Congenital Naevi and Melanocytic Naevi

2

Ranthilaka R. Ranawaka

2.1 Introduction

Classification of congenital naevi is very complex and changing frequently. Although extensive classification is included here, only common naevi are illustrated. This chapter discusses congenital epidermal, pigment cell, melanocytic and connective tissue naevi (Tables 2.1 and 2.2). Also illustrates and discusses naevi in unusual sites, unusual morphology and currently unclassified naevi.

2.2 Congenital Epidermal Naevi

1. Verrucous epidermal naevus
2. Sebaceous naevus
3. Follicular naevi
4. Apocrine naevi
5. Eccrine naevi
6. Inflammatory epidermal naevi
7. Other naevoid epidermal disorders
8. The epidermal naevus syndromes

Congenital epidermal naevi (CEN) are by definition present at birth or become apparent in the first years of life. Become more pronounced with

Table 2.1 Classification of congenital naevi (Kinsler and Sebire 2016)

Clinical phenotypic classification	*Histological classification*
2.1. Congenital epidermal naevi 2.2. Congenital pigment cell naevi 2.3. Congenital connective tissue naevi These can each be divided into: • Cutaneous involvement only—single or multiple lesions • Syndromic—associated with non-cutaneous features	2.1. Within congenital epidermal naevi, there are subclassifications based on the predominant cell type seen on histology: (a) Keratinocytic naevi (b) Sebaceous naevi (c) Follicular naevi (d) Eccrine/apocrine naevi 2.2. Within congenital pigment cell naevi there are subclassifications based on the histology of the pigment cells: (a) Melanocytic naevus (b) Blue naevus (c) Spitz naevus (d) Naevus spilus
Genetic classification For congenital naevi, there are also subclassifications based on the causative genetic mutation	2.3. Within congenital connective tissue, naevi there are subclassifications based on the predominant cell type seen on histology: (a) Collagen naevi (b) Elastic tissue naevi (c) Mucinous naevi (d) Fat naevi

Kinsler VA, Sebire NJ (2016). Congenital Naevi and Other Developmental Abnormalities Affecting the Skin. In: Burns T, Breathnach S, Cox N, Griffiths C (eds) Rook's textbook of dermatology, 9th edn. Wiley Blackwell Science, Oxford, p 75.1

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

R. R. Ranawaka (✉)
General Hospital Kalutara, Kalutara, Sri Lanka

R. R. Ranawaka et al. (eds.), *Atlas of Dermatoses in Pigmented Skin*,
https://doi.org/10.1007/978-981-15-5483-4_2

age. Single CEN lesions can be either round or linear, but larger or multiple lesions are Blaschko linear in distribution. Epidermal naevi are by their nature very superficial and are therefore raised and can have the appearance of being "stuck on" to the surface of the skin rather than intrinsic to it (Figs. 2.1, 2.2, 2.3, and 2.4).

Keratinocytic naevi vary from pale brown and nearly macular with a soft velvety feel, to brown or red, verrucous or hyperkeratotic, and can have a prominent inflammatory component; e.g., ILVEN.

2.2.1 Verrucous Epidermal Naevus (Figs. 2.1, 2.2, 2.3, and 2.4)

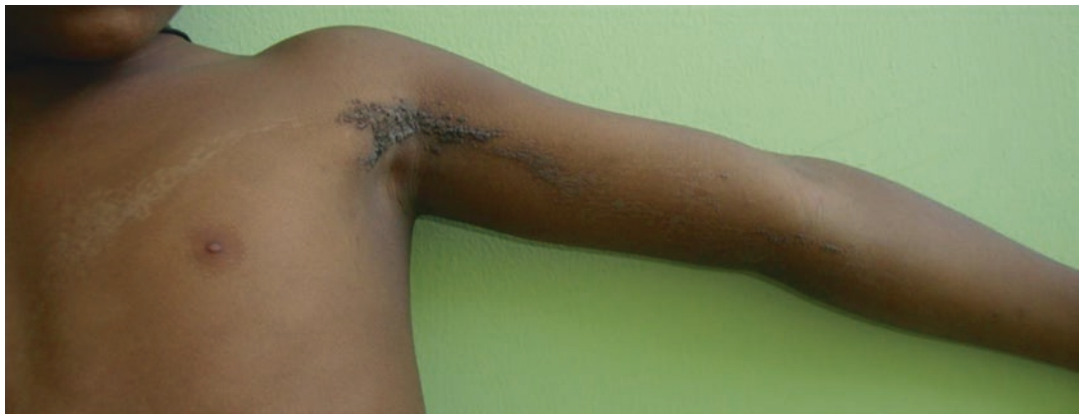

Fig. 2.1 Linear verrucous epidermal naevus on the upper limb in a Blaschko linear distribution in an 8-year-old boy

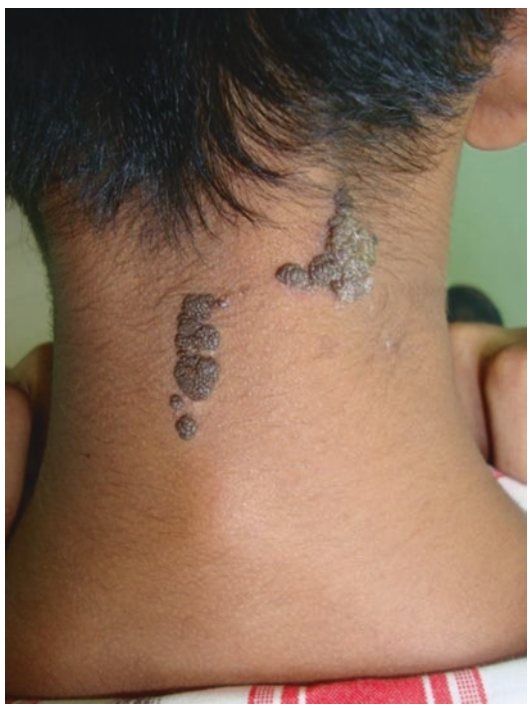

Fig. 2.2 Verrucous epidermal naevus

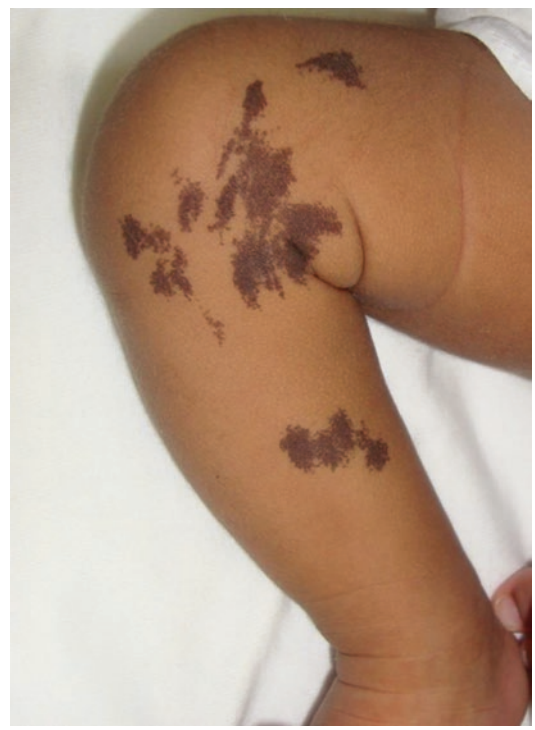

Fig. 2.3 A 38-day-old baby girl with darkly pigmented macular lesions only on the right leg since birth. This may remain macular or may become verrucous later

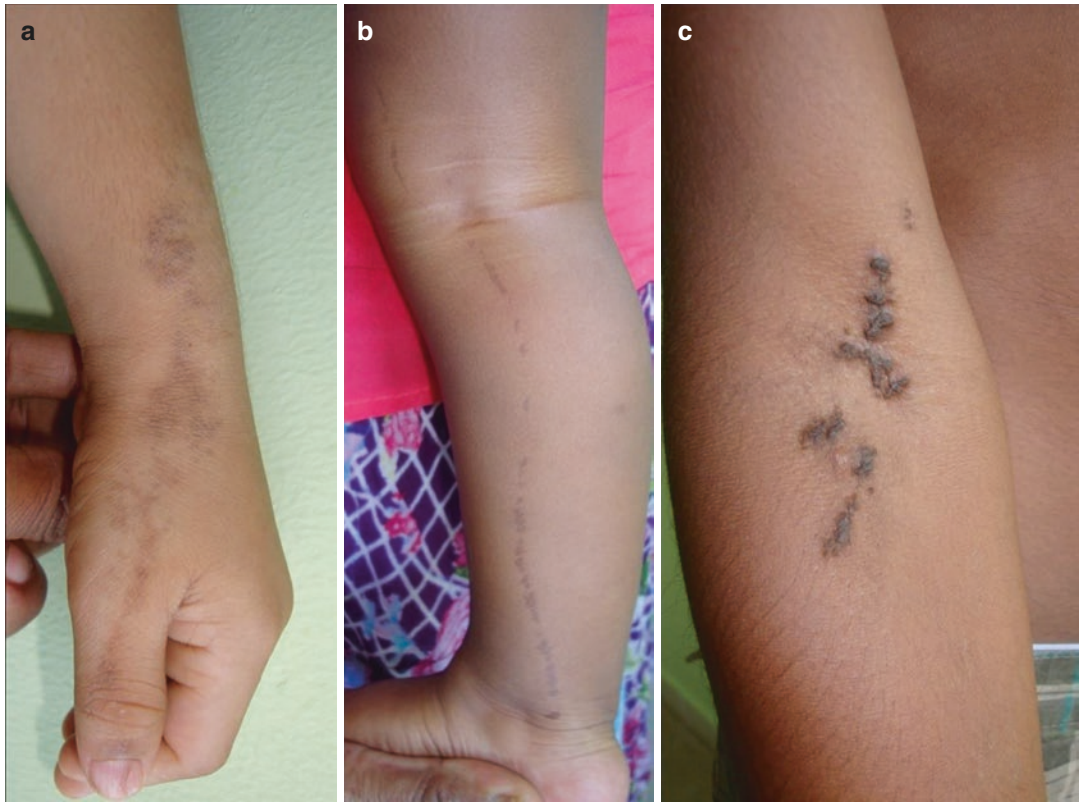

Fig. 2.4 Linear epidermal naevi in children aged (**a**) 1 year, (**b**) 7 months ,and (**c**) 10 years on the limbs in Blaschko linear distribution. Both sex were affected equally

2.2.2 Inflammatory Linear Verrucous Epidermal Naevus (ILVEN)

Commonly appears in the first few years of life rather than at birth and spreads gradually until stabilizing in a classic Blaschko linear distribution. It is characterized clinically by inflamed and hyperkeratotic skin that can be pruritic. It is usually confined to a single limb but can be more extensive. Clinically and histologically, there is a significant overlap between ILVEN and psoriasis (Figs. 2.5 and 2.6).

