eczema or drug rashes, whereas fungal infection or contact dermatitis are 'outside' or exogenous rashes.

Symmetry

As a general rule, most endogenous rashes affect both sides of the body, as in the atopic child, psoriasis on the legs or cutaneous T-cell lymphoma (Figure 1.24). Of course, not all exogenous rashes are asymmetrical. Chefs who hold the knife in their dominant hand can have unilateral disease (Figure 1.25) from metal allergy whereas a hairdresser or nurse may develop contact dermatitis on both hands, and a builder bilateral contact dermatitis from kneeling in cement (Figure 1.26).

Diagnosis

- Previous episodes of the rash, particularly in childhood, suggest a constitutional condition such as atopic dermatitis.

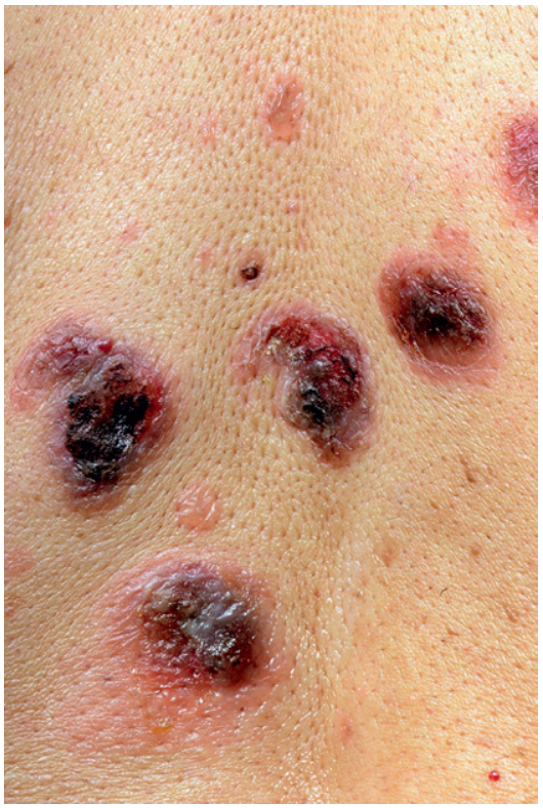


Figure 1.20 Crusted lesions in pemphigus vulgaris.

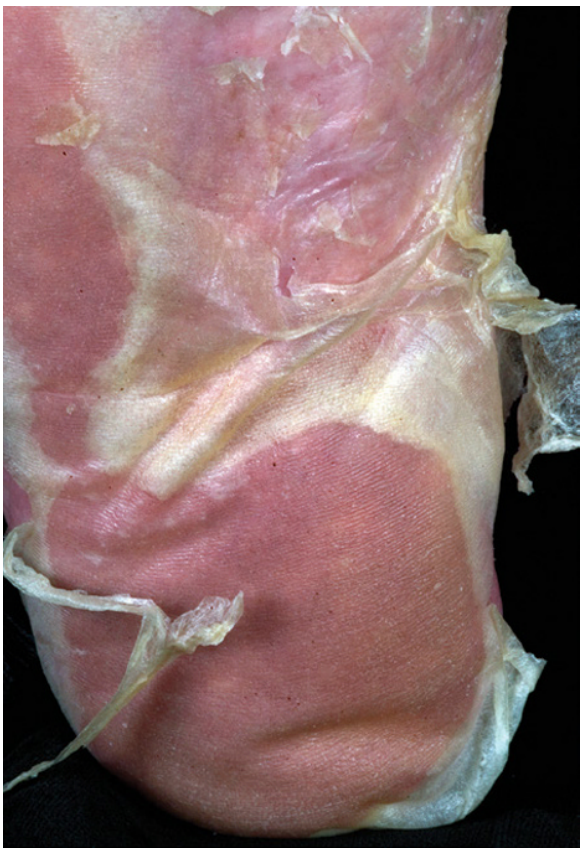


Figure 1.21 Desquamation following a severe drug reaction.

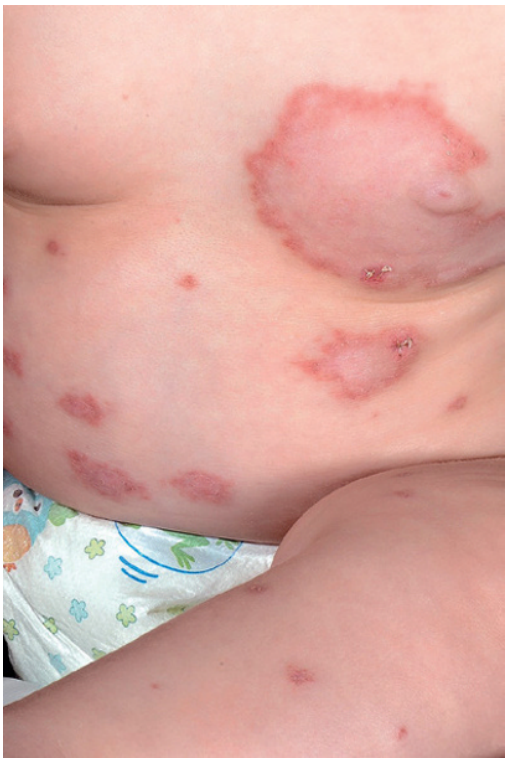


Figure 1.22 Annular (ring-shaped) lesions in neonatal lupus.

