Less Common Papulosquamous Disorders

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Contents

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3.1 Pityriasis Lichenoides

3.1.1 Introduction

Pityriasis lichenoides is a spectrum of self-limiting skin disorders usually lasting for an average of 18–20 months that includes two main clinicopathological subtypes, i.e., pityriasis lichenoides et varioliformis acuta (PLEVA) and pityriasis lichenoides chronica (PLC) [1–3]. Even though its etiopathogenesis is still not completely clear, bacterial and viral agents have been speculated to act as triggers by activating an inflammatory response via T-cell mediation and immune complex induction [1–3]. Both PLEVA and PLC most commonly occur during the first three decades of life without any racial predilection [1–3].

3.1.2 Clinical Presentation

PLEVA typically presents as asymptomatic or burning/itching inflammatory papules whose center tends to become vesiculopustular, hemorrhagic, necrotic, ulcerated, or crusted (Figs. 3.1a, 3.2, 3.3 and 3.4a) [1–3]. Trunk and arms are the most frequently involved areas, while face, palms, and soles are commonly spared [1–3]. Color of the papules usually varies according to phototype, with light- and dark-skinned patients showing a pink/reddish (Figs. 3.1a and 3.2a) and brown shade (Figs. 3.3a and 3.4a), respectively [1–3]. Of note, lesions often regress spontaneously leaving residual hypo/hyperpigmentation or varioliform scars that are more common in skin of color [1–3].

PLC manifests as asymptomatic pink/red or brown (darker phototypes) papules covered by uniform white scaling or a single centrally attached white scale (mica-like scale) commonly falling off spontaneously or by scraping (Figs. 3.5a, 3.6 and 3.7a) [1–3]. Similarly to PLEVA, lesions tend to involve trunk and arms and occur in crops, thus giving rise to a polymorphic appearance with lesions in different phases [1–3]. Hypopigmented macules may be seen after healing of the papules, especially in dark-skinned patients, in whom they may be the sole manifestation of PLC [1–3].

3.1.3 Dermoscopy

Dermoscopic findings of PLEVA remarkably vary based on lesion stage and skin phototype [1, 3–7]. In particular, in fair-skinned patients, early and mature lesions, respectively, feature purpuric areas resulting from erythrocyte extravasation (Fig. 3.1b) and a central amorphous brownish crust due to epidermal necrosis (Fig. 3.2b), whereas healing lesions commonly show a central white area histologically corresponding to fibrosis [1, 3–7]. On the other hand, in darker phototypes, purpuric areas are seen less frequently, and a central necrotic brownish-black crust surrounded by a peripheral white structureless area (sometimes displaying radial white streaks) due to epidermal acanthosis is the main finding (Figs. 3.3b and 3.4b) [1, 3–7]. Moreover, a peripheral scaling collarette with an inner free edge is commonly seen in all lesions stages regardless the phototype (Figs. 3.1b, 3.2, 3.3 and 3.4b), while a peripheral rim of dotted/linear vessels (Fig. 3.2b) and pigmentary structures (bluish-greyish globules/structureless areas corresponding to melanin pigment/melanophages in the

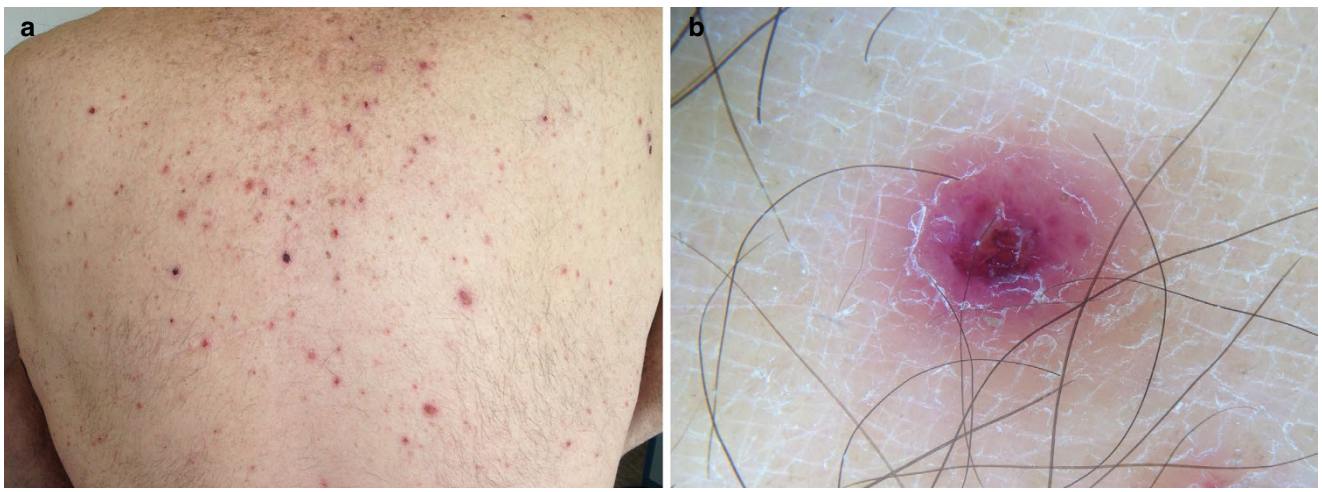

Fig. 3.1 Pityriasis lichenoides et varioliformis acuta in a Caucasian man (**a**). Dermoscopy reveals purpuric areas and a peripheral scaling collarette with an inner free edge (**b**)

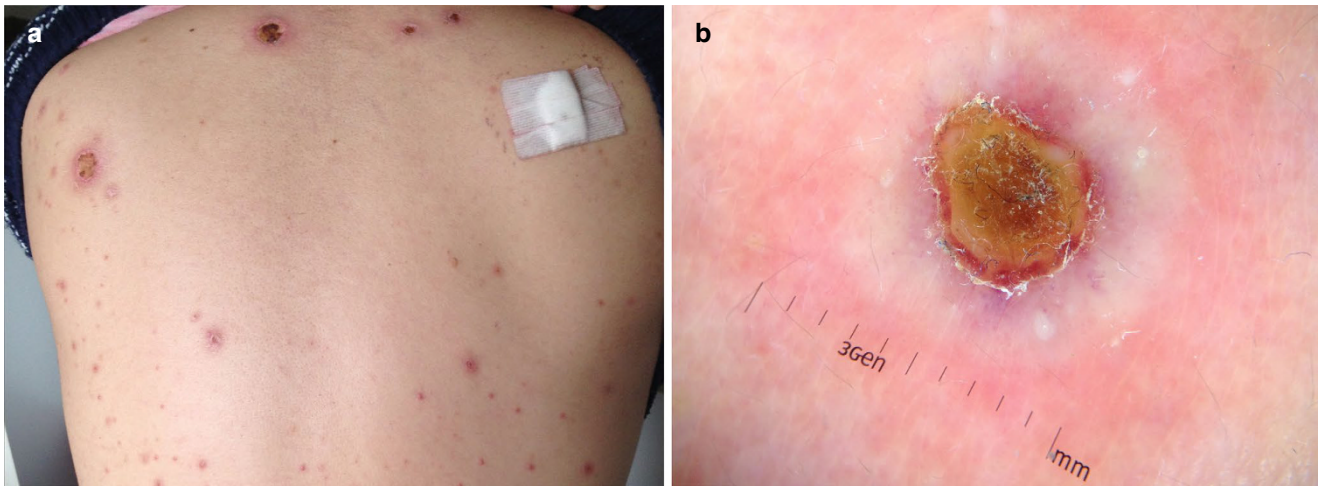

Fig. 3.2 Pityriasis lichenoides et varioliformis acuta in a Caucasian boy (**a**). Dermoscopic examination shows a central amorphous brownish crust with a peripheral scaling collarette with an inner free edge surrounded by dotted vessels (**b**)

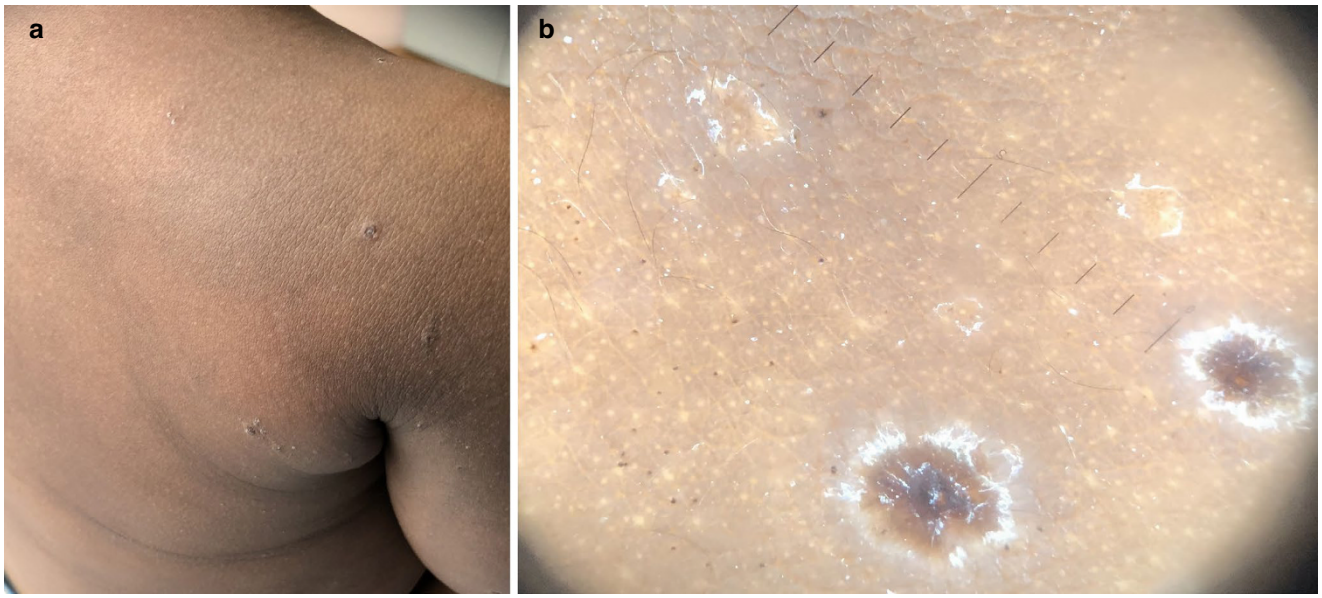

Fig. 3.3 Pityriasis lichenoides et varioliformis acuta in an African boy (**a**). A central necrotic brownish-black crust surrounded by a scaling white collarette with an inner free edge and a peripheral white halo is seen on dermoscopy (**b**)

dermis) (Fig. 3.4b) may be evident mainly in lighter and darker phototypes, respectively [1, 3–7].

The hallmark of PLC in lighter phototypes is represented by the presence of orange structureless areas (corresponding to hemosiderin deposits in the dermis due to erythrocyte extravasation) with or without blurred non-dotted vessels (including globular, linear-irregular, and/or branching vessels) (Fig. 3.5b) [1, 3–7]. Notably, orange areas are difficult to see in dark-skinned patients, who mainly feature a peripheral white scaling collarette with an inner smooth free edge (resulting from hyperkeratosis) (Fig. 3.6b) along with diffuse or focal brown structureless areas (due to basal layer pigmentation and dermal hemosiderin deposits) (Fig. 3.7b) [1, 3–7]. Additional dermoscopic findings include sparse dotted vessels, hemorrhagic spots, central micaceous scale, brown dots (in darker phototypes due to pigment incontinence in the dermis) (Fig. 3.7b), and hypopigmented areas (Fig. 3.7b), with this last feature being more common in longstanding lesions as a result of focal post-inflammatory hypopigmentation [1, 3–7].