2.2.3 Sebaceous Naevi

Sebaceous naevi have a greasy feel and appearance, are often yellowish or pink, but can be deeply pigmented in dark skin (Figs. 2.7 and 2.8). A risk of malignant transformation occur at a rate of approximately 1% (syringocystadenoma papilliferum being the commonest), whereas basal cell carcinoma occurs in less than 1% and squamous cell carcinoma is rarer.

Management: total excision

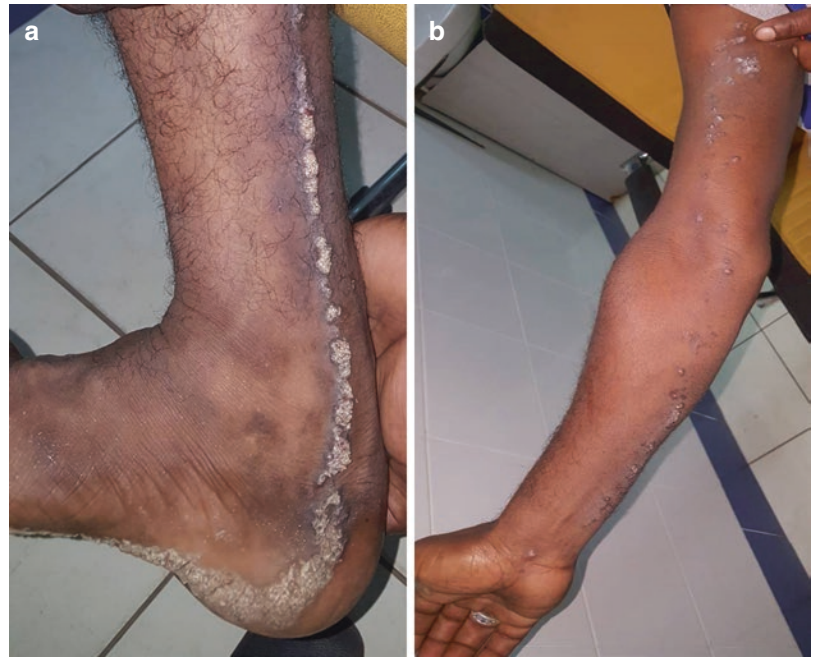

Fig. 2.5 Inflammatory linear verrucous epidermal naevus (ILVEN) (**a**) on the lower limb in a Blaschko linear distribution in an adult man. (**b**) The same patient having similar lesion along the right upper limb in a Blaschko-linear distribution (picture courtesy Dr. Ajith Karawita, consultant venereologist, Teaching Hospital Anuradhapura, Sri Lanka)

2.2.4 Follicular Naevi (Naevus Comedonicus or Acne Naevus)

A hair follicle tumor, an abnormality of the follicular infundibulum. It presents as a group of comedo-like lesions. It is suggested that it is a rare type of epidermal naevus (Calonje 2016). They may be present at birth or develop throughout adult life. No gender predilection (Yigider et al. 2016).

Clinical features Usually occurs on the face, neck, and chest and appears as groups of closely arranged dilated follicular openings with keratin plugs (Figs. 2.9 and 2.10).

Histopathology A rudimentary pilosebaceous follicle is present, with a large overlying keratin-filled crater.

Management For cosmetic reasons; cautery, laser, surgery, daily local application of tretinoin 0.1% gel and corticosteroid ointment (mometasone furoate) given good results (Manola et al. 2003).

2.3 Congenital Pigment Cell Naevi

1. Congenital melanocytic naevi
2. Congenital Spitz naevus
3. Congenital blue naevus
4. Congenital naevus spilus
5. Mongolian blue spot (dermal melanocytosis)

2.3.1 Congenital Melanocytic Naevi

Congenital melanocytic naevi (CMN) are benign, pigmented, melanocytic naevi present at birth. Melanoma occurs more commonly in individuals with CMN. Life time risk of melanoma in all sizes of CMN is 0.1–2%. This risk is variable, being very low for small single lesions, with a range extending up to as high as 10–14% in individuals with naevi of >60 cm projected adult size (PAS). The median age for developing melanoma is approximately 7 years in White skin. For multiple CMN, however, neurological abnormalities are the commonest comorbidity (Figs. 2.11, 2.12, 2.13, 2.14, and 2.15).

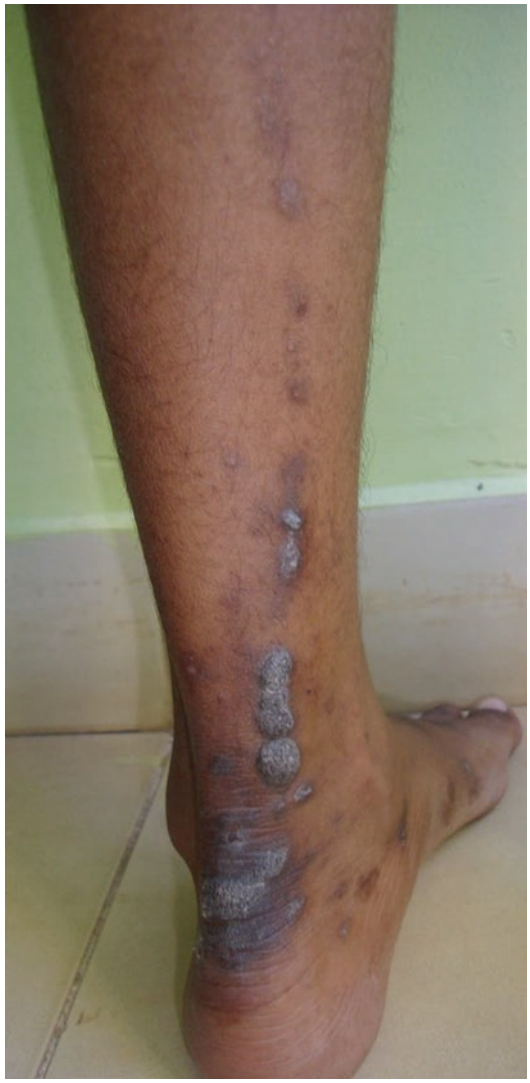

Fig. 2.6 Inflammatory linear verrucous epidermal naevus (ILVEN) in a 23-year-old man

2.3.2 Congenital Spitz Naevus

Congenital Spitz naevi are rare. In later life, they can mimic melanoma. The incidence of malignant transformation in congenital lesions is not known.

2.3.3 Congenital Blue Naevus

Congenital blue naevi are rare. Multiple congenital blue naevi at birth have very rarely been described and can be associated with uveal melanoma and can rarely be familial (Fig. 2.16).

2.3.4 Congenital Naevus Spilus

Congenital naevus spilus (or speckled lentiginous naevi) are pigmented lesions with a cafe-au-lait macule background and superimposed, more darkly pigmented areas (or speckles). The background may not be apparent at birth, and the superimposed speckles may increase in number over time. Occasionally show zosteriform or segmental distributions. Present at birth or appear during childhood. No sex predilection (Figs. 2.17, 2.18, 2.19, and 2.20) (Stefanaki et al. 2016).

Differential Diagnosis cafe-au-lait spot, congenital melanocytic naevus, Becker naevus, agminated naevomelanocytic naevus, and segmental lentiginosis.

Management There have been a few reports of melanoma arising within naevus spilus.

For cosmetic reasons— Excision, ablative and non-ablative lasers and dermabrasion.

2.3.5 Mongolian Spot (Dermal Melanocytosis)

Mongolian spots are congenital macular areas of blue-gray pigmentation of varying size and shape located on the sacral area in normal infants. The lesion develops in utero, increases in depth for a period during infancy, and then diminishes but may occasionally persist into adult life.

