

Keratosis Pilaris Rubra

A Common but Underrecognized Condition

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Background: Keratosis pilaris is a common skin disorder of childhood that often improves with age. Less common variants of keratosis pilaris include keratosis pilaris atrophicans and atrophoderma vermiculata.

Observations: In this case series from dermatology practices in the United States, Canada, Israel, and Australia, the clinical characteristics of 27 patients with keratosis pilaris rubra are described. Marked erythema with follicular prominence was noted in all patients, most commonly affecting the lateral aspects of the cheeks and the proximal arms and legs, with both more marked erythema and widespread extent of disease than in keratosis pilaris. The mean age at

onset was 5 years (range, birth to 12 years). Sixty-three percent of patients were male. No patients had atrophy or scarring from their lesions. Various treatments were used, with minimal or no improvement in most cases.

Conclusions: Keratosis pilaris rubra is a variant of keratosis pilaris, with more prominent erythema and with more widespread areas of skin involvement in some cases, but without the atrophy or hyperpigmentation noted in certain keratosis pilaris variants. It seems to be a relatively common but uncommonly reported condition.

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KERATOSIS PILARIS (KP) IS A common benign disorder of unknown etiology.¹ It typically presents as an eruption of symmetric, asymptomatic, grouped keratotic follicular papules on the extensor and lateral aspects of the proximal extremities and the cheeks.¹⁻⁵ Less often, KP may involve the neck, torso, and buttocks, and, rarely, the eruption may be generalized.⁵ Erythema, when present, is usually mild and localized to the perifollicular skin.⁴ Often, the disease is familial, and it has been suggested that inheritance is autosomal dominant^{1,2} without known predisposition based on race or sex. Keratosis pilaris develops most often in early childhood, with remission by adulthood in many patients.

At least 2 distinct variants of KP have been reported: keratosis pilaris atrophicans (KPA; sometimes referred to as ulerythema ophryogenes) and erythromelanos follicularis faciei et colli (EFFC). We describe 27 patients with another variant of KP, which we have termed *keratosis pilaris rubra* (KPR), that, in our experience, is much more common than either KPA or EFFC. It is characterized by substantial erythema, wide-

spread involvement, and persistence after the onset of puberty. Despite being relatively common, to our knowledge, a detailed case series has not been described in the medical literature to date.

REPORT OF CASES

CASE 1

A 13-year-old boy (patient 1 in the **Table**) presented to the University of California, San Francisco, pediatric dermatology practice with a 1-year history of a mildly pruritic but otherwise asymptomatic rash involving the face, torso, and extremities. The medical history was unremarkable. Before presentation, he had treated the eruption with hydrocortisone cream, which provided some relief of pruritus, and adapalene gel, which did not result in any improvement. Physical examination revealed bilateral erythematous patches with follicular prominence on the cheeks (**Figure 1**) and follicular hyperkeratotic papules with variable erythema on the chest, back, upper arms, and thighs. A punch biopsy specimen from the posterior aspect of the left shoulder showed follicular infundibular plugging with slight

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Table. Clinical Characteristics of 27 Patients

