Dermatology

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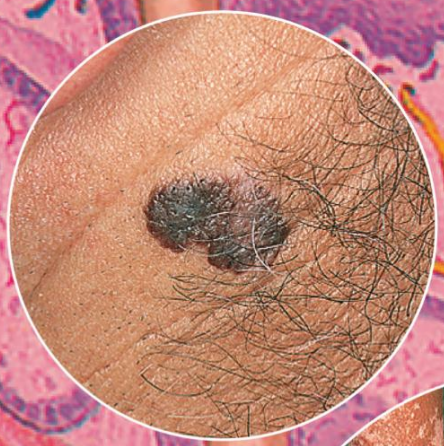

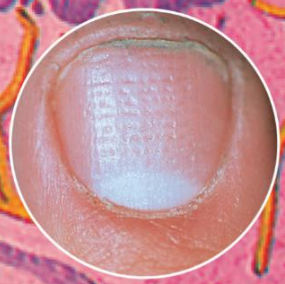

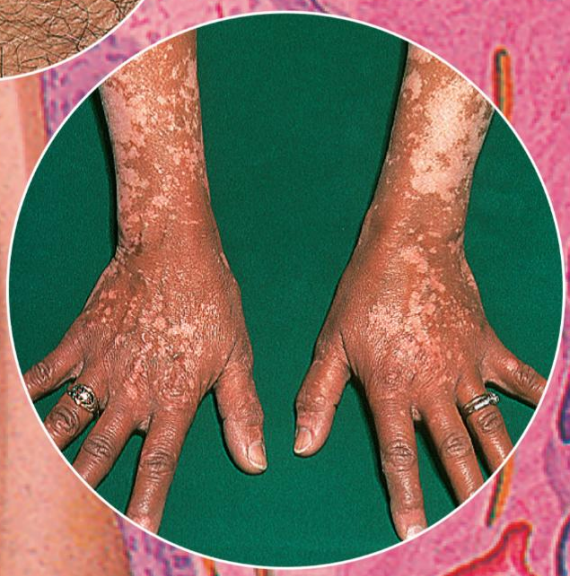

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ILLUSTRATED COLOUR TEXT

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Eighth Edition

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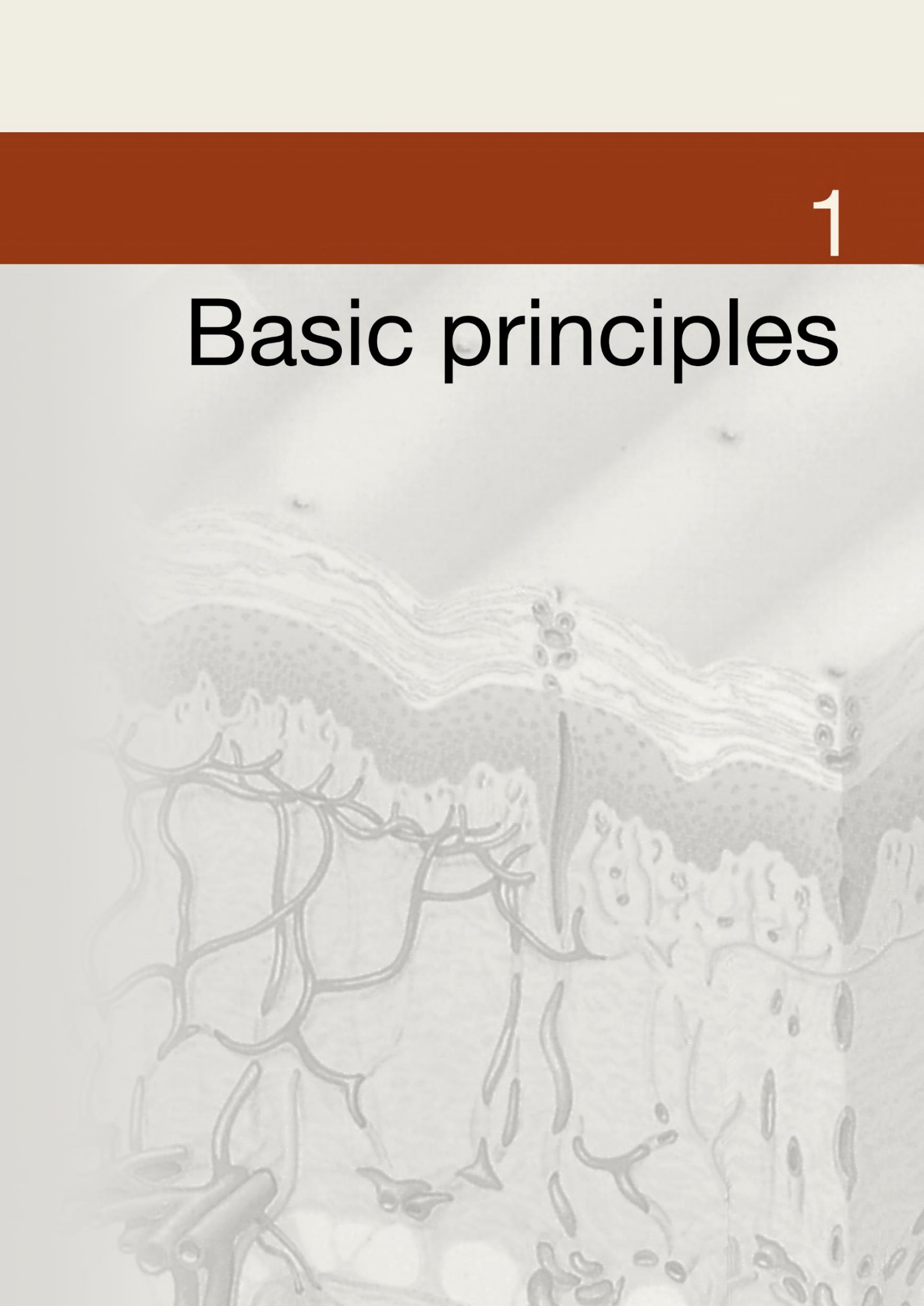
Basic principles



1 Microanatomy of the skin

Introduction

The skin is one of the largest organs in the body, having a surface area of 1.8 m^2 and making up about 16% of body weight. It develops in utero (Box 1.1), and although one of its most important functions is as a barrier to protect the body from noxious external factors, there is increasing recognition of the role of resident immune cells in skin function.

Skin is composed of three layers: the epidermis, the dermis and the subcutis (Fig. 1.1); it supports a complex population of microflora on the surface (skin microbiome) (p. 10).

Epidermis

The epidermis is a stratified squamous epithelium that is about 0.1 mm thick, although the thickness is greater (0.8–1.4 mm) on the palms and soles. Its prime function is to act as a protective barrier. The main cells of the epidermis are *keratinocytes*, which produce the protein keratin. Keratinocytes are squamous cells functionally similar to all other structural epithelial cells as found in the airways and gastrointestinal tract. Keratinocytes differentiate upwards through the epidermis, and their maturation states (p. 6) are divided into four stages (layers) (Fig. 1.2).

Basal cell layer (stratum basale)

The basal cell layer of the epidermis is composed mostly of keratinocytes, of which a small proportion are stem cells that continuously divide. The cells contain keratin tonofilaments (p. 8) and are secured to the basement membrane (see Fig. 1.2) by hemidesmosomes. *Melanocytes* make up 5–10% of the basal cell population. These cells synthesize melanin (p. 8) and transfer it in melanosomes via dendritic processes to neighbouring keratinocytes.

