A case of keratosis pilaris rubra

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Keratosis pilaris is a common mild disorder of keratinisation affecting hair follicles. It is often confused with atopic dermatitis, particularly when it is seen in young children.

Case scenario

A 7-year-old boy was referred to a dermatologist after treatment with 1% hydrocortisone ointment and moisturising creams did not settle his facial dermatitis. The main complaint was persistent, prominent redness on his cheeks that became worse with heat or when he was emotionally active. The dermatitis had started when he was 2 years old and had gradually worsened with time. There was minimal itch. The patient had a past history of mild flexural atopic dermatitis and a current history of mild hayfever. There was also a family history of mild atopy.

On examination he had prominent facial erythema, particularly on the mid to outer cheeks, with a slight papular quality. There was no dermatitis present. He had many small red papules on his outer arms to the elbow and a few on his upper back and thighs. His mother also has a milder version of similar papules on her outer arms and mild facial erythema. Based on the appearance and distribution of the papules, the patient was diagnosed with keratosis pilaris rubra.

Comment

Keratosis pilaris is a common mild disorder of keratinisation affecting the hair follicles.[1,2] There are two main components to the condition, both of which are often annoying: retention of follicular keratin leading to small scaly bumps emerging from the hair follicle openings and increased vascular supply around each hair follicle. This combination leads to the typical clinical profile of papular, rough (sandpaper like) skin and erythema. Each of these components shows considerable variation in extent and severity.

Most commonly keratosis pilaris affects the outer arms (more than forearms), face (more often the mid to lateral cheeks), upper back and anterior thighs, varying from extremely mild to widespread (Figures 1 and 2). It appears during childhood, worsens during adolescence to early adult life and usually improves from middle age onwards. Redness is more confluent on the face where the hairs are close together and there are many discrete small red follicular dots on the body and limbs. It usually becomes worse with heat, exertion or emotional stimuli. Itching is occasionally a problem but is usually mild. Keratosis pilaris is often confused with atopic dermatitis, particularly when it is seen in young children as both conditions are common and may coexist. There is debate about whether keratosis pilaris is seen more commonly in atopic individuals than in nonatopic individuals.

The condition is inherited as an autosomal dominant trait with variable penetrance – around 50% of patients with keratosis pilaris have a family history of

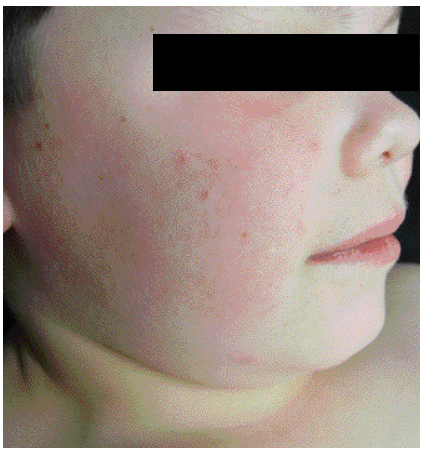

Figure 1. Keratosis pilaris rubra of the face.

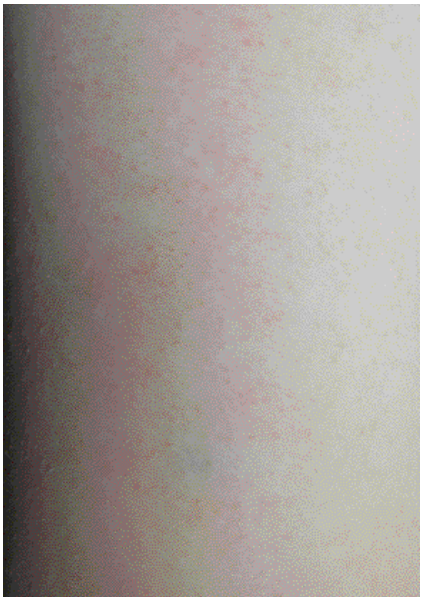

Figure 2. Keratosis pilaris rubra of the outer arms.

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the condition, but it is often not noticed if mild. The nature of the gene defect is not known. Pathology shows a horny plug distending the orifice of the hair follicle, which may contain some twisted hairs. There may be mild inflammation of the surrounding dermis in patients with this condition.

Keratosis pilaris may be initiated or exacerbated by isotretinoin treatment (e.g. for acne). It may also be a part of other conditions (such as ichthyosis vulgaris, Down syndrome, Noonan syndrome, monilethrix and possibly renal insufficiency).

There are different variants of keratosis pilaris. There is a rare form, known as keratosis pilaris atrophicans, in which keratosis pilaris develops initially, followed by atrophy in affected areas. There are various named variants of this process but they may be a spectrum of the same or similar genetic disorders.[3] The perifollicular atrophy in patients with keratosis pilaris atrophicans is often seen on the face or scalp. This is similar to patients with keratosis follicularis spinulosa decalvans (X-linked recessive) in which keratotic follicular papules develop on the scalp in early childhood and erythema and plugging of eyelid follicles may occur. Follicular inflammation often develops leading to variable degrees of scarring alopecia of the scalp. Other features of keratosis follicularis spinulosa decalvans may include focal plantar keratoderma, corneal keratitis and photophobia.