Patient No./ Sex/Race	Age at Disease Onset/ Presentation, y	Symptoms	Medical History	Skin Findings	Treatment
1/M/White	12/13	Mild pruritus	None	Erythematous plaques with follicular prominence on cheeks bilaterally; follicular hyperkeratosis with erythema on chest, back, upper arms, and thighs; no other skin findings	1% Hydrocortisone cream, adapalene gel, 0.05% tazarotene cream
2/M/White	1/4	Mild pruritus	None	Erythema and fine follicular papules on forehead directly above eyebrows, on cheeks and ears bilaterally, and on chin; questionable lateral eyebrow thinning but no follicular atrophy or scarring; KP on upper arms and upper lateral thighs bilaterally; no other skin findings	OTC moisturizers (Eucerin, Cetaphil), 0.3% tacrolimus ointment, ammonium lactate moisturizer
3/M/Asian	8/13	None	Tonsillectomy	Diffuse white and red hyperkeratotic follicular papules on cheeks, chest, abdomen, back, and extensor aspects of upper arms and legs bilaterally, with background of blanchable erythema on cheeks bilaterally, no other skin findings	None
4/M/White	.../15	None	Asthma, benign tremor	Erythema and follicular papules on cheeks and ears; widespread keratotic follicular papules with mild erythema on back and upper arms; other skin findings included midfacial acne	Gentle skin care, trial of 12% ammonium lactate moisturizer
5/M/Hispanic	3-4/14	None	Asthma	Diffuse plaques of dry erythematous papules on forehead, cheeks, ears, arms, and back; no other skin findings	1% Hydrocortisone, 12% ammonium lactate moisturizer
6/F/Chinese	.../12	Occasional pruritus	Warts, allergic rhinitis	Lesions on face and extensor aspects of arms; other skin findings included verrucous vulgaris	0.1% Adapalene gel, 0.1% tazarotene cream
7/F/East Indian	11-12/15	Occasional mild pruritus	Asthma, GERD, atopic dermatitis, acanthosis	Extensive KP with background erythema on face; extensive follicular keratosis with less erythema on extensor aspects of upper arms bilaterally; other skin findings included atopic dermatitis and acanthosis nigricans	Topical corticosteroids, 0.1% adapalene gel
8/F/White	7/17	None	Type 1 diabetes mellitus	Erythema and follicular papules on chin, eyebrows (without hair loss), and glabella; extensive KP with less erythema on extensor aspects of arms, chest, lower abdomen, buttocks, and thighs; no other skin findings	0.1% Adapalene gel
9/M/White	5/16	Recent pruritus	Croup	Erythema and follicular papules on cheeks, chin, and forehead; extensive erythematous follicular papules and pustules on torso, buttocks, and extensor aspects of arms and legs; other skin findings included diffuse eczematous eruption on torso and arms	12% Ammonium lactate moisturizer
10/F/...	9-10/12	None	Juvenile dermatomyositis (in remission), mild acne	Follicular keratotic papules with background macular erythema on cheeks and extensor aspects of arms; other skin findings included mild comedonal acne	0.1% Adapalene gel, 12% ammonium lactate moisturizer
11/M/White	Early childhood/9	None	ADHD, asthma	Prominent erythema and follicular hyperkeratosis of face and outer aspects of arms; lateral eyebrow shedding; no other skin findings	0.05% Tretinoin cream, oral acitretin, urea with lactic acid moisturizer, calcipotriene
12/M/White	7/7	None	None	No description of KP symptoms; no other skin findings	Oral acitretin
13/M/White	Infancy/16	None	None	No description of KP symptoms; no other skin findings	Urea and lactic acid moisturizer, tacrolimus ointment
14/M/White	8/17	None	None	Prominent follicular hyperkeratosis with hyperpigmentation and mild facial erythema; no other skin findings	Urea and lactic acid moisturizer, oral isotretinoin, 20 mg/d, with some improvement
15/F/White	1/10	None	None	Follicular hyperkeratotic papules surrounded by erythema; KP on extensor aspects of arms without erythema; no other skin findings	Tretinoin cream, urea and lactic acid moisturizer
16/F/White	2/3	None	None	Diffuse erythema with hyperkeratotic follicular papules on cheeks and anterior aspects of thighs; no other skin findings	0.025% Tretinoin cream; OTC emollient cream with 20% urea, 12% ammonium lactate, and lactic acid (Axera cream), with some improvement
17/M/White	11/16	None	None	Follicular hyperkeratotic papules surrounded by erythema on back and extensor surfaces of arms with occasional pustules; face not involved; eyebrows normal; no other skin findings	OTC emollient cream with 20% urea, 12% ammonium lactate, and lactic acid (Axera cream)

(continued)

Table. Clinical Characteristics of 27 Patients (cont)