Melanocytes are most numerous on the face and other exposed sites and are of neural crest origin. *Merkel cells* are also found, albeit infrequently, in the basal cell layer. These cells are closely associated with terminal filaments of cutaneous nerves and seem to have a role in sensation. Their cytoplasm contains neuropeptide granules, neurofilaments and keratin. Basal keratinocytes synthesize antimicrobial peptides, which are important in defence against bacteria.

Prickle cell layer (stratum spinosum)

Daughter basal cells migrate upwards to form this layer of polyhedral cells, which are interconnected by desmosomes (the 'prickles' seen at light microscope level). Keratin tonofibrils form a supportive mesh in the cytoplasm of these cells. *Langerhans cells* are mostly found in this layer; these dendritic, immunologically active cells are described fully on page 10.

Granular cell layer (stratum granulosum)

Cells become flattened and lose their nuclei in the granular cell layer. Keratohyalin granules are seen in the cytoplasm together with membrane-coating granules (which expel their lipid contents into the intercellular spaces).

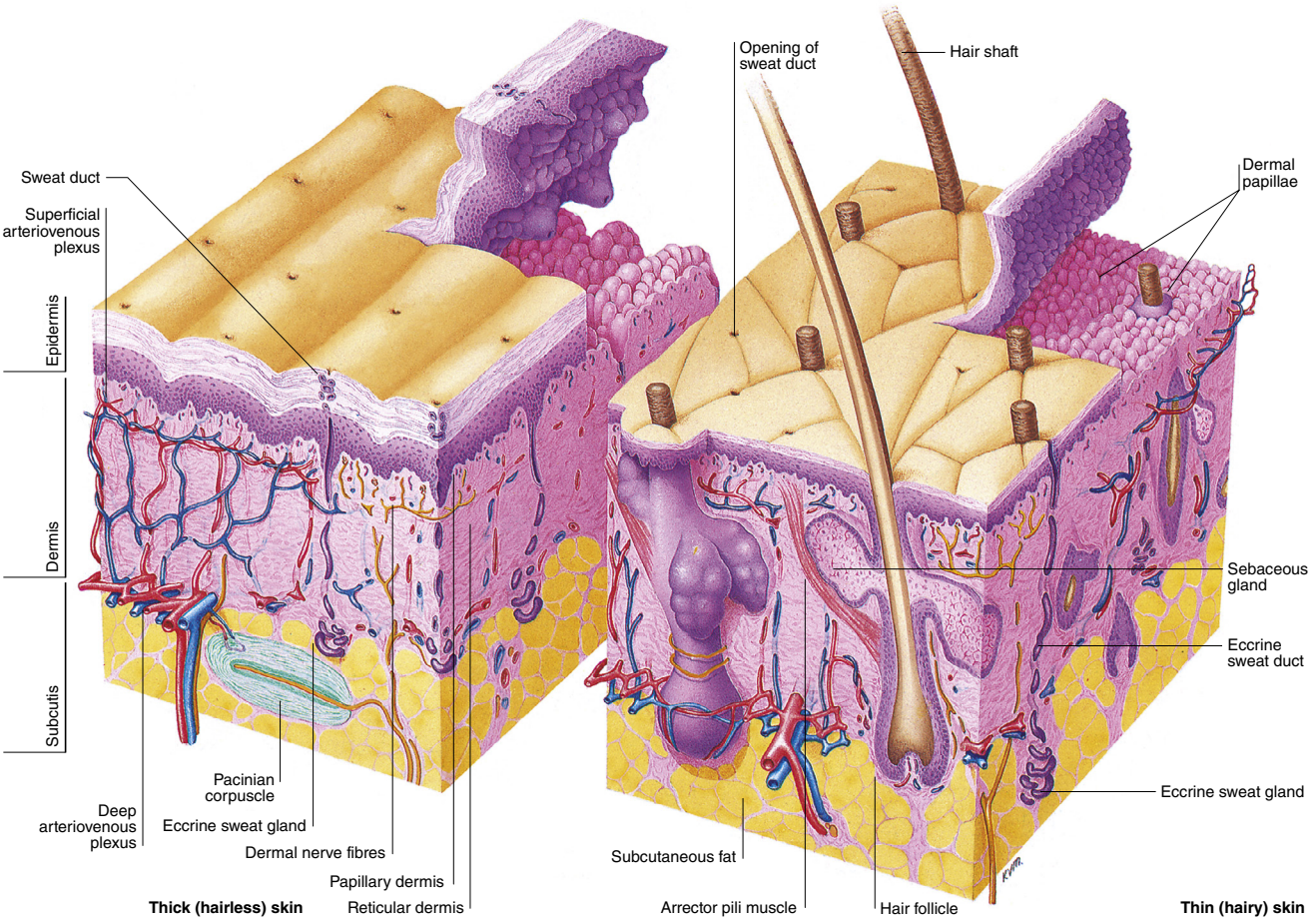


Fig. 1.1 Structure of the skin. The diagram shows a comparison between thick, hairless skin (plantar and planar) and thinner, hirsute skin.

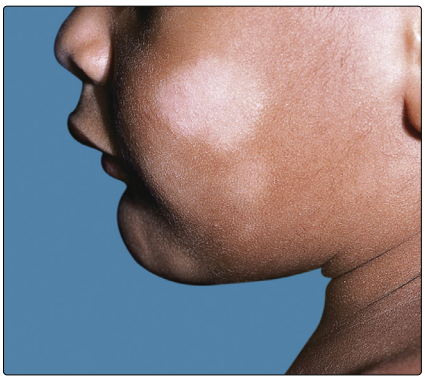


Fig. 70.5 Pityriasis alba. A hypopigmented, slightly scaly patch on the left cheek. *(Courtesy Anthony J Mancini, MD.)*

small roundish, faintly scaly and slightly hypopigmented patches are seen, often on the face or upper arms. It is more frequent in summer and appears to be a mild form of eczema (Fig. 70.5).