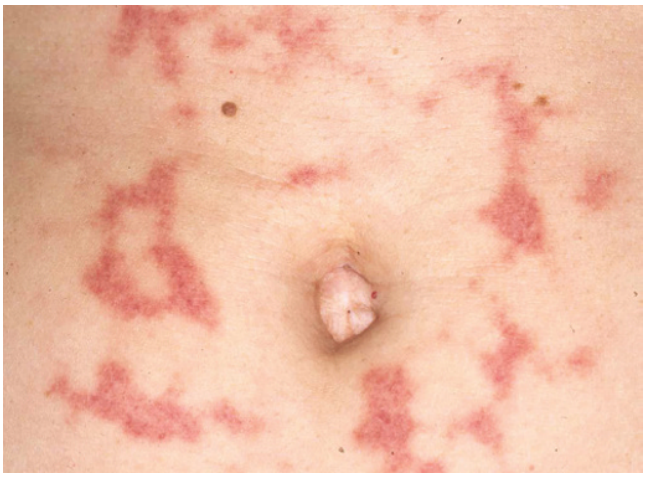


Figure 1.23 Reticulate pattern in vasculitis.

- Recurrences of the rash, particularly in specific situations, suggest a contact dermatitis. Similarly, a rash that only occurs on photo-exposed skin is highly suggestive of UV-driven skin disease (Figure 1.27) such as chronic actinic dermatitis.
- If other members of the family are affected, particularly without any previous history, there may well be a transmissible condition such as scabies.

Distribution

It is useful to be aware of the usual sites of common skin conditions. These are shown in the appropriate chapters. Eruptions that appear only on areas exposed to sun may be entirely or partially due to

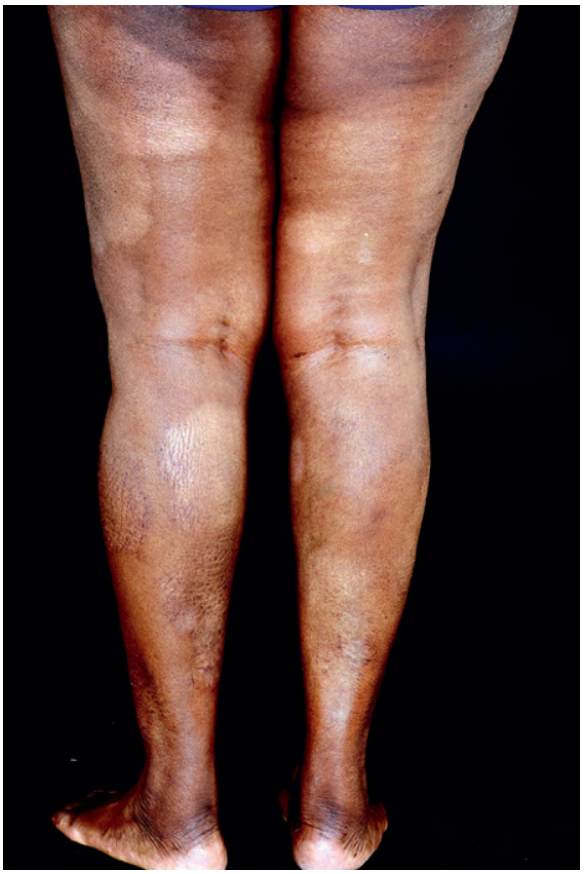


Figure 1.24 Symmetrical hypopigmented plaques of cutaneous T-cell lymphoma.

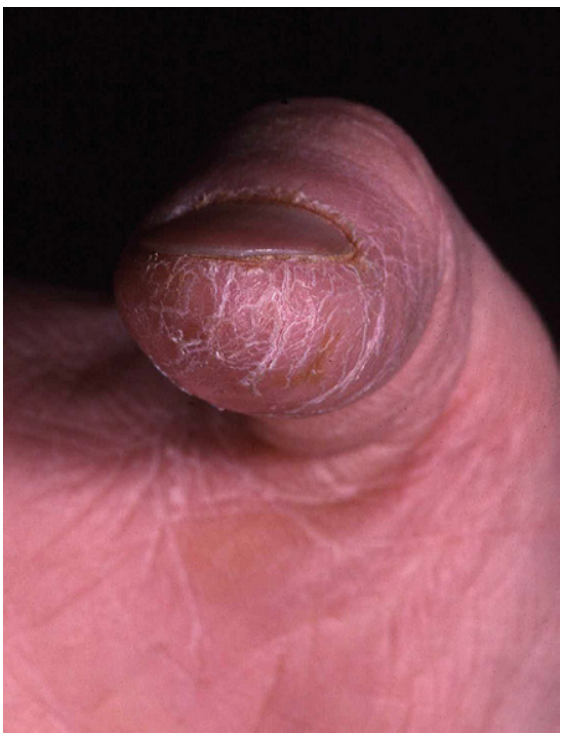


Figure 1.25 Irritant eczema on dominant hand of chef.

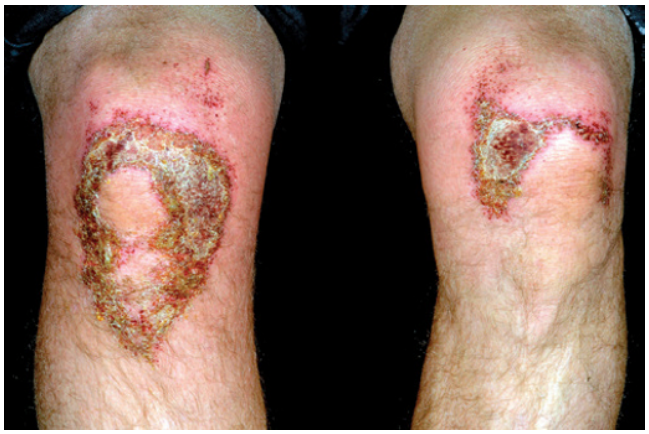


Figure 1.26 Bilateral contact dermatitis to cement.