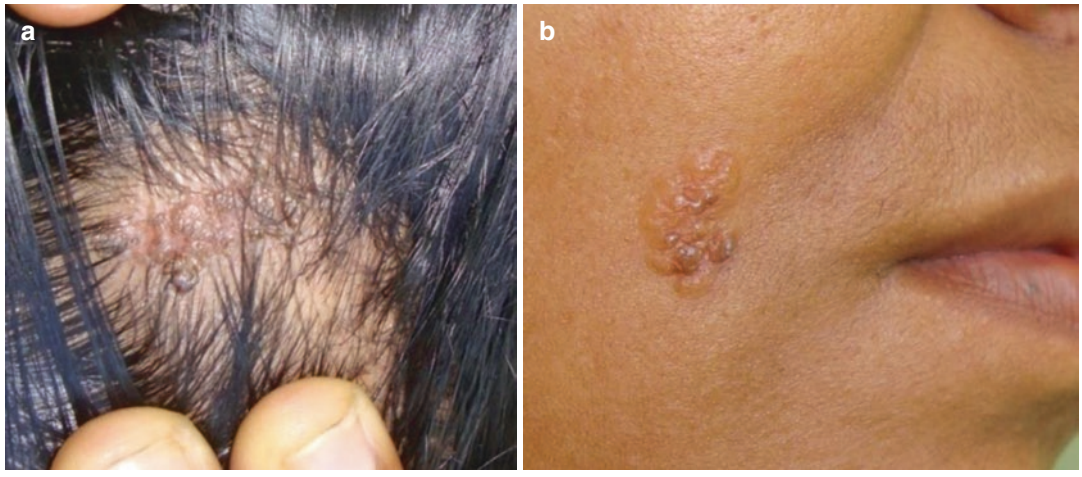

Fig. 2.7 Sebaceous naevus (**a**) on the scalp (the commonest site) and (**b**) on the face of two women

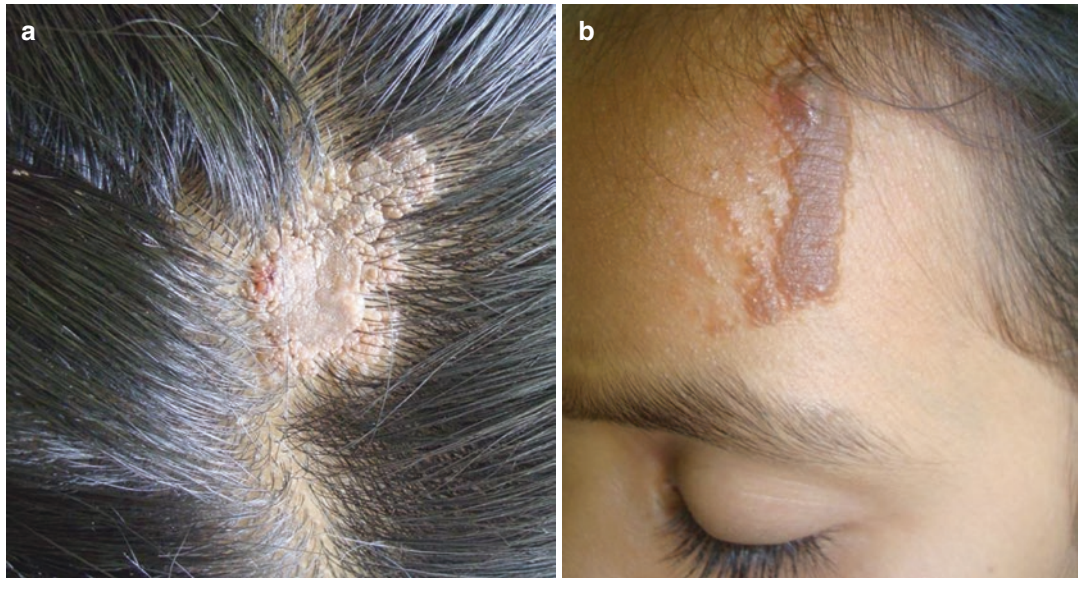

Fig. 2.8 Sebaceous naevus (**a**) on the scalp and (**b**) on the forehead in two girls

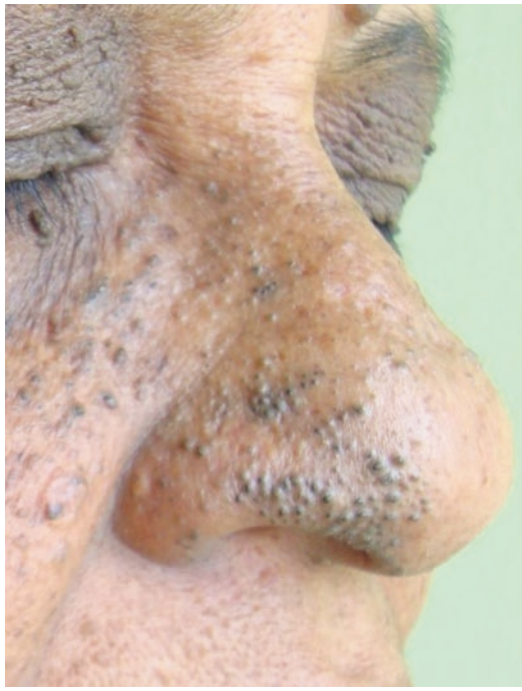

Fig. 2.9 Comedo naevus/naevus comedonicus in a 53-year-old woman. She also has numerous DPN and seborrhoeic keratoses

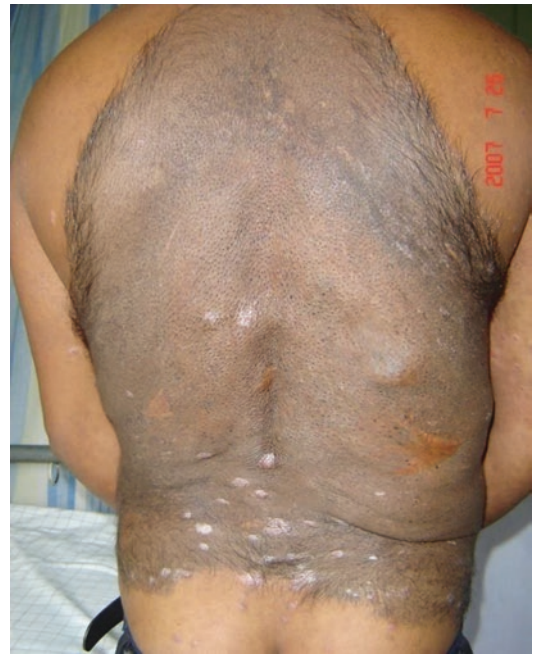

Fig. 2.11 Giant hairy congenital melanocytic naevus

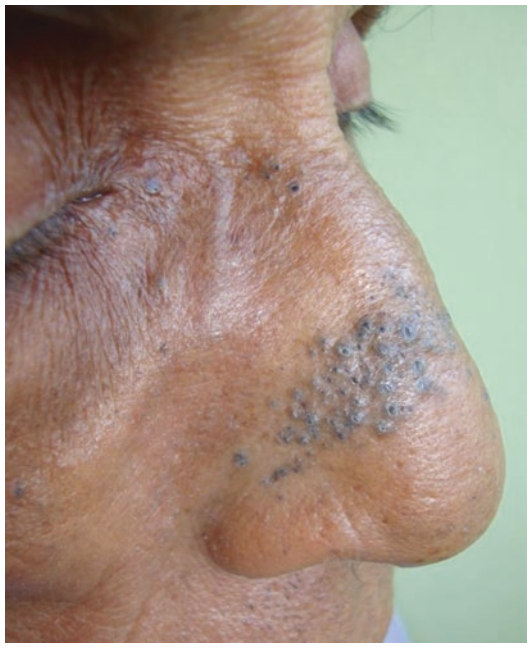

Fig. 2.10 Comedo naevus/naevus comedonicus in a 76-year-old man

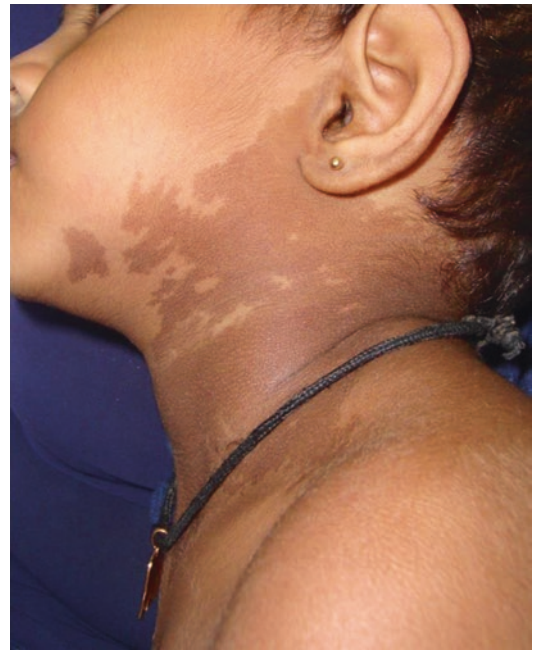

Fig. 2.12 Large congenital melanocytic naevus in a 2½-year-old girl

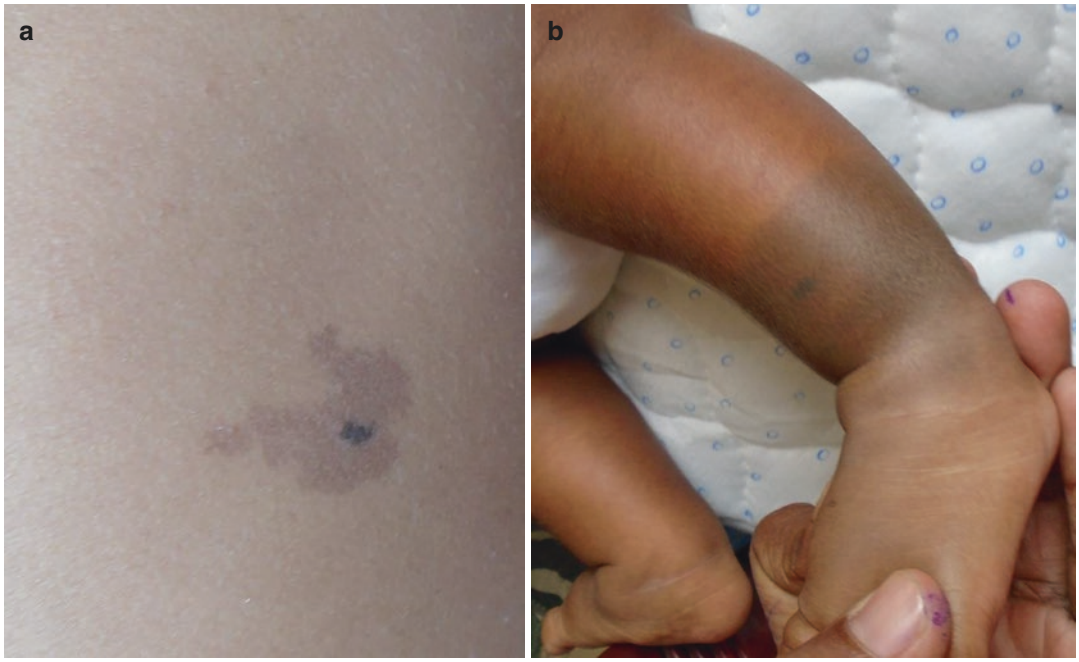

Fig. 2.13 (**a**, **b**) Congenital melanocytic naevi in infants (photographed by Dr. Maduranga Mendis, medical officer, neonatology unit, General Hospital Kalutara, Sri Lanka)

