Dermoscopy of Juvenile Circumscribed Pityriasis Rubra Pilaris

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A 11-year-old girl presented with complaints of mild itchy lesions over bilateral elbows and knees of 1 year duration. There was no history of lesions anywhere else on the body. No other complaints like difficulty in vision or palmoplantar involvement were present. There was no history of atopy. Or any significant family history. Cutaneous examination revealed multiple discrete, erythematous, grouped follicular hyperkeratotic papules with a spiny surfaces and nutmeg grater feel over bilateral elbows, extending slightly over the arms and over bilateral knee joints [Figure 1a and b]. Dermoscopy done with DE-300 polarizing digital dermaoscope (Firefly) having 20× magnification showed whitish follicular keratotic plugs with peripheral yellowish rings, perifollicular erythema, and hair shaft in the center [Figure 2]. Nails, and mucoase were normal. Koebnerization was absent. Skin biopsy from one of the lesions showed acanthosis with broad and short rete ridges, thick suprapapillary plate, focal hypergranulosis, alternating orthokeratosis and parakeratosis, broad follicular infundibulum filled with orthokeratotic plug with perifollicular moderate lymphoplasmacytic inflammatory infiltrates, and perivascular lymphoplasmacytic inflammatory infiltrates in the dermis [Figure 3]. Based on the history, examination, dermoscopy, and histopathological findings, this case was diagnosed as juvenile circumscribed pityriasis rubra pilaris (PRP).

PRP is a rare papulosquamous disorder of childhood, characterized by circumscribed follicular keratoses, branny scale, orange-red erythema, and palmoplantar keratoderma.

PRP was initially classified on the basis of age at onset, behavior, clinical appearance, and prognosis by Griffith in 1980, which continues to be the mainstay in practice for delineating the disease.[1] The exact etiology of PRP is unknown.

Juvenile circumscribed (type-iv) PRP is usually seen in the later part of the first decade of life and is characterized by the

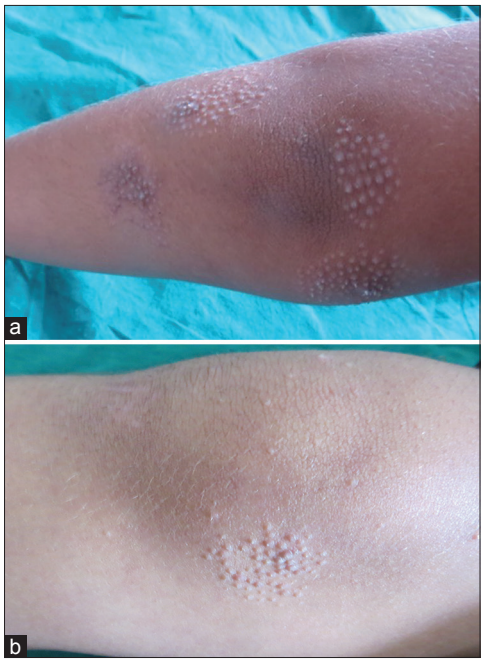

Figure 1: Multiple discrete, erythematous, grouped follicular hyperkeratotic papules over (a) Left elbow, (b) right knee

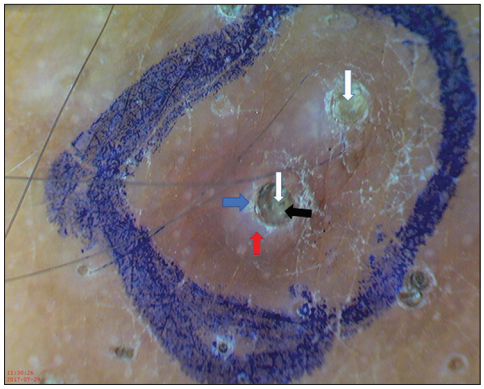

Figure 2: Dermoscopy (DE 300 dermoscope (firefly) 20× magnification with polarized light) showingwhitish follicular keratotic plugs (black arrow) with a peripheral yellowish ring (blue arrow), perifollicular erythema (red arrow), and hair shaft in the center (white arrow)

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Table 1: Differential diagnosis of juvenile circumscribed PRP