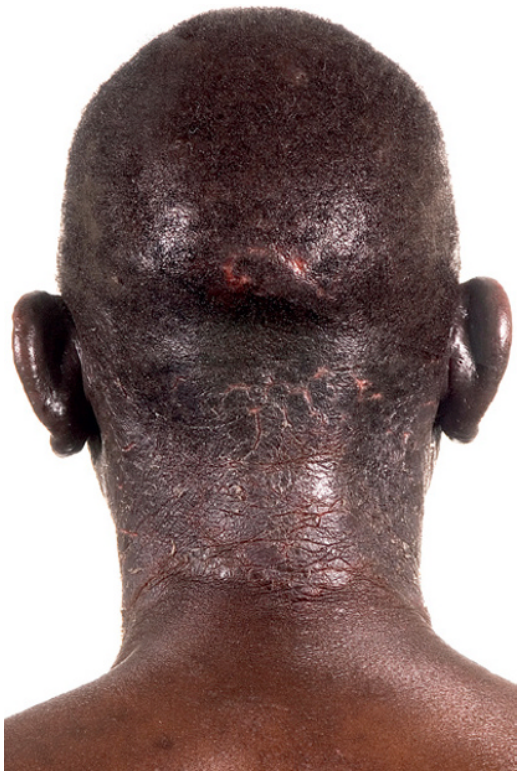


Figure 1.27 Chronic actinic dermatitis.

sunlight. Some are due to sensitivity to sunlight alone, such as polymorphic light eruption, or a photosensitive allergy to topically applied substances or drugs taken internally.

Morphology

The appearance of the skin lesion may give clues to the underlying pathological process.

Changes at the *skin surface* (epidermis) are characterised by a change in texture when the skin is palpated. Visually one may see scaling, thickening, increased skin markings, small vesicles, crusting, erosions, or desquamation. In contrast, changes in the *deeper tissues* (dermis) can be associated with a normal overlying

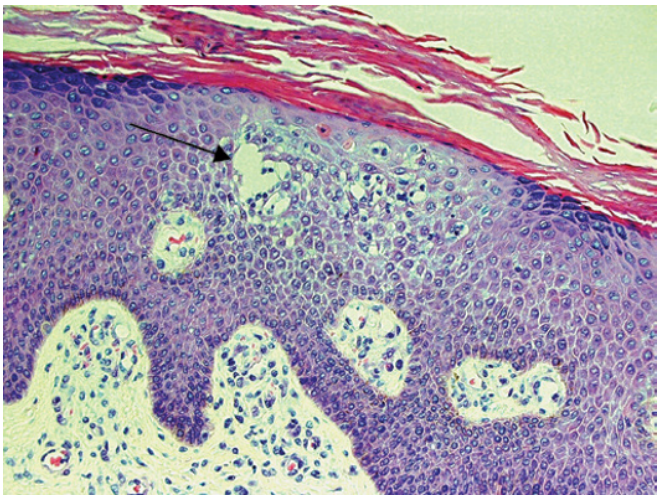


Figure 1.28 Eczema: intraepidermal vesicle (arrow).

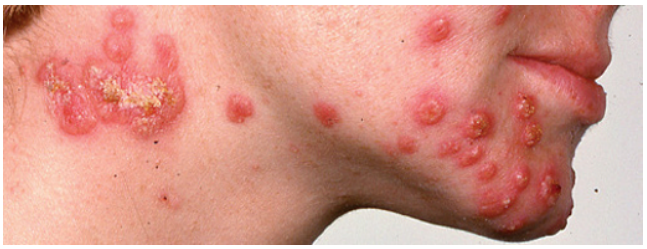


Figure 1.29 Vesicles and bullae in erythema multiforme.

skin. Examples of changes in the deeper tissues include erythema (dilated blood vessels, or inflammation), induration (an infiltrated firm area under the skin surface), ulceration (that involves surface and deeper tissues), hot tender skin (such as in cellulitis or abscess formation), and changes in adnexal structures and adipose tissue.

The *margin* or border of some lesions is very well defined, as in psoriasis or lichen planus, but in eczema it is ill-defined and merges into normal skin.

Blisters or vesicles occur as a result of

- oedema (fluid) between the epidermal cells (Figure 1.28)
- destruction/death of epidermal cells
- separation of the epidermis from the deeper tissues.

There may be more than one mechanism involved simultaneously. *Blisters or vesicles* (Figures 1.29–1.33) occur in

- *viral* diseases such as chickenpox, hand, foot and mouth disease, and herpes simplex
- *bacterial infections* such as impetigo or acute cellulitis
- *inflammatory disorders* such as eczema, contact dermatitis, and insect bite reactions
- *immunological disorders* such as dermatitis herpetiformis, pemphigus, and pemphigoid and erythema multiforme
- *metabolic disorders* such as porphyria.

Bullae (blisters more than 0.5 cm in diameter) may occur in congenital conditions (such as epidermolysis bullosa), in trauma, and as a result of oedema without much inflammation. However, those forming as a result of vasculitis, sunburn, or an allergic reaction may

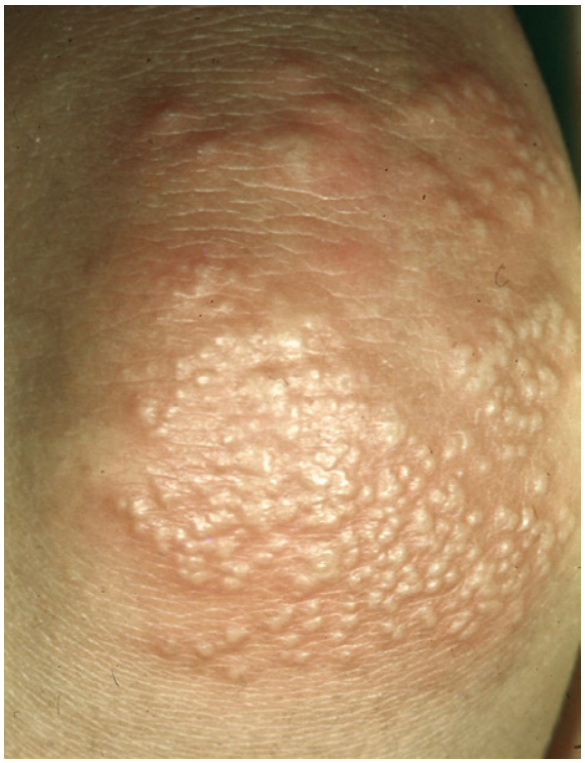


Figure 1.30 Vesicles in herpes simplex.

