Tidsskriftet Tidsskrift for Den norske legeforening

Keratosis pilaris

CLINICAL REVIEW

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Keratosis pilaris, or 'plucked chicken skin', is a very common condition. It is caused by keratin accumulation in the hair follicles. Although mild cases of the condition can be considered to be a normal variant, it can lead to multiple appointments with general practitioners and dermatologists. In rare cases, keratosis pilaris can form part of specific syndromes or be associated with other diseases. The aim of this article is to give an overview of the different variants of keratosis pilaris and discuss the pathogenesis and treatment options.

Keratosis pilaris, often referred to as 'plucked chicken skin' or 'goosebump skin', is a very common condition, and mild cases can be considered to be a normal variant. It is a chronic condition with possible episodic flares, but with a tendency to spontaneous improvement with age (1). Although the physical symptoms are generally modest, the more subjective and cosmetic issues can lead to negative psychosocial consequences (2). In rare cases, the condition can be a sign of an underlying hereditary syndrome or occur as an adverse drug reaction.

There are few Norwegian articles about keratosis pilaris, which is generally treated in the primary care service. This article is targeted at both general practitioners and doctors in the specialist health service and is based on a non-systematic literature search in PubMed and the authors' clinical experience.

Frequency and pathogenesis

Keratosis pilaris is regarded as the most common follicular skin disease in children and occurs to a greater or lesser extent in 50–80 % of adolescents and 40 % of adults (1, 3–5). It must therefore be considered to be a normal variant. Approximately 40 % of patients have a positive family history of the condition, which is thought to have an autosomal dominant inheritance with variable penetrance (1, 6).

There is a strong association with atopy and ichthyosis, which can both be caused by pathogenic variants in genes that code for the skin protein filaggrin (1, 7). Changes in filaggrin can disrupt the skin barrier, cause atrophy and down-regulate sebaceous glands. These can be contributory factors in the development of keratosis pilaris. The pathogenesis is not fully understood, but the main theory is that abnormal follicular keratinisation prevents vellus hairs from exiting, in addition to localised inflammation (1, 7).

Clinical variants and clinical course

The most common variant of keratosis pilaris involves dry, rough skin with small, hard follicular keratoses (bumps) around the hair follicle ostia, sometimes accompanied by perifollicular erythema. This variant occurs most commonly on the extensor aspect of the upper arms (Figure 1), but also on the hips, thighs and face. The condition begins in childhood and often improves spontaneously after puberty (1).