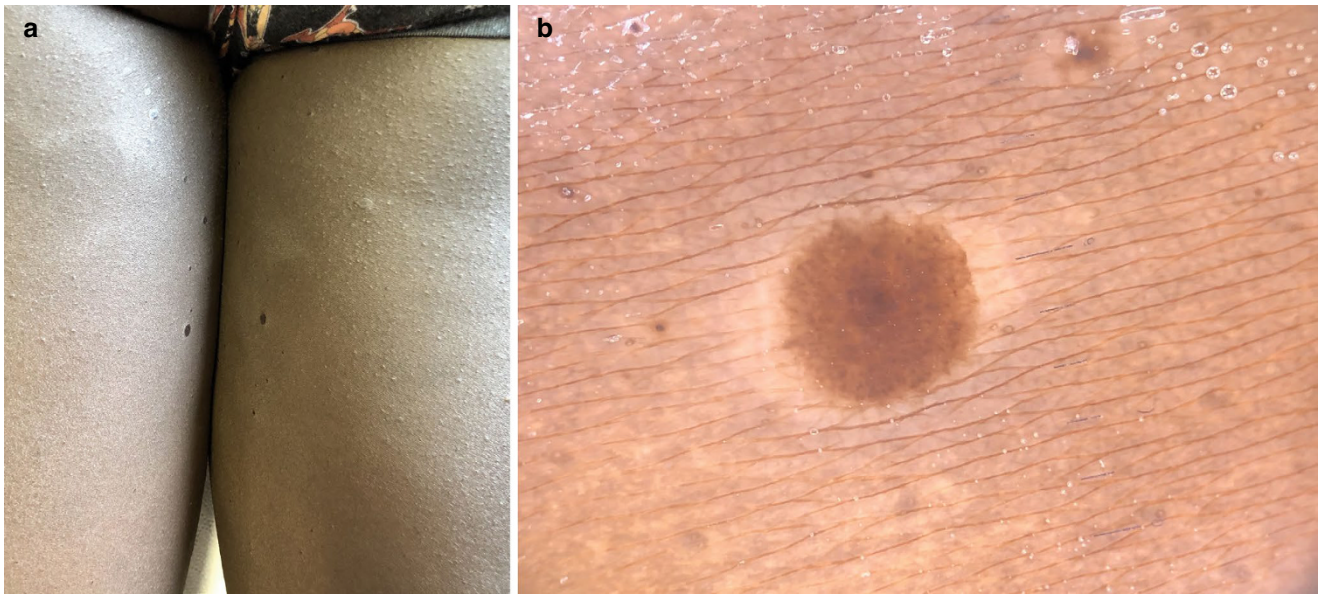

Fig. 3.4 Pityriasis lichenoides et varioliformis acuta in an African girl (**a**). Dermoscopic assessment displays a brown crust containing brown dots and surrounded by a white halo (**b**)

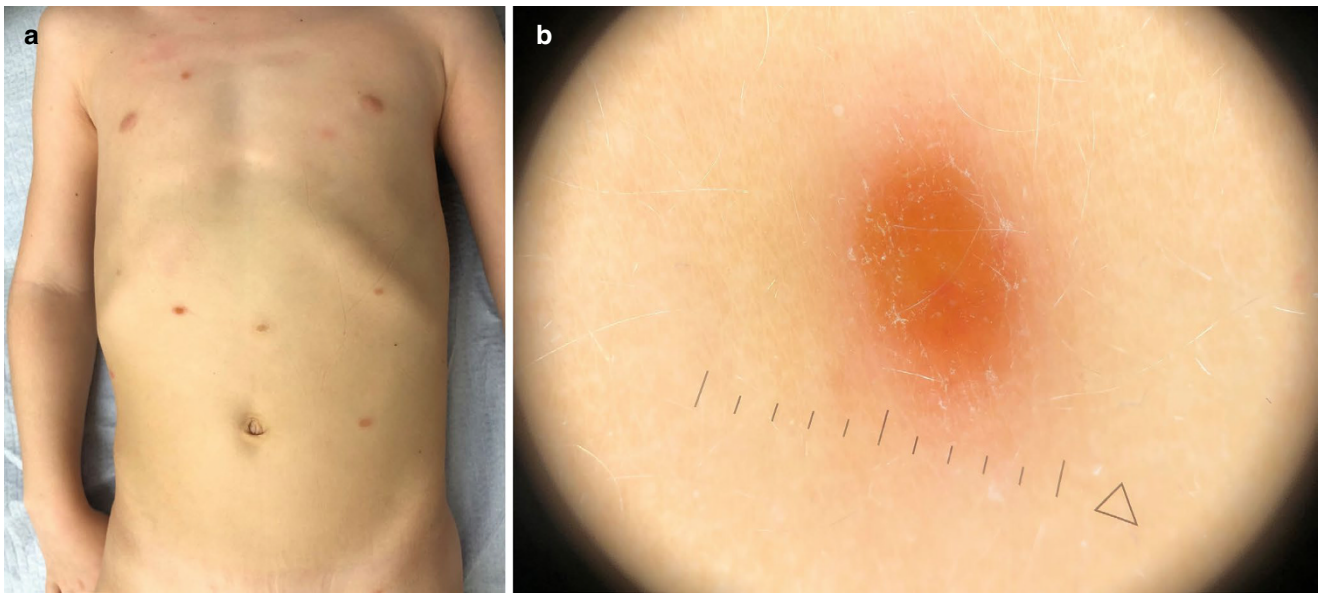

Fig. 3.5 Pityriasis lichenoides chronica in a Caucasian boy (**a**). Dermoscopy shows a diffuse orange structureless area and peripheral white scales (**b**)

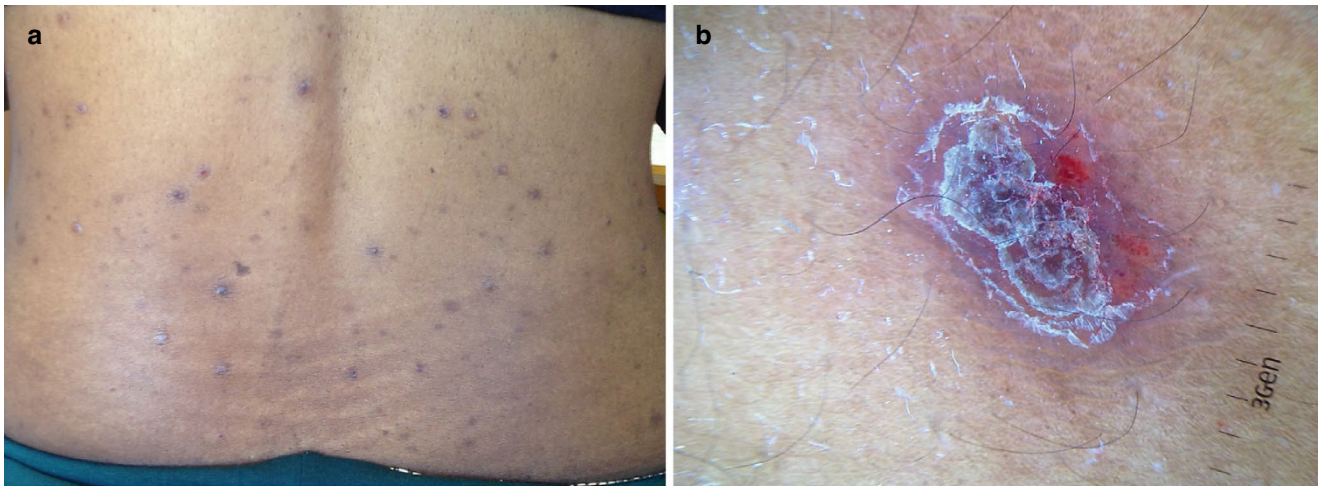

Fig. 3.6 Pityriasis lichenoides chronica in an African girl (**a**). A central white mica-like scale and peripheral smooth white scaling collarette are seen on dermoscopy along with purpuric dots/areas (**b**). (*From "Dermoscopy in General Dermatology for Skin of Color", Errichetti E, Lallas A, eds. CRC Press 2021*)

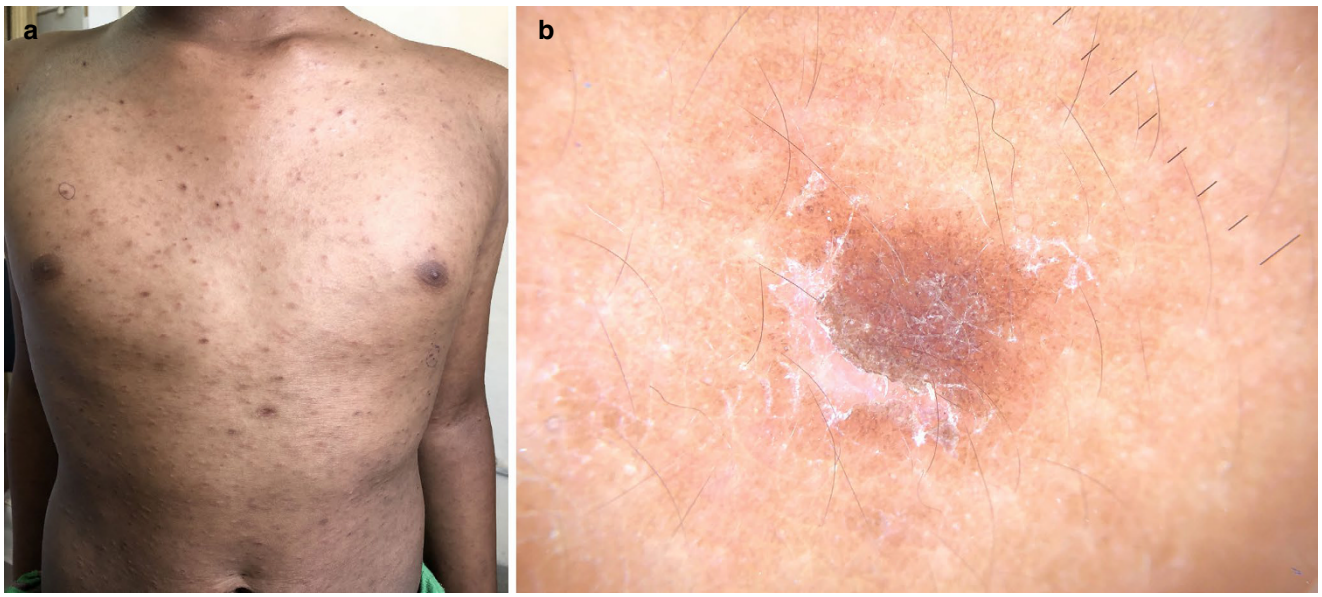

Fig. 3.7 Pityriasis lichenoides chronica in an Indian boy (**a**). Dermoscopic examination shows brown background and brown dots; a peripheral post-inflammatory hypopigmented area is also evident (**b**)

3.2 Pityriasis Rubra Pilaris

3.2.1 Introduction

Pityriasis rubra pilaris (PRP) is a chronic papulosquamous dermatosis with an unknown etiology, though streptococcal infections, vaccinations, and medications have been speculated to be possible antigenic triggers [1, 3, 8]. No gender or racial predilection has been reported; albeit hereditary forms do exist, it is usually sporadic, with a bimodal age distribution, with peaks in the first and fifth decades [1, 3, 8].