Dermatosis	Age of onset	Site	Morphology	Histopathology	Dermoscopy
Juvenile circumscribed PRP	1st decade	Extensors, localized	Islands of sparing, papules with nutmeg grater feel, erythroderma, keratoderma	Follicular plugging, checkerboard pattern of orthokeratosis and hypogranulosis	White keratotic plug, yellow peripheral keratotic ring, perifollicular erythema, and linear vessels mixed with dotted vessel[1]
Follicular psoriasis	2nd decade	Extensors	Scaly papules or plaques with nail changes and scalp involvement	Hyperkeratosis, parakeratosis, hypogranulosis, regular acanthosis, Munromicro abscesses	White-brown homogenous area, terminal hair at the center, perifollicular scaling, multiple red dots/dotted vessels, red globules, twisted red loops, and glomerular vessels/bushy capillaries[2]
Keratosis pilaris	Adolescents, childhood	Extensor surfaces of upper arms, thighs buttocks	Keratotic follicular papules, antenna sign, perifollicular erythema	Distention of the follicular orifice by a keratinous plug that contain one or more twisted hairs	Irregular twisted or coiled vellus hair embedded in the horny layer, perifollicular erythema, scaling, and pigmentation[3]
Phrynoderma	Childhood	Elbow, knee, thigh, and buttocks	Discrete, brown or skin-colored, acuminate, keratotic papules with central keratin plug	Follicular hyperkeratosis with keratin plugging	Follicular-based papules with translucent spines along with multiple perilesional glittering floret like structures. Each floret has 6 heads giving rise to 6-clod dots-like appearance[4]
Lichen spinulosus	Childhood	Neck, buttocks, abdomen, knees, andextensor	Large patches of follicular papules topped by keratotic spines	Hyperkeratosis with follicular plugging	Hypopigmented follicular papules with tiny whitish yellow plugs and perifollicular collarette of scales, hypopigmentation and erythema, and presence of hair present without twisting[4]
Dariers disease	Adults	Seborrheic areas	Greasy, malodorous hyperkeratotic papules Pits on the palm, nail (white, red bands, V nick)	Suprabasal acantholysis, corps, ronds, and grains	Centrally located polygonal, star-like or roundish-oval shaped yellowish/ brownish area, surrounded by a more or less thin whitish halo, overlying a pinkish homogeneous structure less area, with or without whitish scales and dotted and/or linear vessels presenting a whitish halo[5]
Lichen nitidus	Children young adults	Trunk, genitalia	Multiple, small, discrete, shiny, flesh-colored, dome-shaped papules arranged in groups	Clawlike projection of rete pegs with lymphohistiocytic infiltrate expanding the dermal papilla	Round, elevated, shiny, and smooth appearance surrounded by radial rete ridges, and reddish vascular network[5]
Lichen scrofulosurum	Adults orchildren	Trunk, proximal	Asymptomatic, acuminate, grouped scaly lichenoid follicular papules	Tuberculoid granuloma	-

formation of well-circumscribed, erythemato-squamous plaques studded with follicular and/or non-follicular papules with prominent keratotic plugging. Lesions are usually confined to the elbows and knees and is marked by remissions and exacerbations. In atypical cases, PRP has to be differentiated from other conditions which can be a diagnostic challenge and a skin biopsy has to be performed for confirmation.

Dermoscopy is a non-invasive diagnostic tool that allows visualization of many morphologic features not visible to the naked eye especially vascular and pigmented structures, so it represents a link between macroscopic dermatology and pathology.

Dermoscopic features of PRP have been described only in two case-reports. A study conducted by Azim *et al*.[1] showed that most common dermoscopic findings of PRP are yellowish background, whitish keratotic plug, linear vessels either solely or mixed with dotted vessels, arranged mainly peripherally.

Histopathology of PRP is characteristic and shows alternating columns of orthokeratosis and parakeratosis both in vertical and horizontal directions, focal or confluent hypergranulosis, thick suprapapillary plates, broad rete ridges, narrow dermal papillae, and sparse perivascular lymphocytic infiltrate. Plugging of follicular infundibulum by cornified cells is characteristic.

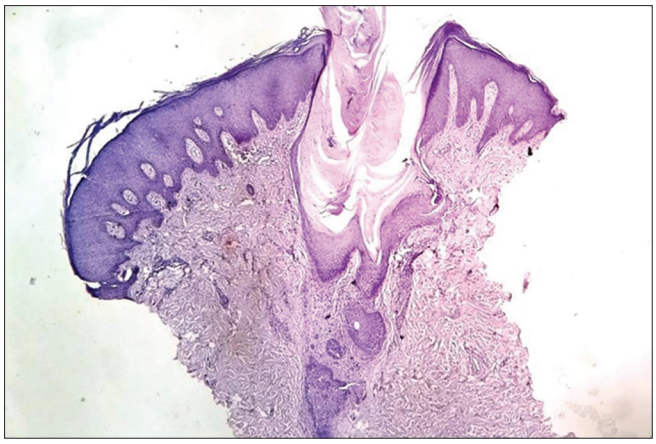

Figure 3: Histopathology showingacanthosis with broad and short rete ridges, thick suprapapillary plate, focal hypergranulosis, alternating orthokeratosis and parakeratosis, broad follicular infundibulum filled with orthokeratotic plug with perifollicularand perivascular lymphoplasmacytic inflammatory infiltrates in the dermis (H and E stain ×40)

Few common follicular and non-follicular dermatoses, which must be considered in the differential diagnosis of PRP are tabulated with their clinical, dermoscopic, and histopathological differences in Table 1.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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