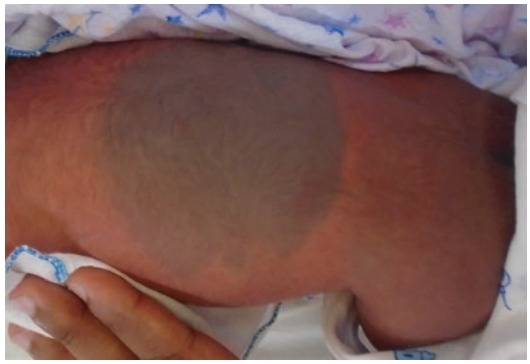

Fig. 2.14 Large congenital melanocytic naevus at the back of the trunk in a new born baby boy (photographed by Dr. Maduranga Mendis, medical officer, neonatology unit, General Hospital Kalutara, Sri Lanka)

Uncommon in white people common in darker skin complexion (Stefanaki et al. 2016).

Management No treatment is required. Persistent cases—Q-switched lasers, intense pulsed light, and bleaching creams (Fig. 2.21).

2.4 Congenital Connective Tissue Naevi

1. Collagen naevus
2. Familial cutaneous collagenoma
3. Elastic tissue naevus
4. Juvenile elastoma
5. Naevus anelasticans
6. Mucinous naevus
7. Mixed connective tissue naevus
8. Congenital lipoma
9. Naevus psiloliparus

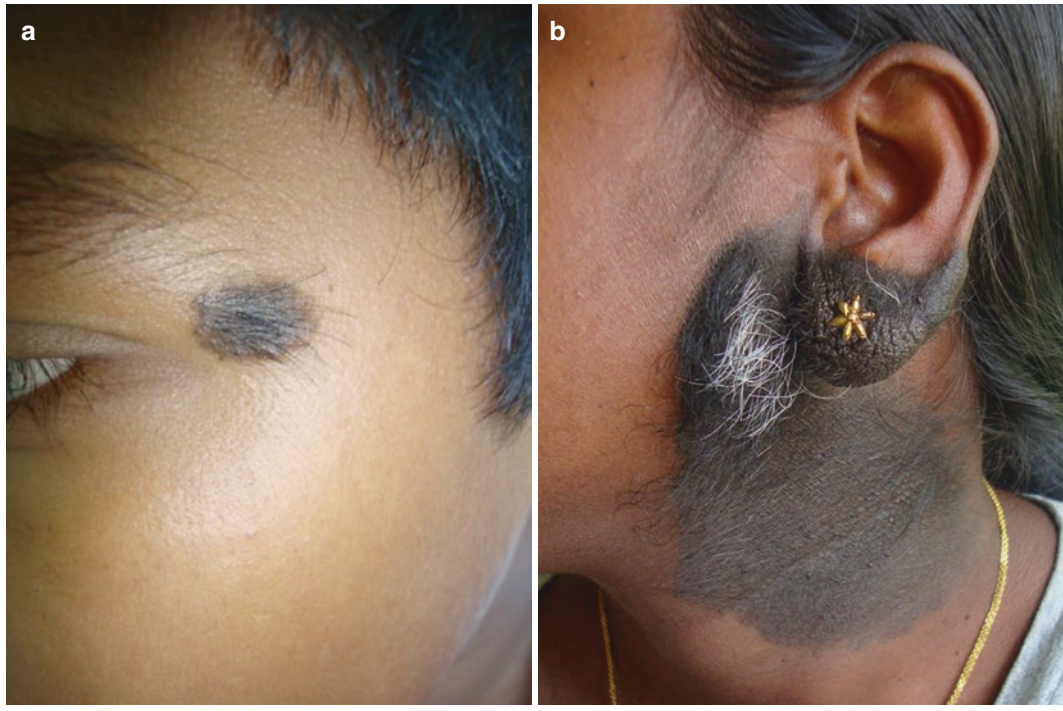

Fig. 2.15 (**a**, **b**) Congenital melanocytic hairy naevus

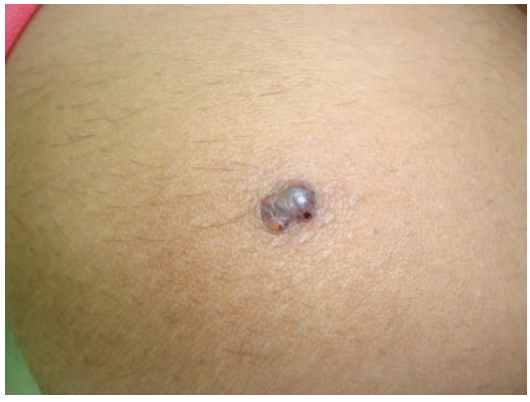

Fig. 2.16 Congenital blue naevus

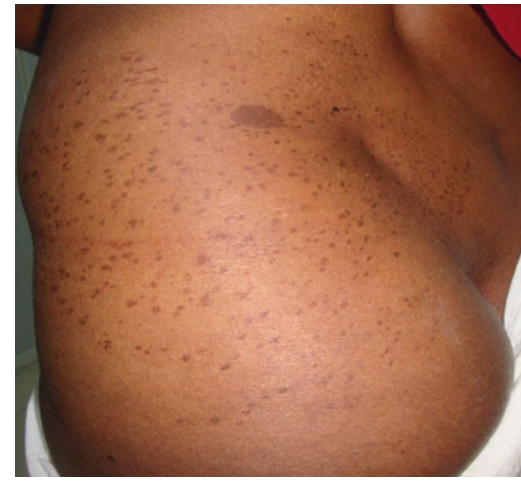

Fig. 2.17 Congenital naevus spilus (or speckled lentiginous naevus) on the right shoulder in a woman

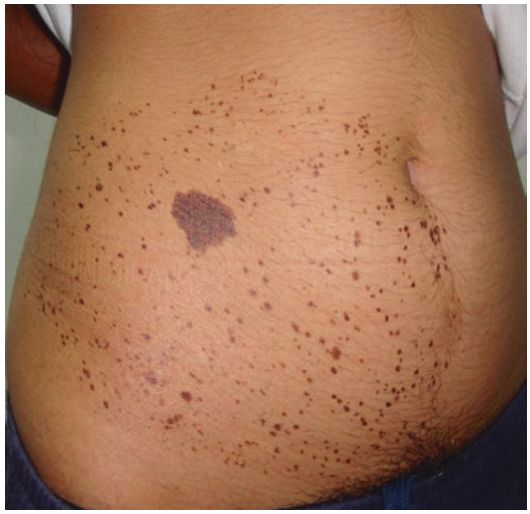

Fig. 2.18 Congenital naevus spilus on the right loin in a woman

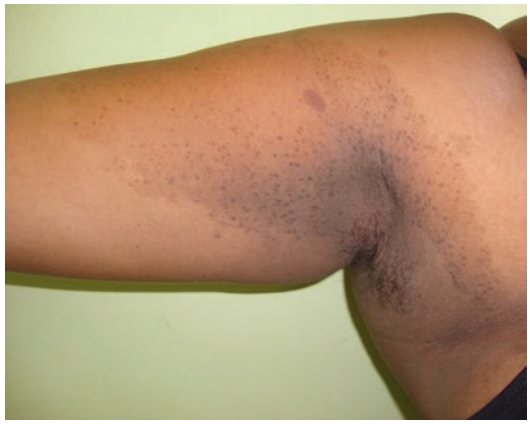

Fig. 2.19 Congenital naevus spilus on the right axilla and upper arm in a woman

Congenital connective tissue naevi are a clinically heterogeneous group of hamartomas, named by the predominant cell type on histology, and therefore usually requiring biopsy for accurate diagnosis. They can be single or multiple and can be associated with extracutaneous features.