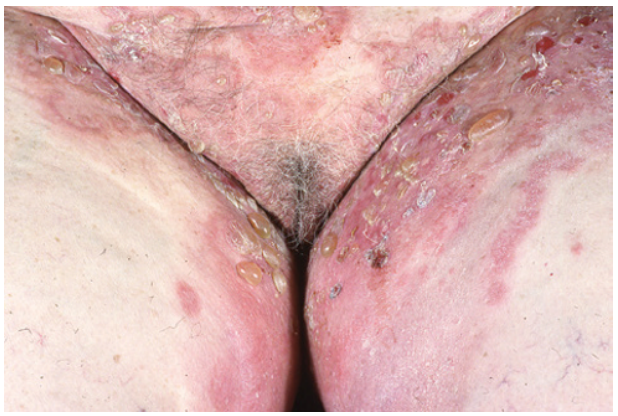


Figure 1.31 Vesicles and bullae in bullous pemphigoid.

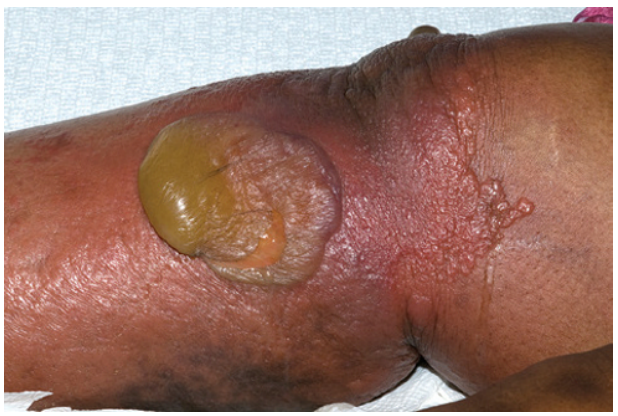


Figure 1.32 Bullae in cellulitis on lower leg.

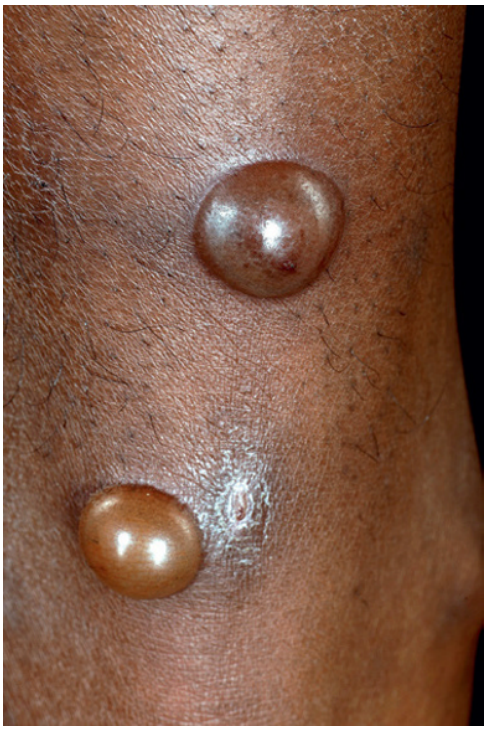


Figure 1.33 Bullae from insect bite reactions.

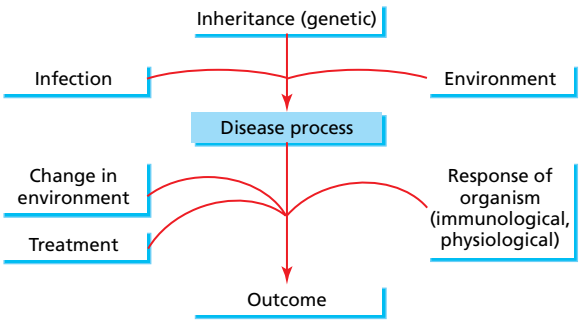


Figure 1.34 Possible precipitating factors in psoriasis.

be associated with pronounced inflammation. Adverse reactions to medications can also result in a bullous eruption.

Induration is the thickening of the dermis due to infiltration of cells, granuloma formation, or deposits of mucin, fat, or amyloid.

Inflammation is indicated by erythema, which can be acute or chronic. Acute inflammation can be associated with increased skin temperature such as occurs in cellulitis and erythema nodosum. Chronic inflammatory cell infiltrates occur in conditions such as lichen planus and lupus erythematosus.

Assessment of the patient

A full assessment should include not only the effect the skin condition has on the patients' lives but also their attitude to it. For example, some patients with quite extensive psoriasis are unbothered while others with very mild localised disease just on the elbows may be very distressed. Management of the skin disease

should take into account the patients' expectation as to what would be acceptable to them.

Fear that a skin condition may be due to cancer or infection is often present and reassurance should always be given to allay any hidden fears. If there is the possibility of a serious underlying disease that requires further investigation, then it is important to explain fully to the patient that the skin problems may be a sign of an internal disease.

