

Dermatoses of the Neonate and Infancy

1

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Introduction

More than 95% of newborns have cutaneous findings, which often are distressing to parents but frequently are benign and self-limited. Among them are milia, cutis marmorata, congenital dermal melanocytosis and the benign neonatal pustular eruptions (Rayala and Morrell 2017; Zuniga and Nguyen 2013). The majority of the dermatologic alterations in neonates are physiological and transient and do not require any treatment; thus the parents can be reassured about the good prognosis (Patrizi et al., 2017). An overview of the most common or important dermatoses presenting in neonatal and infant age group is illustrated in this chapter.

1.



A 6 months old baby was brought with this skin lesion which has been there since birth, the mother is worried that it is being progressively enlarging.

- (a) What is the diagnosis?
- (b) What is the sequelae of the disease?

The clinical photographs in this chapter are photographed by Dr. Ranthilaka R. Ranawaka, consultant dermatologist, General Hospital Kalutara, Sri Lanka

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2.



A 3 months old baby was brought with mildly erythematous hypopigmented patches on the groin, back of the trunk, neck and upper chest. They were mostly asymptomatic. What is the diagnosis?

3.



A 1-year-old child came with the above lesions which were asymptomatic. He was getting these lesions frequently. His 3-year-old sister also was getting few lesions on the face. What is the diagnosis?

4.



A 4-month-old baby was brought with itchy skin lesions on the trunk, buttocks and genitalia for 3 weeks.

- (a) What is the diagnosis?
- (b) Who else do you want to examine?



The mother of this 6 months old baby is worried about these scaly lesions on the scalp which are mildly itchy.

- (a) What is the diagnosis?
- (b) What advice would you give the mother?



A 2-year-old child has a distractible itching more pronounced during the night. On direct inquiry revealed that his whole family has generalized itching without any visible skin rash.

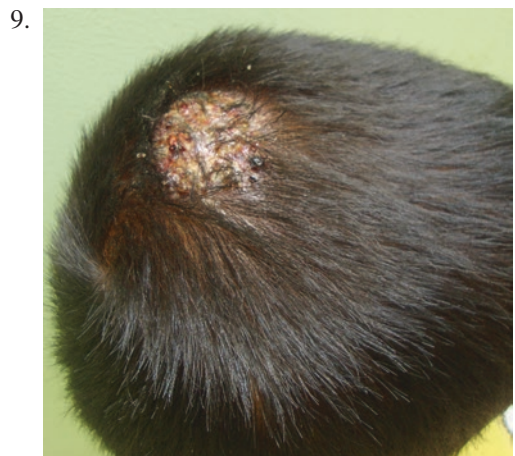
- (a) What is the diagnosis?
- (b) What is the condition shown in this picture?



A 4 days old neonate was brought with this skin eruption. What is the diagnosis?



A 5-year-old boy has these asymptomatic hypopigmented patches on both cheeks for 2 months which are progressively enlarging. What is the diagnosis?



This painless boggy mass has appeared and enlarged within 1 week despite antibiotics prescribed by the family physician. What is the diagnosis?

10.



This 4-year-old child's mother is worried about these asymptomatic lesions which appeared 4 weeks ago.

- (a) What is the diagnosis?
- (b) What advice would you give the mother?

11.



A 6-year-old child developed these patchy hair loss over 3 weeks which were treated with native remedies. What is the diagnosis?

12.



These asymptomatic lesions had appeared over 3 weeks. What is the diagnosis?

13.



A 4-year-old child came with these painful pustules localized to shown area.

- (a) What are the differential diagnoses?
- (b) What was the diagnosis in this child?

14.



A 7-year-old boy had developed these lesions on the face over 3 months.

- (a) What are the possible diagnoses?
- (b) What are the other features you want to look for?
- (c) How do you treat this skin problem?

15.



The mother of this 6 months old baby is worried about these linear hypopigmented patches on the trunk.

- (a) What is the diagnosis?
- (b) What advice would you give the mother?

16.



A newborn had this patch of alopecia.

- (a) What are the possible causes?

17.



The

mother of this 7-year-old child complains foul smell from his feet when he removes socks after school.

- (a) What is the diagnosis?
- (b) What are the predisposing factors?

18.



A 4-year-old child was brought with this nail abnormality. After few weeks these nails fall spontaneously.

- (a) What is the diagnosis?
- (b) What are the predisposing factors?
- (c) How do you manage this?

19.



A 20 days old baby girl who was normal at birth had developed these blisters and erosions for the last 3 days.

- (a) What are the differential diagnoses?

20.



A 5-year-old child developed this skin eruption with fever, malaise and body aches.

- (a) What is the diagnosis?



21.

A 4-month-old baby was brought with this inguinal rash for 1 month.

- (a) What are the differential diagnoses?

22.



A 12-year-old child came with this linear itchy lesion on the dorsum of the hand. They noticed that the lesion is extending forward.

- (a) What is the diagnosis?

Answers

1. Infantile haemangioma (Chap. 3).

It will slowly grow over the first 12 months and then regress gradually over the next 2–3 years which almost disappears leaving some slack skin.

No treatment is given unless it obstructs the function of vital organs. But there is a tendency to easy bleeding and secondary infection.

2. Infantile seborrhoeic dermatitis.
3. Staph impetigo.
4. Tinea infection (Chap. 17).
Close relatives who look after the child, e.g. parents, grandparents, nanny, etc. Usually infants acquire this infection from adults. You should treat the adult who has tinea infection too, to avoid reinfection.
5. Cradle cap.
This will spontaneously clear over months. Treat symptomatically.
6. Scabies (Chap. 20).
Scabietic nodules which have predilection to genitalia, which are very itchy and can persist for weeks to months after successful treatment of scabies.
7. Folliculitis.
8. Polymorphic light eruption (Chap. 10).
9. Kerion (Chap. 17).
10. Granuloma annulare.
These are benign lesions which resolve spontaneously with time.
11. Alopecia areata (Chap. 35).
12. Molluscum contagiosum (Chap. 16).
13. Staph impetigo and herpes infection.
Herpes infection (HSV I).
14. Acneiform eruption due to topical or systemic steroids, drug-induced acne and adenoma sebaceum (angiofibromas) in tuberous sclerosis complex (TCS).
Hypomelanotic macules (ash-leaf macules) and angiofibromas are pathognomonic in TCS, also look for periungual fibromas (Koenen tumours) and shagreen patch (Chap. 4).
This is adenoma sebaceum (angiofibromas) in tuberous sclerosis complex (TCS) which can be treated with electrocautery or laser.
15. Pigmentary mosaicism, streaks and whorls of hypo- or hyperpigmentation following Blaschko's lines. Infants with pigmentary mosaicism should be thoroughly assessed for the development, the internal organs and skeletal and ophthalmological abnormalities.
16. This lesion is circumscribed alopecia of congenital origin. This can be due to several causes, sebaceous naevus, aplasia cutis or sutural alopecia which will be obvious later.
17. Pitted keratolysis.
Hyperhidrosis and frequent emersion of feet in water are the predisposing factors.
18. Onychomadesis (Chap. 37).
This not uncommon in children after viral infection, most commonly hand, foot and mouth disease.
19. Bullous impetigo and dystrophic epidermolysis bullosa (DEB).
In DEB the child is clinically well in spite of large erosions, and lesions are confined to areas of friction.
20. Chickenpox (Chap. 16).
21. Infantile psoriasis, napkin dermatitis and infantile seborrhoeic dermatitis.
Involvement of the napkin area (psoriatic napkin eruption) may be the first presentation of psoriasis in infancy (Burden and Kirby 2016). Edges of the lesions are well defined unless altered by medicaments.
22. Cutaneous larva migrans.

1.1 Inflammatory Conditions

1.1.1 Cradle Cap

Cradle cap may be seen in the neonate; the condition is most common between the ages of 4 and 16 weeks. It can occur in isolation or in association with seborrhoeic dermatitis. Most cases of cradle cap resolve spontaneously after a few weeks (Das and Das 2014).

Clinical presentation: Thick scales adherent to scalp since birth. Mild inflammation with itching in some, but most are asymptomatic (Fig. 1.1).

Management: An emollient to lift the scale, in combination with a shampoo.

1.1.2 Infantile Seborrhoeic Dermatitis

Infantile seborrhoeic dermatitis (ISD) occurs between the ages of 4 and 12 weeks but most commonly before the age of 2 months.