An uncommon variant of keratosis pilaris is erythromelanosis follicularis of the face and neck. This condition was first described in Japanese men but it is also seen in women and people from around the world. There are smooth to papular reasonably well demarcated red brown patches on the mid to outer cheeks spreading to the neck (anterior to the angle of the jaw). More typical keratosis pilaris may be seen on other parts of the patient's body.

Differential diagnoses

Atopic dermatitis

Although atopic dermatitis and keratosis pilaris may overlap, the distinctive distribution, perifollicular roughness and dotty erythema distinguishes keratosis pilaris. Atopic dermatitis is suggested if there is more itch; the patches are dry, more confluent and often with ill-defined margins; there is a typical distribution of the rash and a personal or family history of atopy is present.

Rosacea

Rosacea causes papules and redness of the skin but not usually dryness (although *Demodex* hair follicle mite infestation, which can be part of rosacea, looks like dryness). Small pustules can often be found. There is also an erythematous form with few or no papules. Rosacea is uncommon on areas other than the face and in teenagers.

Telangiectatic erythemas

Patients with telangiectatic erythemas have small telangiectatic vessels often with background erythema but no papules or dryness. Most often this is seen as a result of sun damage in older people, but there are various causes of telangiectatic erythemas in people of all ages.

Treatment

People with keratosis pilaris are often content simply to have the nature of their condition explained. Excessive drying of the skin should be avoided by recommending short (a few minutes) and not too hot showers and soap-free washes. Simple moisturisers often do not work for keratosis pilaris but will help nonspecific dryness of the skin. Exfoliating creams are more effective to smooth the rough skin. Many such formulations are available and are often based on urea, beta hydroxy acids (salicylic acid) or alpha hydroxy acids (fruit acids). They should be applied overnight then washed off in the morning with light abrasive sponging (e.g. with a loofah) until the skin is smooth, then applied as needed. They may irritate a little.

Specific over-the-counter products to treat keratosis pilaris include 2 to 6% salicylic acid in aqueous cream, 20% urea cream, heel balms (including Q.V. Cream, Dermaveen and Eulactol Heel Balm) or alpha hydroxy creams, such as Elucent Skin Refining Night Cream, Lanate Cream or the NeoStrata range (e.g. NeoStrata Skin Smoothing Lotion with 10% alpha hydroxy acid). These products, however, have no effect on the redness. Urea cream at a lower strength (10%) is not keratolytic, only emollient. Redness is more difficult to treat. It can be hidden with camouflaging cosmetics, those with a slight green to yellowish hue hiding the redness more effectively. There are many of these cosmetics on the market, ranging from more opaque concealers to light products such as Avene Diroseal Cream.

Topical retinoids

Topical retinoids such as tretinoin (Retin-A Cream, ReTrieve Cream, Stieva-A 0.05% Cream) applied at night may be helpful. A brief report has also suggested that topical tazarotene (Zorac Cream) is useful.[4] (Treatment should start with a small quantity of the 0.05% strength and the amount applied gradually increased; if not effective and no irritation use 0.1% strength in the same way.) All retinoids are prone to causing dryness and irritation of the skin and could therefore aggravate the redness.

Topical corticosteroids

Topical corticosteroids will help if the skin is mildly itchy. The lowest potency cream or ointment that is needed to have an effect should be used twice daily in short bursts as required to control itch. The vasoconstrictive effect of these agents may temporarily help the redness but their prolonged use will aggravate redness due to their atrophogenic effect.

Laser treatment and intense pulsed light devices

The redness of keratosis pilaris may respond to repeated treatment with various vascular lasers or some intense pulsed light devices. Laser treatments are most effective on the face, and keratosis pilaris erythema on sites other than the face does not usually respond well to this treatment. These treatments are mainly used for patients over 13 years of age.

The literature is scarce, but a potassium titanyl phosphate laser at 532 nm was reported to be effective in one patient with keratosis pilaris rubra after seven treatment sessions at intervals of six to eight weeks.[5] The pulsed tunable dye laser at 585 nm was effective in 12 patients with keratosis pilaris atrophicans after two to eight treatmentsessions.[6] This laser, however, has the disadvantage of causing post-treatment bruising.

Several Australian dermatologists have noted that patients with facial keratosis pilaris rubra may respond well to vascular lasers or second generation intense pulsed light devices used monthly for at least three sessions. For lasers, higher fluences are required so treatments are not overlapped, meaning there will be an uneven result until subsequent sessions lead to more even fading. Recurrence of erythema seems uncommon. Second generation intense pulsed light devices are used by some dermatologists and cosmetic doctors. MT

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