3.2.2 Clinical Presentation

Five clinical variants are classically reported, including classical (adult onset [type I] and juvenile onset [type III]), atypical (adult onset [type II] and juvenile onset [type V]), and circumscribed juvenile (type IV) [1, 3, 8]. Classical subtypes manifest as follicular hyperkeratotic papules and scaly inflammatory patches spreading in a cephalocaudal pattern (Figs. 3.8a and 3.9a) [1, 3, 8]. Erythroderma typified by areas of uninvolved skin (islands of sparing), palmoplantar keratoderma, and nail changes are other possible manifestations [1, 3, 8]. Atypical

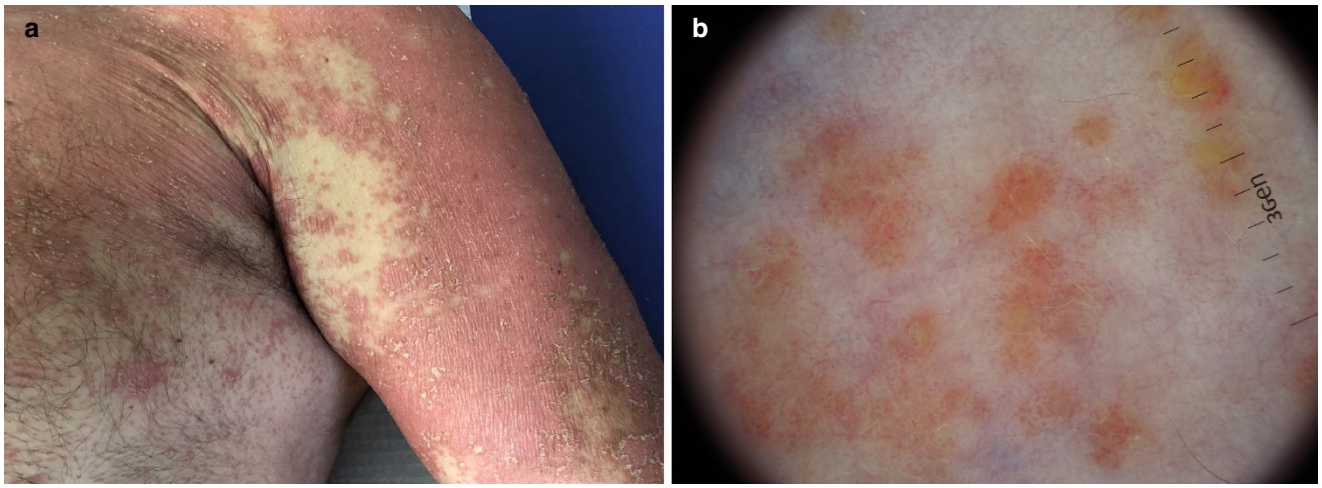

Fig. 3.8 Pityriasis rubra pilaris (classical adult type) in a Caucasian man (**a**). Dermoscopy displays multiple orange structureless areas surrounded by dotted and linear vessels (**b**)

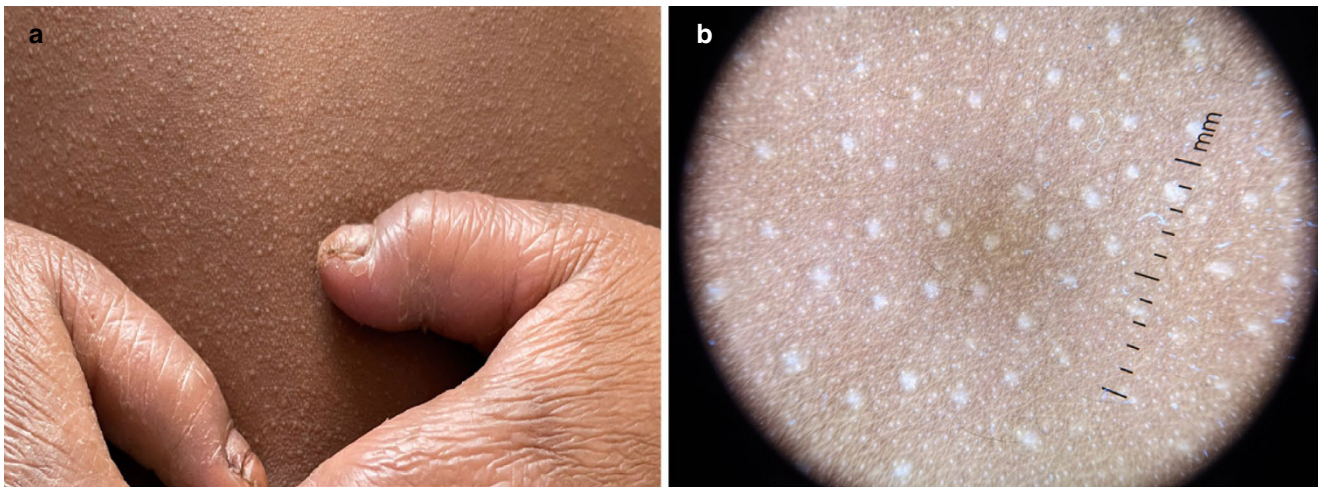

Fig. 3.9 Pityriasis rubra pilaris (classical juvenile type) in an African boy (**a**). Dermoscopic examination reveals white follicular plugs (**b**)

forms show follicular hyperkeratosis and either ichthyosiform lesions on the legs along with sparse scalp hair (adult onset) or scleroderma-like change of the hands and feet (juvenile onset) [1, 3, 8]. Finally, circumscribed juvenile PRP presents as scaly erythematous (psoriasis-like) plaques involving elbows and knees, with or without palmoplantar keratoderma (Figs. 3.10a and 3.11a) [1, 3, 8]. The most remarkable difference between fair- and dark-skinned patients is the diverse shade of erythema, being salmon-colored in the former and violaceous/brown-black in the latter (Figs. 3.8a, 3.9, 3.10 and 3.11a) [1, 3, 8].

3.2.3 Dermoscopy

The main dermoscopic finding of PRP consists of follicular scaling/plugs histologically corresponding to follicular hyperkeratosis (Figs. 3.8b, 3.9, 3.10 and 3.11b) [1, 3, 4, 6, 7]. A perifollicular yellow-orange halo or orange focal structureless areas due to dermal hemosiderin deposits may also be evident in lighter phototypes (Figs. 3.8b and 3.10b), whereas they are less commonly seen in very dark skin (Figs. 3.9b and 3.11b) [1, 3, 4, 6, 7]. However, orange structureless areas may be found on palms and soles of dark-skinned patients in case of keratoderma as palmoplantar areas show a lighter tone compared to other anatomical sites [1, 3, 4, 6, 7]. Notably, characteristic islands of non-erythematous (spared) skin displaying reticular vessels may be observed in erythrodermic forms in fair skin [1, 3, 4, 6, 7]. Other less specific dermoscopic findings include patchy whitish scales and scattered dotted/linear vessels (fair skin) (Fig. 3.8b and 3.10b) [1, 3, 4, 6, 7].

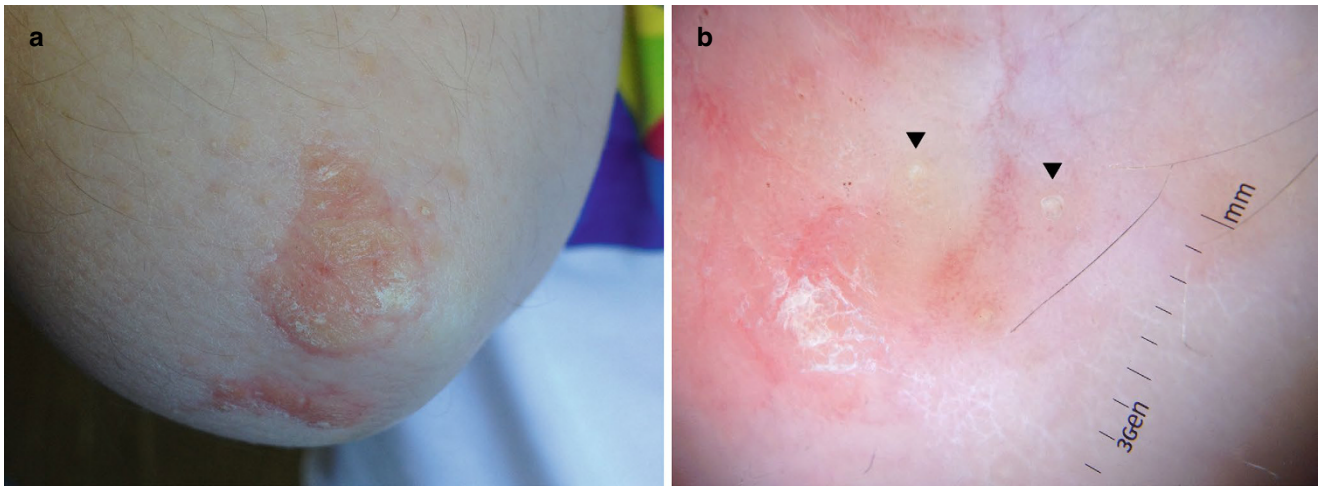

Fig. 3.10 Circumscribed juvenile pityriasis rubra pilaris in a Caucasian boy (**a**). Follicular plugs with perifollicular yellow-orange halo (arrowheads) along with patchy white scales and some dotted/linear vessels are evident on dermoscopy (**b**)

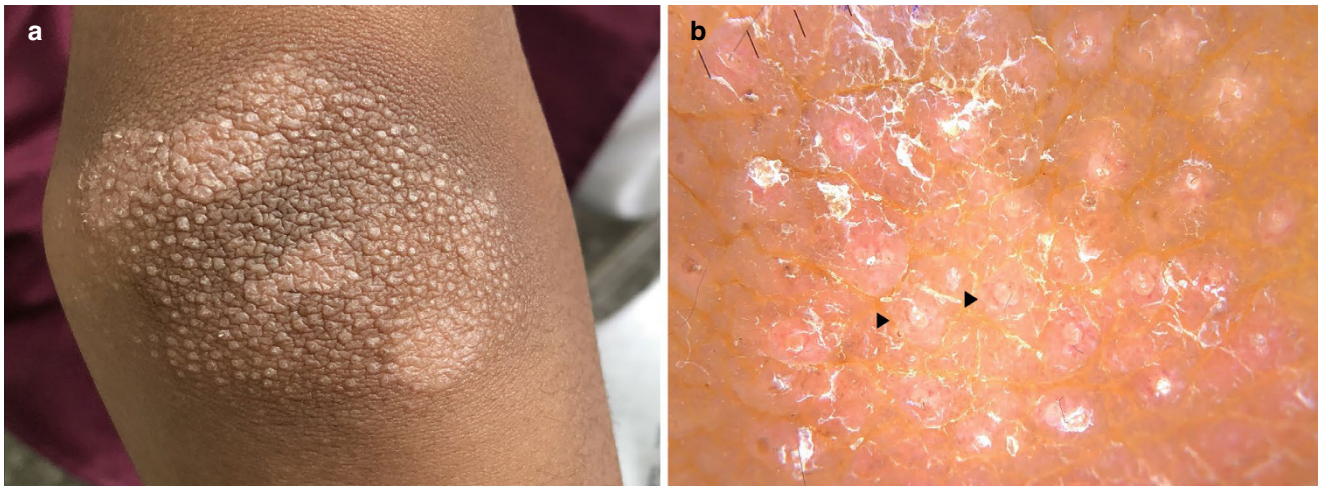

Fig. 3.11 Circumscribed juvenile pityriasis rubra pilaris in an Indian boy (**a**). Follicular plugs with subtle perifollicular yellow halo (arrowheads) along with some patchy white scales (**b**)

3.3 Porokeratosis

3.3.1 Introduction

Porokeratosis is a term encompassing a heterogenous group of genodermatoses histologically typified by the so-called cornoid lamella clinically resulting in a peripheral keratotic rim [1, 3, 9]. Porokeratosis is more commonly seen in fair than dark skin [1, 3, 9].