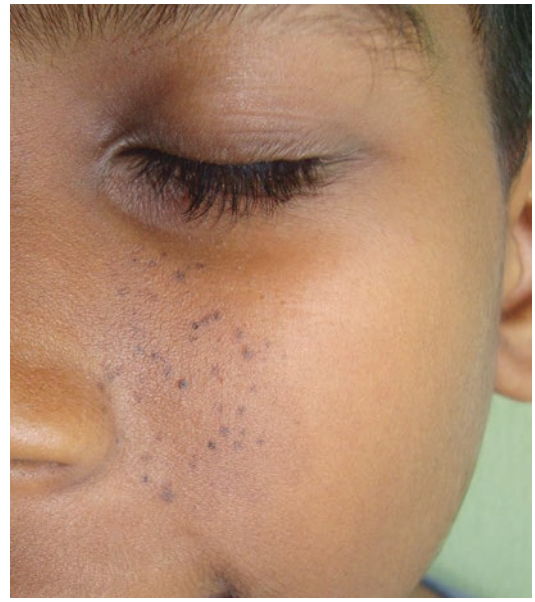

Fig. 2.20 Congenital naevus spilus on the face of a 10-year-old boy

2.5 "Naevoid" Entities and Currently Unclassifiable Naevi

1. Becker naevus
2. Linear atrophoderma of Moulin
3. Lichen striatus

2.5.1 Becker Naevus

The genetic basis of these naevi is not yet known. Becker naevus (or Becker melanosis) is a relatively common hyperpigmented, generally nonlinear lesion with an incidence of around 0.25% and is commoner in males than females. It is only rarely congenital, with the majority of lesions appearing in the first two decades, classically at puberty. It is frequently but not always hypertrichotic and is commonest on the upper trunk (Kinsler and Sebire 2016) (Figs. 2.22, 2.23, 2.24, 2.25 and 2.26).

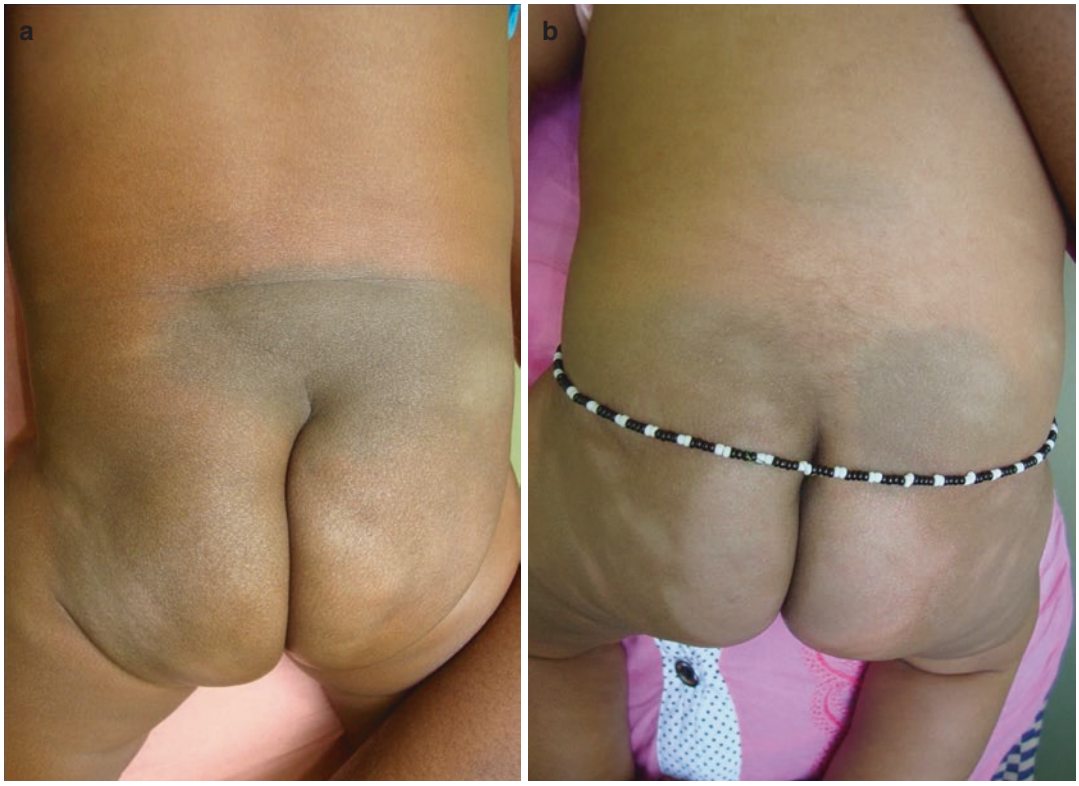

Fig. 2.21 (**a**, **b**) Mongolian spots on sacral area in two infants

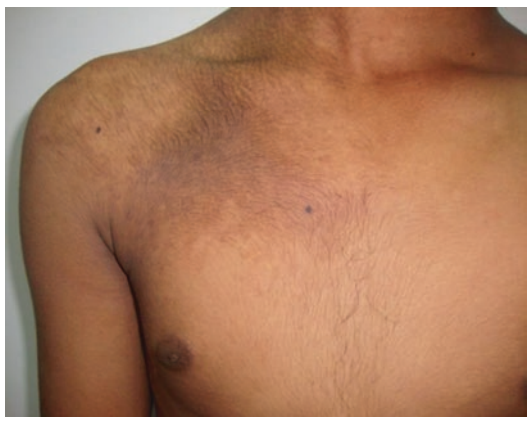

Fig. 2.22 Becker naevus in a 19-year-old boy with hypertrichosis

Becker naevus is not uncommonly associated with extracutaneous abnormalities, then termed Becker naevus syndrome, which can involve underlying structures, namely, aplasia or hypoplasia of the underlying breast tissue (Fig. 2.27), or pectoralis major muscle (or sometimes shoulder muscles) or lipoatrophy. Other extracutaneous associations described are ipsilateral limb growth disturbance, supernumerary nipples, and scoliosis.

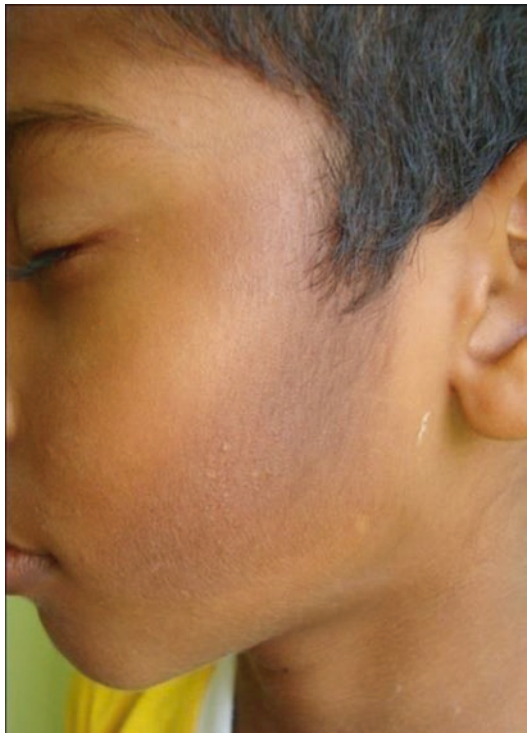

Fig. 2.23 Becker naevus. Early pigmentation in a 9-year-old boy, mild hypertrichosis is apparent even at this stage

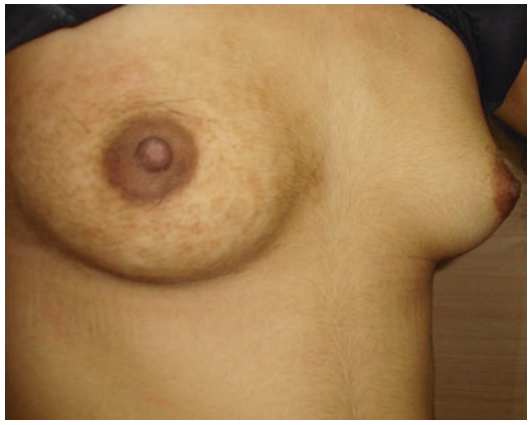

Fig. 2.24 Becker naevus involving unilateral breast in a 22-year-old woman

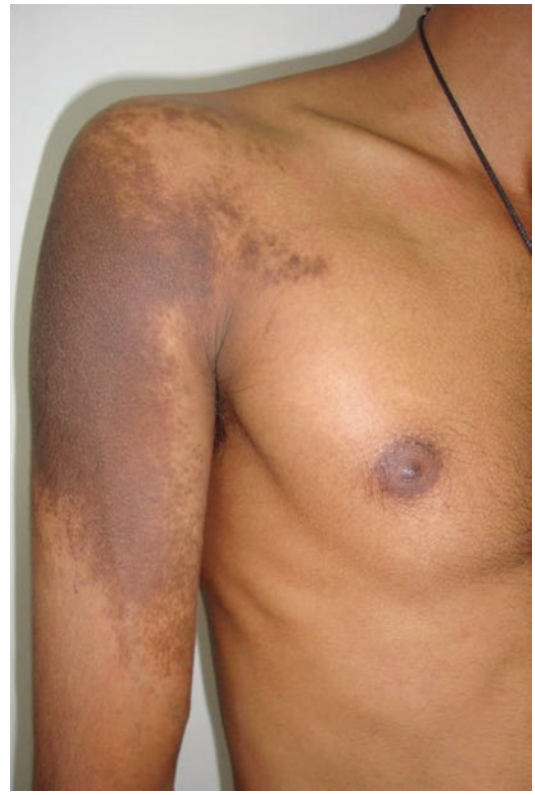

Fig. 2.25 Becker naevus in a 17-year-old boy

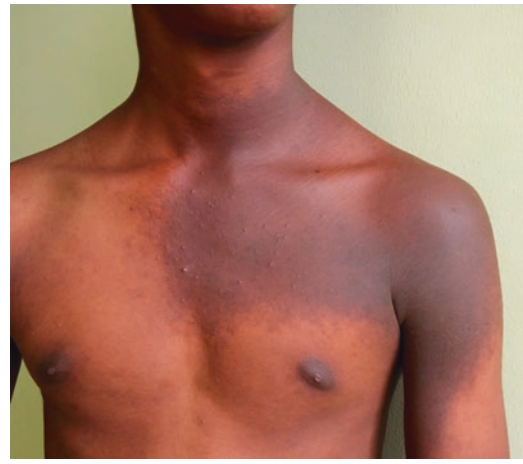

Fig. 2.26 Becker naevus in an 18-year-old man

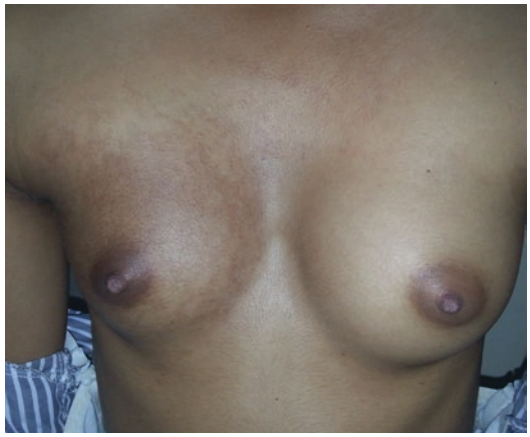

Fig. 2.27 Becker naevus syndrome; a 20-year-old woman with Becker's naevus and ipsilateral breast hypoplasia (photographed by Dr. Amitha Chandima Ranasinghe, Senior Registrar Dermatology, Colombo North Teaching Hospital, Ragama, Sri Lanka)