Clinical presentation: macerated erythema on the skin folds, neck, axillae, inguinal region, elbows and skin creases. ISD may associate with

cradle cap, rarely symptomatic (Sarkar and Garg 2010). Typically inflammation resolves with postinflammatory hypopigmentation which is marked in pigmented skin and is a great concern to parents (Figs. 1.1, 1.2, 1.3, 1.4 and 1.5).

Differential diagnosis: napkin dermatitis, atopic eczema, Langerhans cell histiocytosis (Leung et al. 2019).

Management: a combination of steroid–anti-fungal creams in cases where inflammation is marked, intermittently for short periods. Hypopigmentation resolves spontaneously within weeks to months (Victoire et al. 2019).

1.1.3 Atopic Eczema

In infants, AE characteristically begins on the face with subsequent spread to involve the torso and limbs. A more nummular (discoïd) pattern occurs, particularly on the back and legs, especially in toddlers. AE in the majority of infants clears over time. Nearly half of children with early AE are in complete remission by age 3 years. A significant number of children were found to develop AD shortly after their ISD diagnosis. This finding demonstrates a strong association in the clinical course between the two diseases or indicates that the two diseases may be in the same clinical spectrum (Alexopoulos et al. 2014).

Clinical presentation: Mildly itchy scales mainly on the cheeks and on extensors, which change to flexural involvement when the child grows (Figs. 1.6 and 1.7).

Management: Exclusive breastfeeding for the first 6 months is advisable. This is a chronic recurrent problem in many. Topical steroid should be used sparsely with frequent emollients.

1.1.4 Napkin Dermatitis

Prolonged contact with urine induces an irritant erythema, which may break down to form erosions if untreated.

Clinical presentation: dermatitis is limited to the areas in contact with the irritant, while the skin folds may be spared (in contrast to sebor-

rhoic dermatitis or infantile psoriasis where skin folds are affected) (Figs. 1.8, 1.9 and 1.10).

Differential diagnosis: allergic contact dermatitis, intertrigo, psoriasis and atopic and seborrhoic dermatitis (Tüzün et al. 2015).

Management: the mainstay of management is to keep the skin dry and use barrier creams or emollients to restore normal epidermis. Mild topical steroids are used if inflamed or itchy. Treat secondary bacterial or candida infection appropriately. Advise to wear disposable diapers which are much more absorbent, especially during the night.

1.1.5 Infantile Psoriasis

Napkin psoriasis with dissemination is the most common pattern in infants (question 21). All patterns of psoriasis have been described in children: guttate, chronic plaque, pustular and erythrodermic, but severe disease and joint involvement are relatively rare (Burden and Kirby 2016).

Differential diagnosis: napkin dermatitis, infantile seborrhoic dermatitis and irritant contact dermatitis.

Treatments: mild topical steroids, often in combination with an anticandidal agent, and emollients usually suffice in most children.

1.1.6 Miliaria

Miliaria is due to blockage of eccrine sweat ducts. Immature sweat ducts are an important factor in neonates although high levels of heat and humidity are important at any age. It is subdivided into three subtypes dependent on the level of blockage: miliaria crystallina (stratum corneum), miliaria rubra (mid-epidermal) and miliaria profunda (dermal–epidermal junction) (Paige 2016; Dixit et al. 2012).

Clinical Presentation

Miliaria crystallina presents as crops of clear, thin-walled, superficial vesicles 1–2 mm in diameter, without associated erythema, resembling drops of water. These are exceedingly delicate and gener-

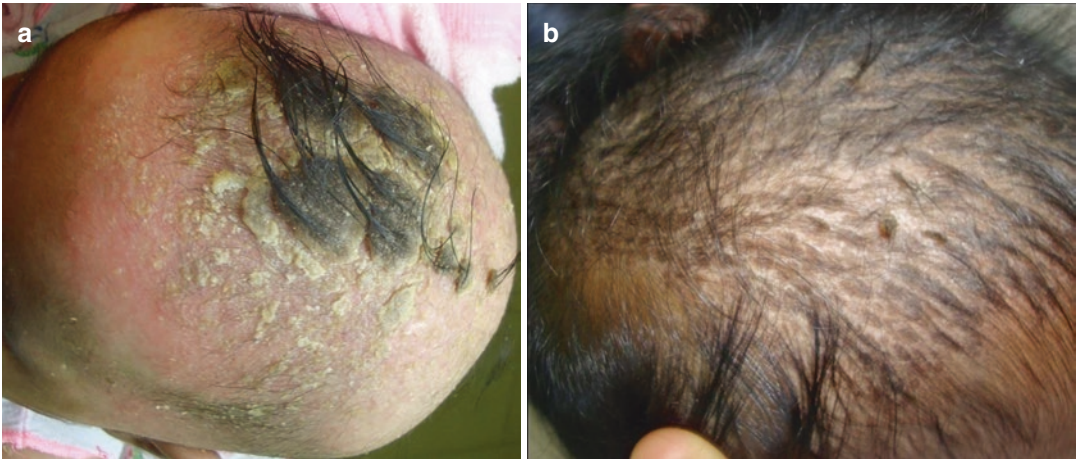


Fig. 1.1 (a, b) Cradle cap. Large flakes of brownish scale are seen on the scalp, especially over the vertex and frontal regions, and may become matted into large plaques of

crust. There is usually minimal inflammation, but the eyebrows may be involved



Fig. 1.2 Infantile seborrhoeic dermatitis. More macerated erythema occurs in the skin folds, especially the neck and the inguinal regions



Fig. 1.3 Infantile seborrhoeic dermatitis. Typically the inflammation resolves with transient hypopigmentation, which is pronounced in children with pigmented skin



Fig. 1.4 Infantile seborrhoeic dermatitis. Note skin folds are affected (in contrast to napkin dermatitis where skin folds are spared)



Fig. 1.5 Infantile seborrhoeic dermatitis. Post-inflammatory depigmentation is marked since this child's skin colour is very dark



Fig. 1.6 (a, b) Atopic eczema. In infants, atopic eczema characteristically begins on the face



Fig. 1.7 (a, b) Atopic eczema on flexures in a 9-month-old baby girl



Fig. 1.8 Napkin dermatitis in a 3-month-old infant. Note skin folds are spared in contrast to seborrhoeic dermatitis or infantile psoriasis. Dermatitis can spread up to lower back or abdomen when infants sleep on a rubber mattress



Fig. 1.9 Napkin dermatitis in an infant. This dermatitis is usually confined to nappy areas



Fig. 1.10 Napkin dermatitis with candida superinfection in a preterm neonate (photographed by Dr. Maduranga Mendis, medical officer, neonatology unit, General Hospital Kalutara, Sri Lanka)

ally rupture within 24 h and are followed by bran-like desquamation. Lesions are asymptomatic.

Miliaria rubra (prickly heat, sweat rash) comprises erythematous papules and papulovesicles about 1–4 mm in diameter, on a background of macular erythema. Lesions can be itchy or sore.

Miliaria pustulosa: Some of the miliaria rubra lesions are pustular, but not infected.

Miliaria profunda (granulomatous giant centrifugal variant) is very uncommon in neonates as it usually occurs in adults where there have been repeated episodes of miliaria rubra.

Management: Miliaria crystalline improves spontaneously when sweat ducts are mature. Remove from conditions of high heat/humidity and any occlusive clothing or bedding (Figs. 1.11, 1.12 and 1.13).



Fig. 1.11 Miliaria crystallina resembling drops of water in a 10-day-old baby on the face and the trunk



Fig. 1.12 Miliaria crystallina in a new born baby



Fig. 1.13 Miliaria pustulosa in an older child

1.2 Infections and Infestations

1.2.1 Hand, Foot and Mouth Disease

Coxsackie A viral infection, most usually A16, and infection with A6, Coxsackie B and enterovirus 71 have also been described. Highly contagious and widespread outbreaks occur every year.

Clinical presentation: Mild malaise, oral ulcers and hand and foot vesicular eruption. Crops of vesicles are grouped in elbows and knees. Very rarely, a widespread vesicular eruption occurs over the buttocks, trunk and perioral area. In some, the eruption may be papular or maculopapular without vesicles. When it resolves it may form marked scaling or postinflammatory hyperpigmentation which settles spontaneously (Sterling 2016; Higgins and Glover 2016) (Figs. 1.14, 1.15, 1.16 and 1.17).



