Case 19

A 12 year-old Thai girl from Nakornsawan

Chief complaint:

Red face and progressive roughness of skin at both arms and thighs since infancy

Present illness:

From 8 months of age, she gradually developed rash involving both cheeks, arms, upper thighs and legs. The intensity of erythema and roughness pronounced with increasing ages without any symptoms.

Past history:

She was otherwise healthy and had normal growth and development.

Family history:

Her mother had similar but less severe eruption on both arms.

Skin examination:

Numerous follicular papules with background erythema on both cheeks, widespread hyperkeratotic erythematous spiny follicular papules on extensor aspects of arms, thighs and legs.

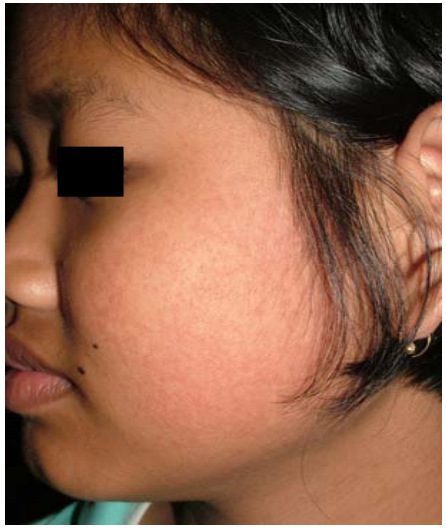

Fig. 19.1

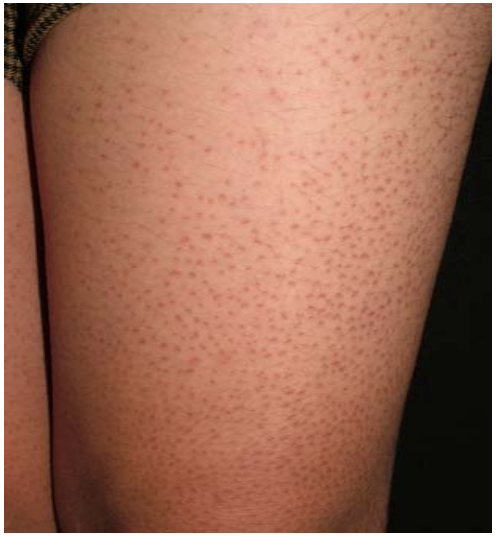

Fig. 19.2

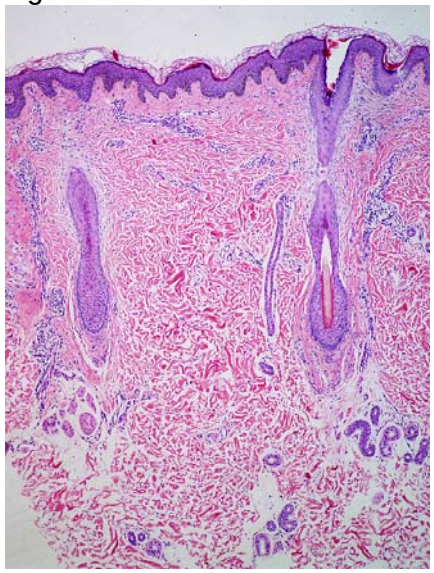

Fig. 19.1

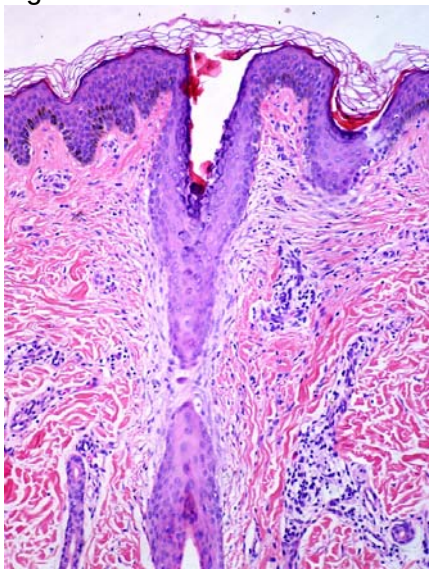

Fig. 19.2

Histopathology: (S08-17588) (Fig. 19.3, 19.4)

- perivascular and perifollicular infiltrate of lymphocytes
- follicular fibrosis with atrophy of follicular epithelium

Diagnosis: Keratosis pilaris rubra

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Discussion:

Keratosis pilaris (KP) is a common benign skin disorder develops most often in childhood and improves with age.[1] The etiology is still unknown. It is characterized by an eruption of symmetric, asymptomatic, grouped keratotic follicular papules on extensor and lateral aspects of proximal extremities and cheeks.[2] Erythema is sometimes present in KP, but it usually mild and limited to the perifollicular skin. There are 3 distinct variants of KP have been reported:

1. Keratosis pilaris atrophicans (KPA)
2. Erythromelanosis follicularis faciei et colli (EFFC)
3. Keratosis pilaris rubra (KPR)

KPA is often localized on the face and progresses to scarring involving eyebrows.[3] EFFC and KPR, some authors suggest that they are likely part of the same disease spectrum. Features that differentiate EFFC from KPR are lack of reported involvement on the torso and the presence of hyperpigmentation.[4]

Keratosis rubra pilaris (KPR) is characterized by substantial erythema, widespread involvement, and persistence after puberty. The mean age of onset is 5 years old and the earliest presents at birth.[5] The pathogenesis of KPR is not well understood. The erythema present in KPR fluctuates, and can present even in areas without significant keratotic papules in some patients. The question of whether flushing via autonomic dysregulation may play a role in the clinical presentation can be raised. Various treatments has been attempted with unsatisfactory results. These include topical corticosteroid, emollients, keratolytic agents, tretinoin cream, oral isotretinoin. However,there are few cases report of successful treatment of erythema component by pulsed dye laser[6] and potassium titanyl phosphate laser.[7]

References:

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3. Callaway SR, Lesher JL. Keratosis pilaris atrophicans: case series and review. Pedriatr Dermatol. 2004; 21: 14-7.
4. Stephen F, Ayoub N, et al. Erythromelanosis follicularis faciei et colli. Ann Dermatol Venereol. 2002; 129: 63-5.
5. Marqueling AL, Gilliam AE, et al. Keratosis pilaris rubra: a common but underrecognized condition. Arch Dermatol. 2006;142(12): 1611-6.
6. Kurita M, Momosawa A, et al. Long-pulsed dye laser for the treatment of erythromelanosis follicularis faciei: report of 2 clinical cases. Dermatol Surg. 2006; 32: 1414-7.
7. Dawn G, Urcelay M, et al. Keratosis rubra pilaris responding to potassium titanyl phosphate laser. Br J Dermatol. 2002; 147: 822-4.