Other childhood dermatoses

Some uncommon but characteristic eruptions are found in childhood. These include:

- Measles
- Urticaria pigmentosa
- Langerhans cell histiocytosis
- Linear immunoglobulin A disease (p. 106)
- Kawasaki disease and other viral infections (p. 71)
- Ichthyosis (p. 120)
- Epidermolysis bullosa (p. 121)

Measles

Measles is a serious generalized childhood infection due to an RNA morbillivirus (p 71). Widespread vaccine use from the 1960s meant that cases became uncommon, but the recent reduction in vaccination coverage has allowed measles to become resurgent in Europe, with attendant mortality and morbidity. After a short upper respiratory prodrome, with conjunctivitis and fever, the morbilliform rash starts on day 4, typically first behind the ears, then spreading generally (Fig. 70.6). White-on-red punctate Koplik spots on the buccal mucosa are pathognomonic. Pneumonia and encephalitis are complications, though most make a full recovery.

Urticaria pigmentosa

Urticaria pigmentosa is characterized by multiple red-brown macules or papules on the trunk and limbs of an infant (Fig. 70.7). It is associated with c-*KIT* gene mutations. The lesions may become red, swollen and itchy after a bath or when rubbed, and blistering may occur. Histologically, there are accumulations of mast cells in the dermis. The disorder normally resolves spontaneously before adolescence. There is a form with a later onset, usually beginning in adolescence or adult life, which rarely resolves and may involve internal organs—something that is uncommon in the childhood variety.

