Dermatoses with "collarette of skin"

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A collarette in dermatology refers to *"a narrow rim of loosened keratin overhanging the periphery of a circumscribed skin lesion, attached to the normal surrounding skin."*[1] The outer margin of the "collarette" is adherent while the inner margin is free. In practice, however, the skin forming the collarette may be scaly (e.g., pityriasis rosea, keratolysis exfoliativa, etc.) or thick and ridge-like, forming a rim around the lesion (e.g., acral fibrokeratoma, viral warts, etc.). While many disorders primarily exhibit such a morphology [Table 1], it may be a secondary or a nonspecific manifestation in certain others.

Disorders of Keratinization

Pityriasis rosea

Pityriasis rosea is a common, acquired, self-limiting papulosquamous disorder commonly affecting children and young adults. A possible infective role has been speculated in its pathogenesis but has not been proven unequivocally. Classically, the disease begins as an erythematous macule with a peripheral rim of fine scaling that enlarges over the next few days into an annular to oval patch ("herald patch," "mother patch"), commonly involving the trunk. The scaling appears as a collarette and characteristically lags behind the outer erythematous edge of the patch – "trailing scale" or the "hanging curtain sign" [Figure 1]. In the next 1–2 weeks, smaller multiple pruritic lesions, similar to the herald patch, develop predominantly on the trunk extending up to the proximal arms. On the back, the long axes of these lesions are aligned parallel to the relaxed skin tension lines giving a "Christmas tree" appearance.[2,3] The disease is self-limiting and resolves spontaneously in approximately 6 weeks.[2] Atypical forms have been described. Although innocuous, pityriasis rosea occurring in the first trimester of pregnancy may account for an increased risk of spontaneous abortion, preterm delivery and infantile hypotonia and hyporeactivity.[4,5] A pityriasis rosea-like drug eruption can occur with certain medications as well.[6]

Keratolysis exfoliativa

Keratolysis exfoliativa is a common, noninflammatory, focal or diffuse peeling of the skin of palms and less commonly of the soles. It is usually asymptomatic, commonly affecting healthy individuals. It is common in summer and worsened by warm climate. Clinically, it is characterized by discrete air-filled tiny vesicles that rupture and extend in an irregular annular or circinate pattern for up to 10 mm, with a characteristic peripheral collarette of scaling [Figure 2]. Keratolysis exfoliativa is believed to be a milder variant of dyshidrotic eczema or is associated with atopy. Chang *et al.* suggest the disorder to be a distinct peeling disorder occurring mainly due to premature corneodesmolysis.[7-9]

Ichthyosis bullosa of Siemens (superficial epidermolytic ichthyosis)

Ichthyosis bullosa of Siemens is an autosomal dominant disorder of keratinization that is regarded as a superficial variant of bullous congenital ichthyosiform erythroderma (epidermolytic hyperkeratosis). It occurs due to mutation in the *keratin 2e* gene. Clinically, it is characterized by superficial trauma-induced blisters that spontaneously rupture leaving behind denuded skin with annular collarette-like scaling at the margins, classically referred to as *mauserung* phenomenon (epidermal moulting). The flexures, lower abdomen and the pretibial areas are commonly involved which may also exhibit a grayish, rippled, scaly hyperkeratosis. The disease usually begins

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Table 1: Dermatoses exhibiting a collarette of scaling

Category	Dermatoses with scaly collarette	Dermatoses with thick keratotic collarette
Keratinization disorders	Pityriasis rosea Keratolysis exfoliativa Ichthyosis bullosa of Siemens	Porokeratosis
Infectious disorders	Cutaneous candidiasis Bullous impetigo Secondary syphilis Bacillary angiomatosis Staphylococcal blepharitis Pitted keratolysis	Plantar warts
Vesico-pustular disorders	Transient neonatal pustular melanosis Subcorneal pustular dermatosis Pustular psoriasis Acute generalized exanthematous pustulosis	
Gyrate erythemas	Erythema annulare centrifugum Erythema gyratum repens	
Cutaneous tumors	Clear cell acanthoma Pyogenic granuloma	Acral fibrokeratoma Supernumerary digit Cutaneous horn Subungal exostosis Eccrine poroma

in the neonatal period and extends into the childhood, and may occasionally persist until adulthood. Histopathologic features are similar to those in epidermolytic hyperkeratosis, but are confined to the superficial epidermis. Vacuolated keratinocytes in the upper spinous layer are an important diagnostic clue.[10,11]