2.5.2 Linear Atrophoderma of Moulin

This entity describes an acquired hyperpigmented atrophy of subcutaneous tissue rather than a naevus but in a Blaschko linear distribution. It generally appears in the first two decades of life and usually stabilizes after its initial development. Most commonly lesions are truncal and unilateral.

2.5.3 Lichen Striatus

Self-limiting and asymptomatic inflammatory dermatosis characterized by skin colored or hypopigmented papules in pigmented skin in a

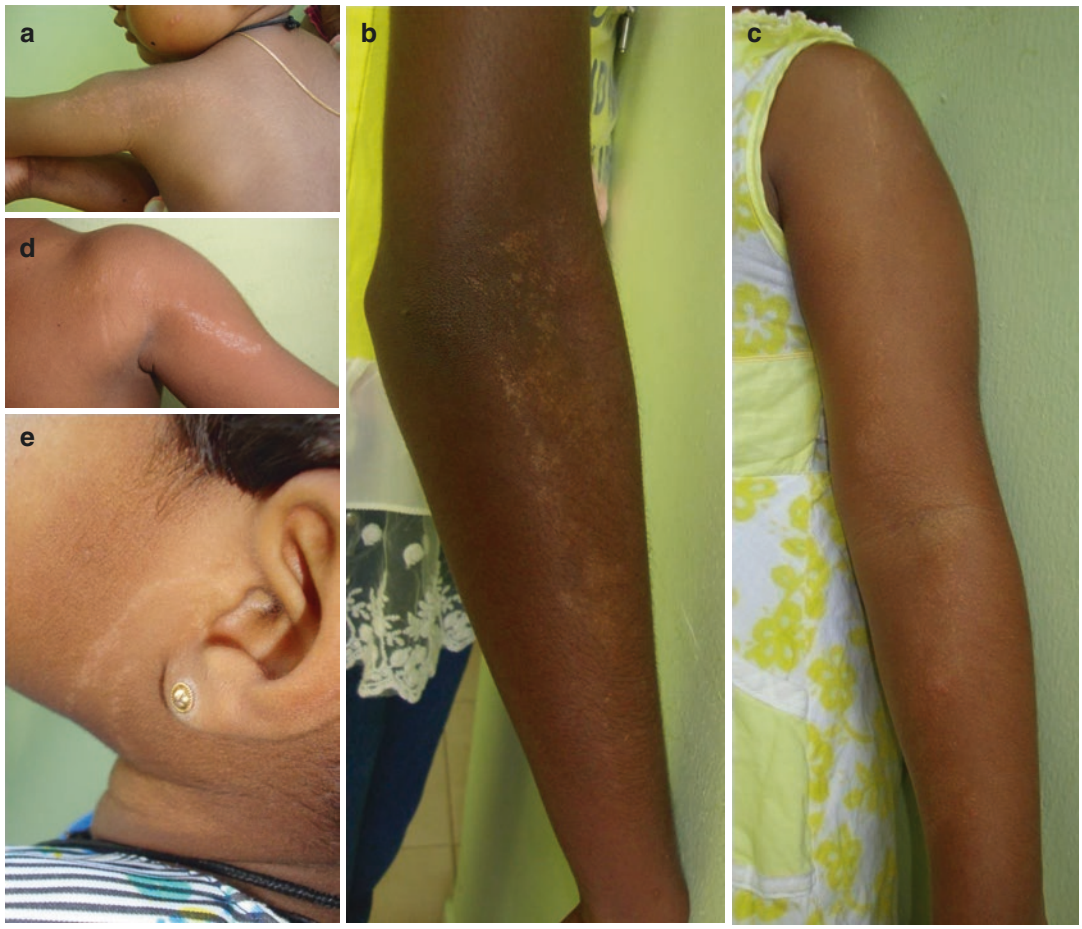

Fig. 2.28 (**a**–**e**) Lichen striatus in children. Skin colored or mildly hypopigmented papular linear distribution that develop in the lines of Blaschko

linear distribution that develop in the lines of Blaschko. These usually occur as isolated lesions on the limbs in children aged 5–15 years (Graham and Hossler 2016). Females are affected approximately two to three times as frequently as males.

Clinical Presentation Skin colored or hypopigmented papules appear in a linear distribution within days. Mostly asymptomatic, but pruritus may occur in adults. The area affected reaches its maximum extent within 2–3 weeks, but gradual extension can continue for several months. Spontaneous resolution within 6–12 months in most cases, but some lesions may persist for over a year. Resolution may be followed by hypopigmentation or rarely hyperpigmentation (Fig. 2.28) (Piguet et al. 2016).

Differential Diagnosis Linear psoriasis, linear Darier disease, linear lichen planus, linear porokeratosis, and inflammatory linear verrucous epidermal naevus (ILVEN), during hypopigmented stage linear vitiligo or nevoid hypomelanosis (Stolze and Hamm 2018).

Management Observation and reassurance, topical corticosteroids in patients with troublesome itch (usually adults), topical tacrolimus, topical pimecrolimus, and photodynamic therapy.

2.6 Melanocytic Naevi (Table 2.2)

2.6.1 Naevus of Ota

A naevus of Ota is an extensive, bluish, patchy, dermal melanocytosis that affects the sclera and the skin adjacent to the eye, distributed along the first and the second branches of the trigeminal nerve (Fig. 2.29). Most lesions present at birth, minority of cases around puberty. Commonly occur in darkly pigmented individuals, Asian, and Black people.

Table 2.2 Classification melanocytic naevi (Stefanaki et al. 2016)

Dermal melanocytic lesions	*Naevi with unusual morphology*
Mongolian spot	Combined melanocytic naevi
Naevus of Ota	Recurrent melanocytic naevi
Naevus of Ito	Halo naevus
Congenital melanocytic naevi	Meyerson naevus
Speckled lentiginous naevus	Cockade naevus
Common acquired naevi	Targetoid haemosiderotic naevus
Acquired melanocytic naevi	*Other naevi*
Naevi in unusual sites	Spitz naevus
Melanocytic naevi of the genital area	Blue naevus and variants
Acral naevi	Malignant blue naevus
Conjunctival naevi	Clinically atypical naevi
Naevi of the nail matrix or nail bed	

Stefanaki I, Christina Antoniou C, Stratigos A. Benign Melanocytic Proliferations and Melanocytic Naevi. In: Burns T, Breathnach S, Cox N, Griffiths C (eds) Rook's textbook of dermatology, 9th edn. Wiley Blackwell Science, Oxford, p 132.1

Management Unlike Mongolian spots, it does not disappear with time.

1. Q-switched lasers are the first line treatment.
2. Cosmetic camouflage.

Any new subcutaneous nodule arising on a naevus of Ota should be further investigated histologically to exclude the possibility of melanoma.

2.6.2 Naevus of Ito

Naevus of Ito is a dermal melanocytosis involving the acromioclavicular region and the upper chest. Presents as a unilateral, blue-greyish macular discoloration commonly in Chinese and Japanese people.

Differential Diagnosis Becker naevus

Management Benign lesion does not disappear with time. Pigment targeting Q-switched laser is the treatment of choice.