The significance of occupational factors must be considered. In some cases, such as an allergy to hair dyes in a hairdresser, it may be impossible for the patient to continue his or her job. In other situations, the allergen can be easily avoided.

Patients often want to know why they have developed a particular skin problem and whether it can be cured. In many skin diseases these questions are difficult to answer. Patients with psoriasis, for example, can be told that it is part of their inherent constitution but that additional factors can trigger clinical lesions (Figure 1.34). Known trigger factors for psoriasis include emotional stress, local trauma to the skin (Koebner's phenomenon), infection (guttate psoriasis), and drugs (β-blockers, lithium, antimalarials).

Skill in recognition of skin conditions will evolve and develop with increased clinical experience. Seeing and feeling skin rashes 'in the flesh' is the best way to improve clinical dermatological acumen (Box 1.2).

Box 1.2 **Examination of skin lesions – key points**

Distribution
Examine all the skin for clues. For example, there are many possible causes for dry thickened skin on the palms, and finding typical psoriasis on the elbows, knees, and soles may give the diagnosis.

Morphology
Are the lesions dermal or epidermal? Macular (flat) or forming papules? Indurated or forming plaques? Well defined or indistinct? Forming crusts, scabs, or vesicles?

Pattern
The overall morphology and distribution of the rash – for example, an indeterminate rash may be revealed as pityriasis rosea when the 'herald patch' is found.

Further reading

Graham-Brown, R., Harman, K., and Johnson, G. (2016). *Lecture Notes: Dermatology*, 11e. New York: Wiley-Blackwell.

Wolff, K., Johnson, R.A., and Saavedra, A.P. (2013). *Fitzpatrick's Colour Atlas and Synopsis of Clinical Dermatology*, 7e. Oxford: McGraw-Hill Medical.

CHAPTER 2

Psoriasis

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OVERVIEW

- Psoriasis is a complex immune-mediated disease that manifests with chronic inflammatory lesions in the skin and systemic manifestations.
- Psoriasis has been shown to be an independent risk factor for cardiovascular disease.
- Specific biological therapies are transforming the management of psoriasis and psoriatic arthropathy.
- Clinical presentations can be variable, from chronic stable plaques, to pustules on the hands and feet, to unstable erythroderma.

Introduction

Psoriasis is now considered to be a genetically determined inflammatory systemic autoimmune disease. It is characterised by plaques of diseased skin often at sites of minor friction (elbows/knees), which occur next to areas of clear 'normal' skin. Plaques of psoriasis are clinically well-demarcated and are erythematous (dilated dermal blood vessels) with white surface scale (rapid keratinocyte proliferation). Psoriasis not only affects the skin but can also lead to seronegative arthritis in approximately 8–30% of patients. However, there is an increasing body of evidence that psoriasis is also associated with other important comorbidities such as type 2 diabetes (1.4-fold increased risk), cardiovascular disease (CVD), metabolic syndrome, obesity, non-alcoholic fatty liver disease (NAFLD), depression, and reduced quality of life.

The pathogenesis of psoriasis is complex; nonetheless, it is largely accepted that the disease is mediated by the dysregulation of T-helper lymphocytes (Th1/Th17). The development of psoriasis is multifactorial, with multiple potential susceptibility factors in a genetically at-risk individual. This combination of susceptibility factors and genetic predisposition results in an interactive web of immune cells/chemical cytokines impacting on skin cells and leading to disease. Increased understanding of these complex cellular changes has led to the introduction of multiple targeted biological therapies that are now used to manage severe psoriasis and psoriatic arthritis (PA).

Globally 1–2% of the population is affected by psoriasis (125 million people in UK/USA/Japan alone). A child who has one parent with psoriasis has a one in four chance of developing the disease. If one identical twin has psoriasis, there is a 70% chance that the other will also be affected; however, only a 20% chance exists in dizygotic twins. Linkage and genome-wide association studies have started identifying some of the important susceptibility factors leading to the inheritance of psoriasis and psoriatic arthropathy. The first and arguably one of the most important psoriasis susceptibility loci identified is the so-called *PSORS1* found on chromosome 6p21.3. This region of the chromosome contains several genes which may be important in the inheritance of psoriasis, including HLA-C (human leukocyte antigen-C), CCHCR1 (coiled-coil α-helical rod protein 1), and CDSN (corneodesmosin). Subsequently, multiple susceptibility loci on several chromosomes have now been identified including 1q21, 3q21, 4q, 7p, 8, 11, 16q, 17q, and 20p. Recently, a loss of function mutation in a gene encoding an IL-36 receptor antagonist has been shown to be associated with the development of palmo-plantar pustular psoriasis.

Plaques of psoriasis are highly infiltrated with CD3+ T-cells and CD11c+ dendritic cells which produce pro-inflammatory cytokines including tumour necrosis factor alpha (TNF-α), interferon gamma (INF-γ), and interleukin 17 (IL-17), IL-22/23/12/1β, which activate keratinocytes and other skin cells. Keratinocytes are the skin cells that predominate in the epidermis; they grow from the basal layer and slowly migrate to the surface (Figures 2.1 and 2.2). In normal skin, this process of cell turnover takes about 23 days; however, in psoriasis cell turnover is rapidly accelerated, taking only three to five days for cells to reach the surface and accumulate in large numbers (hyperkeratosis). Keratinocytes normally lose their nuclei as they move to the skin surface; however, in psoriasis they move so quickly that the cells retain their nuclei throughout the epidermis, seen as parakeratosis histologically. This rapid turnover and failure of proper maturation result in defective keratinocytes, which are poorly adherent and easily scraped off ('Auspitz sign') revealing underlying dilated blood vessels.