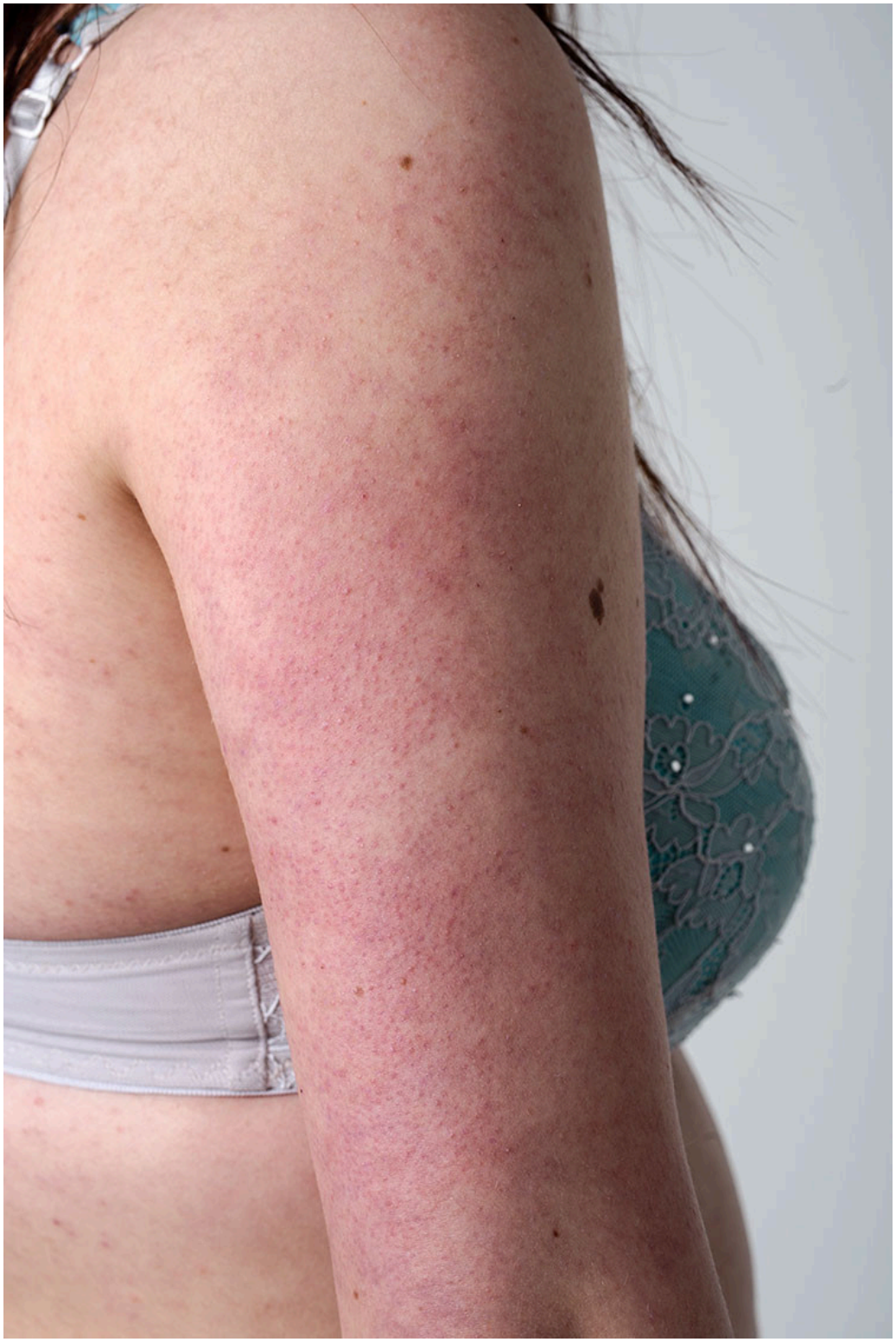

Figure 1 Keratosis pilaris on the upper arm.

Keratosis pilaris rubra refers to cases with predominant facial erythema, particularly on the lateral aspects of the cheeks (Figure 2), which can be cosmetically disfiguring (8). This condition can both worsen and persist after puberty (1). A rarer variant is erythromelanosis follicularis faciei et colli. This condition begins in adolescence and is characterised by erythema, hyperpigmentation and small hyperkeratotic papules on the cheeks, temples and neck, in addition to small papules on the extensor aspect of the upper arms (1). The clinical course is usually prolonged (1). Commonalities of the three aforementioned variants are that they are common and benign, and can be treated if necessary at the patient's request (2).