Fig. 1.14 Hand, foot and mouth disease showing small vesicles with surrounding erythema on the sole and palms



Fig. 1.15 (a, b) Hand, foot and mouth disease, vesicles are commonly crops around knees and elbows, but it can spread to all over the body

Diagnosis: Clinical.

Differential diagnosis: Oral lesions may suggest herpes simplex or Stevens–Johnson syndrome and, if widespread, chickenpox or Kaposi varicelliform eruption.

Management: Symptomatic treatment only is required in many children. In very extensive



Fig. 1.16 Hand, foot and mouth disease, vesicular eruption on the trunk, upper limbs and palms

eruption, oral acyclovir is given three times daily for 3 days.

1.2.2 Varicella/Chickenpox

The incubation period is 14–21 days. Small erythematous papules and vesicles appear on the trunk, scalp or genital regions. The onset of the rash may be preceded by 1–2 days of malaise and fever. Lesions in different stages of evolution are evident. Centripetal pattern. The child should be isolated until the lesions have crusted over (Fig. 1.18).

1.2.3 Impetigo

Caused by *Staphylococcus aureus* but less frequently *Streptococcus pyogenes*. Honey-coloured



Fig. 1.17 Resolving lesions in hand, foot and mouth disease showing postinflammatory hyperpigmentation (a) and scaling (b). Scaling is more common



Fig. 1.18 Chickenpox, a characteristic feature, is the presence of lesions at different stages in each site. Fever and malaise precede vesicular eruption by a day or 2



Fig. 1.19 Non-bullous impetigo

crusts appear on a background of erythema. Bullous and non-bullous forms exist (Hay and Morris-Jones 2016; Ghosh 2015) (Figs. 1.19, 1.20, 1.21 and 1.22).

1.2.4 Staphylococcal Scalded Skin Syndrome

Causative organisms: *Staphylococcus aureus* strains producing exfoliative toxins.

It is an exfoliative dermatosis in which most of the body surface becomes tender and erythematous and the superficial epidermis strips off. Children (under the age of 6 years) and neonates are most commonly affected by the generalized form of SSSS; rarely adults may be affected (Leung et al. 2018).

Differential diagnosis: Toxic epidermal necrolysis (TEN).

Treatments: Depend on the antibiotic sensitivity • flucloxacillin • clindamycin • temocillin



Fig. 1.20 Bullous impetigo

• tigeicycline • daptomycin, if MRSA is suspected, • vancomycin • tobramycin (Figs. 1.23 and 1.24).

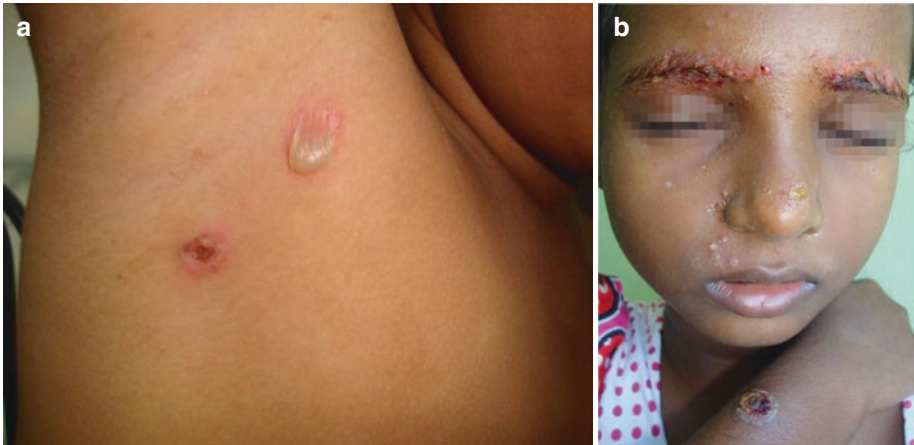


Fig. 1.21 (a, b) Bullous impetigo showing flaccid bullae filled with pus



Fig. 1.22 Folliculitis in a 2-week-old baby. He was treated with topical fucidin cream twice daily and reviewed in 3 days. Note complete clearance of the lesions leaving mild postinflammatory hypopigmentation

1.2.5 Pitted Keratolysis

Causative organisms: *Corynebacterium* spp.

Clinical presentation: Numerous superficial punched out erosions on soles, undersurfaces of the toes and rarely palms. But pressure-bearing or friction areas are most common. Hyperhidrosis is often associated, sometimes with maceration, stickiness and a foul odour. Soaking the feet in water for 15 min causes swelling of the horny layer and accentuates the lesions. Irritation is minimal, and in most cases, patients are unaware of the condition (Makhecha et al. 2017; Pranteda et al. 2014) (Fig. 1.25).

Differential diagnosis: Tinea pedis.

Management: Treat hyperhidrosis with potassium permanganate soaks, aluminium chloride and iontophoresis, if resistant botulinum toxin. Topical fucidin ointment, imidazoles such as clotrimazole are also effective.

1.2.6 Cutaneous Fungal Infections

Candida infection: Secondary colonization of eroded or macerated skin in intertriginous areas, especially the napkin region, may occur but is much less common since the decline in diaper



Fig. 1.23 Staphylococcal scalded skin syndrome in an infant. A 2-month-old baby came with fever and generalized erythema. On examination scaling of skin was noted which was marked around the orifices and flexures. Initial

widespread erythematous eruption may not be obvious in dark-skinned infants as seen in this baby. That may lead to late diagnosis unless vigilant

dermatitis. Satellite pustules are characteristic. If children get recurrent candida infection, underlying predisposing factors should be sought; inherited immune deficiency, or acquired habit disorder (Figs. 1.26, 1.27 and 1.28).

Tinea corporis: nearly always acquired from an older child or from an infected adult (mother, grandmother, caretaker, etc.) (Figs. 1.29 and 1.30).

Tinea capitis: mildly scaly and mildly itchy well-demarcated patch on the scalp (Figs. 1.31 and 1.32).

1.2.7 Scabies

Infestation with the mite *Sarcoptes scabiei*. In infants, burrows may be seen on the palms and soles more characteristically than in the finger webs, and the wrists are also commonly involved. Burrows can appear quite inflammatory with sur-

rounding secondary eczematous changes. Nodular lesions develop in the axillae and genital region if the infection is untreated. Post-scabietic pruritus may be prolonged and symptomatic treatment is usually required (Figs. 1.33, 1.34 and 1.35).

1.2.8 Cutaneous Larva Migrans

Creeping or migrating lesion due to the presence of moving parasites in the skin. These are all hookworms of mainly dogs and cats. The lesion is exceedingly itchy, slightly raised, skin coloured or pink and forms bizarre, serpentine patterns.

Causative organisms: *Ancylostoma braziliense*: Dog and cat hookworm is the commonest cause of creeping eruption in humans. • *Ancylostoma caninum*: Dog hookworm.

Treatment: Ivermectin 200 µg/kg orally once daily for 1–2 days or albendazole 400 mg orally for 3 days (Fig. 1.36).



Fig. 1.24 A 3-year-old child with *Staphylococcus* Scalded Skin Syndrome (SSSS). Generalized erythema, oedema and scaling in a 3-year-old ill child. Note peri-

ificial involvement, generalized desquamation, superficial blister formation and positive Nikolsky sign

1.2.9 Molluscum Contagiosum

Characterized by discrete, pearly, umbilicated papules 2–5 mm in diameter. The eruption is usually self-limiting but is often a source of great parental concern.

Management: cryotherapy (if tolerated) or topical treatment with either hydrogen peroxide creams or commercially available potassium hydroxide solutions can speed resolution (Fig. 1.37).

1.3 Reactive Conditions

1.3.1 Acute Haemorrhagic Oedema in Infancy

Benign, cutaneous, leukocytoclastic vasculitis, arising after respiratory infection, medication administration or immunization. It is thought

likely to be an immune complex-mediated vasculitis. The condition affects children between the ages of 4 months and 2 years, with males being affected twice as frequently as females. Fever is mild and systemic disturbance is minor. Resolution occurs within 3 weeks, and recurrences are not a feature (Fig. 1.38).

1.3.2 Papular Urticaria

Papular urticaria arises as a result of a hypersensitivity reaction to insect bites.

