Keratosis Pilaris

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**Abstract**

Keratosis pilaris is a common dermatological condition and is sometimes considered as a normal variant of the skin. It is also referred to as follicular keratosis. It is multifactorial and innocuous, affecting the extensor surfaces of the proximal extremities. Keratosis pilaris first appears in early childhood and progresses to become extensive during the second decade of life. Keratosis pilaris is also associated with many other conditions, including atopic dermatitis, ichthyosis vulgaris, obesity, diabetes mellitus, and malnutrition, as well as in conjunction with syndromes like Down syndrome and Noonan syndrome. Although the disease may improve over time, there are still some treatments available, including emollients, exfoliants, anti-inflammatory medication, phototherapy, and laser therapy.

Keywords: Keratosis pilaris; follicular keratosis; Darier's disease; keratosis follicularis.

**Introduction**

Keratosis pilaris is a skin disease, involving the keratinization of hair follicles. This disease must be differentiated from keratosis follicularis, Darier-White disease or Darier's disease, which contrast with 50% to 80% of incidence in adolescence[1]. Males and females are equal to inherit the disease, as about 30-50% of them have family history. Treatment is always available and required if the condition exacerbates or the lesions have become reddish, swollen and itchy upon sweat or frictional irritation. The dark-colored protruding particles in hair follicles are more obvious during winter. Although the disease is actually milder in summer, the patient may experience more rash or itchiness. And because of wearing short-sleeved clothes that expose their skin, the patients are more likely to seek treatment. Generally speaking, the patients have good overall health, despite some of them may have ichthyosis, xeroderma and other keratin abnormalities, or atopic dermatitis, allergic rhinitis, and asthma, even though there is no evidence of direct causal relationship. Common sites are the side of the cheeks, the outside of the arms, and the front and outside of the thighs. In severe cases, the lesions may extend to the neck, the back, the buttocks, and the calves. Black and hard keratinized particles can be seen at the opening of the pores, protruding from the skin surface. They are sometimes accompanied with slightness red rash, especially on the cheeks, which is commonly known as keratosis pilaris rubra. Physical examination will require the check of the clogged pores for the hair that is trapped inside. In some way, keratosis pilaris can be thought as acnes, but the cause and the signs are totally different. External exfoliating ointment, retinoid ointment, and anti-inflammatory cream are the choices of treatment[2-4].

**Pathophysiology of Keratosis Pilaris**

Keratosis pilaris is a constitutional disease. The pathophysiological mechanism of keratinization in the pores is not yet clear but may be related to the defect of keratins in the epidermis. It is also related to the mutation of *FLG* gene, insulin resistance or other genetic and metabolic abnormalities. The thickening of keratin in the hair follicles will block the opening and result in small bumps like acnes. Some patients with atopic dermatitis and ichthyosis are also subjected to this disease. The symptoms are worse in winter, since the pores and the cutin will thicken in a drier and colder climate, making the signs more apparent. There are also reports of its relatedness to vitamin A deficiency, since it is known to cause body-wide dryness, which may indeed aggravate the condition[5,6]. Keratosis pilaris shows an autosomal dominant pattern of inheritance, as there is evidence of mutation in filaggrin genes that leads to the abnormal keratinization of hair follicles. The gene mutation and the abnormal transcription of Ras gene also suggest the co-occurrence of atopic dermatitis. Despite that there is no consensus on the pathophysiology of keratosis pilaris, the most widely accepted theory is the abnormal keratinization at the hair follicles and epithelial cells of the skin, which results in a funnel-shaped obstruction of the pores. The blockage will cause erythema and scaling of the skin around the opening, manifesting as inflamed papules that are the characteristic sign of keratosis pilaris. In addition, curly vellus hair is still present in the inflamed lesion.

**Epidemiology of Keratosis Pilaris**

Keratosis pilaris is actually quite common among adolescents, reaching a percentage of 50 to 80 percent of the population. The disorder is also common in adults, affecting 40 percent of the population. However, keratosis pilaris is often underreported as a disease that it is suspected to have a much higher prevalence rate around the globe. Ethnicity and sex do not predispose an individual to the disease.

**Differential Diagnosis of Keratosis Pilaris**

It is characterized by keratinized follicular papules of 1 mm in size, combined with varying degrees of erythema around the follicles, mainly affecting the cheeks, upper limbs or thighs, and the extensor side of the buttocks. Children and adolescents are mostly affected on the face and upper limbs, while the adults have the extensor side of the limbs being affected. Keratosis pilaris rubra is a subgroup of the disease, in which erythema is more prominent and extends beyond the skin adjacent to the hair follicle. This disease mainly affects the cheeks, forehead, and neck[7]. Erythromelanosis follicularis is another similar disease, but there is pigmentation in the front of the ears and the upper jaw that is symmetrically distributed. This disease is more common in male adolescents and adults. A thorough history and physical exam are required to diagnose keratosis pilaris. The physician should investigate the onset, appearance, location, and symptoms of the epidermal lesions. Inquiring about home treatments and how the condition affects the patient are also important steps in managing keratosis pilaris. Given its nature to affect mostly teenagers, it is also correlated with atopic dermatitis and these patients will often complain about red bumpy skin but without pain or pruritus. These asymptomatic eruptions are generally on the extensor surfaces of the proximal upper and lower extremities, as well as on the buttocks. However, the face, trunk, and distal extremities may also be involved. The physician should look for numerous small inflammatory papules with follicular involvement in these typical locations. Erythema and edema may accompany some lesions if the patient tries to pop the papules to remove the keratinized material within. Psychological distress from the appearance of the lesions, instead of being affected by the symptoms, may prompt an individual to seek medical assistance. Keratosis pilaris is a clinical diagnosis based on history and physical examination. And dermatoscope may aid in the evaluation, in which the abnormalities of the hair follicles can be clearly visualized for identification. Hair shafts are observed to be short, thin and coiled in the layer of stratum corneum. Scaling and erythema are also clearly seen in dermoscopy. As mentioned before, skin biopsy, although helpful, is not necessary for diagnosis[8,9].