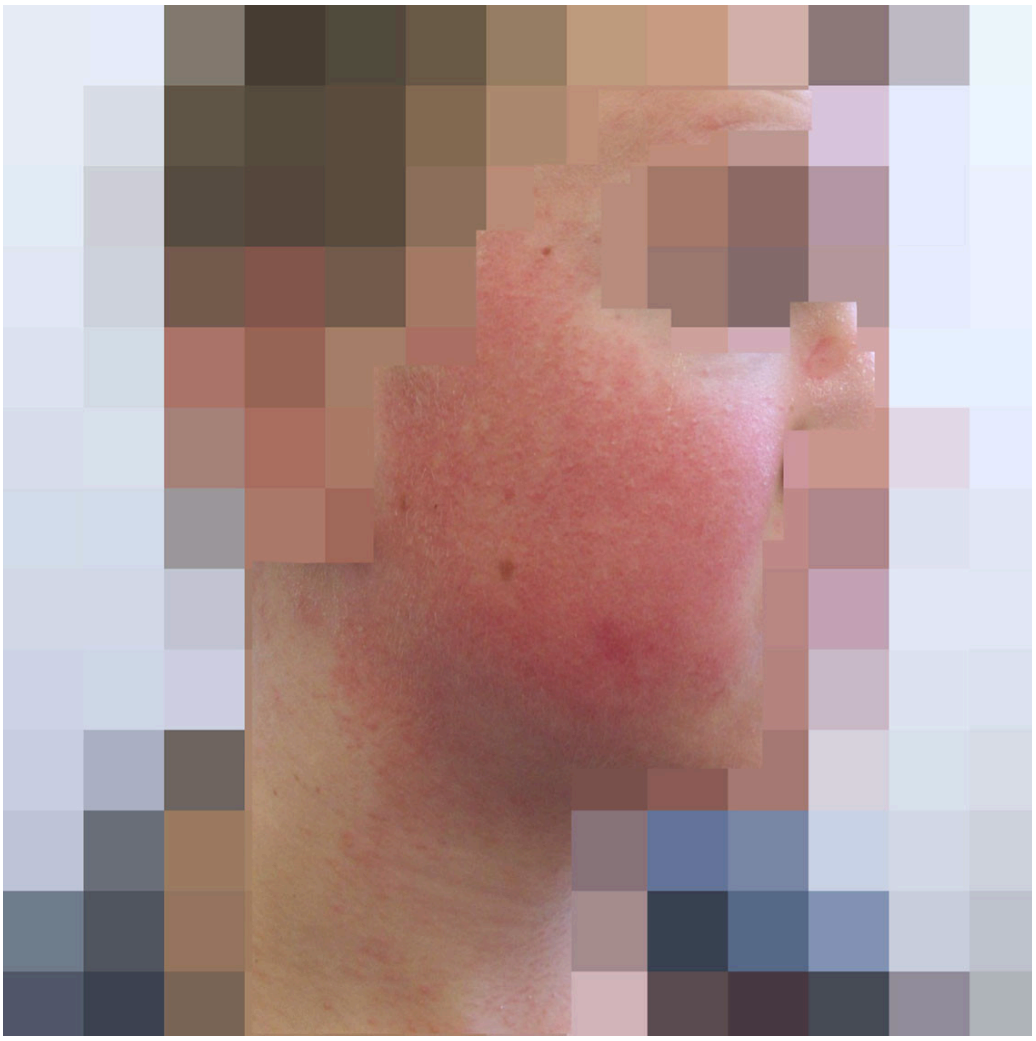

Figure 2 Keratosis pilaris rubra in a patient with Juberg-Marsidi syndrome, an X-linked genetic syndrome with a pathogenic variant in *HUWE1*.

There are rarer variants that have varying degrees of inflammation and atrophy. The umbrella term for these variants is keratosis pilaris atrophicans (Figure 3). In contrast to the other variants, these variants entail a risk of both scarring and hair loss, and in rare cases, extracutaneous manifestations, such as blepharitis, keratitis and dental enamel hypoplasia (1).

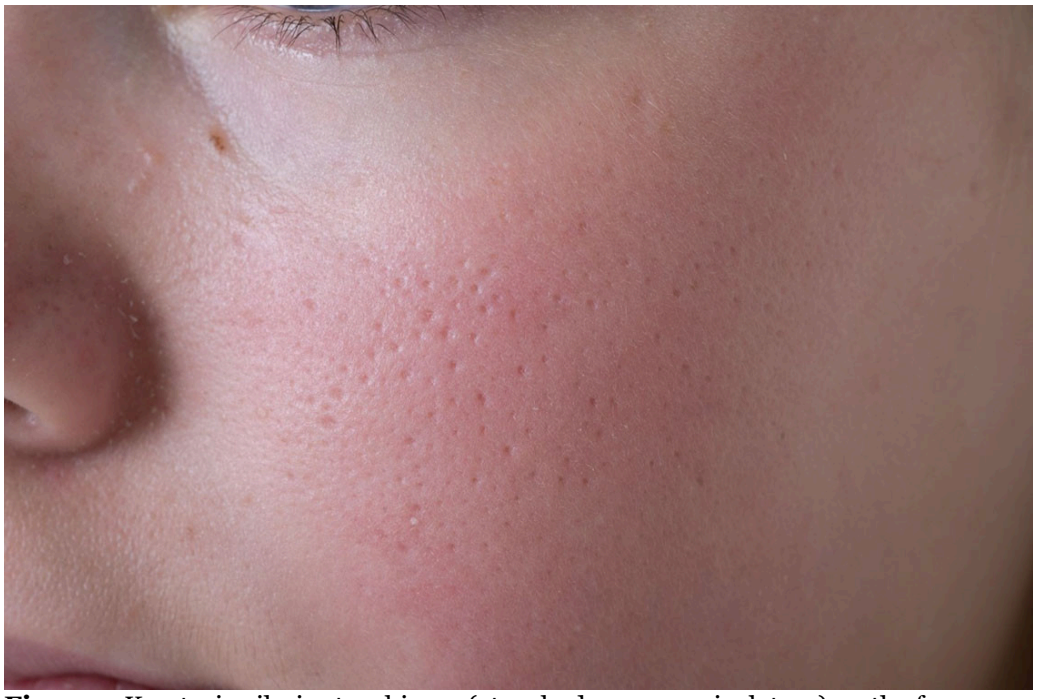

Figure 3 Keratosis pilaris atrophicans (atrophoderma vermiculatum) on the face.