Clinical presentation: crops of more or less symmetrically distributed, itchy papules and papulovesicles most frequently on exposed areas of the extremities. They are often heavily excoriated, and secondary bacterial infection is common.

Management: topical steroids and systemic antihistamines. Avoid insect bites (Fig. 1.39).



Fig. 1.25 Pitted keratolysis in a 12-year-old child who had hyperhidrosis



Fig. 1.27 Candida intertrigo in a 3-month-old baby, note satellite lesions which are characteristic



Fig. 1.28 Candida finger webs in a 2-year-old child due to frequent handling of water, identified as an obsessive compulsive disorder



Fig. 1.26 Candida intertrigo in a preterm neonate (photographed by Dr. Maduranga Mendis, medical officer, neonatology unit, General Hospital Kalutara, Sri Lanka)



Fig. 1.29 Tinea corporis in a 9-month-old baby. Note the well-marginated inflammatory edge. This baby got wide-spread tinea infection from her untreated mother



Fig. 1.30 Tinea corporis in a 4-month-old baby. This can easily misdiagnose to nummular eczema



Fig. 1.31 (a, b) Tinea capitis. Note well-demarcated mildly scaly patch. This can appear as multiple scaly patches

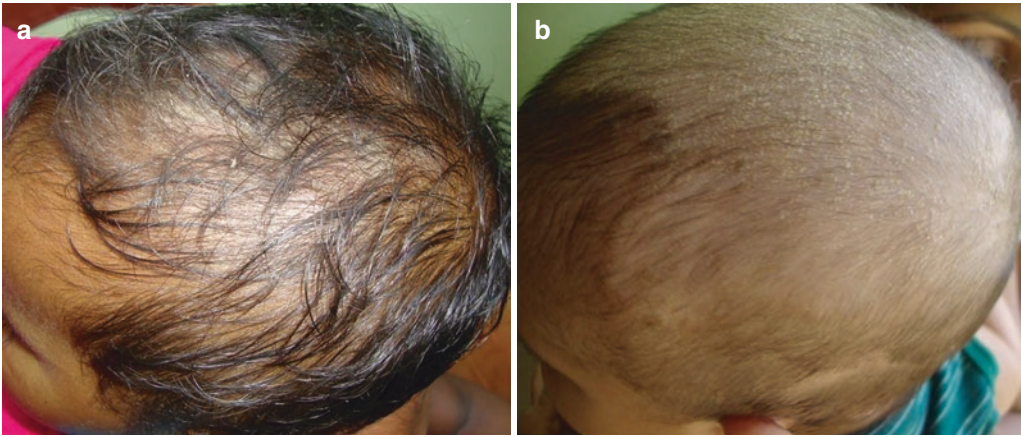


Fig. 1.32 (a, b) Tinea capitis can be widespread scaling. Note well-demarcated lesion



Fig. 1.33 A 1-month-old baby got scabies from her mother. Note mother has itchy papules and burrows on finger webs and dorsum of hand



Fig. 1.34 Scabies in a 6-month-old baby. Burrows are clearly visible on the trunk

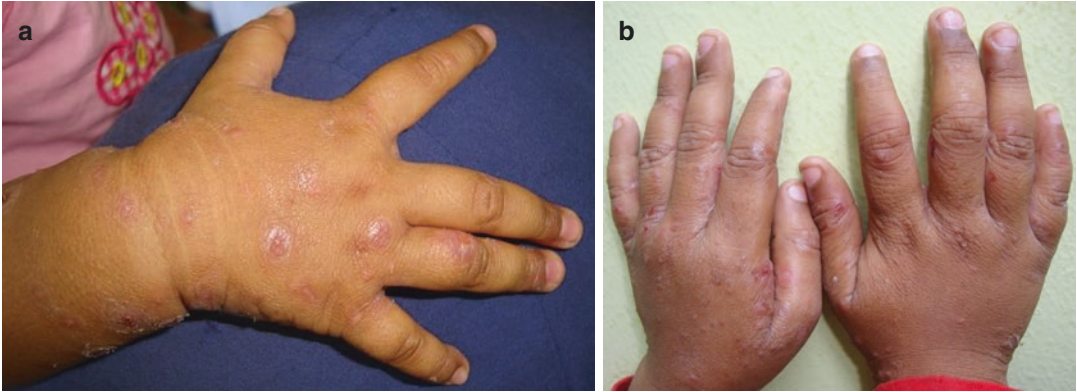


Fig. 1.35 (a, b) Scabies in two siblings aged 9 months and 2 years

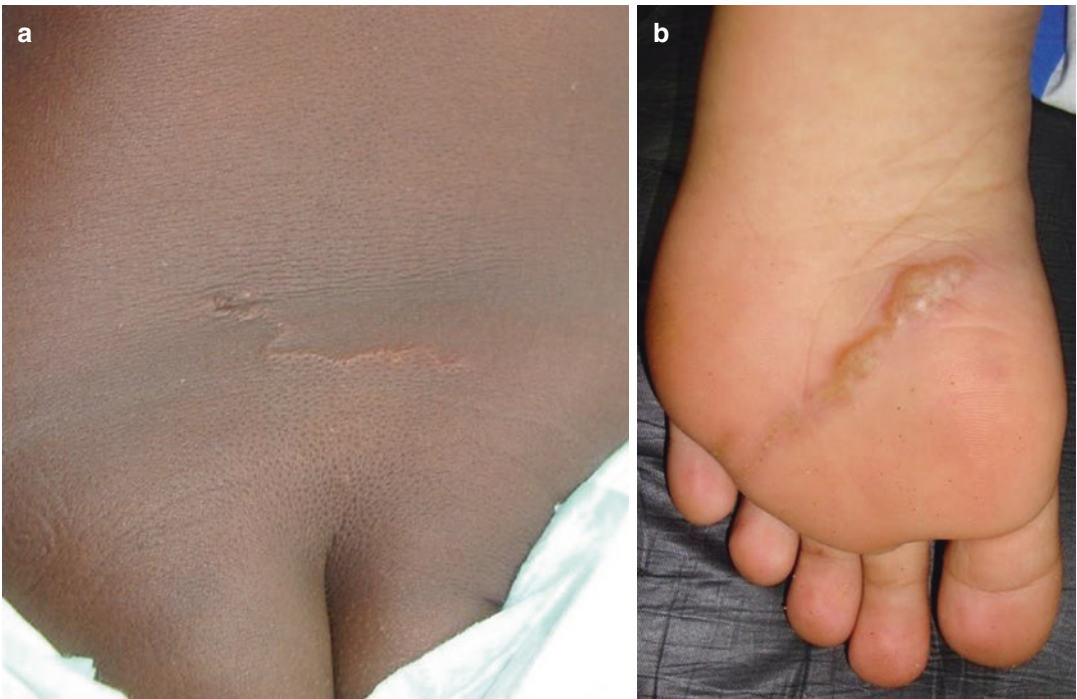


Fig. 1.36 (a, b) Cutaneous larva migrans. These are exceedingly itchy and serpiginous and creep or migrate due to the moving parasites in the skin. On the sole blisters has formed along the migrating pathway due to thick stratum corneum



Fig. 1.37 (a, b) Molluscum contagiosum in a 6-month-old baby and his sibling 7-year-old brother



Fig. 1.38 Acute haemorrhagic oedema in infancy. These lesions resolved spontaneously within 1 week (picture courtesy Dr. Anoma Weerasinghe, consultant paediatrician, General Hospital Kalutara, Sri Lanka)

1.4 Developmental Problems

1.4.1 Pigmentary Mosaicism

Streaks and whorls of hypo- or hyperpigmentation following Blaschko's lines. Infants with pigmentary mosaicism should be thoroughly assessed with particular attention to development, the internal organs and skeletal and ophthalmological abnormalities (Figs. 1.40, 1.41 and 1.42).

1.5 Genetic Disorders

1.5.1 Epidermolysis Bullosa

Epidermolysis bullosa comprises a group of genetically determined skin fragility disorders characterized by blistering of the skin and mucosae following mild mechanical trauma (McGrath 2016). Epidermolysis bullosa is categorized according to inheritance, clinical presentation and level of cleavage (Figs. 1.43, 1.44, 1.45, 1.46 and 1.47).

1.5.2 Inherited Generalized Cutis Laxa

Cutis laxa is characterized clinically by lax pendulous skin that only slowly recoils when pulled.

