KERATOSIS PILARIS ATROPHICANS: A CASE SERIES

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Summary

Introduction: *Keratosis pilaris (KP) and keratosis pilaris atrophicans (KPA) are follicular keratoses with inflammatory and sometimes atrophic evolution.*

Clinical cases: *We present a case series of patients diagnosed with different subtypes of KPA.*

Conclusions: *The differential diagnosis of various KPA subtypes is a clinical one, with particular importance regarding the evolution and therapeutical approach of these patients.*

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Introduction

Keratosis pilaris (KP) and keratosis pilaris atrophicans (KPA) are follicular keratoses with inflammatory and sometimes atrophic evolution. Histologically, they are defined by orthokeratotic plugs in the follicular orifice.

Clinical cases

We present a series of clinical cases with different subtypes of KPA.

Clinical case 1: We report the case of an 11-year-old female patient who was consulted for the presence of keratotic papules that appeared several months after birth. Dermatological examination highlighted the presence of hyperkeratotic follicular papules surrounded by an erythematous halo located on the eyebrows, cheeks, extension surfaces of the arms, thighs and buttocks, with associated scarring alopecia of the eyebrows [Fig. 1]. The general condition of the patient was good, without other dermatological conditions. The patient's mother had identical, but less severe skin lesions [Fig. 2]. Taking into account the clinical appearance and family history, a diagnosis of keratosis pilaris atrophicans faciae was established.

Clinical case 2: A 17-year-old patient was consulted for the presence of hyperkeratotic papules located on the hairy scalp. The lesions started at the age of 6 and showed progressive evolution. Cutaneous examination identified keratotic follicular papules and crusts on the hairy scalp [Fig. 3], as well as keratotic follicular papules on the eyebrows, trunk, upper and lower limbs [Fig. 4]. Histologically, corneous plugs were observed around the follicular openings. The diagnosis of keratosis follicularis spinulosa decalvans was established and treatment with isotretinoin 20 mg/day was started for 6 months with improvement of the lesions.

Clinical case 3: A 32-year-old woman was consulted for symmetric reticular atrophy of the face. Skin lesions appeared at the age of 12 and progressed until the age of 19. Dermatological examination revealed a seborrheic and erythematous face, well defined and atrophic scars of irregular shape, with a diameter of

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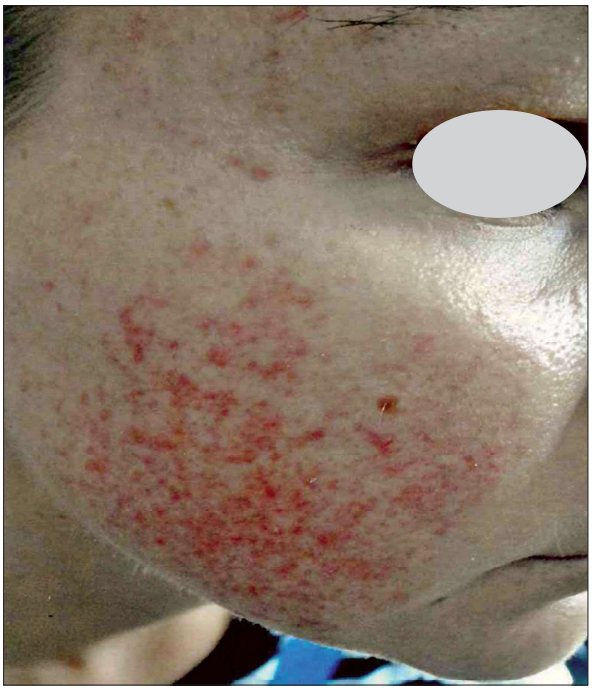

Fig. 1. Keratosis pilaris atrophicans faciae. Erythematous hyperkeratotic and atrophic papules located on the face and eyebrows.

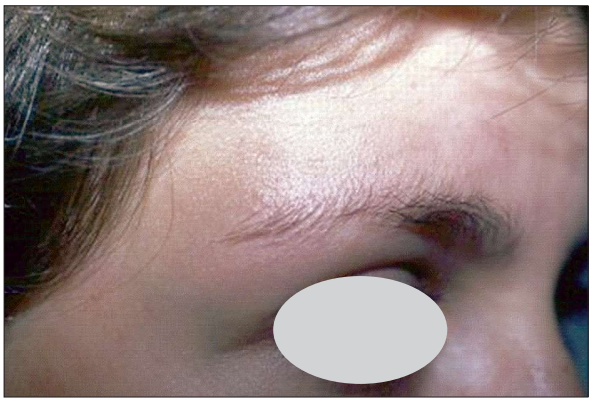

Fig. 2. Keratosis pilaris atrophicans faciae. Discrete follicular papules located on the eyebrows and forehead. Lateral and external atrophic alopecia of the eyebrow, as seen at the mother of the patient.