Porokeratosis

Porokeratosis is a group of keratinization disorders characterized by a common manifestation of marginate keratotic collarette that histopathologically corresponds to vertical columns of parakeratosis (coronoid lamellae). Different clinical forms of porokeratosis can be broadly grouped into disseminated (actinic, of childhood and of the immunosuppressed) and localized (of Mibelli, linear, punctate, giant and palmoplantar) forms. The disseminated actinic type is the commonest form of porokeratosis. Except for the punctate form, all the types exhibit a common pathognomonic rim of a keratotic ridge surrounding a central reddish, dry and atrophic skin [Figure 3]. The peripheral ridge may be furrowed as well. This distinct feature of porokeratosis is conspicuously visualized by dermoscopy, which circumvents the need for a biopsy even in clinically atypical cases. Squamous cell carcinoma may rarely develop in long-standing lesions, with a greater frequency in the linear and giant forms.[12-14]

Infectious Disorders

Cutaneous candidiasis

Candidiasis refers to an infection caused commonly by the yeast *Candida albicans*. Cutaneous candidiasis can manifest as congenital and neonatal candidiasis, diaper dermatitis and candidal intertrigo. Clinically, the lesions appear as erythematous eroded patches with marginal and satellite pustules. The pustules rupture to form a fringed irregular scalloped edge with a collarette of scaling [Figure 4]. The anogenital region is commonly involved and lesions involving the crural folds invariably exhibit deep central clefting.[15,16]

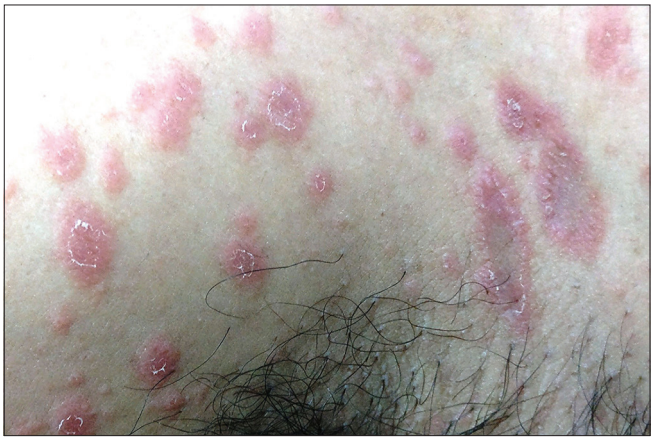

Figure 1: Pityriasis rosea

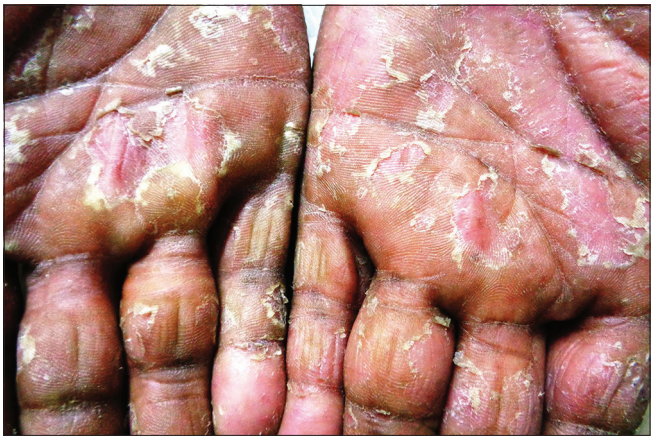

Figure 2: Keratolysis exfoliativa

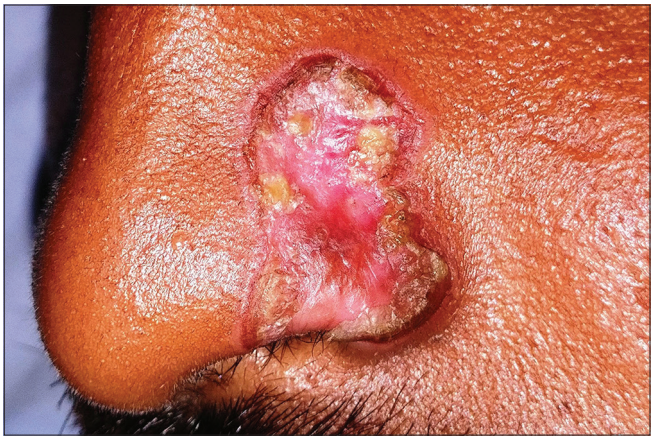

Figure 3: Porokeratosis

Bullous impetigo

Impetigo refers to a superficial bacterial infection, mainly occurring in two clinical forms: non-bullous and bullous. While the former is caused by both Staphylococci and Streptococci, the latter is caused by the epidermolytic toxin producing strains (group II, phage types 71, 55, 3A and 3C) of *Staphylococcus aureus* that hydrolyze desmoglein 1 leading to flaccid intraepidermal blistering. Bullous impetigo commonly affects children aged between 2 and 5 years. Although any site including the palms and soles may be affected, the flexures are frequently involved where the lesions begin as tiny vesicles enlarging to form flaccid bullae measuring up to 2 cm. Initially, the bullae contain clear fluid that later becomes purulent. Following the rupture of the bullae, raw erosions and crusting is seen with the reminder of the blister forming a pathognomonic collarette at the margin of the erosion [Figure 5].[17-19]