In addition, inflammatory polymorphs infiltrating the epidermis lead to swelling (oedema), inflammation, and erythema. These

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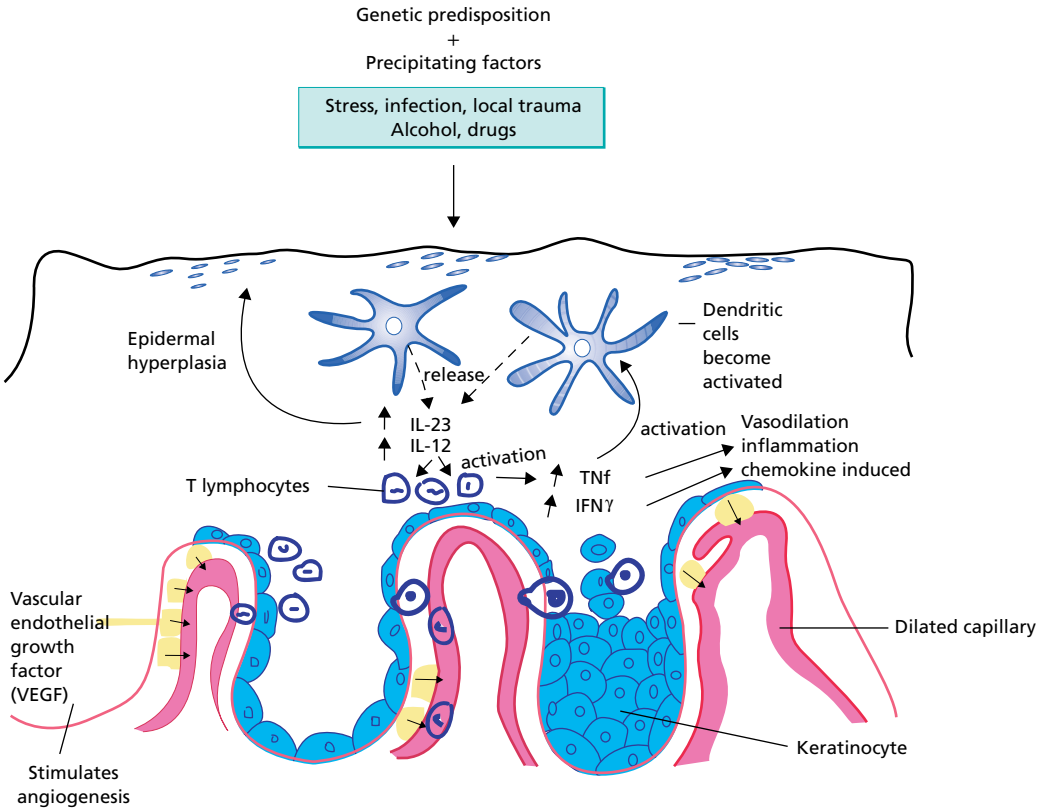


Figure 2.1 Pathophysiological mechanisms involved in the development of psoriasis.

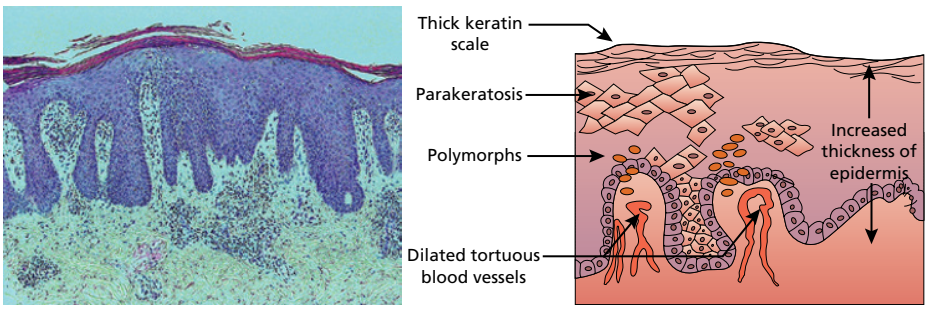


Figure 2.2 (Diagram/histology composite) Increased epidermal proliferation.

inflammatory cells may occur in such large numbers that they form collections of sterile pustules at the skin surface. These are most commonly seen in palmo-plantar pustulosis, a variant of psoriasis affecting the palms and soles.

The cellular abnormalities in the skin of patients with psoriasis can occur in the nails, and many patients will therefore have additional nail changes.

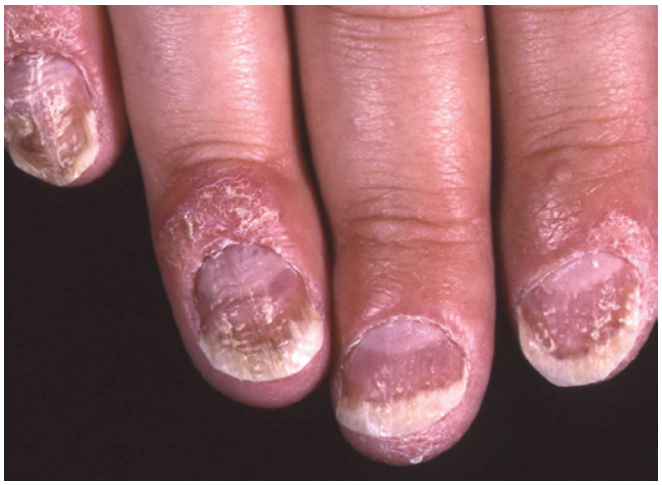
Psoriatic nail dystrophy is characterised by:

- *onycholysis* (lifting of the nail plate off the nail bed) due to abnormal cell adhesion; this usually manifests as a white or salmon patch on the nail plate (Figure 2.3a pitting and onycholysis of nails)
- *subungal hyperkeratosis* (Figure 2.3b) (accumulation of chalky looking material under the nail) due to excessive proliferation of the nail bed that can ultimately lead to onycholysis
- *pitting* (very small depressions in the nail plate) which result from parakeratotic (nucleated) cells being lost from the nail surface
- *Beau's lines* (transverse lines on the nail plate) due to intermittent inflammation of the nail bed, leading to transient arrest in nail growth
- *splinter haemorrhages* (which clinically look like minute longitudinal black lines) due to leakage of blood from dilated tortuous capillaries.