3.3.2 Clinical Presentation

Four main clinical subtypes are classically recognized, i.e., disseminated actinic porokeratosis, porokeratosis of Mibelli, linear porokeratosis, and disseminated porokeratosis, with the first two forms being the most frequently encountered ones [1, 3, 9]. All such variants are typified by slightly raised papules/plaques featuring a peripheral keratotic rim (Figs. 3.12a, 3.13, 3.14, 3.15 and 3.16a) [1, 3, 9]. Lesions are single or few in porokeratosis of Mibelli (Figs. 3.15a and 3.16a) and multiple in the other forms (Figs. 3.12a, 3.13 and 3.14a), with restriction to sun-exposed areas in actinic form and distributed along Blaschko lines in linear variant (due to a mosaicism) [1, 3, 9]. While an erythematous background is often visible in lighter phototypes (Fig. 3.12a), in dark skin patients, lesions often show a hyper- or hypopigmented appearance (Figs. 3.14a and 3.16a) [1, 3, 9].

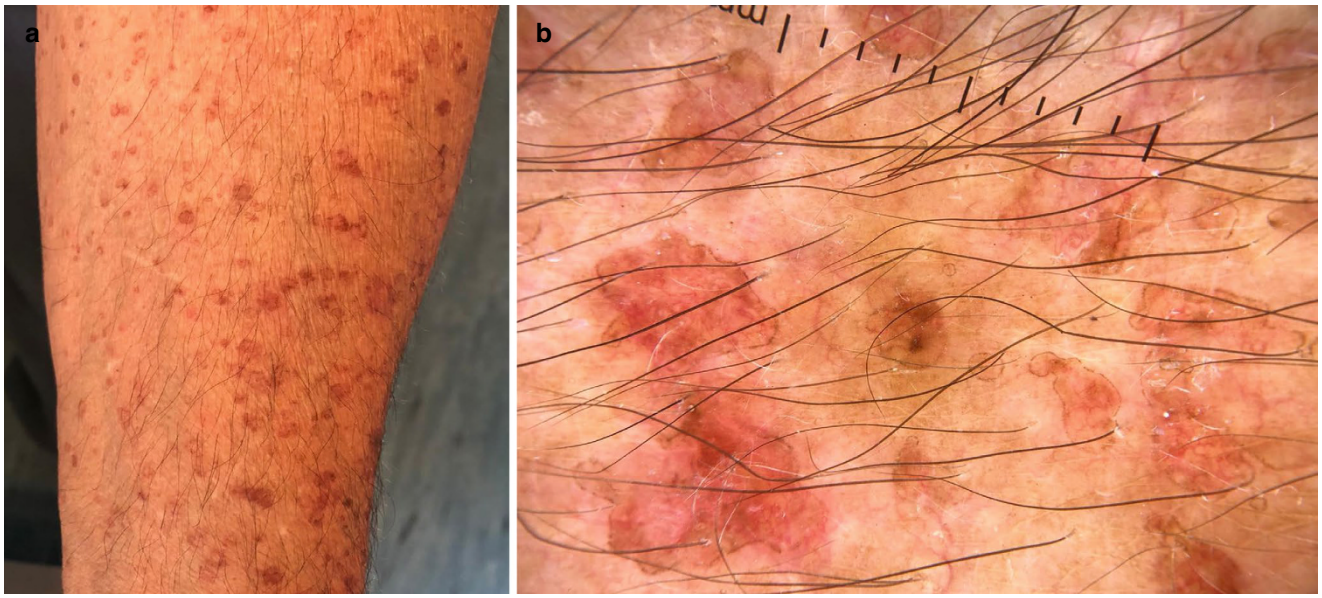

Fig. 3.12 Disseminated actinic porokeratosis (inflammatory lesions) in a Caucasian lady (**a**). Dermoscopy reveals a brownish keratotic collarette with a double free edge along with central erythema and some unfocused vessels (**b**)

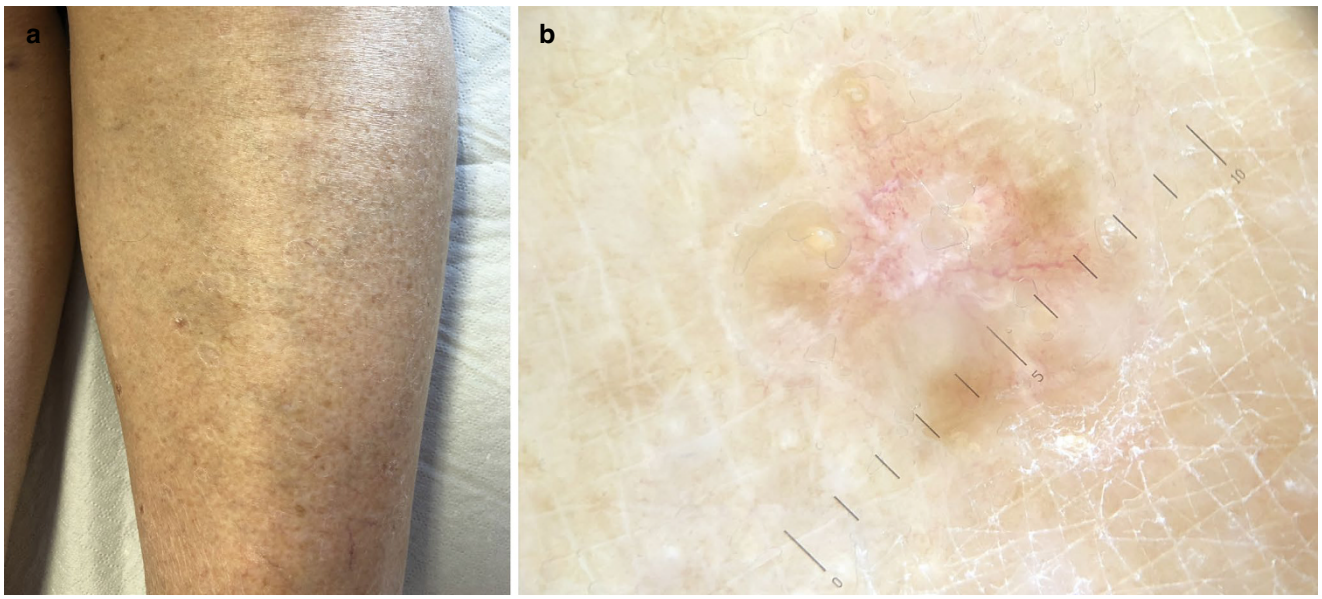

Fig. 3.13 Disseminated actinic porokeratosis (non-inflammatory lesions) in a Caucasian lady (**a**). Dermoscopic assessment shows a white keratotic collarette with a double free edge along with central white areas and linear/dotted/linear-curved vessels (**b**)

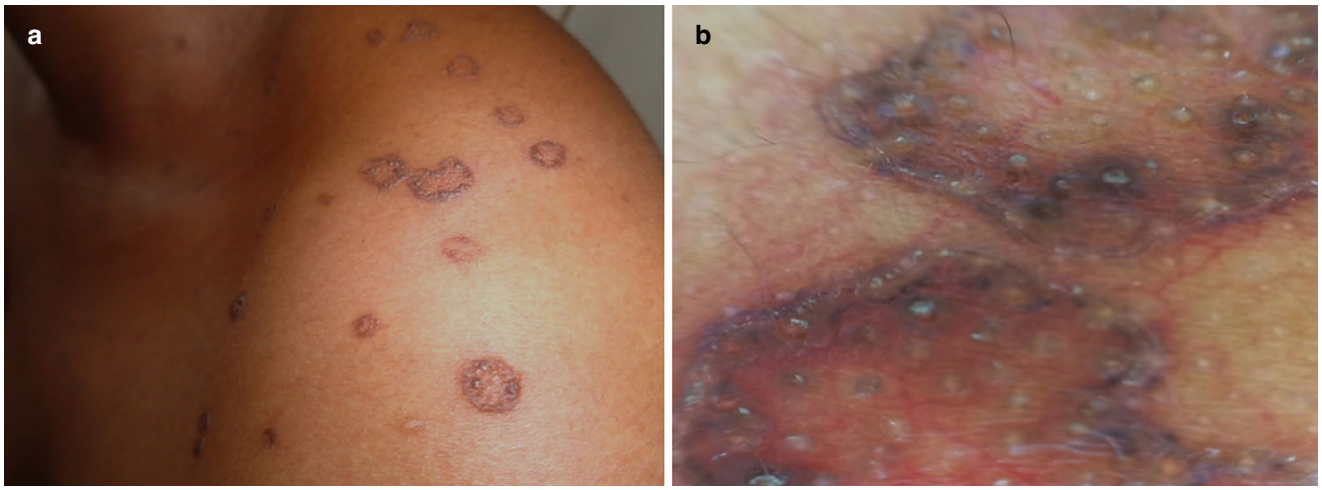

Fig. 3.14 Disseminated actinic porokeratosis in an African man (**a**). The typical brownish keratotic collarette with a double free edge is evident on dermoscopy along with keratotic follicular plugs (**b**)