Keratosis pilaris is primarily a clinical diagnosis, but dermoscopy or skin biopsy can be helpful in case of doubt. Dermoscopic findings include mounds at follicular ostia, dilated follicular ostia with hyperpigmentation and erythema, dermal vascular dilatation, and short, thin hair shafts (1).

Associated conditions

Keratosis pilaris may be associated with overweight, diabetes and pregnancy (1), but since keratosis pilaris is so common, it is uncertain if there is any causal relationship. An association with several syndromes has also been reported, including Down's syndrome, Noonan syndrome and cardiofaciocutaneous syndrome (1, 9). The latter two belong to the RASopathies and are caused by mutations in the genes involved in the RAS (*rat sarcoma virus*) signalling pathway, which is essential for regulation of cell growth and division. Keratosis pilaris is a significant finding in these syndromes, and keratosis pilaris atrophicans has been proposed as a potential marker for Noonan syndrome (1, 9, 10). In these cases, there will usually be symptoms from other organ systems in addition to the skin. Consideration should be given to genetic testing if the condition occurs in combination with extracutaneous manifestations and an underlying syndrome is suspected.

B-Raf protein is a proto-oncogene involved in the RAS signalling pathway. Onset of keratosis pilaris has been reported following treatment of melanoma with dabrafenib and vemurafenib, both of which inhibit the B-Raf protein (1). Therefore, it is possible that an abnormal *RAS* gene may be a contributory factor in the pathogenesis of keratosis pilaris. Adverse reactions to other drugs, such as cyclosporine and tyrosine kinase inhibitors, may also be involved (1).

Treatment

Patients with keratosis pilaris should be treated in the primary care service, with a few exceptions. Many patients will be satisfied to learn that the condition is common and does not need to be treated. Patients who find it troublesome will benefit from the application of moisturising creams, keratolytic agents and prescription-only creams.

Moisturising creams strengthen the skin barrier and add moisture to the skin. Unscented creams should be chosen to avoid the development of contact allergy to fragrances. Creams containing lactic acid, propylene glycol, salicylic acid and urea are particularly effective because they also have a keratolytic action (1, 11).

Camouflage creams containing green pigment may be effective for camouflaging the erythema in keratosis pilaris rubra. Dry brushing, exfoliating gloves or pumice can be used to remove bumps and hard skin. Climatotherapy, with exposure to sunlight and saltwater, has been shown to help. Therefore, the condition is often found to improve in summer and worsen in winter (1). Treatment with shortwave ultraviolet light (UVB) can be effective.

In severe cases, prescription-only drugs such as topical retinoids and calcineurin inhibitors can be tried. Retinoids have a comedolytic and anti-inflammatory effect, but can cause erythema and skin irritation. Calcineurin inhibitors, in the form of pimecrolimus or tacrolimus, work by reducing the amount of pro-inflammatory cytokines in the skin, but can cause a transient burning sensation at the application site. The products are registered for the indication of atopic eczema. Local corticosteroids are not indicated, apart from for eczematous skin resulting from scratching (1, 11). Topical vitamin D derivatives have no effect (1).

The patient should be referred to the specialist health service if the diagnosis is uncertain, symptoms are severe or it is suspected that keratosis pilaris is part of a syndrome. It has recently been reported that local treatment with sirolimus, an mTOR inhibitor, is effective in keratosis pilaris rubra (12). Sirolimus inhibits T-lymphocyte-mediated immune reactions and also has an antiproliferative effect on blood vessels. It is not available as a topical drug, but has been compounded and used successfully off-label for angiofibroma associated with tuberous sclerosis (13, 14).

Laser treatment, primarily Q-switched neodymium-doped yttrium aluminium garnet (Nd:YAG) laser or pulsed dye laser, can also be effective in keratosis pilaris that is resistant to other treatment, particularly keratosis pilaris rubra and keratosis pilaris atrophicans (2, 15). These types of laser are effective in treating erythema, while fractional CO_2 laser will work better for atrophy and scarring (15). Adverse reactions are transient erythema and oedema following treatment, as well as the risk of post-inflammatory pigment changes (2). Public funding for this type of laser treatment is not available for this indication in Norway.

Summary

Keratosis pilaris is very common. Many patients will benefit from regular application of moisturising creams and keratolytic agents. In particularly disfiguring cases, laser treatment and topical treatment with sirolimus have shown promising results.

The individuals depicted have consented to the publication of the article.

The article has been peer-reviewed.

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Publisert: 16 March 2023. Tidsskr Nor Legeforen. DOI: 10.4045/tidsskr.22.0513
Received 12.8.2022, first revision submitted 19.10.2022, accepted 28.11.2022.