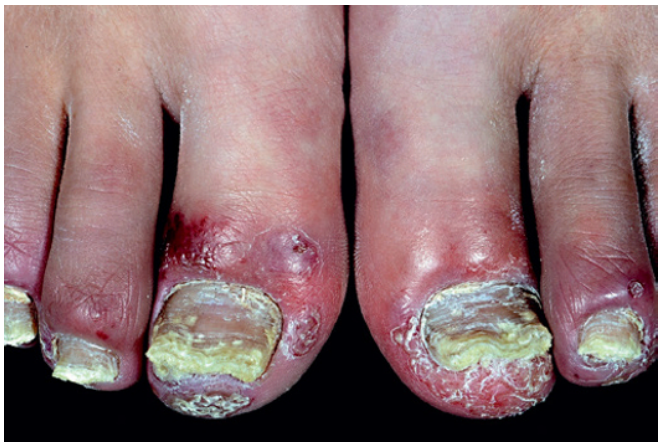
Clinical appearance

The main clinical features of psoriasis reflect the underlying pathological processes (as described above). Patients characteristically have the following:

- *Plaques* which are well-defined raised areas of psoriasis. These may be large or small, few or numerous and scattered over the trunk and limbs (Figures 2.4 and 2.5).



(a)



(b)

Figure 2.3 (a) Pitting and onycholysis of the nails. (b) Hyperkeratotic nail plates in psoriasis.

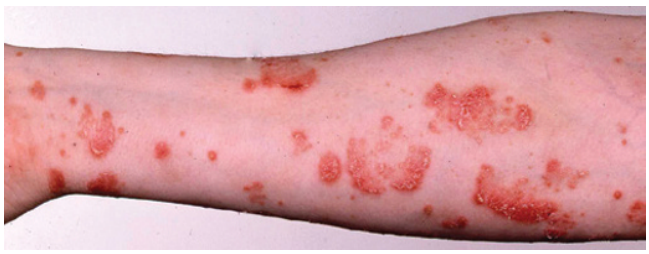


Figure 2.4 Multiple small plaques.

- *Scaling* may be very prominent causing plaques to appear thickened with masses of adherent and shedding white scales. Scratching the surface produces a waxy appearance – the 'tache de bougie' (literally 'a line of candle wax').
- *Erythema* or redness of the affected skin may be very marked, especially in the flexures. Erythema is a prominent feature in patients with erythrodermic psoriasis (who have >90% of their body surface involved).
- *Pustules* are commonly seen in *palmo-plantar pustulosis*, where deep-seated yellowish sterile pustules are often the dominant feature of this chronic condition. However, if pustules develop around the periphery of chronic plaques of psoriasis or sheets of monomorphic pustules appear more generally in the context of psoriasis, this is a sign of unstable disease – a dermatological emergency.

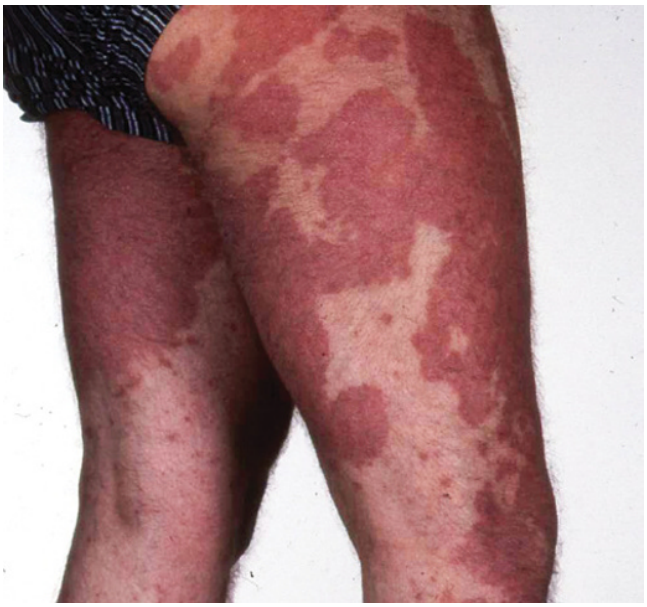


Figure 2.5 Large chronic plaques.

The typical patient

Psoriasis is reported to affect approximately 2% of the US population. The median age of onset is 28 years; however, it can present from infancy to old age, when the appearance may be atypical.

The following factors in the history may help in making a diagnosis:

- Family history of psoriasis; 16% of children will have psoriasis if a single parent is affected and 50% if both parents are affected.
- Trigger factors include stress, infections, trauma, or childbirth.
- Lesions may first appear at sites of minor skin trauma – Koebner's phenomenon.
- Lesions usually improve in the sun.
- Psoriasis is usually only mildly itchy.
- Arthropathy may be associated.