Secondary syphilis

Syphilis is a sexually transmitted disease caused by the spirochete *Treponema pallidum* occurring in primary, secondary and tertiary stages, with a long and chronic course in the untreated. Papular lesions are the most prominent cutaneous manifestations of the secondary stage of syphilis that follow an evanescent coppery-red macular rash. The papular lesions involve the trunk and extremities, including the palms and soles. They are characterized by erythematous or brownish-red 0.5–2 cm discrete papules often demonstrating a pathognomonic collarette of scale on the surface [Figure 6a] which is more conspicuous on the palmoplantar lesions. This collarette of scaling was first described by a Swiss-born French dermatologist Laurent-Théodore Biett and is named after him as "Biett's collarette" or "Biett's sign." Dermoscopically, the collarette appears as a continuous white ring of scales encircling a central erythematous papule with a surrounding rim of erythema [Figure 6b].[20-22]

Bacillary angiomatosis

Bacillary angiomatosis is primarily a vasoproliferative disease, seen in human immunodeficiency virus infected patients (with a CD4+ cell count of <50 mm^{-3}) caused by *Bartonella henselae* and *Bartonella quintana.* Cutaneous bacillary angiomatosis manifest in various forms. Disseminated angiomatous papules resembling pyogenic granuloma are the commonest. They are tender, often pedunculated and surrounded by a collarette of scales.[23-26] Similar lesions may involve the viscera as well.[27] Skin biopsy reveals aggregates of plump endothelial cells with luminal differentiation in a pale stroma. The pathognomonic feature is the presence of neutrophilic infiltrate throughout the lesion associated with smudgy amphophilic areas representing the organisms which stain with Giemsa, Warthin-Starry and Grocott's methenamine silver stains.[26,28]

Staphylococcal blepharitis

Staphylococcal blepharitis is commonly caused by *Staphylococcus aureus* and *Staphylococcus epidermidis*. There is usually a unilateral localized erythema and edema of the anterior lid margin associated with the pathognomonic brittle fibrinous scales forming collarette around the base of the cilia. As the eyelashes grow, the adherent scales are lifted up. Other features that may be seen include dilated blood vessels on the lid margin arranged in rosettes, madarosis, localized ulceration and/or poliosis. The infection may extend to the posterior lid margin, and then onto the conjunctiva and periphery of the cornea, manifesting as follicular conjunctivitis and marginal keratitis.[29-31]

Pitted keratolysis

Pitted keratolysis is a superficial bacterial infection caused by a variety of organisms (*Kytococcus sedentarius, Dermatophilus congolensis, Actinomyces* and *Corynebacterium* spp.) that commonly affects the weight-bearing areas of soles in hot and humid climates. The lesions at these sites are seen as malodorous discrete superficial pits that may coalesce into larger noninflammatory erosions. Lesions can uncommonly occur on the palms and nonweight-bearing areas of the feet where they typically appear as collarette of scales rather than pits resembling keratolysis exfoliativa (see above).[32-35]

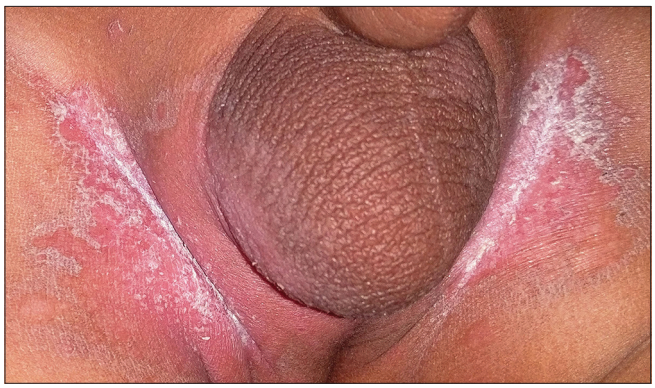

Figure 4: Candidal intertrigo. Note the characteristic scalloped border with collarette of scales