Histology: there is loss and fragmentation of elastic tissue in the dermis (Fig. 1.48).

1.5.3 Hypohidrotic Ectodermal Dysplasia

X-linked hypohidrotic ectodermal dysplasia (Freire-Maia 1-2-3-4) is the most common

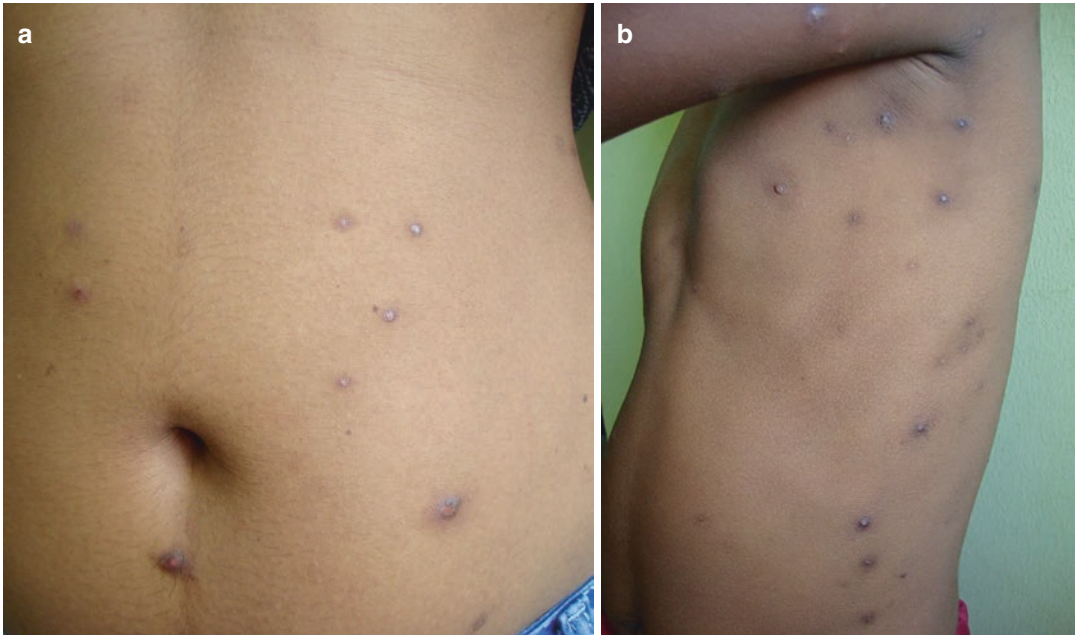


Fig. 1.39 (a, b) Papular urticaria

of the ectodermal dysplasias. The disorder is characterized by hypotrichosis with fine, slow-growing scalp and body hair, sparse eyebrows, hypohidrosis, nail anomalies and hypodontia.

Clinical presentation: The hair is sparse, dry and lusterless with a light colour. Eyebrows are scanty or absent occasionally; just the outer two-thirds are missing. Eyelashes may be normal, sparse or completely absent. Hair on the torso and extremities is usually absent. Secondary sexual hair in the beard, pubic and axillary regions is variably present and may be normal.

Peg-shaped primary and secondary teeth are typical.

Decreased sweating leads to heat intolerance and enhances dryness of the skin. Sweating is often severely diminished or absent due to a paucity or absence of eccrine glands. An absence of sweating leads to an inability to thermoregulate by evaporative cooling, and hyperthermia can occur with physical exertion or in a warm environment (Itin 2016).

Disease course and prognosis: Early diagnosis is important to avoid life-threatening complications induced by hyperthermia and infections.

Avoiding heat and physical overexertion is the most important preventative measure to recommend. Cooling the body with wet clothing and cool drinks is the only efficient way of treating hyperthermia. Orthodontic intervention is necessary (Fig. 1.49).

1.5.4 Acrodermatitis Enteropathica

Autosomal recessive disorder affecting the intestinal absorption of zinc. The resulting zinc deficiency leads to dermatitis, alopecia and diarrhoea. This is very rare (approximately 1: 500,000). Zinc is a cofactor for many enzymes. The skin, intestine and immune system are most severely affected because of their rapid cell turnover.

Clinical features: Patients present with apathy or irritability and a rash around the mouth and anus and on the hands and feet. Symptoms start after weaning in breastfed babies and at 4–10 weeks of age if they are formula-fed. Erythema progresses to vesicles, bullae, pustules, desquamation and crusting. There is alopecia and frequently blepharitis, conjunctivitis and photophobia. Infections are common, including secondary infections of the



Fig. 1.40 (a, b) Pigmentary mosaicism showing streaks and whorls of hypo- or hyperpigmentation following Blaschko's lines



Fig. 1.41 Congenital vitiligo in a 9-month-old baby



Fig. 1.42 Lichen striatus along the Blaschko's lines on the trunk

skin, for example, with *Candida albicans*. Wound healing is poor, and many patients have diarrhoea and growth faltering. Without treatment, the condition can be fatal, but some patients survive into adulthood (Morris 2016).

Investigations: The serum zinc concentration is usually low but can be normal. Decreased zinc absorption can be demonstrated using radioisotopes. But it is easier to undertake a trial of zinc therapy: patients respond within a week.



Fig. 1.43 Epidermolysis bullosa simplex



Fig. 1.45 Extensive erosions in a newborn with generalized junctional epidermolysis bullosa



Fig. 1.46 A 20-day-old baby with epidermolysis bullosa (photographed by Dr. Maduranga Mendis, medical officer, neonatology unit, General Hospital Kalutara, Sri Lanka)



Fig. 1.44 Generalized junctional epidermolysis bullosa. Since we do not have facilities for molecular diagnostics, these babies were diagnosed based only on clinical features



Fig. 1.47 An 8-month-old baby girl with generalized intermediate junctional epidermolysis bullosa. Eroded areas are healed with atrophic scarring. Post-inflammatory hypopigmentation or depigmentation present in areas of scarring



Fig. 1.48 (a, b) Inherited generalized cutis laxa in two infants



Fig. 1.49 Hypohidrotic ectodermal dysplasia. Hypotrichosis with fine, slow-growing scalp and body hair, sparse eye-brows and sparse eye lashes. This baby's nails were normal



Fig. 1.50 Acrodermatitis enteropathica. A 5-month-old baby before Zn supplements, showing the rash around (a) the anus and (b) the mouth



Fig. 1.51 Acrodermatitis enteropathica. Four days after Zn supplements. Response to the trial of Zn therapy is very dramatic and is diagnostic

Management: Patients respond to oral zinc sulphate within a few days. The normal dose is 150–400 mg/day in childhood; a lower dose may suffice after puberty. A 400–500 mg/day is needed during pregnancy. Monitoring for copper deficiency should be undertaken (Figs. 1.50, 1.51, 1.52 and 1.53).

1.6 Miscellaneous Conditions

1.6.1 Hair Loss in Infancy

Occipital hair loss: Shedding of hair occurs during the 7th to 8th month in utero in all areas except the occiput, where shedding is delayed until 2–3 months postpartum, leading to the normal occipital alopecia in this age group. Trauma due to repeated friction also may contribute (Brar et al. 2019) (Fig. 1.54).

Alopecia areata: Is relatively rare in the 1st year of life, and early onset tends to indicate a poor prognosis (Fig. 1.55).

1.6.2 Circumscribed Alopecia of Congenital Origin

Differential diagnosis. Alopecia areata and the acquired cicatricial alopecias.

Sebaceous naevus (organoid naevus). Sebaceous naevus usually presents in childhood as a hairless patch on the scalp, often in a linear configuration. The affected skin may have a yellowish hue (Figs. 1.56 and 1.57).

Aplasia cutis. Aplasia cutis is a heterogeneous group of conditions characterized by the localized absence of a portion of skin at birth. It most

Fig. 1.52 Acrodermatitis enteropathica in an 8-week-old baby



Fig. 1.53 Five days after Zn supplements. Response to the trial of Zn therapy is very dramatic and is diagnostic



Fig. 1.54 Occipital alopecia in a 3-month-old baby is normal at this age and self-limiting



Fig. 1.55 A patch of alopecia areata in a 1-year-old child



Fig. 1.56 Naevus sebaceus on the scalp in a new born



Fig. 1.57 Naevus sebaceus in a new born appears as patchy alopecia. Note the yellowish hue in affected skin (photographed by Dr. Maduranga Mendis, medical officer, neonatology unit, General Hospital Kalutara, Sri Lanka)

commonly presents as a single patch on the scalp vertex. At birth the affected skin may be superficially or deeply ulcerated or may have healed leaving an atrophic scar.