Clinical presentation

Classically psoriasis patients present with plaques on the elbows, knees, and scalp (Figure 2.6). Lesions on the trunk are variable in size and are often annular (Figures 2.7–2.9). Psoriasis may develop in scars and areas of minor skin trauma – the so-called Koebner's phenomenon (Figure 2.10). This may manifest as hyperkeratosis on the palms, associated with repetitive trauma from manual labour (Figure 2.11). Scalp scaling, which affects 50% of patients, can be very thick, especially around the hairline, but may be more confluent, forming a virtual 'skull cap' (Figure 2.12). Erythema often extends beyond the hair margin. The nails show 'pits' and

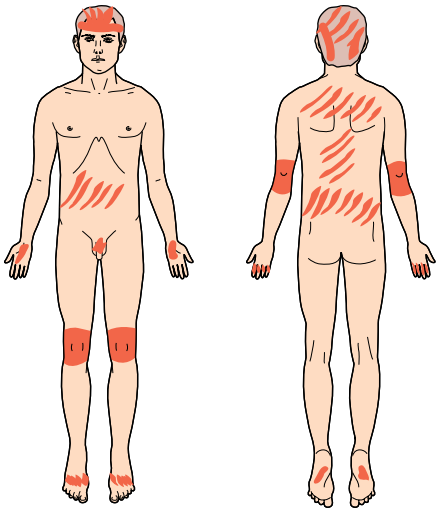


Figure 2.6 Common patterns of distribution of psoriasis.

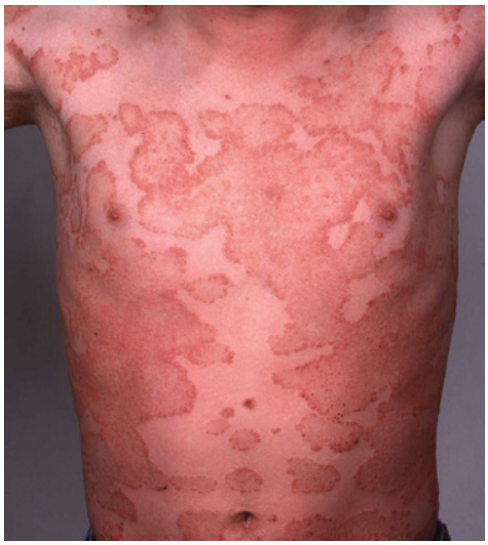


Figure 2.7 Generalised plaques.

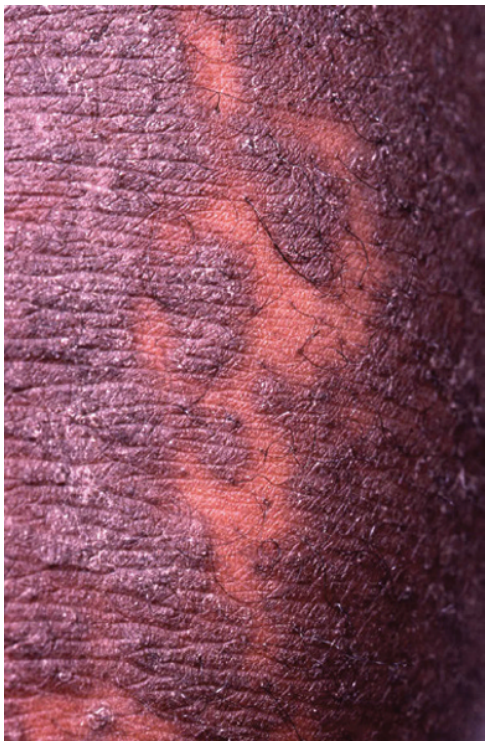


Figure 2.8 Psoriatic plaques on the trunk.

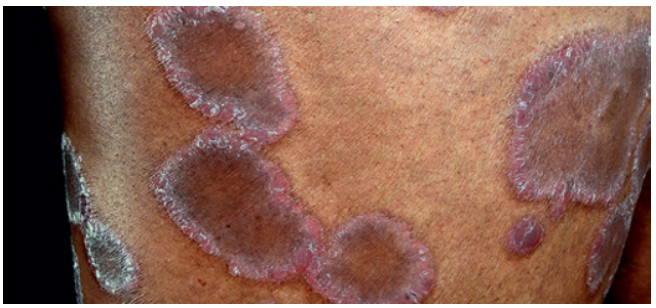


Figure 2.9 Annular plaques.

also thickening with separation of the nail from the nail bed (onycholysis) (Figure 2.13).

Guttate psoriasis – from the Latin *gutta*, a drop – consists of widespread small plaques scattered on the trunk and limbs (Figure 2.14). Adolescents are most commonly affected and there is often a preceding sore throat with associated group β-haemolytic streptococcus. There is frequently a family history of psoriasis. The sudden onset and widespread nature of guttate psoriasis can be very alarming for patients, fortunately it usually resolves completely, but can be recurrent or herald the onset of chronic plaque psoriasis.

Palmo-plantar pustular psoriasis (PPPP) is characterised by multiple sterile pustules on the palms and soles. Pustules first appear as yellowish monomorphic lesions that turn a brown colour with chronicity (Figure 2.15) and associated scaling. Most patients with PPPP are smokers.

Generalised pustular psoriasis (GPP) is uncommon as it is usually an indicator of severe and unstable psoriasis (Figure 2.16). Mutations in IL36RN gene that encodes IL-36Ra have recently been identified in a proportion of patients with GPP. Clinically the skin becomes acutely erythematous and tender, with sheets of monomorphic sterile pustules, which can develop over a few hours/days. It may be precipitated by the patient taking systemic steroids or using potent topical steroids. The pustules usually occur initially at the peripheral margin of plaques, which are often sore and erythematous. Pustules eventually dry and the skin desquamates.

Acropustulosis is a rare variant of psoriasis that usually occurs in young children. Here pustules appear around the nails and the fingertips, associated with brisk inflammation.

Flexural psoriasis produces well-defined erythematous areas in the axillae, groin, natal cleft, beneath the breasts, and in skin folds. Scaling is minimal or absent (Figure 2.17). It should be distinguished from a fungal infection and if there is any doubt a specimen for mycology should be taken.