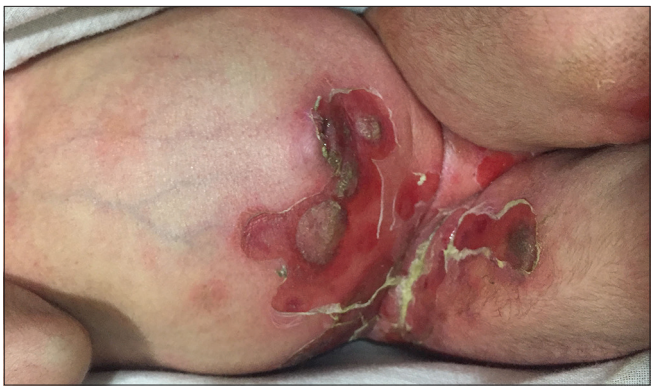

Figure 5: Bullous impetigo

Plantar warts

Plantar warts are commonly caused by the human papilloma virus 1. They may be difficult to distinguish at times from corns and callosities. However, plantar warts obliterate the dermatoglyphics and demonstrate a soft core (as opposed to the hard core in corns) with pin-point bleeding spots or intracapillary thrombosis on superficial pairing of the lesion (not seen with corns and calluses). Plantar warts are sharply demarcated and exhibit a collarette of raised normal surrounding skin. Superficial pairing of the wart surface through this collarette shows abrupt ending of the epithelial ridges of the normal plantar skin at this point [Figure 7]. A thick raised keratinous collarette may also be seen surrounding an old wart elsewhere on the body too.[4,36-38]

Vesico-pustular Disorders

Transient neonatal pustular melanosis

Transient neonatal pustular melanosis is one of the transient neonatal pustuloses, characterized by superficial noninflammatory vesicles or pustules that rupture spontaneously leaving behind a characteristic collarette of white scales. These scales are later replaced by postinflammatory hyperpigmentation that resolves gradually over several weeks. The head and neck region and/or the lower extremities are commonly involved, and the condition is almost exclusive to full-term neonates of African descent. Some authors believe it to be a variant of erythema toxicum neonatorum; however, most of the lesions are seen at birth, and the sterile pustules are composed of neutrophils as opposed to the eosinophils in erythema toxicum neonatorum.[39,40]

Subcorneal pustular dermatosis (Sneddon-Wilkinson disease)

Subcorneal pustular dermatosis is an uncommon, chronic, remitting and relapsing pustular dermatosis commonly affecting the middle-aged females. Clinically, the lesions appear as flaccid sterile pustules on either normal or erythematous skin. Lesions usually favor the flexures and appear in crops with a tendency to coalesce forming various patterns – annular, polycyclic, serpiginous or circinate. A typical feature is the formation of a fluid level within the blister as the pus accumulates in the dependent portion and the clear fluid on top. The lesions rupture in a few days and dry up to form crusts and collarette scaling. Histopathologically, there is accumulation of neutrophils just beneath the stratum corneum.[41,42]

Pustular psoriasis

Pustular psoriasis, either generalized or localized, is characterized by development of sterile pustules on a patchy or confluent erythema. The pustules begin as discrete tiny lesions that may coalesce to form lakes of pus. In a few days, they rupture and dry up, leaving behind erosions surrounded by collarette of scaling [Figure 8]. In the generalized forms (von Zumbusch and impetigo herpetiformis), the skin lesions are preceded by fever and systemic complaints. The pustules favor the trunk and extremities, and typically appear in waves accompanied by fever. The localized form of pustular psoriasis commonly involves the hands and feet (palmoplantar pustulosis and acrodermatitis continua of Hallopeau) and although not associated with any systemic effects, the chronic and persistent nature of the disease significantly affects the patients' quality of life [Figure 9].[43,44]

Acute generalized exanthematous pustulosis

Acute generalized exanthematous pustulosis is an acute febrile exanthematous pustulosis, commonly occurring as an adverse cutaneous drug reaction. Aminopenicillins, sulfonamides, quinolones, hydroxychloroquine and calcium channel blockers are the frequently implicated drugs. Lesions generally appear in 24–48 h following the intake of the offending drug, frequently in the flexures as tiny nonfollicular pustules on an erythematous base that gradually extend onto the trunk and extremities. The lesions involute spontaneously in a couple of weeks leaving behind the typical collarette of scaling. The disorder is usually a benign one, though secondary infection of the lesions can complicate the otherwise uneventful course.[45-47]