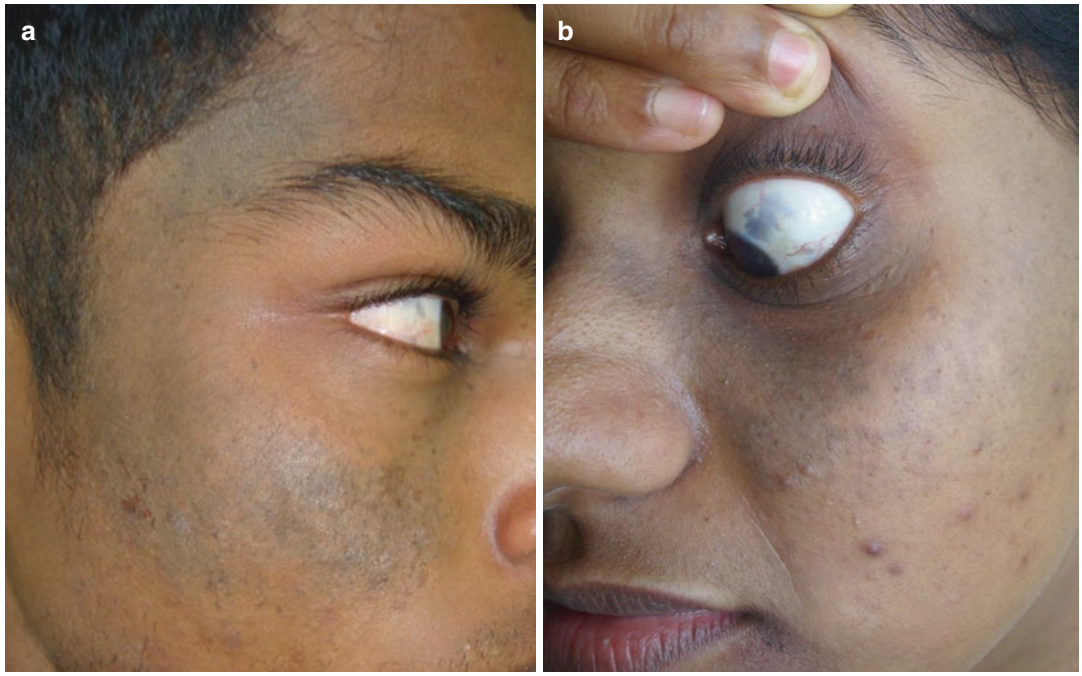

Fig. 2.29 (**a**, **b**) Naevus of Ota, an extensive, bluish, patchy, dermal melanocytosis that affects the sclera and the skin adjacent to the eye, distributed along the first and the second branches of the trigeminal nerve

2.7 Naevi in Unusual Sites (Stefanaki et al. 2016)

1. Melanocytic naevi of the genital area
2. Acral naevi
3. Conjunctival naevi
4. Naevi of the nail matrix or nail bed

2.7.1 Melanocytic Naevi of the Genital Area

Despite their benign course, these lesions are often difficult to differentiate from vulval melanoma (Fig. 38.48)

2.7.2 Acral Melanocytic Naevi

Usually macular or slightly elevated, uniformly pigmented lesions with irregular and sharp borders distributed along the parallel furrows of acral skin. The most common dermoscopic pattern of acral naevi is the parallel furrow pattern, followed by the lattice-like and fibrillar patterns.

In contrast melanomas are situated along the ridges of the palms and soles. Since acral lentiginous melanoma is the commonest melanoma in Asians and Africans, these lesions should arose suspicion (Figs. 2.30, 2.31, and 2.32).

2.7.3 Conjunctival Melanocytic Naevi

There are no specific clinical signs that predict the transformation of a conjunctival naevus to melanoma (presumed to occur in less than 1% of naevi). Attachment to the sclera, extension into the cornea, and development of "feeder" vessels upon slit lamp examination represent worrisome changes (Fig. 2.33).

Fig. 2.30 Large melanocytic naevus on the sole. Histopathology excluded acral lentiginous melanoma (picture courtesy Dr. Kanishka de Silva, Consultant Oncological Surgeon, National Cancer Institute Maharagama, Sri Lanka)

2.7.4 Melanocytic Naevi of the Nail Matrix or Nail Bed

Longitudinal, parallel, and homogeneous pigmentation ranging from light brown to dark brown to black on the underside of the nail plate. A pseudo- Hutchinson sign presenting as visible pigmentation through a relatively translucent cuticle is characteristically seen in nail matrix naevi.

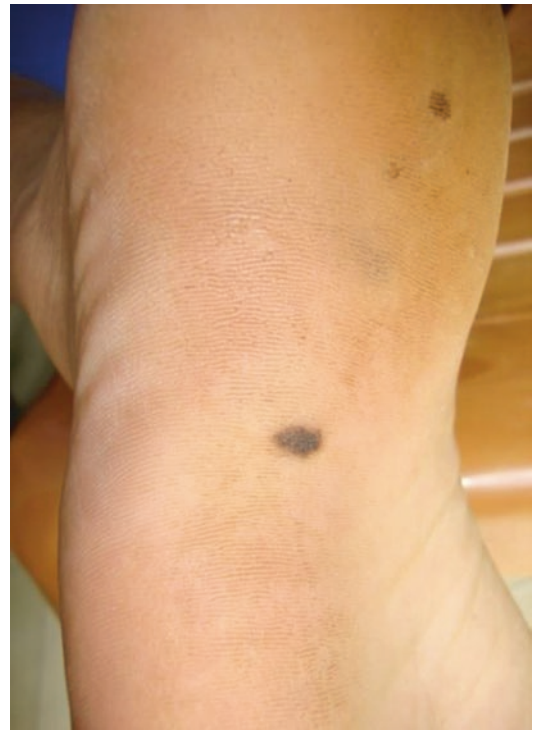

Fig. 2.31 Acral melanocytic naevus on the sole

The distinction from early subungual melanoma is essential (Figs. 2.34 and 2.35).

Longitudinal melanonychia involving single nail should arouse the suspicion of acral lentiginous melanoma. In both, these patients' lesions were confirmed benign melanocytic naevi by histopathology.

2.8 Naevi with Unusual Morphology (Stefanaki et al. 2016)

1. Combined melanocytic naevi
2. Recurrent melanocytic naevi
3. Halo naevus
4. Meyerson naevus
5. Cockade naevus
6. Targetoid haemosiderotic naevus

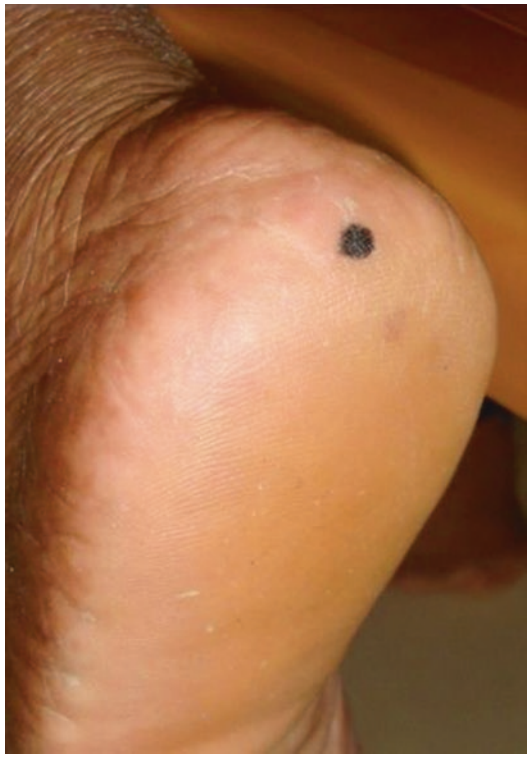

Fig. 2.32 Acral lentiginous melanoma. A 67-year-old woman came with this pigmented lesion which had appeared over 8 months. Histopathology confirmed malignant melanoma

2.8.1 Halo Naevus

The development of a halo of hypomelanosis around a central cutaneous tumor. This tumor is usually a benign melanocytic naevus (rarely primary or secondary malignant melanoma).

Disease Course and Prognosis The naevus tends to flatten and may disappear completely. The depigmented areas often persist but may pigment after many years (Cohen et al. 2016; Margileth 2018).

Management Normally none is required (Figs. 2.36 and 2.37).