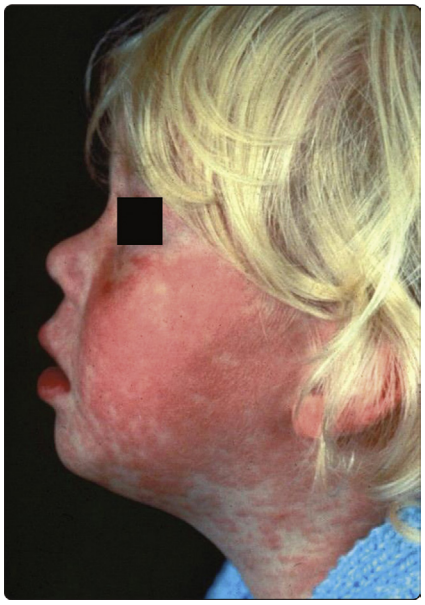


Fig. 70.6 Measles, showing the morbilliform eruption. *(From James WD, Berger TG, Elston DM. Andrews' Diseases of the Skin. 11th ed. Saunders; 2011.)*

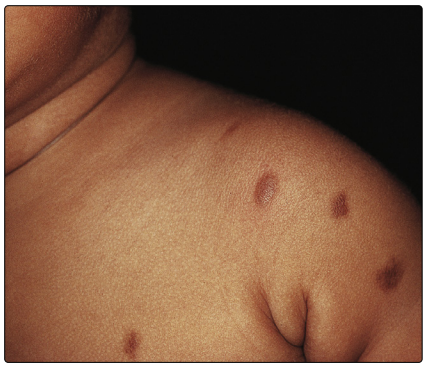


Fig. 70.7 Mastocytosis nodules of juvenile urticaria pigmentosa. Scattered nodules on the chest and shoulder are evident. They show some hyperpigmentation and slight surrounding erythema. Darier's sign of urtication on rubbing a lesion is virtually pathognomonic. *(From Bolognia JL, Jorizzo JL, Schaffer JV. Dermatology. 3rd ed. Saunders; 2012.)*

Langerhans cell histiocytosis (histiocytosis X)

Langerhans cell histiocytosis is a rare and serious condition that normally involves internal organs. The skin signs are prominent, variable and include a seborrhoeic-like dermatitis, papules or pustules on the trunk, and ulceration, particularly of the flexures (Fig. 70.8). The skin, abdominal organs, lungs and bones are infiltrated by clonal myeloid precursors that differentiate into $CD1a^+/CD207^+$ cells, now believed to represent inflammatory myeloid neoplasia. Skin biopsy is usually diagnostic. The prognosis is poorer when the onset is <2 years of age.

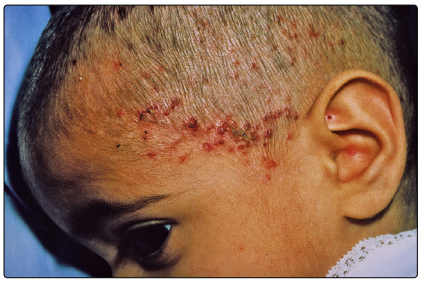


Fig. 70.8 Langerhans cell histiocytosis. The disease may present with a seborrhoeic dermatitis–like eruption on the scalp, but papules and crusting are more prominent than would be anticipated in an eczema. *(From James WD, Berger TG, Elston DM. Andrews' Diseases of the Skin. 11th ed. Saunders; 2011.)*

Paediatric dermatology—eczemas and other childhood dermatoses

Disorder	Age at onset	Clinical features
Napkin dermatitis	First few weeks–12 months	Glazed erythema that spares body folds
		Erosions may occur
Infantile seborrhoeic eczema	First few weeks	Moist scaly erythema
		Flexures and scalp affected
Candidiasis	Infancy	Erythema, with scaling and pustules
		Flexures affected
		Secondary infection found
Juvenile plantar dermatosis	School age to middle teens	Glazed red fissured skin on weightbearing forefeet and soles
Measles	Infancy and childhood	Upper respiratory prodrome, morbilliform rash, Koplik spots
Urticaria pigmentosa (cutaneous mastocytosis)	Mostly at 3–9 months	Red-brown macules or papules on trunk, which urticate when rubbed
Langerhans cell histiocytosis	All ages (different types)	Seborrhoeic-like dermatitis, papules/pustules, ulceration

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