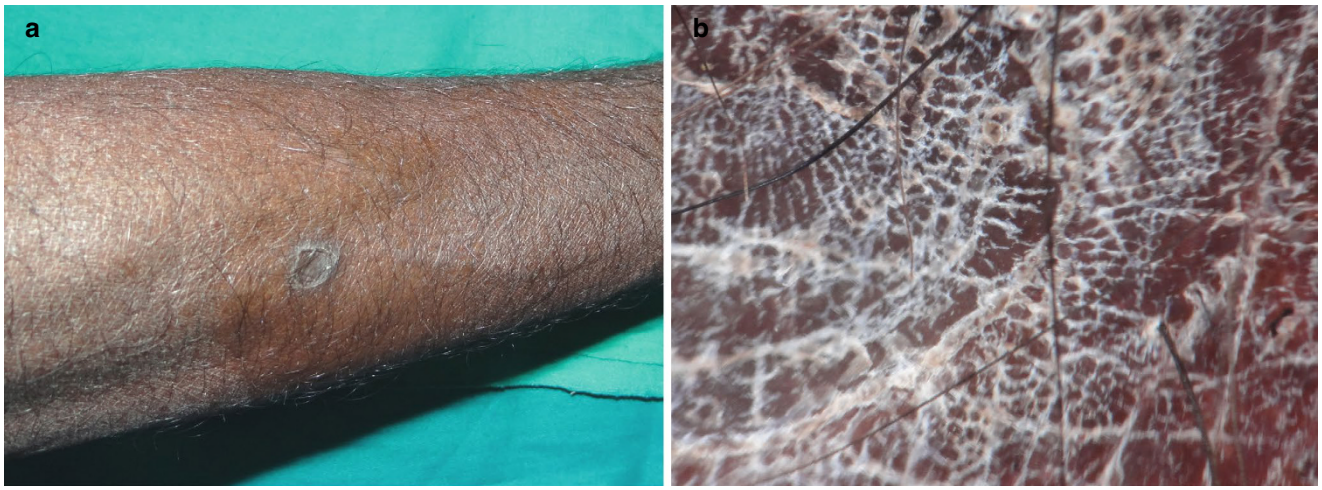

Fig. 3.15 Porokeratosis of Mibelli of the forearm of an Indian man (**a**). Dermoscopy reveals a peripheral gray keratotic collarette with a double free edge which is barely detectable as there is also remarkable hyperkeratosis (**b**)

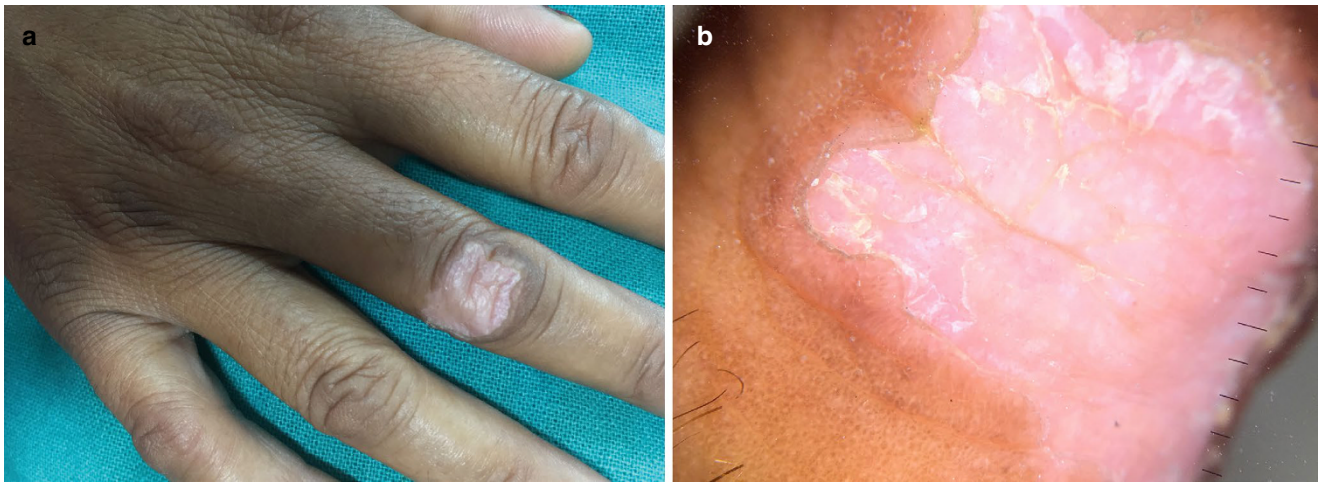

Fig. 3.16 Porokeratosis of Mibelli of the dorsal aspect of the hand in an Indian girl (**a**). Dermoscopic examination shows the typical peripheral brownish keratotic collarette with a double free edge and a central white background (**b**)

3.3.3 Dermoscopy

Dermoscopy is a key diagnostic tool in porokeratosis, especially in dark-skinned patients in whom the peripheral keratotic margin is not always easy to identify on clinical ground [1, 3, 6, 7, 10, 11]. The dermoscopic hallmark consists of a peripheral keratotic rim featuring a double free edge representing the cornoid lamella on histology (Figs. 3.12b, 3.13, 3.14, 3.15 and 3.16b) [1, 3, 6, 7, 10, 11]. Its color may be either white/gray or brown, with the latter being more frequently seen in darker phototypes [1, 3, 6, 7, 10, 11]. Additionally, several less specific findings may be observed in the center of the lesions, including white scales, dotted/reticular vessels (especially fair skin), keratotic plugs, white or brown structureless areas, and brown dots (especially dark skin) (Figs. 3.12b, 3.13, 3.14, 3.15 and 3.16b) [1, 3, 6, 7, 10, 11].

3.4 Lichen Nitidus

3.4.1 Introduction

Lichen nitidus (LN) is an inflammatory condition typified by a self-limiting course in a few years that has been supposed to be triggered by allergens causing epidermal and dermal antigen-presenting cells to activate a cell-mediated response [1, 3]. No racial predilection has been reported in the literature, despite the general impression of a greater involvement of dark-skinned patients compared to lighter phototypes [1, 3].

3.4.2 Clinical Presentation

It typically presents as 1–2 mm in diameter, skin-colored or hypopigmented (particularly in dark-skinned patients), asymptomatic, shiny papules (Figs. 3.17a, 3.18 and 3.19a) most commonly affecting the abdomen, chest, extremities, and genitalia (especially in men) [1, 3]. Koebner phenomenon with lesions arranged in a linear pattern is quite common in LN [1, 3]. Several less frequent clinicopathological subtypes have been described, such as actinic, linear, perforating, purpuric/hemorrhagic, and vesicular [1, 3].

3.4.3 Dermoscopy

The typical dermoscopic pattern of LN in both fair and dark skin consists of round, homogeneous, sharply demarcated, white areas (one for each papule) characteristically devoid of skin markings (Figs. 3.17b and 3.19b); an orange hue may also be seen in lighter phototypes (Fig. 3.18b) [1, 3, 4, 6, 7, 12]. From a dermoscopic-pathological correlation point of view, these areas correspond to the dense lymphohistiocytic inflammatory cell infiltrate located in close proximity to the thinned epidermis and enveloped by bordering elongated rete ridges ("claw clutching a ball" appearance), while the loss of skin markings would be the result of the epidermal-dermal junction flattening (due to the pressure by the underlying dermal inflammatory cell infiltrate) [1, 3, 4, 6, 7, 12]. Dermoscopy may also come in handy to show clinically unnoticeable linear arrangement of some lesions ("micro-Koebner phenomenon") (Fig. 3.18b) [1, 3, 4, 6, 7, 12].

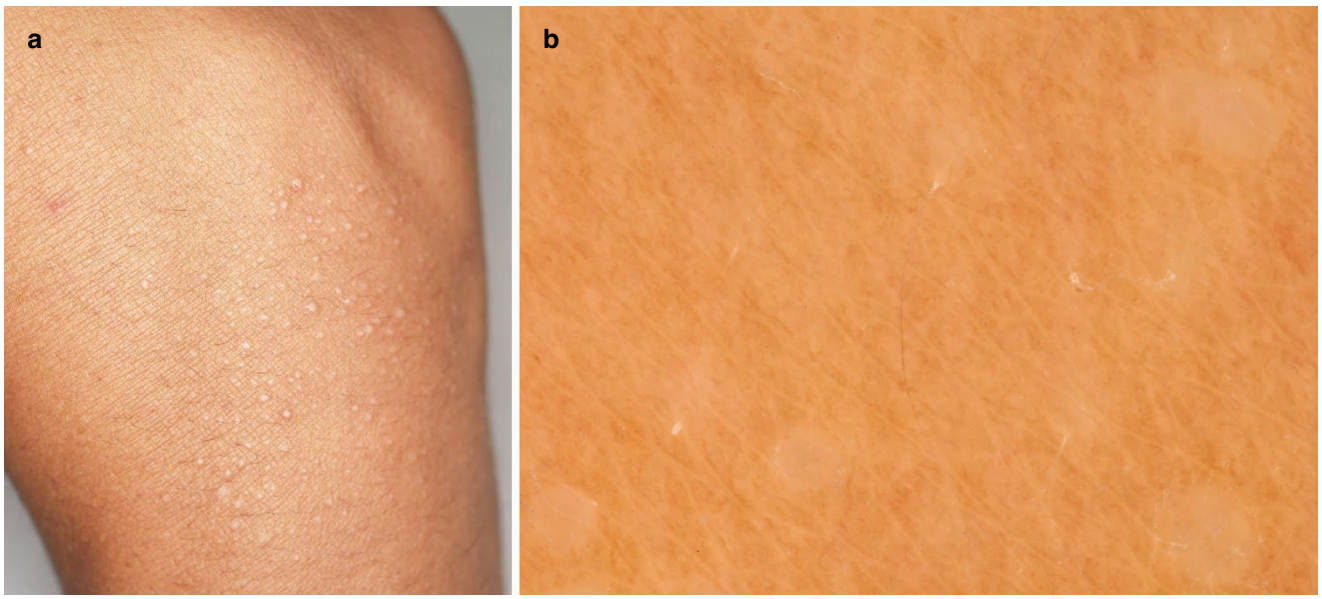

Fig. 3.17 Lichen nitidus (hypopigmented papules) in a Caucasian girl (**a**). Dermoscopy displays round white areas devoid of skin markings (**b**)

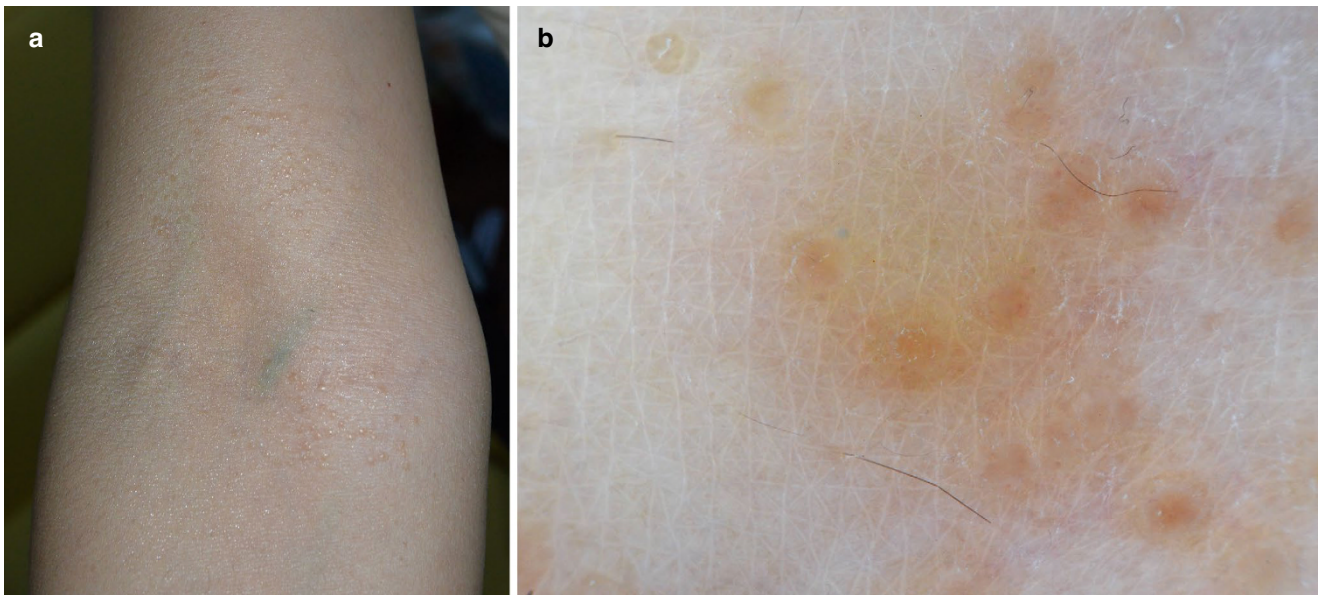

Fig. 3.18 Lichen nitidus (skin-colored papules) in a Caucasian girl (**a**). Round orange areas are evident on dermoscopic assessment; some of them are arranged in a linear pattern ("micro-Koebner phenomenon") (**b**)