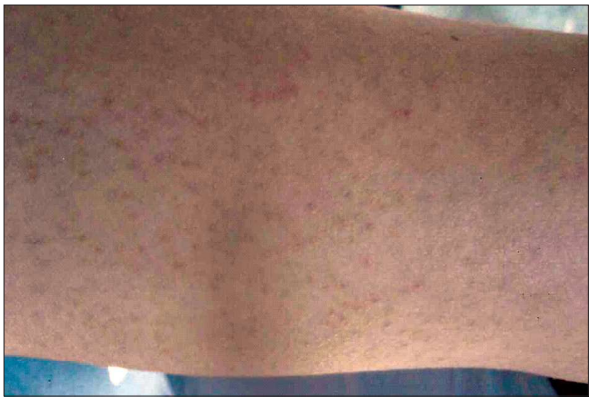

Fig. 4 Keratosis follicularis pilaris rubra on the forearm of the same patient.

1-3 mm, and separated by narrow bridges of epidermis giving a honeycomb appearance . The lesions were located symmetrically on the forehead, cheeks, nose, chin and preauricular area [Fig. 5,6]. The diagnosis of atrophodermia vermiculata was established.

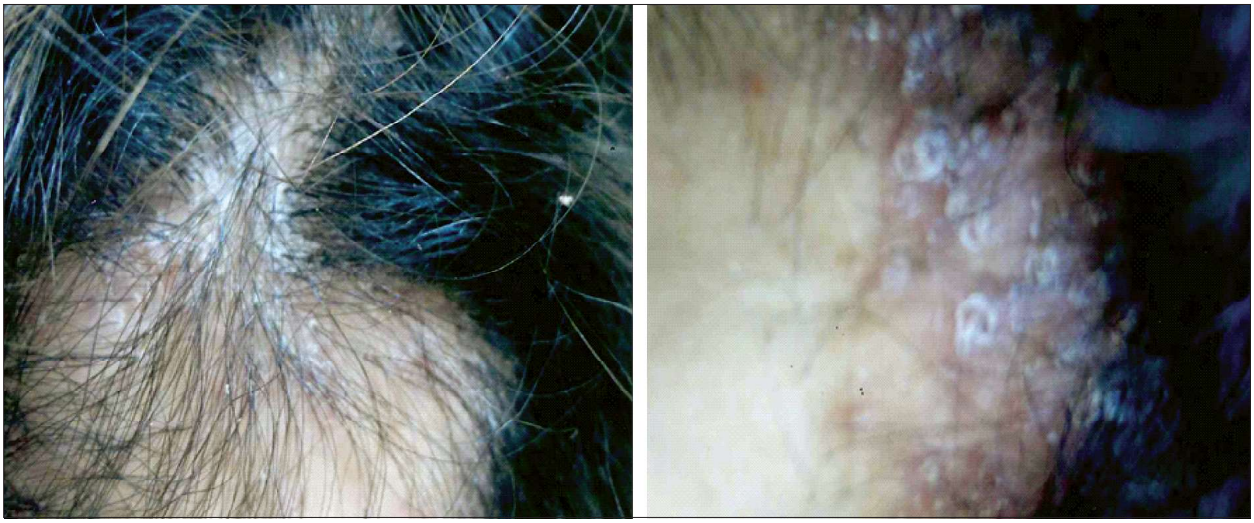

Fig. 3 Keratosis follicularis pilaris decalvans. Crusts and scarring alopecia of the scalp.

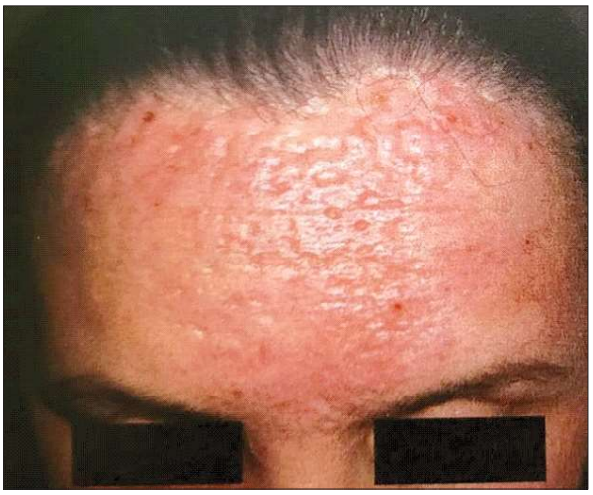

Fig. 5 Atrophodermia vermiculata. Reticular atrophy of the face, with honeycomb appearance.

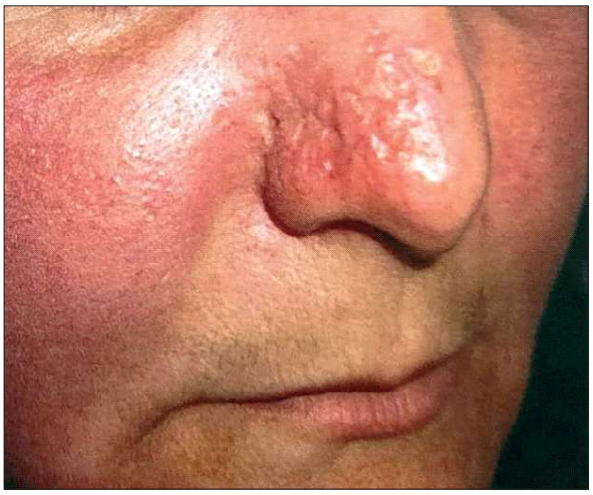

Fig. 6 Hyperseborrheic skin. Atrophic pitted scars of irregular shape on the nasal bridges.