Gyrate Erythemas

Erythema annulare centrifugum

Erythema annulare centrifugum is a reactive migratory erythema that has been speculated to be associated with certain infections (Epstein-Barr virus infection, herpes zoster, molluscum contagiosum, candidiasis, dermatophyte infections, etc.), drugs (piroxicam, hydroxychloroquine sulfate, hydrochlorothiazide, amitriptyline, etc.) and certain other disorders such as sarcoidosis, hepatic and thyroid disorders, hematological and solid organ malignancies. However, none of these associations have been proven beyond doubt. It is divided into a superficial type (of Colcott Fox) and a deep type (of Darier) with distinct clinical and histopathological features. The disease commonly affects adults and the lesions favor the buttocks, thighs and trunk. The lesions begin as erythematous papules that slowly migrate and enlarge centrifugally reaching up to 10 cm in diameter with central clearing. The peripheral margin is raised enough to be just palpable or thick cord-like. The superficial variant exhibits the typical trailing collarette scales (as in pityriasis rosea and histologically demonstrates spongiosis, parakeratosis and superficial perivascular mononuclear infiltrate. The deep variant lacks the scaling, and histopathologically is typified by the dense superficial and deep perivascular infiltrate (giving a "coat-sleeve" appearance) without any epidermal changes.[48-51]

Erythema gyratum repens

Erythema gyratum repens is a paraneoplastic inflammatory gyrate erythema occurring most commonly in association with carcinoma of lung. It is seen more commonly in males, beyond the fourth decade. It has also been reported with other hematological and solid organ malignancies. Clinically, the trunk, neck and extremities are commonly

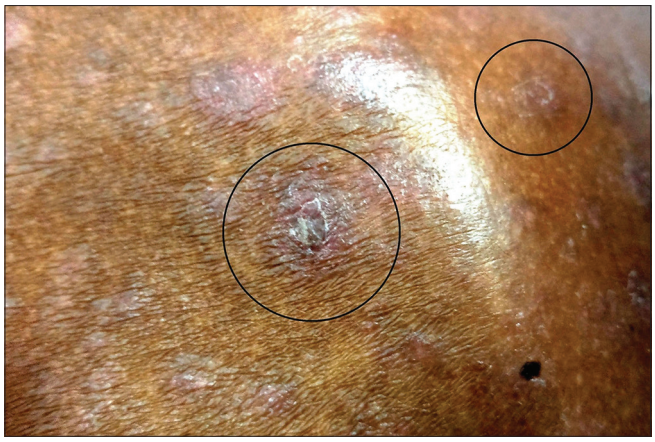

Figure 6a: Collarette scales on top of erythematous papules in secondary syphilis (black circles)

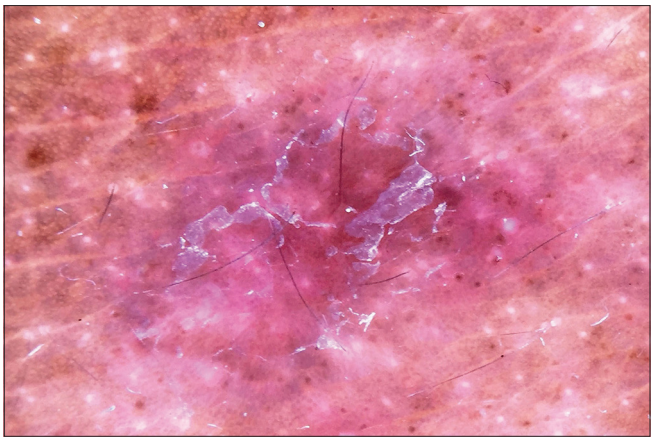

Figure 6b: The "collarette" morphology of the scale is conspicuously visualized on dermoscopy (×10)

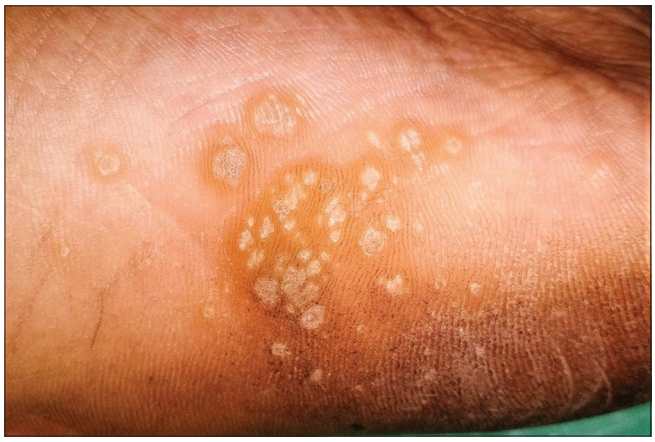

Figure 7: Plantar warts with colarette of raised surrounding normal skin. Note the abrupt termination of dermatoglyphics at the inner margin of the collarette.