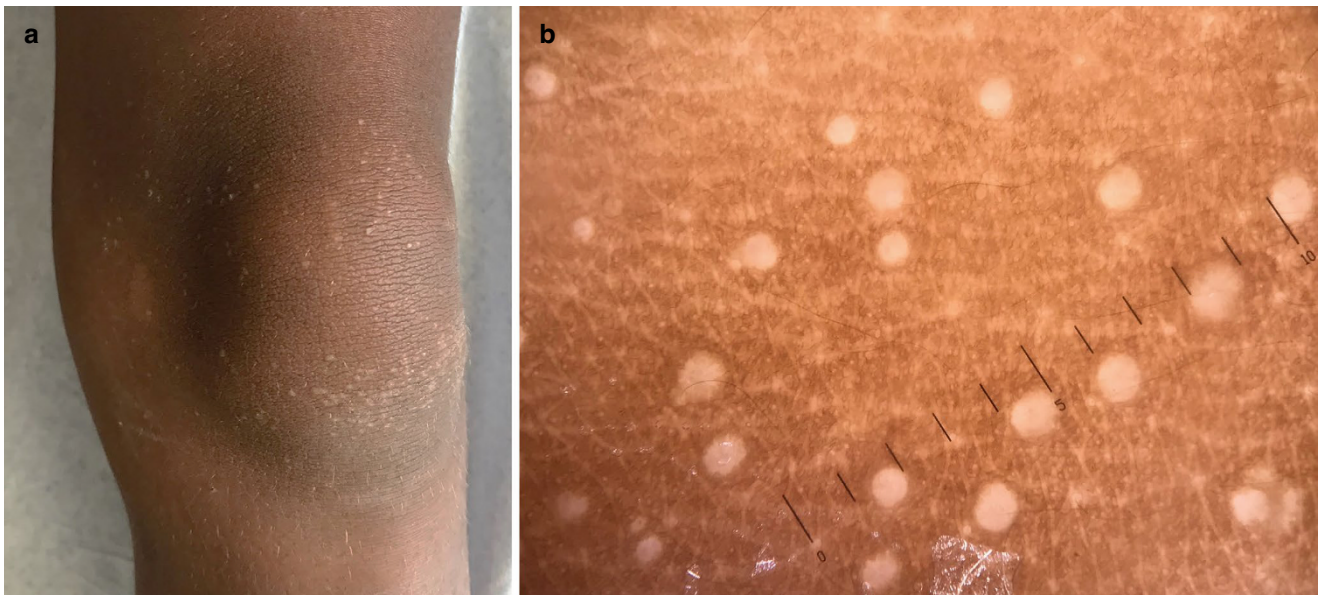

Fig. 3.19 Lichen nitidus (hypopigmented papules) in an African boy (**a**). Dermoscopy reveals very well-demarcated round white areas devoid of skin markings (**b**)

3.5 Lichen Striatus

3.5.1 Introduction

Lichen striatus (LS) is a self-limiting inflammatory condition of unknown etiology manifesting with lesions following Blaschko's lines [3, 13]. Although it may occur at any age, it tends to be more common in children, with a spontaneous healing within 1 year, yet it may last longer [3, 13].

3.5.2 Clinical Presentation

LS usually manifests as skin-colored, erythematous, or hypopigmented (especially in darker phototypes) small (1–3 mm) papules often coalescing into linear plaques following lines of Blaschko of an extremity or trunk (Figs. 3.20a and 3.21a) [3, 13]. Post-inflammatory hypopigmentation is quite frequent especially in dark-skinned patients and may last for long time (Figs. 3.22a and 3.23a), yet post-inflammatory hyperpigmentation is more uncommon and visible nearly exclusively in skin of color (Fig. 3.24a) [3, 13]. Onychodystrophy may also be seen less commonly, and it is often limited to a single nail [3, 13].

3.5.3 Dermoscopy

Dermoscopic features of LS significantly vary according to patient's phototype [3, 14]. In detail, the main findings in fair skin are poorly specific and include vessels of mixed morphology (dotted and linear-irregular) and white scaling (Fig. 3.20b), though ill-defined white areas may also be seen in hypopigmented variants or post-inflammatory hypopigmentation (Fig. 3.22b) [3, 14]. On the other hand, whereas the abovementioned vascular structures may also be detected in skin of color (Fig. 3.21b), LS in darker phototypes is mainly typified by non-follicular, ill-defined,

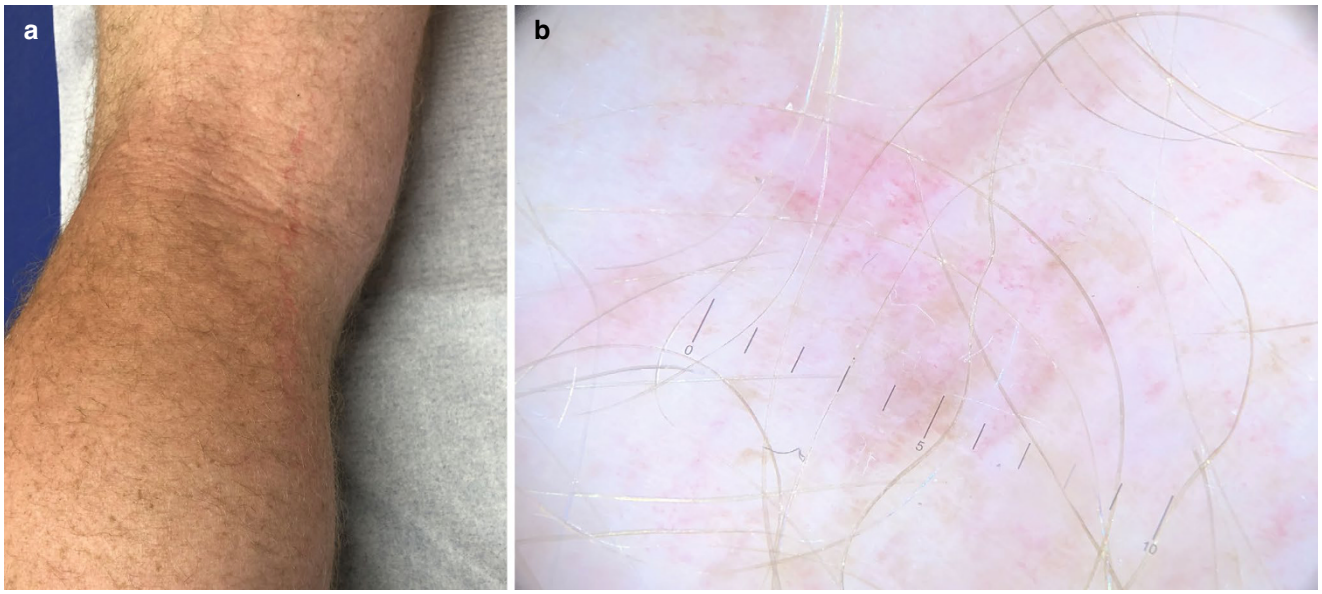

Fig. 3.20 Lichen striatus (papular erythematous lesions) in a Caucasian man (**a**). A mixed vascular pattern (dotted and linear vessels) is evident on dermoscopy (**b**)

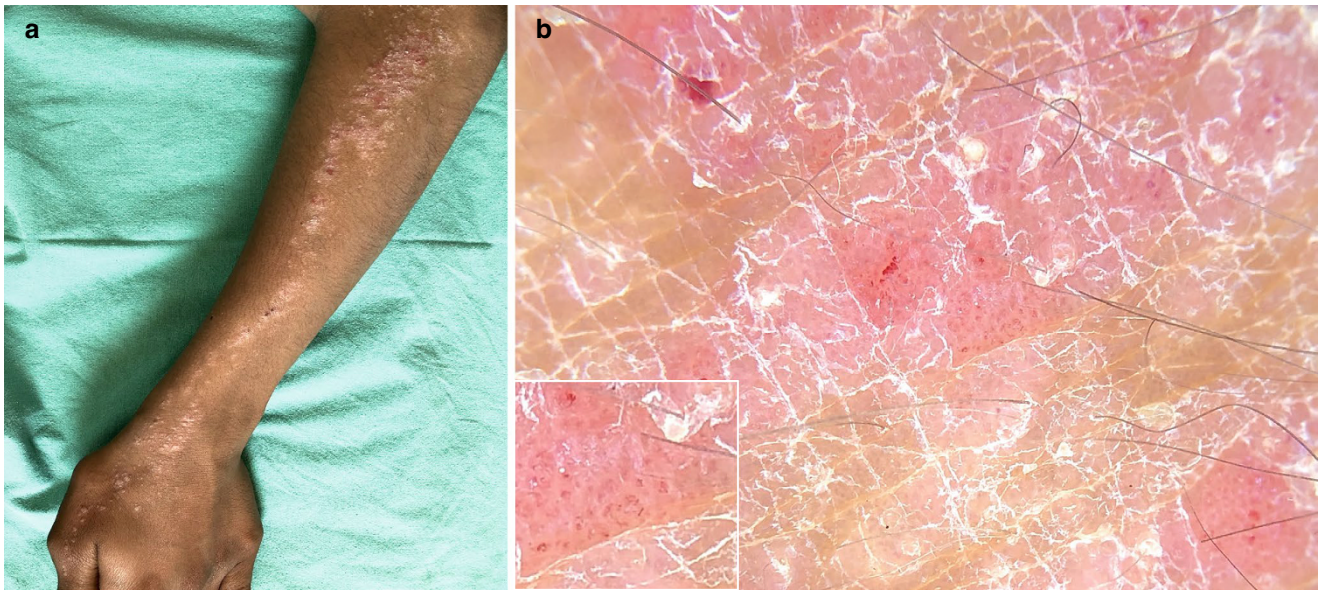

Fig. 3.21 Lichen striatus (papular hypopigmented lesions) in an Indian boy (**a**). Dermoscopic examination reveals white scales as well as dotted and linear vessels (magnification in the inset) (**b**)

structureless white areas often coalescing to form polycyclic structures, with or without white or brown scales and brown dots (Fig. 3.23b) [3, 14]. From a histological point of view, white areas would be related to epidermal acanthosis and/or inflammatory reduction of the melanin content, while white and brown scaling would correspond to hyperkeratosis (with melanin exfoliation for brown scales) [3, 14].