Discussions

Keratosis pilaris (KP), also known as lichen pilaris, is the most common form of follicular keratosis. Being hereditarily transmitted, probably polygenic [1], it presents as follicular hyperkeratotic papules surrounded by a follicular erythema, usually localized on the extensor surface of the extremities and the gluteal region and rarely on the face and trunk. It appears gradually in the first decade of life and becomes more severe at puberty, later showing a slow improvement. It can be associated with atopic dermatitis or ichthyosis vulgaris, metabolic (malnutrition, obesity, diabetes) or endocrine (hypothyroidism, hyperandrogenism and hyperadrenocorticism) conditions [2] and can be a part of Noonan, Greither and Down syndromes. As for the differential diagnosis, lichen spinulosus (skin-colored hyperkeratotic papules, without erythematous halo, frequently associated with HIV [3]), pityriasis rubra pilar (follicular corneous papules, orange-colored plamo-plantar keratoderma, erythematous and scaly lesions with islands of spared skin), phrynoderma (monomorphic pigmented papules, located on the extensor surfaces of the limbs and trunk and associated with vitamin A deficiency) or lichen scrofulosorum (scaly lichenoid follicular papules, asymptomatic, located on the trunk and root of the limbs) should be taken into account.

Keratosis pilaris atrophicans (KPA) is phenotypically related to KP. It has three subtypes: keratosis pilaris atrophicans faciae, keratosis follicularis spinulosa decalvans and atrophodermia vermiculata. All subtypes initially involve the cheeks or lateral edges of the eyebrows, leading to atrophy, scarring or alopecia. It can present as an independent condition or can be a part of another syndrome.

Keratosis pilaris atrophicans faciae (KPAF), described by Wilson in 1878, appears in the first months of life. Clinically, it presents as keratotic follicular papules surrounded by an erythematous halo located on the lateral edge of the eyebrows and cheeks, with progressive evolution towards eyebrows' alopecia, cheeks and less often of the forehead. The lesions stop evolving at puberty. KPAF is considered a marker of Noonan syndrome [4], cardio-facial-cutaneous syndrome [5], Rubinstein-Taybi syndrome [6] and Cornelia de Lange syndrome [7].

Keratosis follicularis spinulosa decalvans (KFSD), described by Siemers in 1926, is a disease with recessive transmission linked to p21.13-p22.2 locus. The clinical picture includes hyperkeratotic papules, initially located on the cheeks and nose with extension to the eyebrows, hairy scalp, nape, and trunk. Subsequent scarring alopecia of the eyelashes, eyebrows and hairy scalp may occur. Focal plantar keratoderma, blepharitis, conjunctivitis, corneal dystrophy, dental anomalies, deafness, atopic dermatitis,

cutis laxa, syndactyly or Noonan syndrome can be associated. The differential diagnosis must be carried out with folliculitis spinulosa decalvans (erythema, pustules, and crusts) and Quinquaud's epilating folliculitis (follicular pustules evolving to scarring alopecia; Staphylococcus Aureus is present in 80% of cases).

Atrophodermia vermiculata (AV), also known as honeycomb atrophy, occurs more often in childhood between the ages of 5 and 12, but can also be found in adults. The patients present with hyperkeratotic papules and characteristic reticular atrophic scars located on the cheeks, above the lip and forehead and "pitted" with a diameter of 2 mm and a depth of 1 mm divided by narrow epidermal bridges, that give a honeycomb appearance. Evolution is slow until puberty, after which the lesions may even regress. It can be associated with oral leukokeratosis or Down, Marfan [8], Rombo or Nicolau-Băluș syndromes. The differential diagnosis should be made with atrophia maculosa varioliformis cutis, in which mildly, not preceded by inflammation or trauma, facial scarring is evident.

The lesions are due to hyperkeratosis of the follicular orifice. The histology of KPA depends on the stage of the disease. As such, initial stages are defined by keratotic plugs located in the follicular openings and a lymphocytic infiltrate with few neutrophils around the hair follicle in the upper dermis. In the atrophic stage, hair follicle atrophy, dermal sclerosis, corneous cysts, dilated lymphatic and vascular vessels should be observed, while, in advanced stages, scarring alopecia will be present.

Treatment of KP and KPA is done in all forms with topical keratolytics, such as urea and retinoids [9], and systemic retinoids (acitretin, isotretinoin) and dapsone in severe forms such as KFSD. Topical and systemic antibiotics are ineffective. In AV, dermabrasion and/or injections with fillers, CO2 or Nd:YAG laser should be considered .

Conclusions

The differential diagnosis of the three forms of KPA is a clinical one, with particular importance regarding the evolution and therapeutical approach of these patients. Also, a careful, complete and multidisciplinary examination of such cases is required, taking into account that this condition can be a part of several syndromes.

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Conflict of interest
NONE DECLARED

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