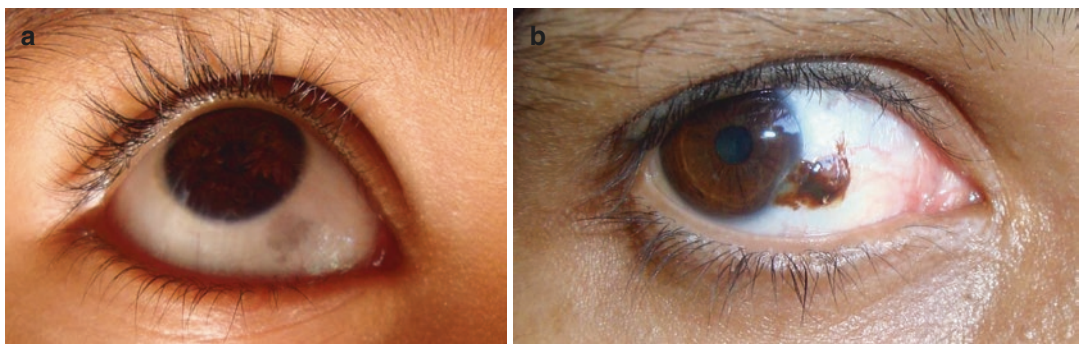

Fig. 2.33 (**a**, **b**) Conjunctival melanocytic naevi

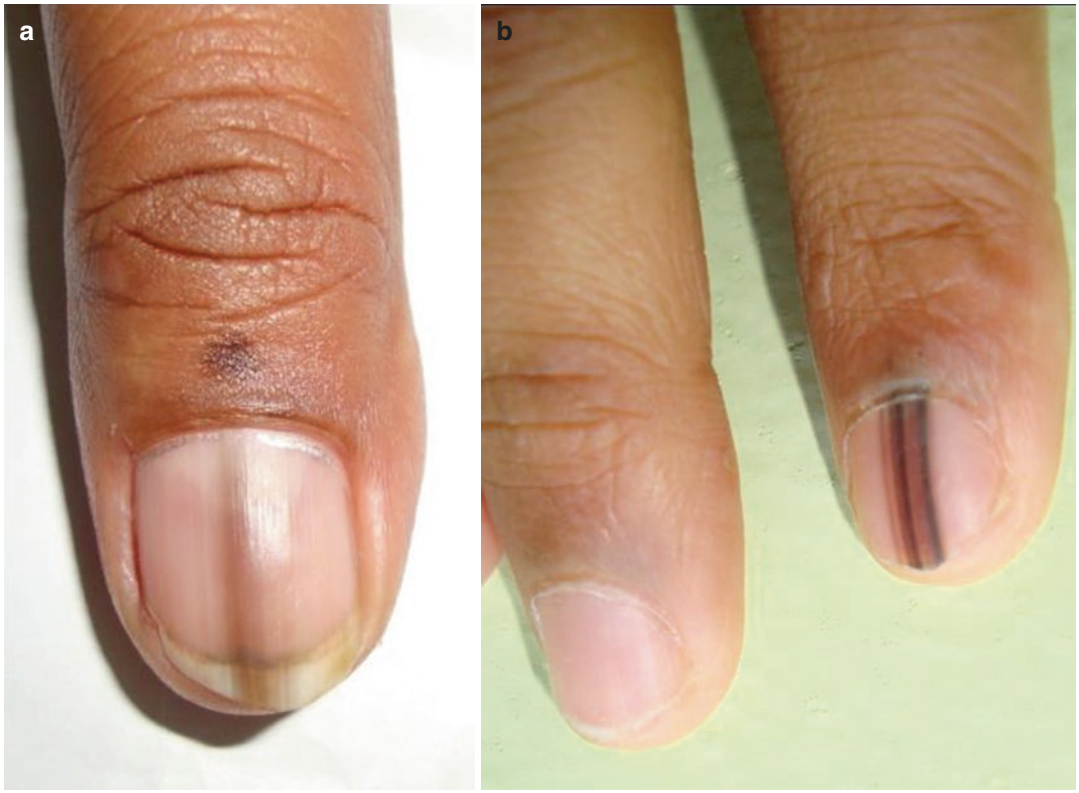

Fig. 2.34 (**a**, **b**) Melanocytic naevi of the nail matrix or nail bed

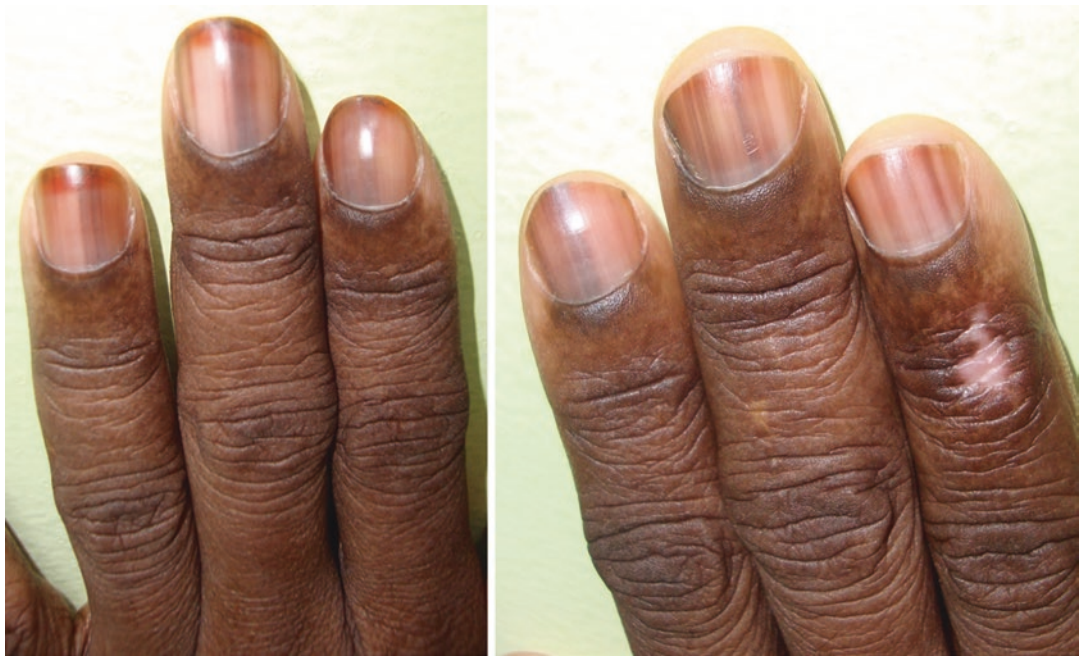

Fig. 2.35 Racial pigmentation which is common in dark skin individuals. Note this pigmentation is seen in all the 20 nails

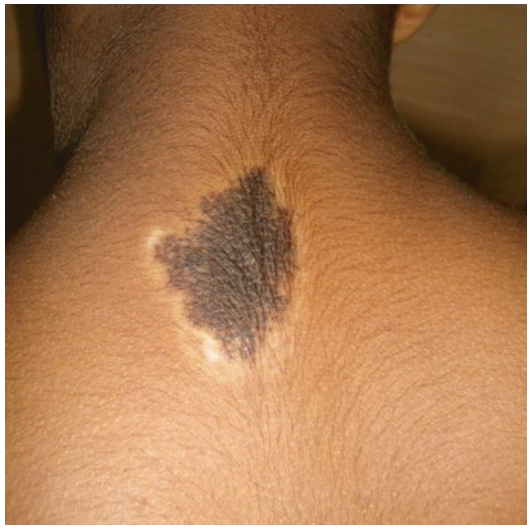

Fig. 2.36 Recent onset of depigmented halo around a congenital epidermal naevus

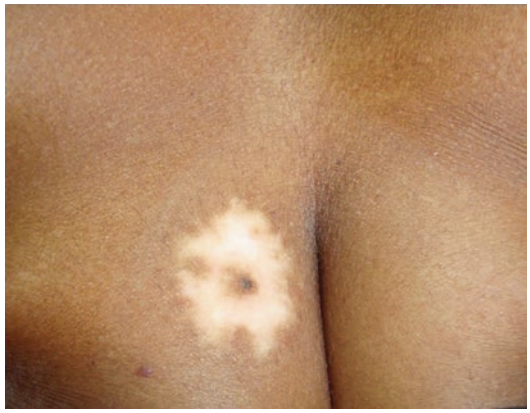

Fig. 2.37 Halo naevus. Depigmented halo around an intradermal naevi

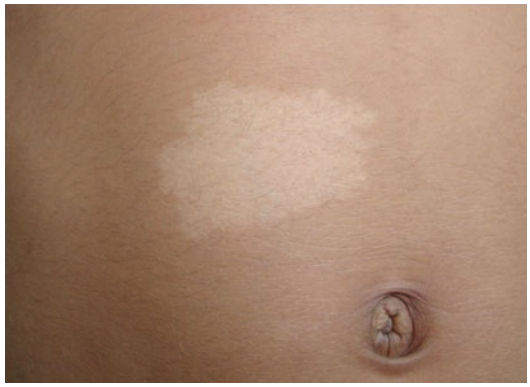

Fig. 2.38 A solitary congenital depigmented naevus

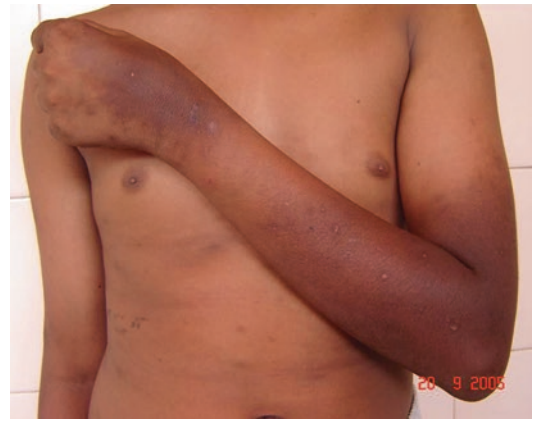

Fig. 2.39 This 18-year-old boy has multiple pigmented naevi on his body

References

Calonje E (2016) Tumours of skin appendages. In: Burns T, Breathnach S, Cox N, Griffiths C (eds) Rook's textbook of dermatology, 9th edn. Wiley-Blackwell Science, Oxford, pp 138.1–138.44

Cohen BE, Mu EW, Orlow SJ (2016) Comparison of childhood vitiligo presenting with or without associated halo nevi. Pediatr Dermatol 33(1):44–48. https://doi.org/10.1111/pde.12717

Graham JN, Hossler EW (2016) Lichen striatus. Cutis 97(2):86;120;122

Kinsler VA, Sebire NJ (2016) Congenital Naevi and other developmental abnormalities affecting the skin. In: Burns T, Breathnach S, Cox N, Griffiths C (eds) Rook's textbook of dermatology, 9th edn. Wiley Blackwell Science, Oxford, p 75.3

Manola I, Ljubojević S, Lipozencić J, Pustisek N (2003) Nevus comedonicus--case report and review of therapeutical approach. Acta Dermatovenerol Croat 11(4):221–224

Margileth AM (2018) Spontaneous regression of large congenital melanocytic nevi, with a halo rim in 17 children with large scalp and trunk nevi during 45 years: a review of the literature. Clin Pediatr (Phila) 3:9922818816421. https://doi.org/10.1177/0009922818816421

Piguet V, Stephen M, Breathnach SM, Cleach LL (2016) Lichen planus and lichenoid disorders. In: Burns T, Breathnach S, Cox N, Griffiths C (eds) Rook's textbook of dermatology, 9th edn. Wiley-Blackwell Science, Oxford, p 37.18

Stefanaki I, Antoniou C, Stratigos A (2016) Benign melanocytic proliferations and melanocytic naevi. In: Burns T, Breathnach S, Cox N, Griffiths C (eds) Rook's textbook of dermatology, 9th edn. Wiley Blackwell Science, Oxford, p 132.17

Stolze I, Hamm H (2018) Lichen nitidus and lichen striatus. Hautarzt 69(2):121–126. https://doi.org/10.1007/s00105-017-4100-7

Yigider AP, Kayhan FT, Yigit O, Kavak A, Cingi C (2016) Skin diseases of the nose. Am J Rhinol Allergy 30(3):83–90. https://doi.org/10.2500/ajra.2016.30.4318