Patient No./ Sex/Race	Age at Disease Onset/ Presentation, y	Symptoms	Medical History	Skin Findings	Treatment
18/M/White	Lifetime/. . .	None	Splenomegaly and fever	Keratotic follicular papules with strikingly ruddy erythema on lateral cheeks (in triangular patches), glabella, pinnae, eyebrows (without hair loss), extensor aspects of arms, lateral back, and posterior aspects of legs; no other skin findings	Dove soap, 12% ammonium lactate moisturizer, 20% urea and urea with hydrocortisone moisturizers; PDL, 595 nm, 9.0 J/cm ² , 7 mm
19/F/White	2½/2½	None	Allergic rhinitis, midfacial acne	Keratotic follicular papules and moderate erythema on lateral cheeks (in triangular patches); patchy appearance of keratotic papules and erythema on extensor aspects of the arms; other skin findings included midfacial acne vulgaris	OTC moisturizer (Eucerin), 12% ammonium lactate moisturizer, 20% urea moisturizer, urea and glycolic acid cream, 0.1% adapalene, 6% salicylic acid cream
20/M/White	Birth/15	None	Transposition, pneumonia	Follicular keratotic papules with underlying intense erythema of cheeks, chin, eyebrows, ears (pinnae), extensor aspects of the arms, lateral chest, and back; no other skin findings	12% Ammonium lactate moisturizer; PDL, 595 nm, 36 pulses to the cheeks
21/M/White	Birth/15	Psychosocial problems, irritation on shaving	Atopic dermatitis, asthma, allergic rhinitis	Keratotic papules with erythematous background and light brown hyperpigmentation on lateral cheeks; keratotic follicular papules with background erythema on extensor aspects of arms and legs, back, and buttocks; other skin findings included atopic dermatitis (antecubital)	40% Urea moisturizer; 12% ammonium lactate moisturizer; 0.01% fluocinolone acetonide, 4% hydroquinone, and 0.05% tretinoin (Tri-Luma cream); azelaic acid cream; oral beta carotene, 5000-8000 IU/d, with slight improvement
22/M/White	11-12/14	None	Asthma	Folliculocentric papules with background erythema on cheeks; folliculocentric keratotic papules with erythema on extensor aspects of arms and legs, chest, and upper back; other skin findings included congenital melanocytic nevi on left arm and back, scattered nevi	12% Ammonium lactate cream
23/F/White	8/13	Initially pruritic	None	Erythematous keratotic follicular papules on cheeks, arms, legs, and buttocks; no other skin findings	6% Salicylic acid cream, 12% ammonium lactate moisturizer
24/F/Hispanic	1/13	None	None	Widespread hyperkeratotic spiny follicular papules on cheeks, predominantly skin-colored or erythematous, some pigmented; widespread hyperkeratotic spiny follicular papules on extensor aspects of the arms and legs and on buttocks; other skin findings included nevus spilus and other benign nevi	12% Ammonium lactate moisturizer
25/M/White	8-9/12	None	None	Erythema and roughness on cheeks bilaterally, with KP on arms; no other skin findings	PDL, 1 treatment with excellent response
26/F/White	Early childhood/4	Irritation, redness	Molluscum contagiosum	Erythema on eyebrows, cheeks, outer aspects of upper arms, and back; significant KP on limbs and torso; no other skin findings	20% Lactic acid, 10% propylene glycol, sorbolene cream, tazarotene
27/M/Asian	Early childhood/8	None	Pustular acne at age 8 y	Follicular papules with erythema on back, arms, and thighs; no other skin findings	None

Abbreviations: ADHD, attention-deficit/hyperactivity disorder; GERD, gastroesophageal reflux disease; KP, keratosis pilaris; OTC, over-the-counter; PDL, pulsed-dye laser; ellipses, not reported.

perifollicular inflammation and fibrosis, consistent with KP. The serum vitamin A level was normal (40 µg/dL [1.40 µmol/L]). Prescribed 0.05% tazarotene cream caused peeling and a sensation of burning, with minimal improvement.

CASE 2

A 4-year-old boy (patient 2 in the Table) presented to the University of California, San Francisco, pediatric dermatology practice with a 3-year history of facial erythema. The intensity of the erythema waxed and waned but never completely resolved. The child had mild pruritus in the areas of erythema, with no other symptoms. There was no response to treatment with over-the-counter moisturizing

creams or lotions [Eucerin (Beiersdorf, Inc, Wilton, Conn), Cetaphil [Galderma Laboratories, Fort Worth, Tex], ammonium lactate moisturizer (Lac-Hydrin [Westwood-Squibb Pharmaceuticals, Inc, Princeton, NJ]), or 0.3% tazarotene ointment. Medical history and family history were unremarkable. Physical examination revealed erythema and fine follicular papules on the forehead directly above the eyebrows and on both cheeks. The chin also exhibited erythema and follicular papules with a rough, sandpaper quality. There was questionable lateral eyebrow thinning bilaterally, but no follicular atrophy or scarring was noted. Both ears were erythematous but felt less rough than other affected areas. Typical areas of KP were present on the upper arms and upper lateral surfaces of the thighs (**Figure 2**).

METHODS

Another 25 cases were collected during a 3-month period from physicians who received an e-mail via the Society for Pediatric Dermatology ListServe requesting data on cases similar to our index cases. Responses were received from the United States, Canada, Israel, and Australia. Physicians were sent a data collection sheet asking for information including patient age, sex, and race; age at onset of the disease; age when first seen by a dermatologist; symptoms; other skin conditions; other medical conditions; physical examination findings including location and description of the involved areas, extent of erythema, and other cutaneous findings; pathologic findings if a biopsy specimen was obtained; treatment; and response to treatment. Approval for the study was granted by the University of California, San Francisco, Committee on Human Research.