Sutural alopecia. Multiple patches of hair loss overlying the cranial sutures (Fig. 1.58).

Triangular alopecia. This usually presents during childhood although often referred to as congenital it is often not noticed at birth.

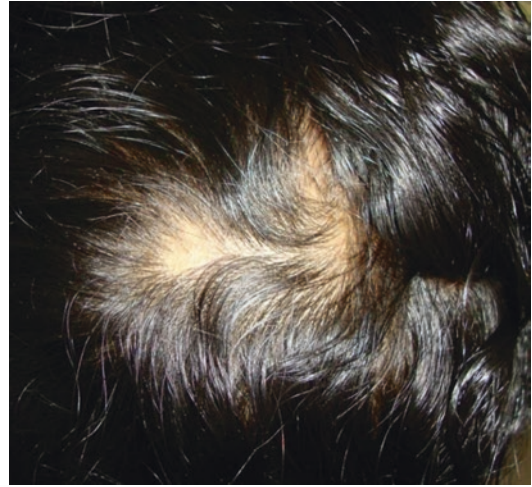


Fig. 1.58 Sutural alopecia. Multiple patches of hair loss overlying the cranial sutures

1.6.3 Congenital Localized Hypertrichosis

Congenital localized hypertrichosis refers to both areas of excess hair present at birth and hamartomas that may have a delayed clinical presentation. They may occur in isolation as an area of increased hair density or in association with congenital melanocytic naevi, Becker naevi, spinal dysraphism or neurofibromas (Messenger et al. 2016) (Fig. 1.59).

Congenital melanocytic naevi and neurofibromas. Congenital melanocytic naevi are often associated with prominent terminal hairs. The hair may be present from infancy or may develop after puberty.

Lumbosacral hypertrichosis. Lumbosacral hypertrichosis or the faun tail sign is a tuft of hair in the lumbosacral region that is often associated with spina bifida or diastematomyelia.

Naevoid hypertrichosis. Naevoid hypertrichosis refers to a localized area of hypertrichosis. This is usually well circumscribed (Fig. 1.60).

1.6.4 Juvenile Xanthogranuloma

Juvenile xanthogranuloma is a well-circumscribed yellowish nodule with an erythematous margin. Localized cutaneous JXG heals spontaneously, sometimes leaving atrophic scars.



Fig. 1.59 Congenital naevoid hypertrichosis

Diagnosis: clinical, can confirm by histopathology (Figs. 1.61, 1.62 and 1.63).

1.6.5 Anetoderma

Anetoderma is a circumscribed area of slack skin associated with a loss of dermal substance on

palpation and a loss of elastic tissue on histological examination.

Primary anetoderma remains unchanged throughout life, and new lesions often continue to develop for many years.

Secondary anetoderma is seen in association with systemic or chronic cutaneous lupus erythematosus and lupus profundus.

Investigations: In patients with primary anetoderma, it is important to test for antiphospholipid syndrome and treat appropriately, e.g. with aspirin or warfarin.

Management: No specific treatment exists (Figs. 1.64 and 1.65).

1.6.6 Mastocytosis

Mastocytosis in infancy is usually limited to the skin, with three distinct clinical presentations: maculopapular (formerly urticaria pigmentosa), diffuse cutaneous mastocytosis and solitary mastocytoma.

Clinical presentation: Present with patchy hyperpigmentation which urticate on rubbing.

Investigations: A skin biopsy is usually required to confirm a clinical diagnosis of cuta-



Fig. 1.60 Congenital hypertrichosis on both pinnas of the ears in a baby boy since birth (hairy tragus)

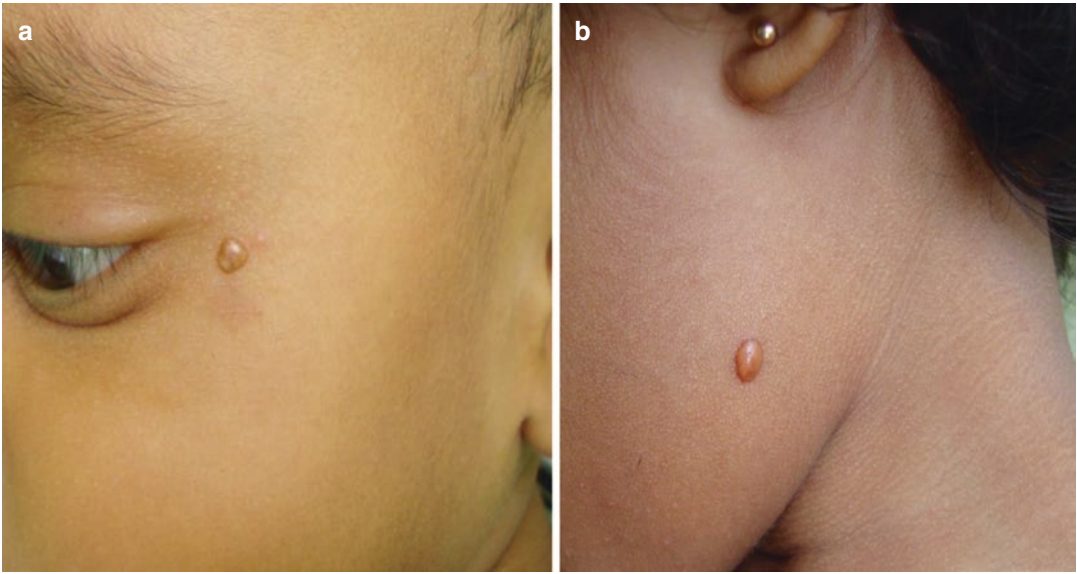


Fig. 1.61 (a, b) Solitary juvenile xanthogranuloma

Fig. 1.62 Juvenile xanthogranuloma is characterized by a dense infiltrate of small histiocytes in the dermis, which stain positively for factor XIIIa, CD68, CD163, CD14 and fascin. H & E Stain X 100 (picture courtesy Dr. Priyanka H. Abeygunasekara, Consultant Pathologist, National Cancer Institute Maharagama, Sri Lanka)

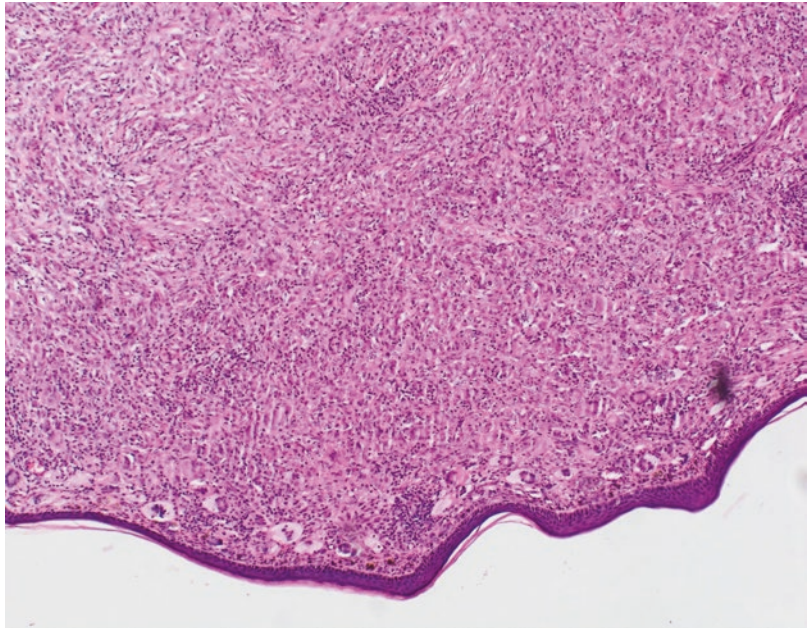


Fig. 1.63 Juvenile xanthogranuloma, Touton giant cells, seen in 85% of JXG cases, can be distinguished by the characteristic wreath of nuclei around a homogenous eosinophilic centre and prominent xanthomatization in the periphery H & E stain X 400 (picture courtesy Dr. Priyanka H. Abeygunasekara, Consultant Pathologist, National Cancer Institute Maharagama, Sri Lanka)