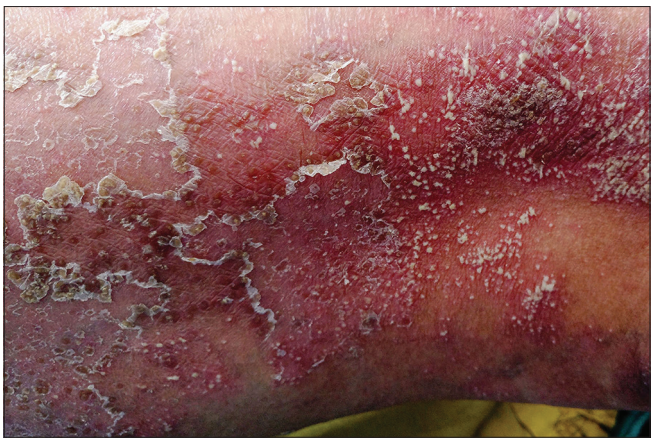

Figure 8: Pustular psoriasis

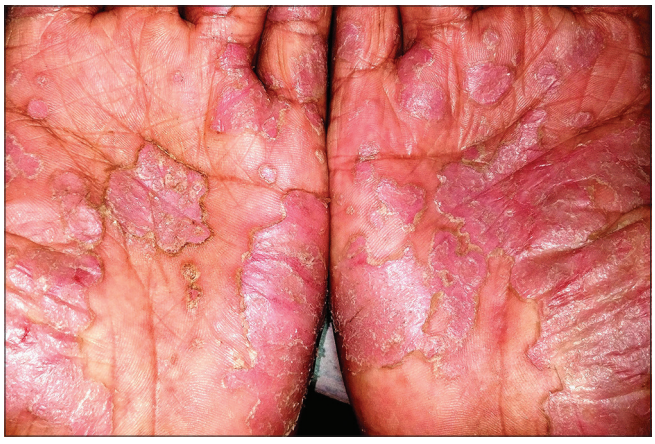

Figure 9: Palmar pustulosis

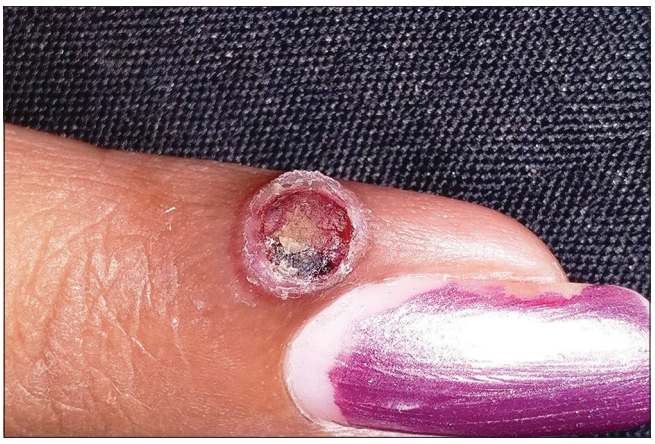

Figure 10: Pyogenic granuloma

involved and the lesions are characterized by intensely pruritic migratory erythematous concentric wave-like bands. The characteristic feature is the rapid migratory rate of the leading edge (1 cm/day) and development of new gyrate lesions within preexisting ones, giving an appearance of concentric bands resembling wood grain. The outermost edge shows collarette scales which may be trailing the edge. Although histopathological features of erythema gyratum repens are imprecise, the distinctive clinical characteristics, however, warrant evaluation of the patient for an underlying neoplasia. Resection of the tumor results in resolution of the lesions.[52-54]