**Management of Keratosis Pilaris**

Generally, the disease will improve by itself after puberty. However, one third of the population may carry the condition well into their adulthood. The disease is benign, and treatment can relieve symptoms, but it cannot be completely cured. Treatments include using moisturizing or exfoliating drugs (such as urea/lactic acid/salicylic acid) or topical A acid. Keratosis pilaris is an asymptomatic condition that generally improves over time. As a result, the treatment of the disease is not required. However, patient may speed up the improvement by maintaining hygiene and using hypoallergenic soap, as well as refraining from popping the papules. For patient who is interested in treatment, topical medication can be used to treat keratosis pilaris and these include emollients and topical keratolytics. 6% Salicylic acid lotion or 20% urea cream will improve the skin texture. Other alternatives involve laser treatment, retinoids, and vitamin D3 derivatives. Although these treatments may provide a cosmetic benefit, there are no controlled clinical trials to support their therapeutic effect or prove them as cure for keratosis pilaris. Some case reports have found success with the use of topical retinoids and 0.01% tazarotene. When applied nightly, keratosis pilaris may recede in two weeks and completely resolved by four to eight weeks. Also, skin peeling by using 70% glycolic acid for 5 to 7 minutes has been proven useful for cosmetic improvement on the personal appearance. Several case studies have also described the use of laser in treatment, especially showing success with pulsed dye laser, alexandrite laser, Nd: YAG laser, and fractional CO2 laser. It is crucial for the physician to clearly explain the expected treatment outcome and duration, where patient must understand the process as difficult and tedious but the condition will surely improve over time as the patient ages[10].

**Conclusion**

The physician should devise the therapy in stepwise approach when treating the disease. Initial treatment includes the use of emollient and topical keratolytics. If the patient shows no response to a trial of treatment, the physician should inform and switch to another option, such as retinoids, topical anti-inflammatories, and Vitamin D3 derivatives. Phototherapy and several laser modalities are also therapy options. And if the patient continues to respond poorly to treatments, the physician may consider a dermatologist referral. The general role of a physician to the patient is to assist and provide counseling and monitoring of the treatment progress. Patient must inform the physician of any possible issues. As the patient switches to different treatment options, pharmacist may also be involved to verify the medication and the administration approach to make sure no interference on the patient’s treatment regimen, especially when it is the retinoid therapy. Although the disease is relatively benign and self-limiting, it is still helpful to take an interdisciplinary team approach to directly manage and optimize the treatment result. Lastly, it is imperative to educate patient of the condition. Despite the numerous treatments, they are not preferrable to deal with the disease, because keratosis pilaris is not life-threatening that patient needs to know the treatments may do more harm than good.

**Reference**

1. Haber RN, Dib NG. Management of darier disease: A review of the literature and update. Indian Journal of Dermatology, Venereology and Leprology 2021; 87(1): 14-21.
2. Maghfour J, Ly S, Haidari W, Taylor SL, Feldman SR. Treatment of keratosis pilaris and its variants: a systematic review. Journal of Dermatological Treatment 2020: 1-12.
3. Cohen L, Seminario-Vidal L, Lockey RF. Dermatologic problems commonly seen by the allergist/immunologist. J Allergy Clin Immunol Pract. 2020; 8(1): 102-12.
4. Maghfour J, Ly S, Haidari W, Taylor SL, Feldman SR. Treatment of keratosis pilaris and its variants: a systematic review. Journal of Dermatological Treatment 2020: 1-12.
5. Thomas M, Khopkar US. Keratosis pilaris revisited: is it more than just a follicular keratosis? International Journal of Trichology 2012; 4(4): 255.
6. Castela E, Chiaverini C, Boralevi F, Hugues R, Lacour JP. Papular, profuse, and precocious keratosis pilaris. Pediatric Dermatology 2012; 29(3): 285-8.
7. Hosking AM, Elsensohn A, Makdisi J, Grando S, de Feraudy S. Keratosis pilaris rubra with mucin deposition. J Cutan Pathol. 2018; 45(12): 958-61.
8. Wang JF, Orlow SJ. Keratosis pilaris and its subtypes: associations, new molecular and pharmacologic etiologies, and therapeutic options. American journal of clinical dermatology 2018; 19(5): 733-57.
9. Sonthalia S, Bhatia J, Thomas M. Dermoscopy of keratosis pilaris. Indian Dermatology Online Journal 2019; 10(5): 613-4.
10. Reddy S, Brahmbhatt H. A narrative review on the role of acids, steroids, and kinase inhibitors in the treatment of keratosis pilaris. Cureus 2021; 13(10).