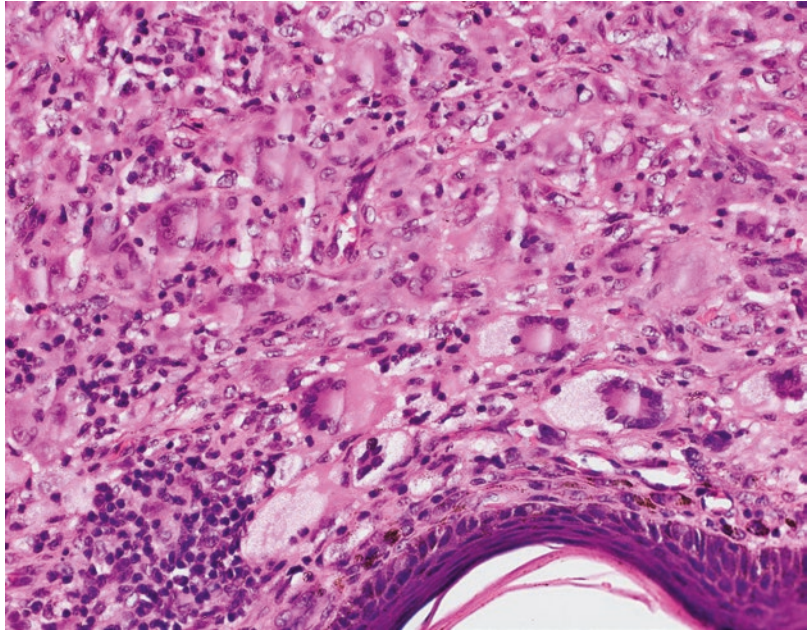


Fig. 1.64 Anetoderma in a 3-month-old baby

neous mastocytosis. Solitary mastocytoma can be diagnosed clinically.

Histopathology: The epidermis is normal apart from an increase in melanin. Mast cell numbers are increased in the dermis of all types of mastocytosis. They are well demonstrated by toluidine blue, Giemsa, tryptase or chloroacetate esterase stains in formalin-fixed biopsies.

Disease course and prognosis: Most mastocytomas resolve in childhood. Around 50% of children with urticaria pigmentosa clear by adolescence.

Management: Parents of infants with extensive skin involvement should be given advice on the avoidance of factors known to stimulate mast cell degranulation, including aspirin, nonsteroidal anti-inflammatory drugs, codeine, opiates, polymyxin B and intravenous radiograph contrast fluids and MRI contrast media. (Figs. 1.66, 1.67, 1.68, 1.69, 1.70 and 1.71).

1.6.7 Milia

Small, firm, white papules, predominantly occurring on the face of newborn babies and infants, are common and harmless. These are follicular epidermal cysts. If milia are exceptionally extensive or persistent, or whose distribution is atypical, may associate with syndromes.

Management: this disappears spontaneously.

1.6.8 Neonatal Lupus Erythematosus (see Chap. 31)

Well-defined areas of macular or slightly elevated erythema, frequently annular, occurring predominantly on the face, particularly the forehead,



Fig. 1.65 Localized anetoderma in an otherwise normal baby

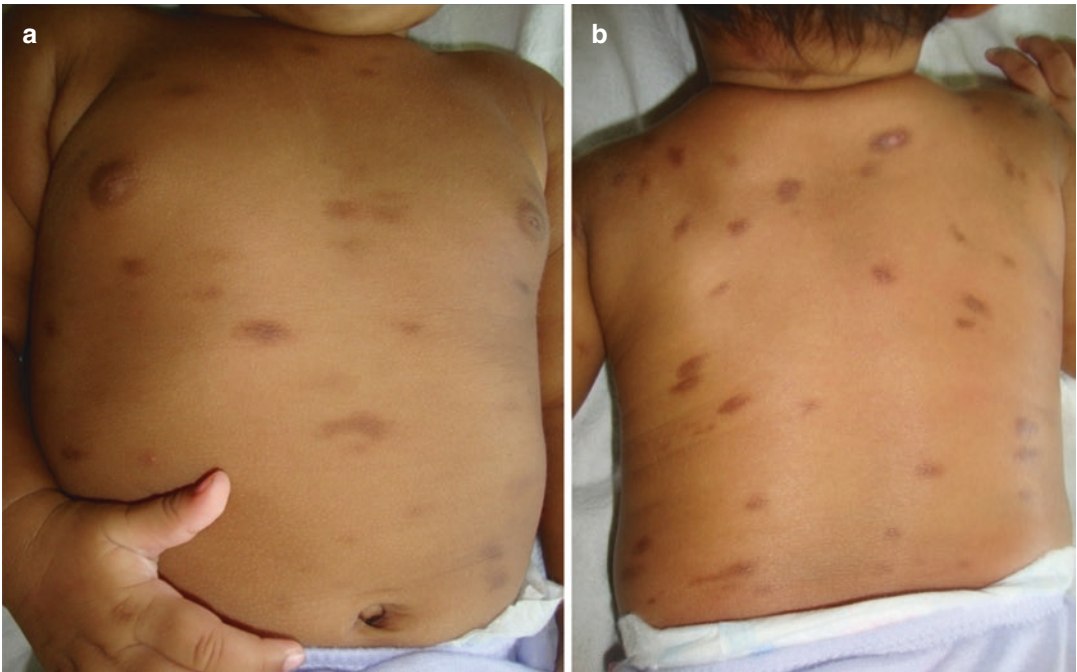


Fig. 1.66 Diffuse and extensive maculopapular mastocytosis (formerly urticaria pigmentosa) in a 2-month-old baby (a) front and (b) back



Fig. 1.67 Diffuse and extensive maculopapular mastocytosis in a 7-month-old baby girl



Fig. 1.68 Acute urticaria in a 7-month-old baby



Fig. 1.69 (a, b) Solitary mastocytoma in infants

Fig. 1.70 Mastocytosis (H & E stain X 400). The epidermis is normal apart from an increase in melanin. The mast cells are usually oval or spindle-shaped. Mast cell infiltrates are predominantly found around blood vessels and skin appendages in the papillary dermis (picture courtesy Dr. Priyanka H. Abeygunasekara, Consultant Pathologist, National Cancer Institute Maharagama, Sri Lanka)

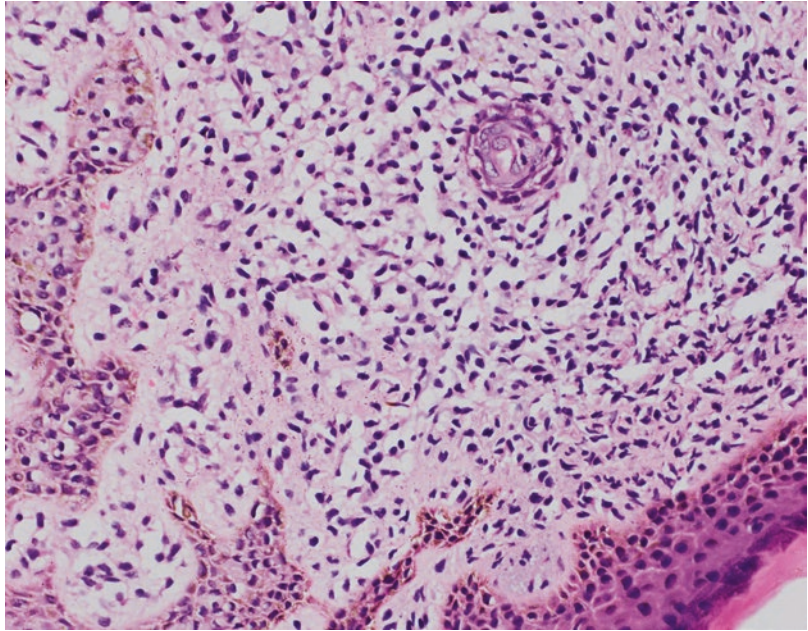
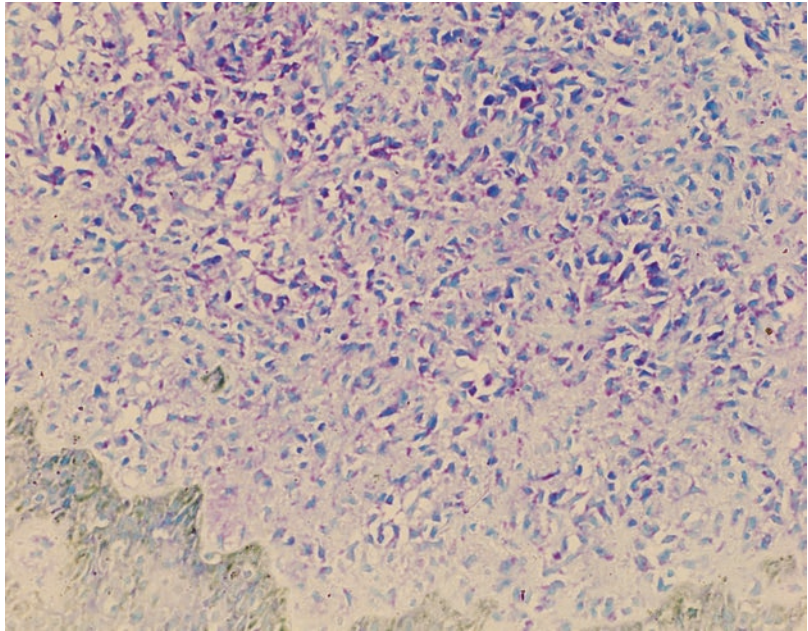