RESULTS

Of the 27 patients, 17 (63%) were boys and 10 (37%) were girls. The mean age at onset of disease was 5 years (range, birth to 12 years; SD, 4 years). Twenty patients (74%) were white. In most patients the eruption was asymptomatic, although a few patients noted occasional



Figure 1. Case 1. Note the erythematous patches with follicular prominence on the cheeks.



Figure 2. Case 2. Note the erythema and fine follicular papules on the cheeks (A) and the forehead directly above the eyebrows (B). There is questionable lateral eyebrow thinning but no follicular atrophy or scarring.

pruritus. Six patients (22%) reported a history of asthma, and 3 (11%) had a history of allergic rhinitis. Of 23 patients with sufficient descriptions of involved anatomic sites, 21 (91%) had substantial involvement of the face and body. Lesions on the face were typically erythematous keratotic papules with follicular accentuation, without evidence of scarring or hair loss, except in 1 patient (patient 11), who had hair loss of the eyebrows. Facial involvement included the cheeks in 17 patients (74%), the eyebrows in 6 (26%), the ears in 5 (22%), the chin in 4 (17%), the forehead in 3 (13%), and the glabella in 2 (9%). The specific facial location was not described in 4 patients (17%). Other sites of involvement included the lateral or extensor aspects of the arms in all patients, the torso in 13 (57%), the legs in 12 (52%), and the buttocks in 5 (22%). Most of the 27 patients in this case series had no other skin findings, but those noted included acne in 3 patients (11%), atopic dermatitis or an eczematous eruption in 3 (11%), various nevi in 2 (7%), and acanthosis nigricans, molluscum contagiosum, and warts in 1 each (4%). Except for atopic disease (asthma, atopic dermatitis, or allergic rhinitis) in 6 patients, no other consistent medical conditions were noted.

Treatments given for KPR in these patients included emollients; emollients containing urea, lactic acid, topical corticosteroids, or a combination of these ingredients; topical agents containing cholecalciferol, topical or systemic retinoid agents; topical corticosteroids; topical salicylic acid; and pulsed-dye laser therapy. In most patients, there was no substantial improvement with these treatments; 1 patient had a partial response to oral isotretinoin.

COMMENT

Keratosis pilaris is a common condition that has been seen in association with several disorders, including ichthyosis vulgaris and ichthyosis-like phenotype accompanying dry skin and atopy,⁶ cardiofaciocutaneous syndrome,⁷ metabolic disturbances (eg, malnutrition and hypovitaminosis A),³ Noonan syndrome, Down syndrome, diabetes mellitus, and obesity.^{8,9} Given its frequency, some of these associations could be coinciden-

tal. Treatments for KP commonly include emollients, keratolytic agents, topical corticosteroids, and topical retinoids, but these are often ineffective in diminishing the appearance of KP.⁴

Erythema is sometimes present in KP, but is usually mild and limited to the perifollicular skin.⁴ When perifollicular erythema is more noticeable, the disorder has been called KPR by some authors.^{1,4,5} The only study that examined KPR in detail was observational.¹⁰ Voss¹⁰ studied a large number of patients with KP, but he used the term *keratosis follicularis*, which is also used as a synonym for Darier disease. He differentiated 2 forms, *keratosis follicularis alba* and *keratosis follicularis rubra*. The alba form is described as manifesting with follicular papules without erythema, most commonly in children younger than 10 years, with both sexes affected equally, and decreasing in frequency with increasing age. The rubra form is described as having erythematous follicular-based papules. It was noted to increase in frequency with increasing age, was most common in patients 20 to 40 years of age, and was twice as common in women than in men. X-linked dominant inheritance of the rubra form was suggested. Although the clinical description of *keratosis follicularis rubra* by Voss parallels that in our patients with KPR, nearly two thirds of our patients were boys. In addition, our patients were younger when seen by a physician, with ages ranging from birth to 12 years. Since our cases were acquired primarily from pediatric dermatology practices, however, the lack of older patients may be an ascertainment bias. Voss¹⁰ also emphasized the commonness of this condition; in his series, the rubra variant occurred in 25% of the patient population studied.