Of note, in very dark phototypes, white eccrine sweat gland openings surrounded by brown circles may be also visible in the abovementioned white areas as a result of the typical arrangement of the dense inflammatory infiltrate around the eccrine sweat glands and ducts (Fig. 3.24b) [3]. Additionally, brown network-like structures may also be seen in post-inflammatory hyperpigmented lesions (Fig. 3.24b) [3, 14].

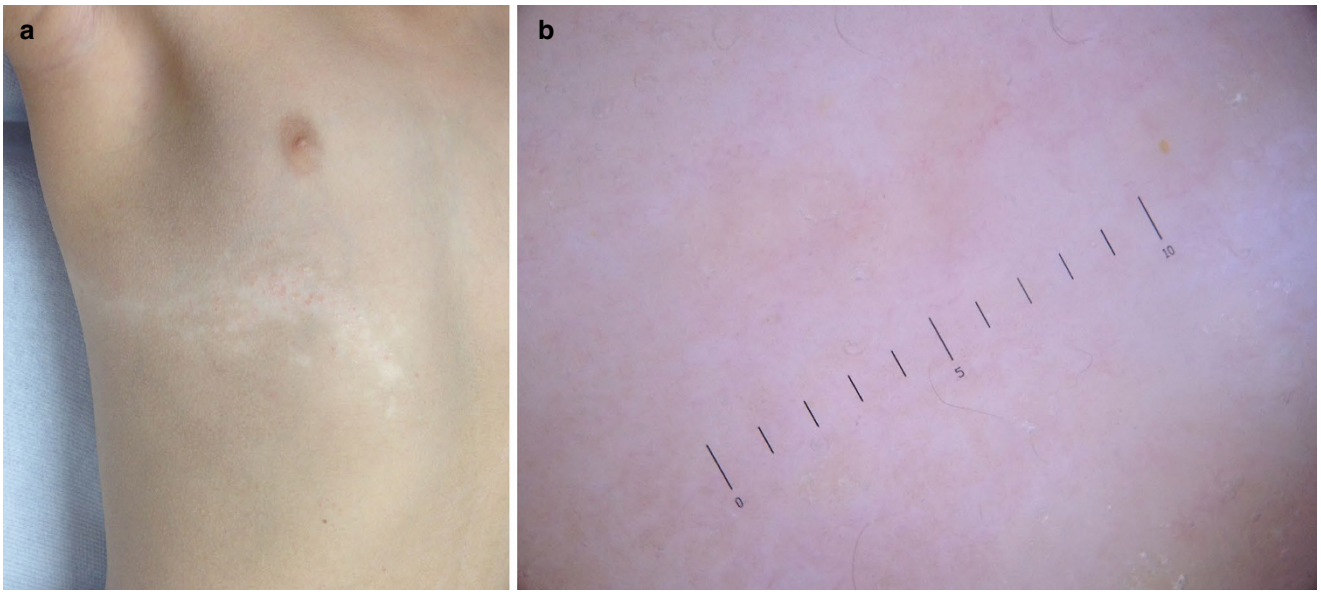

Fig. 3.22 Lichen striatus (hypopigmented post-inflammatory lesions) in a Caucasian girl (**a**). Ill-defined confluent white areas are seen on dermoscopy (**b**)

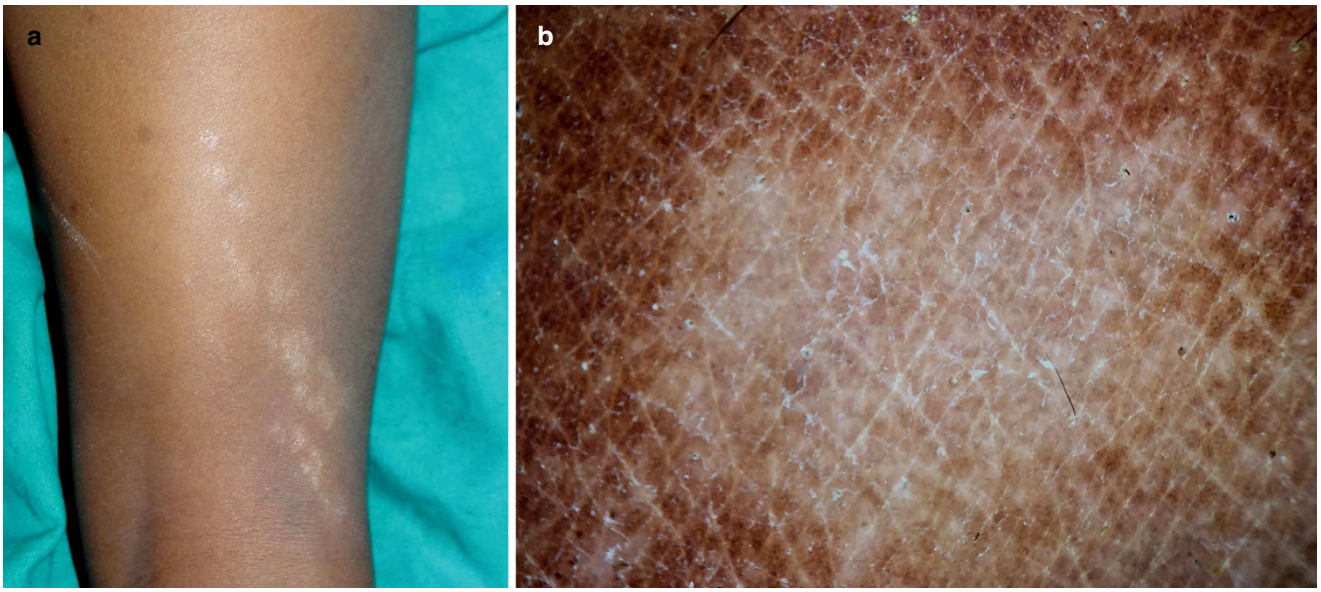

Fig. 3.23 Lichen striatus (hypopigmented post-inflammatory lesions) in an Indian boy (**a**). Dermoscopy shows ill-defined confluent white areas with white scales and a few brown dots (**b**). (*Courtesy of Balachandra Ankad, MD – Bagalkot, India*)

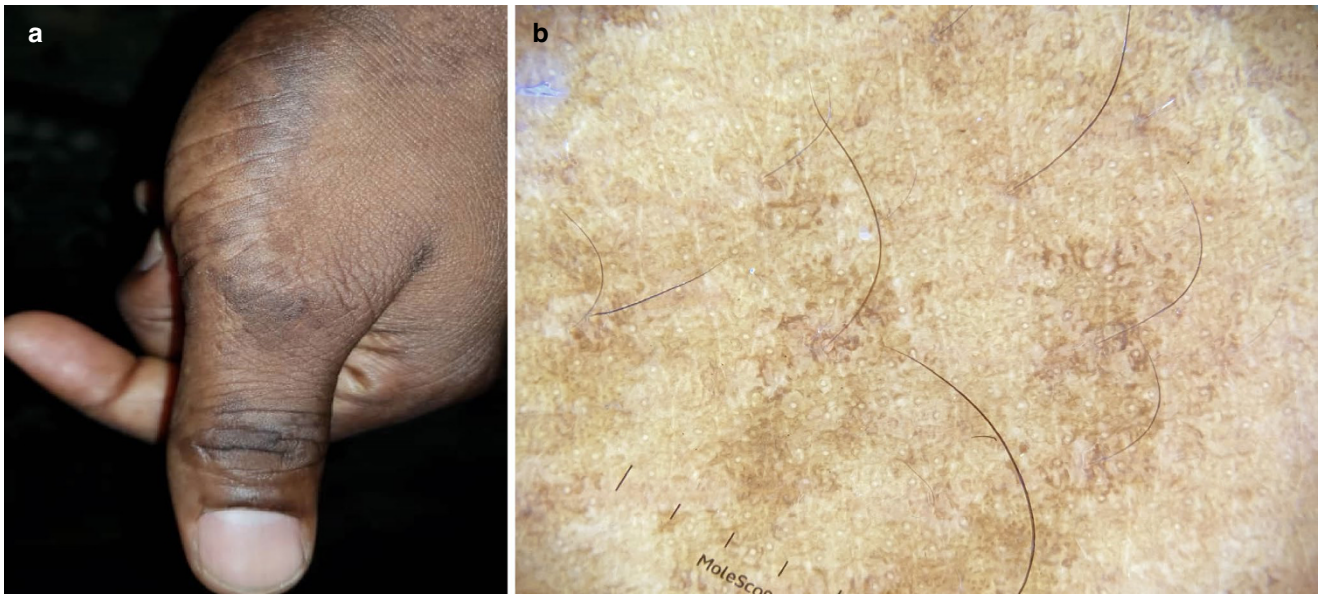

Fig. 3.24 Lichen striatus (hyperpigmented post-inflammatory lesions) in an African man (**a**). Dermoscopy displays brown network and brown circles around white eccrine sweat gland openings (**b**). (*Courtesy of Ibrahima Traoré, MD – Conakry, Guinea*)

3.6 Lichen Spinulosus

3.6.1 Introduction

Lichen spinulosus is a follicular keratotic dermatoses most commonly seen in children, though it may also be encountered in adults [15, 16]. Albeit its precise etiopathogenesis is yet to be clear, some authors have speculated it might be an abnormal reaction pattern of follicles to various triggering factors, including among the others vitamin A deficiency, exposure to toxins, infections (e.g., HIV), and autoimmune diseases (e.g., Crohn's disease) [15, 16].

3.6.2 Clinical Presentation

It presents with follicular papules of 1–3 mm in diameter associated with thorny and hair-like spines that cluster into more or less symmetrical plaques (Figs. 3.25a and 3.26a) [15, 16]. Lesions are usually localized, though generalized forms have been reported; trunk, neck, acral areas, buttocks, abdomen, and extensor arms are among the most commonly involved areas [15, 16]. Lichen spinulosus is usually asymptomatic and aflegmasic, yet pruritus and erythema may be observed [15, 16]. Of note, in darker phototypes, lesions are typically more keratotic, and erythema is not found (Fig. 3.26a). Evolutions are variable, with some cases showing self-resolution and others persisting for many years [16].

3.6.3 Dermoscopy

Dermoscopic pattern of lichen spinulosus consists of equidistant follicular plugs surrounded by a white halo (Figs. 3.25b and 3.26b), histologically corresponding to follicular hyperkeratosis and perifollicular acanthosis; hairs are usually present [15]. Notably, the color of the plugs may vary based on the phototype, with white and brown hue being more commonly observed in fair and dark skin, respectively. This difference is due to the melanin exfoliation that is associated with follicular hyperkeratosis in dark-skinned patients [6]. Additionally, plugs tend to be more pronounced in skin of color because of the higher tendency to follicular reactions, whereas a perifollicular erythematous halo may unfrequently be found in lighter phototypes [6]. Lastly, perifollicular scaling (Fig. 3.25b) and coiled hairs may also sometimes be evident, though the latter finding is usually focal (and not diffuse as seen in keratosis pilaris) [15].