Fig. 1.71 Urticaria pigmentosa, mast cells stained with Giemsa X 400 (picture courtesy Dr. Priyanka H. Abeygunasekara, Consultant Pathologist, National Cancer Institute Maharagama, Sri Lanka)



temples and upper cheeks and on the scalp and neck (A ‘spectacle-like’ distribution of lesions around the eyes is especially characteristic. It is now accepted that this disease is provoked in the fetus or newborn infant by maternal IgG autoantibodies that have crossed the placenta.

Management: The skin lesions of neonatal LE require no treatment, but sun protection is essential.

1.6.9 Collodion Baby (see Chap. 7)

Highly characteristic clinical entity present at birth where a child is born with an ‘extra’ skin resembling a shiny membrane or collodion. Almost 90% of collodion babies will go on to develop a severe form of autosomal recessive ichthyosis in the first few weeks of life: lamellar ichthyosis and non-bullous ichthyosiform erythroderma (syn. congenital ichthyosiform erythroderma) are the most common (Oji et al. 2016).

1.6.10 Harlequin Ichthyosis (see Chap. 7)

Harlequin ichthyosis has much thicker skin, which typically encases the baby like a suit of armour with deep fissures. Harlequin ichthyosis shows almost fusion of the fingers and toes with thick palmoplantar skin. Finally, the ears are usually bound down to the scalp in harlequin ichthyosis, whereas they are normally ‘free’ in collodion babies.

1.6.11 Neonatal Purpura Fulminans

Potentially lethal disorder characterized by progressive haemorrhagic necrosis of the skin associated with cutaneous vascular thrombosis.

1.6.12 Median Raphe Cysts

Cystic or nodular and linear swellings of the ventral penis occur near the glans. In adolescence or

adulthood, they may become traumatized or infected with staphylococci, gonococci or *Trichomonas* and present as tender, erythematous and purulent nodules.

Histology: they are either dermoid or mucoid, depending on their embryology or epithelial lining. Very rarely, the basal epithelial lining of the cysts may contain melanocytes, imparting a brown-black pigment to the lesion.

Congenital cystic median raphe anomalies may remain unobtrusive until adulthood (Fig. 1.72).

1.6.13 Granuloma Annulare

This is most common in children and young adults but can occur at any age.

Clinical features: Four clinical variants identified, localized GA (commonest type in children), generalized or disseminated GA (mostly in adults), perforating GA (in all ages, infancy and



Fig. 1.72 Congenital cystic median raphe anomaly in a 2-year-old boy



Fig. 1.73 Granuloma annulare, localized GA. Typically presents as a ring of small, smooth, skin coloured or erythematous annular plaques. Stretching the skin enables the plaques to be seen more readily



Fig. 1.74 Granuloma annulare, localized GA. Dorsa of the hands, knuckles, fingers and feet are the commonest sites although it can appear at any site

Fig. 1.75 Histopathology of Granuloma annulare H & E X100. Necrobiotic palisading granuloma showing necrobiosis surrounded by a palisade of histiocytes (picture courtesy Dr. Anuruddha Galappatti, consultant histopathologist, General Hospital Kalutara, Sri Lanka)

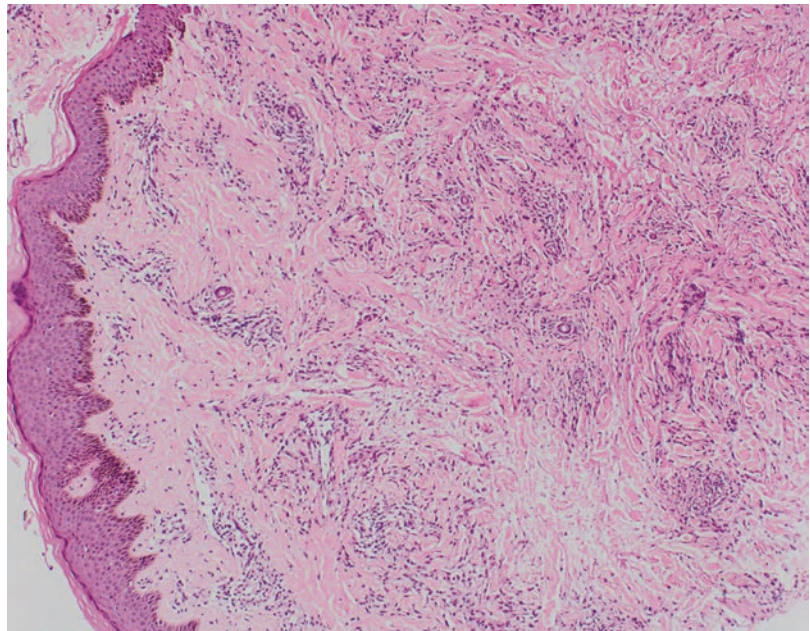
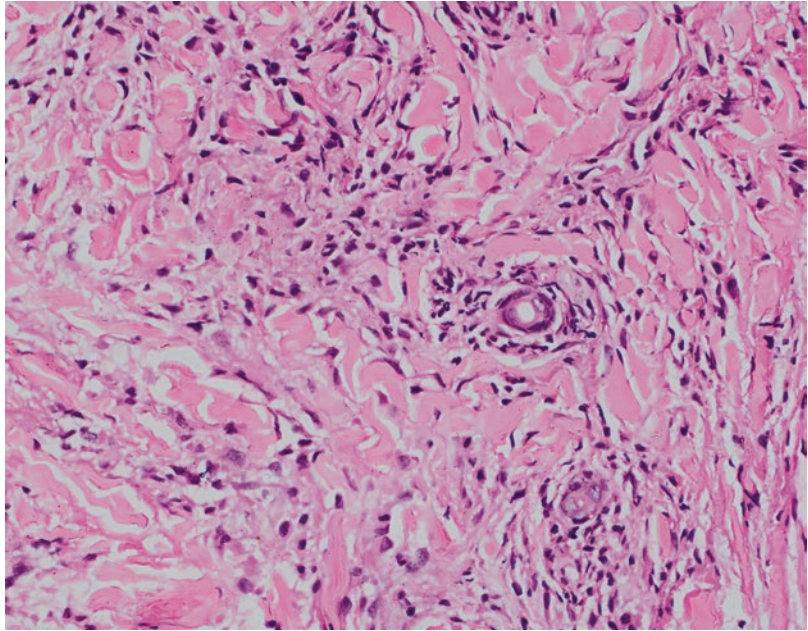


Fig. 1.76 Histopathology of Granuloma annulare H & E X400. Close-up view showing necrobiotic collagen and multinucleated giant cells (picture courtesy Dr. Anuruddha Galappatti, consultant histopathologist, General Hospital Kalutara, Sri Lanka)



in HIV) and subcutaneous GA (uncommon but mostly in children).

Histopathology: The most characteristic histological lesion in GA is the necrobiotic granuloma. **Management:** No treatment and reassurance of eventual resolution in localized GA. For generalized GA PUVA, laser, systemic therapies have tried with variable success (Bourke 2016) (Figs. 1.73, 1.74, 1.75 and 1.76).

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