Using multiple search strategies on both the PubMed and EMBASE databases (search terms included *keratosis pilaris rubra*, *keratosis pilaris*, *keratosis pilaris atrophicans*, *keratosis pilaris faciei*, and *keratosis follicularis*) and review of major dermatology textbooks, we found that the term *keratosis pilaris rubra* has appeared in a few publications,^{1,4,5} but there was only 1 detailed case description of KPR.¹¹ This patient had rosy cheeks since infancy, with progressive erythema during childhood. At age 15 years, she had erythema and small papules on the cheeks and chin, without atrophy; sparse eyebrows; and erythema and KP on the extensor aspects of the arms and thighs. Potassium titanyl phosphate laser therapy resulted in improvement of symptoms.

Several other conditions with clinical features that overlap with KPR have been described. That which most closely approximates KPR is EFFC, which typically develops later in life, most often in the second decade, and is more common in male patients.^{12,13} The condition is also characterized by fine follicular papules with perifollicular erythema involving the cheeks, forehead, and neck. The eruption is usually symmetric. Concomitant KP on the arms has been noted in a few patients with EFFC.¹⁴⁻¹⁶ Features that differentiate EFFC from KPR are lack of reported involvement on the torso and the presence of hyperpigmentation. Erythromelanosus follicularis faciei et colli and KPA, another KP variant, have been considered by some authors to be variants of the same condition,¹² but EFFC lacks scarring. The similarities be-

tween KPR and EFFC, however, are striking, and photographs of EFFC in some reports show findings that are virtually identical to those in our cases,¹⁶ which suggests that they are likely part of the same disease spectrum. The hyperpigmentation noted in EFFC may, at least in part, be related to skin pigmentation type, with darker skin types showing more evidence of hyperpigmentation. Two of the 27 patients in our case series had some degree of hyperpigmentation on the cheeks but were included because of the additional finding of more widespread involvement, not solely involvement of the face and neck. This, too, suggests that KPR and EFFC may be forms of the same condition.

Two other conditions with considerable clinical overlap with KPR are KPA and its more severe variant, atrophoderma vermiculata.¹⁷ The erythematous component of KPR persists or even worsens at puberty, without progression to scarring, whereas KPA results in scarring.^{1,2,18} In addition, the more widespread areas of skin involvement in KPR are less common in KPA, in which involvement is often localized to the face. *Keratosis pilaris atrophicans* is likely inherited in an autosomal dominant fashion with incomplete penetrance.^{19,20} *Keratosis follicularis spinulosa decalvans*, a rare condition with an X-linked dominant inheritance, can also be considered in the differential diagnosis. It is morphologically similar to KPA but is more widespread, resulting in a scarring alopecia involving the eyebrows, eyelashes, and scalp.^{2,21}

Keratosis pilaris has been reported as a feature of cardiofaciocutaneous syndrome. This condition is characterized by congenital heart defects, characteristic facial anomalies, and ectodermal abnormalities including sparse and woolly hair, hyperkeratotic skin lesions, and a generalized ichthyosis-like condition.^{22,23} Mutations in 2 genes, Kirsten rat sarcoma viral oncogene homolog (KRAS) and v-*raf* murine sarcoma viral oncogene homolog B1 (BRAF), have recently been identified in patients with cardiofaciocutaneous syndrome,²³ but only mutations of BRAF have been associated with the hyperkeratosis noted in this condition. This genetic abnormality suggests at least 1 possible putative locus for KP and may provide clues for future genetic studies.

A lack of reliable response to treatment is shared by all of the KP variants. Most of our patients received several treatments, without significant improvement. With the exception of patients (including one of ours) who have responded to systemic retinoid agents^{24,25} and the previously reported patient whose symptoms improved with potassium titanyl phosphate laser therapy, these treatments had minimal to no efficacy. Patients, especially those with prominent facial involvement, are often quite disturbed by the appearance of KPR. Because no treatment is uniformly effective, the potential risks and benefits of various therapies must be considered and explained to patients.

As with other forms of KP, the pathogenesis of KPR is not well understood. The erythema present in KPR fluctuates, and in some patients it is present even in areas without significant keratotic papules. This raises the question of whether flushing via autonomic dysregulation may have a role in the clinical manifestations.

In summary, we describe 27 patients with KPR, a variant of KP that has not been previously emphasized in the medical literature. We believe that KPR, while not so common as KP, is much more common than other KP variants such as KPA. Keratosis pilaris rubra shares many features with EFFC and may be part of the same disease spectrum. Treatments to date have had limited efficacy.

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