Figure 11: Acral fibrokeratoma

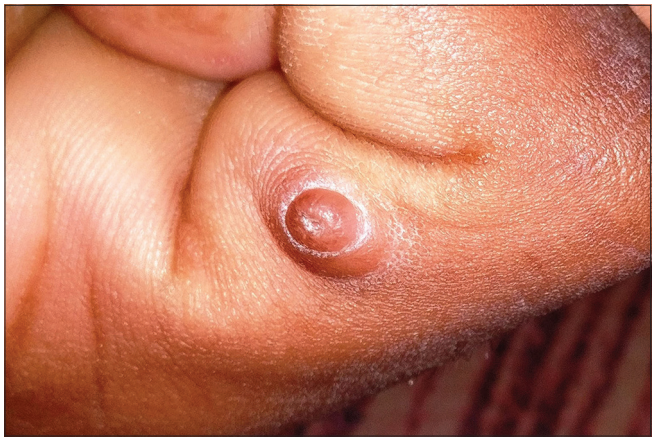

Figure 12: Rudimentary supernumerary digit

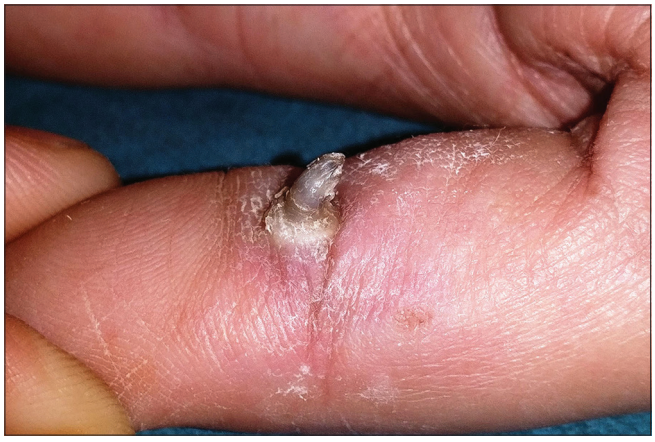

Figure 13: Cutaneous horn

Cutaneous Tumors

Clear cell acanthoma (pale cell acnathoma, degos tumor)

Clear cell acanthoma was first described by Degos *et al*. in 1962.[55] The tumor commonly affects the legs and feet of middle-aged men and women. It usually appears as a solitary circumscribed dome-shaped papule or nodule with pigmented scaly surface and a characteristic collarette of white scales surrounding the lesion. Multiple, polypoid and giant forms have also been described. Dermoscopy of the tumor shows central blood vessels oriented perpendicular to the skin surface and a peripheral squamous collarette. Similar features may be seen in psoriasis and in Bowen's disease and hence, a clinical correlation and histopathological examination is necessary for definitive diagnosis. The distinctive histopathological features of Degos tumor include an acanthotic epidermis with abundant clear cells, spongiosis, hypo- or agranulosis and elongation of rete ridges. The clear cells are polyhedral and larger than the keratinocytes and show positivity and sensitivity to periodic-acid Schiff staining and diastase reaction, respectively.[56,57]

Table 2: Dermatoses exhibiting a collarette of scaling inconsistently or nonspecifically

Category	Disorders
Keratinization disorders	Kyrle's disease [Figure 14] Flegel's disease
Infectious disorders	Erythema induratum
Vesico-pustular disorders	Pemphigus foliaceus
Gyrate erythemas	Necrolytic migratory erythema
Cutaneous tumors	Acral fibromyxoma Stucco keratosis Juvenile xanthogranuloma

Pyogenic granuloma

Pyogenic granuloma is a common acquired benign vascular tumor appearing mostly as rapidly growing, solitary, friable sessile or pedunculated vascular papules or nodules. A surrounding collarette of scale is seen at the base of the lesion in majority of the cases [Figure 10]. Pyogenic granuloma is seen commonly in children and young adults with frequent involvement of the extremities and face. Trauma is the most frequent implicated factor in its etiopathogenesis. Lobular proliferation of capillary-like vessels in a loose stroma is the pathognomonic histopathological feature.[28,58,59] Dermoscopy shows central vascular area and a peripheral collarette. The central vascular area may show different patterns – red homogenous area, ulceration with hemorrhagic crusts or white "rail lines" on a reddish homogenous background intersecting the lesion.[60]

Acral fibrokeratoma

Acral fibrokeratoma (acquired digital fibrokeratoma) is a solitary benign tumor of adults frequently affecting the fingers and toes. The distinctive feature of the tumor is the moat-like collarette of thick raised skin surrounding a central dome-shaped papule or nodule with a warty surface [Figure 11]. Dermoscopic features include a central homogenous pale yellow area with marginal white scaly collarette. Acral fibrokeratoma clinically resembles various other cutaneous tumors such as eccrine poroma, pyogenic granuloma [Figure 10], a rudimentary supernumerary digit [Figure 12], viral wart [Figure 7], dermatofibroma and a cutaneous horn [Figure 13]. Therefore, histopathological