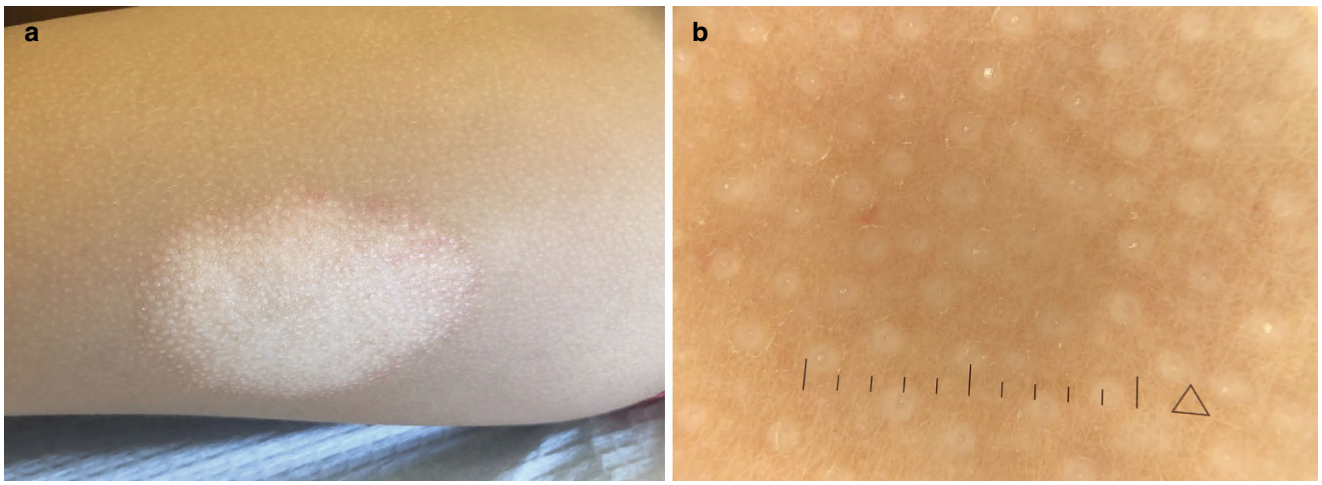

Fig. 3.25 Lichen spinulosus in a Caucasian child (**a**). Dermoscopy reveals white follicular plugs surrounded by a white halo (**b**)

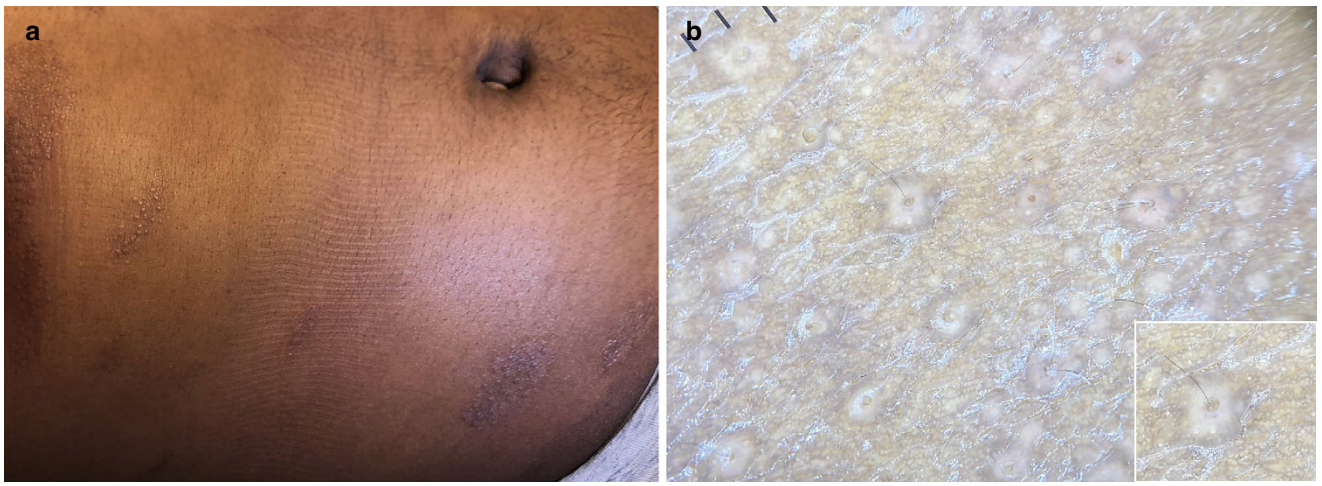

Fig. 3.26 Lichen spinulosus in an African woman (**a**). Dermoscopic examination shows brownish follicular plugs with a white halo (**b**) (*Courtesy of Rosana Buffon, MD – Vicenza, Italy*)

3.7 Keratosis Pilaris

3.7.1 Introduction

Keratosis pilaris is a common condition that has been related to filaggrin loss-of-function mutations, atopic dermatitis, and ichthyosis vulgaris; such a condition is seen in every skin type [17, 18].

3.7.2 Clinical Presentation

It typically manifests as more or less diffuse follicular scaly papules that are usually asymptomatic (Figs. 3.27a and 3.28a) [17, 18]. The main anatomical sites involved include the extensor surfaces of the proximal extremities, though involvement of the lower limbs or cheeks is not so rare [17, 18]. Lesions in light phototypes show a reddish hue (Fig. 3.27a), whereas they are brownish in skin of color (Fig. 3.28a).

3.7.3 Dermoscopy

Dermoscopic examination of keratosis pilaris usually reveals coiled and twisted hairs which are surrounded/embedded by white scales [15]. Notably, a perifollicular erythematous halo is visible in fair-skinned patients, while a brownish halo (diffuse pigmentation or reticular pigmentation) is evident in skin of color [15]. Moreover, a few dotted vessels may also sometimes be observed around the follicles in lighter phototypes [15].

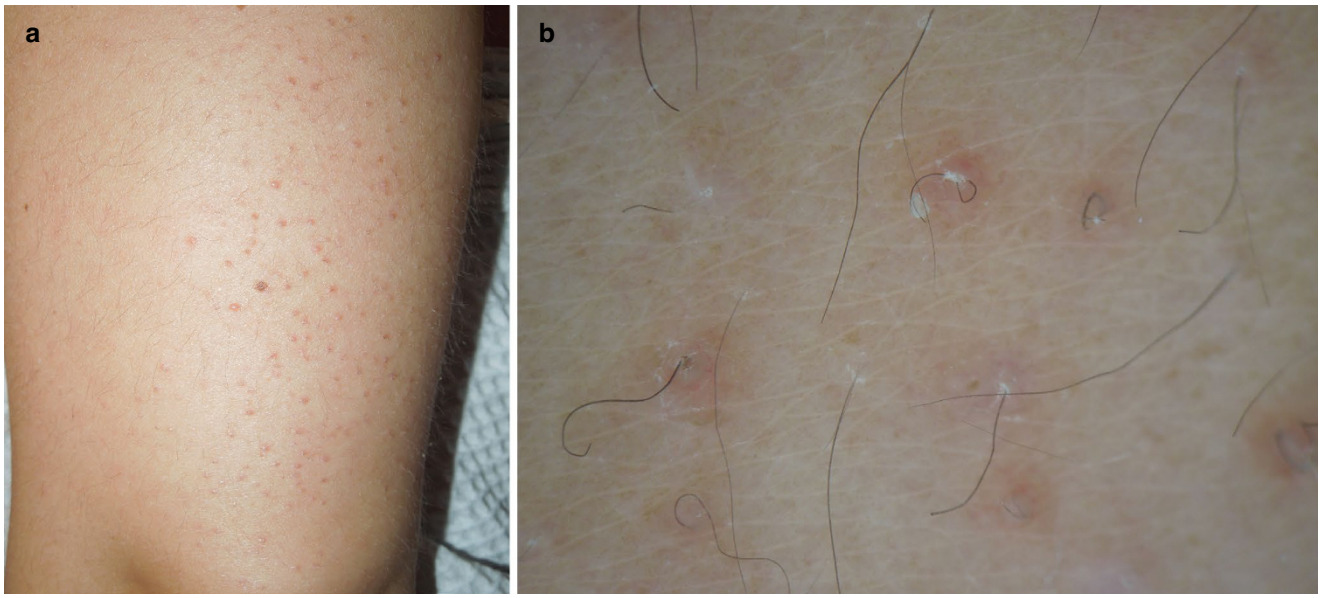

Fig. 3.27 Keratosis pilaris in a Caucasian boy (**a**). Coiled hairs along with perifollicular white scaling surrounded by an erythematous halo are evident on dermoscopy (**b**)

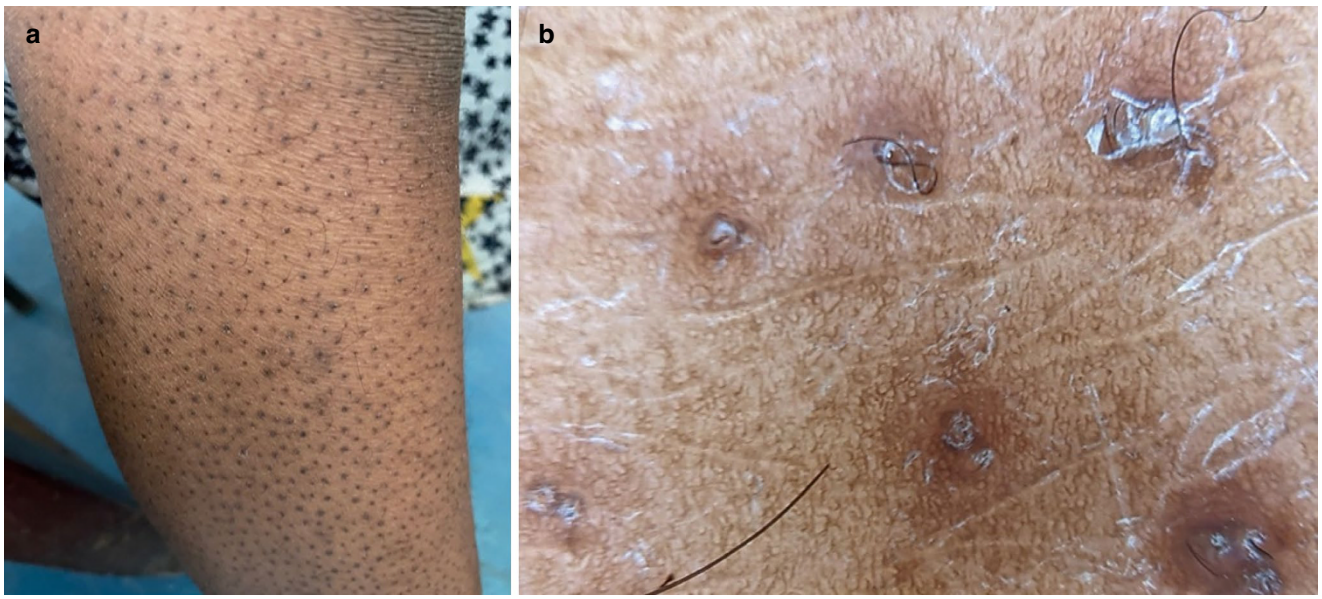

Fig. 3.28 Keratosis pilaris in an African woman (**a**). Dermoscopy displays coiled and twisted hairs embedded by white scaling along with a brown reticular perifollicular pigmentation (**b**)

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