Our Dermatology Online

Case Report

Generalized keratosis pilaris rubra

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ABSTRACT

Generalized Keratosis Pilaris Rubra is a rare dermatosis that is related to Keratosis Pilaris group of disorders. We are reporting a case of Generalized Keratosis Pilaris Rubra in a young Arabic male whom treatment options failed to improve his condition.

Key words: keratosis pilaris; generalized keratosis pilaris; keratosis pilaris atrophicans

INTRODUCTION

Keratosis pilaris (KP) is a cutaneous disorder of unknown etiology. It presents as symmetric, keratotic follicular papules on the extremities and the cheeks [1]. Rarely, it can be generalized. It usually develops in early childhood, with remission by adulthood.

There are three distinct variants of KP that have been identified: keratosis pilaris atrophicans, erythromelanosis follicularis faciei et colli and keratosis pilaris rubra (KPR). KPR is characterized by significant widespread erythema and persistence after the onset of puberty.

CASE REPORT

18-year-old Arabic male presented to the outpatient clinic in the dermatology department at King Fahd Hospital of the University with a 2-year history of mildly pruritic facial erythema. The intensity of the erythema waxed and waned but never completely resolved. After few months, the facial erythema became persistent irrespective of the season. Besides, there was generalized skin eruption involving the trunk and upper and lower extremities. There was no history of photosensitivity. Past medical and family history was unremarkable. Physical examination revealed facial erythema affecting mainly the cheeks and ears (Fig. 1). Follicular hyperkeratotic papules with a rough, sandpaper quality and variable erythema on the chest, back, upper limbs and lower limbs (Fig. 2). His complete blood count, liver function and renal function tests were normal. Antinuclear antibodies, anti double stranded DNA, anti Ro and anti Jo were negative. A punch biopsy specimen from the posterior aspect of the left shoulder showed follicular infundibular plugging with slight perifollicular lymphocytic inflammatory infiltrate consistent with KP (Fig. 3).

The patient's informed consent was obtained.

Prior to the study, patient gave written consent to the examination and biopsy after having been informed about the procedure.

The patient was treated with different therapeutics including topical steroid of moderate potency for 3 months, topical etretinoin for 4 months, oral isotretinoin 20 mg daily for six months and acitretin 25 mg daily for 3 months but with no significant improvement. Two sessions of Pulse dye laser was done for the facial lesions but with slight improvement which was discontinued upon the patient request. So we discontinued all medications and kept the patient on regular follow up plan every 12 weeks.

DISCUSSION

Keratosis pilaris (KP) is a common benign condition that can be seen in association with several disorders,

How to cite this article: Bukhari IA, Al Ghamdi N, Al Zahrani A, Al Shawarby M. Generalized keratosis pilaris rubra. Our Dermatol Online. 2015;6(2):187-189.
Submission: 08.11.2014; **Acceptance:** 26.01.2015
DOI: 10.7241/ourd.20152.50

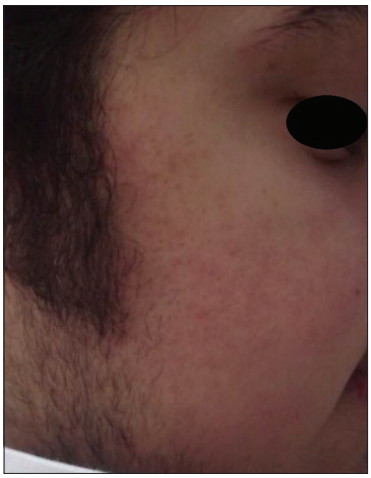

Figure 1: Erythematous patches involving the right cheek.

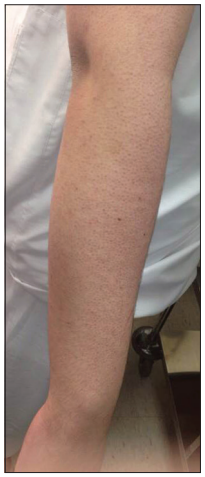

Figure 2: Note the erythematous patches with follicular prominence on the arm.

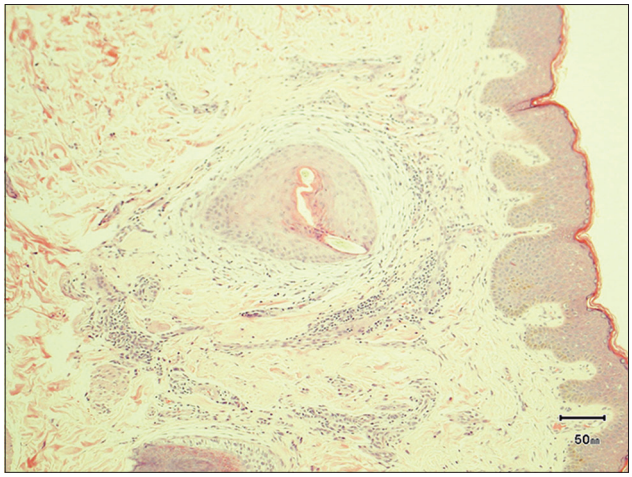

Figure 3: Section showing follicular plug with surrounding dermal perivascular lymphocytic infiltrate. Note also mild acanthosis. H&E, X100.

including ichthyosis vulgaris [2], cardiofaciocutaneous syndrome [3], metabolic disturbances (e.g. malnutrition and hypovitaminosis A), Noonan syndrome, Down syndrome, diabetes mellitus, and obesity [1,4].While Generalized Keratosis Pilaris (GKP) is a rare cutaneous disorder which presents with generalized mildly pruritic symmetric follicular-based papules affecting the face and extremities. Voss [5] studied a large number of patients with KP. He differentiated two forms, keratosis follicularis alba and keratosis follicularis rubra. The rubra form occurred in 25% of the patients with age range of 20 to 40 years and female to male ratio of 2:1. X-linked dominant inheritance of the rubra form was suggested. In Marqueling et al case series [1], 27 cases of KP rubra were reviewed, 63% were males and the maximum age of presentation was 17 years starting mainly during childhood. Pruritus was the main complaint. Our patient is considered the 28th case of generalized KPR reported in the medical literature. The pathogenesis of KPR is not well understood but since the erythema fluctuates and in some patients it is presents without significant keratotic papules. This raises the question of whether flushing via autonomic dysregulation may have a role in the clinical manifestations. Features that differentiate erythromelanosis follicularis faciei et colli from KPR include: erythromelanosis follicularis faciei et colli typically develops in the second decade [6], it lacks the involvement of the torso and there is hyperpigmentation. Treatment for KPR commonly include emollients, keratolytic agents, topical corticosteroids and topical retinoids, but they are of limited efficacy. Pulse Dye Laser and Potassium titanyl phosphate laser were reported to be effective as single case reports [1,7].

In summary, we describe generalized Keratosis pilaris rubra which is a rare entity of KP. Further reports of similar condition will help elucidate its pathogenesis in the future.

CONSENT

The examination of the patient was conducted according to the Declaration of Helsinki principles. Written informed consent was obtained from the patient for publication of this article

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Source of Support: Nil, **Conflict of Interest:** None declared.