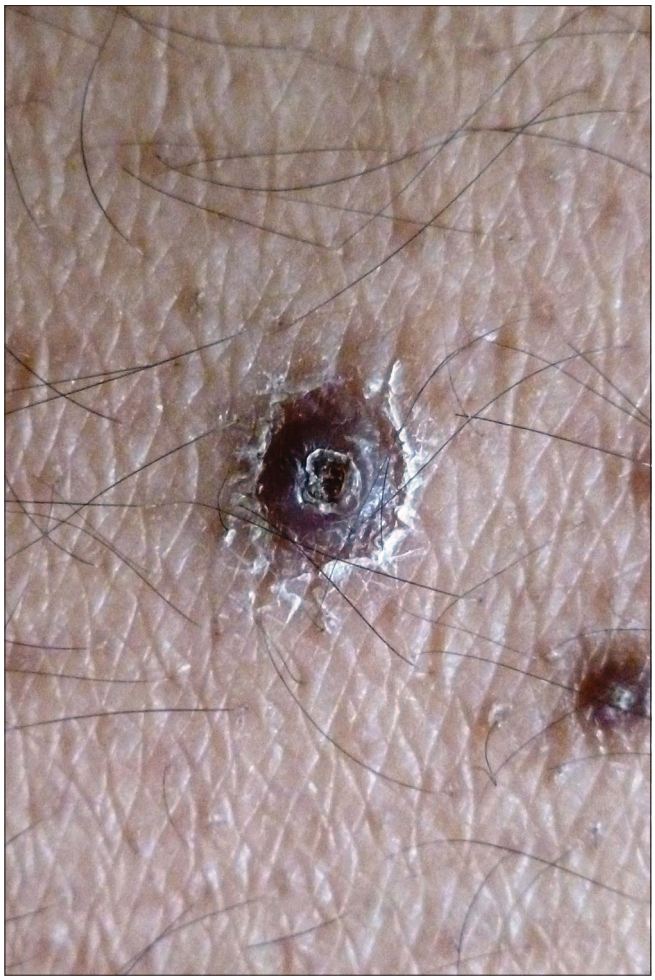

Figure 14: Kyrle's disease

analysis is required for definitive diagnosis which shows a central area of increased vertically oriented collagen fibers with interspersed blood vessels underlying a hyperkeratotic and acanthotic epidermis with wide, elongated, branching rete ridges. Acral fibrokeratoma and rudimentary supernumerary digit are indistinguishable clinically. The latter however is congenital, seen at the base of little finger and histologically shows increased nerve bundles at the base of the lesion.[61-63]

Cutaneous horn

Cutaneous horn is a firm keratotic horny excrescence commonly involving the face and dorsum of the hands of elderly. The lesion may have a keratotic rim at the base [Figure 13]. Cutaneous horn usually overlies a benign (seborrheic keratosis, verruca vulgaris), premalignant (actinic keratosis) or a malignant lesion (squamous cell carcinoma). Histopathologically, the lesions are composed of amorphous or lamellated keratin with features of the underlying disease at the base.[64,65]

Subungual exostosis

Subungual exostosis is typically seen arising from underneath the anteromedial region of the tip of great toes in young individuals. Trauma is the most common implicated inciting factor and hence it was previously thought to be a reactive benign osteochondral outgrowth. However, it is now classified as a true neoplasm demonstrating a pathognomonic translocation t(X; 6)(q22;q13-14). It arises most frequently from the hyponychium or the lateral sulcus as a porcelain-white firm papule with surface telangiectasia and a surrounding collarette. As it enlarges, the surface becomes hyperkeratotic. The tumor is moderately painful and associated with onycholysis and predisposition to secondary infection. Radiologically, it appears as a trabeculated bony outgrowth with an enlarged distal portion covered with radiolucent fibrocartilage. Histopathologically, mature trabecular bone showing endochondral ossification with a cap of proliferating mature cartilage is seen. Simple excision is curative.[66-69]

Eccrine poroma

Eccrine poroma is a benign tumor arising from the acrosyringium. Eccrine poromas are frequently seen on the palms and soles and appear as solitary sessile pinkish-red papules or nodules often surrounded by a moat-like collarette of thick skin. Multiple lesions associated with chemotherapy and bone marrow transplantation have also been described, so have been the lesions occurring at atypical sites. Histologically, they are composed of proliferating cuboidal cells with prominent nuclei and scanty cytoplasm showing ductal differentiation (showing immunohistochemical positivity for carcinoembryonic antigen). The tumor is well-circumscribed with a clear demarcation between the lesion and the adjacent normal epidermis. Eccrine poromas are frequently misdiagnosed as acral fibrokeratoma or pyogenic granulomas because of the similar clinical appearances.[70-75]

Other Disorders

A collarette of scaly skin may also be seen in several other disorders although not as a characteristic or consistent feature. In certain conditions such a feature may be a secondary phenomenon (e.g., collarette of scales seen on removal of lesions of stucco keratosis or crusting in impetigo). Table 2 enlists such disorders that inconsistently or nonspecifically exhibit collarette scaling as per the same categorization in Table 1.[14,42,76-81]

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Conflicts of interest

There are no conflicts of interest.

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