

WHAT IS YOUR DIAGNOSIS?

Case for diagnosis

Caso para diagnóstico

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HISTORY OF THE DISEASE

Eleven-year-old boy with rough skin on the facial region associated with darkening and gradual loss of the eyebrow since he was three years old. Within this period similar lesions, on the shoulders and arms, associated with pruritus and xerosis appeared. Emollients were used presenting partial improvement of xerosis and pruritus but without improvement of hyperpigmentation. Exam showed keratosis pilaris on the frontal region, temporal, malar and of the neck associated with brownish-erythematous lesions delimited on the facial region

(Figure 1). The patient also presented alopecia of the eyebrows with discrete papules without atrophy or scar on the area. (Figure 2). In the deltoid and tricipital regions there was keratosis pilaris with hyperpigmentation (Figure 3). Histopathologic exam (Figure 4) revealed orthokeratotic hyperkeratosis, follicular "plug" and brownish pigmentation on the basal layer. On the papillary dermis there are some dilated vases, rare melanophages and superficial perivascular inflammatory infiltrate formed by lymphocytes and histiocytes.



FIGURE 1: Brownish-erythematous hyperpigmentation associated with keratosis pilaris and temporal region



FIGURE 3: Keratotic papules in the posterior region of the left arm



FIGURE 2: Eyebrow alopecia with discrete papulous lesions

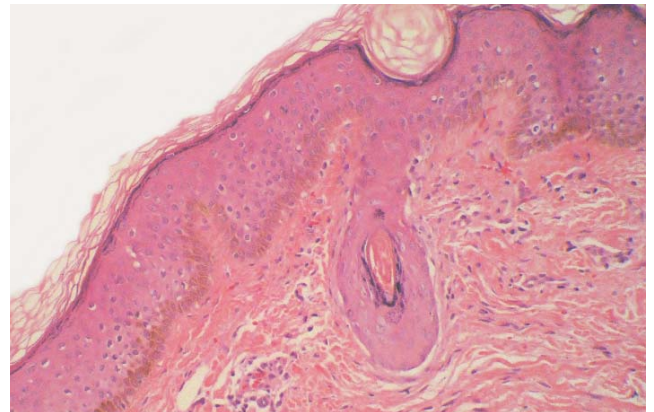


FIGURE 4: It is observed orthokeratotic hyperkeratosis, follicular plug and clear brownish pigmentation on the basal layer. There are some dilated vases in the papillary dermis, and rare melanophages and superficial perivascular inflammatory infiltrate consisting of lymphocytes and histiocytes

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COMMENTS

Erythromelanosis follicularis faciei et colli is a pigmentary disease associated with erythema and involvement of the pilary follicle. Primarily, it is observed in adolescents. However, it also affects children and young adults, with preference for the male sex. Bilateral distribution is the main characteristic but unilateral cases were described.¹ It was first described in 1960, by Kitamura and collaborators in Japan. Its origin is unknown but there is an hereditary component (autosomal recessive) that seems to be interfering with its genesis.²

Clinically it presents three characteristic findings: delimited erythema, hyperpigmentation and follicular papules. Brownish delimited hyperpigmentation occurs in preauricular and maxillary areas symmetrically, affecting also the region of the neck and the temporal region. Follicular papules³ occur associated with pigmentary alteration erythema. In some cases diascopy evidences brownish colour caused by compression of telangiectasia. However, the absence of such vascular alterations does not influence the diagnosis of EFFC. Another common characteristic is alopecia in the region of the vellus hair which does not occur so frequently in the terminal hair areas such as beard and scalp.⁴ Associated keratosis pilaris on the arms and shoulders can be found and to some authors the EFFC would be a variant of Keratosis pilaris rubra.^{4,5}

Differential diagnoses include Athrophoderma Vermiculatum, "ulerythema ophryogenes", poikiloderma of Civatte, and keratosis pilaris.^{4,6,7} Athrophoderma Vermiculatum is characterized by atrophy with aspect of honeycomb in the malar regions. "Ulerythema ophryogenes" presents erythema with alopecia, atrophy and scar, involving the eyebrows. Both conditions differ from EFFC due to the presence of atrophy and scar.⁶ Poikiloderma of

Civatte is observed in middle aged women as reticulated dyschromia with atrophy and erythema affecting preferably photoexposed areas and sparing the submental region. The absence of follicular papules associated with starting age and lesions distribution is different from EFFC. Keratosis pilaris faciei does not present well delimited area of hyperchromia although erythema can be seen around the follicles.

Histopathology is not diagnostic although it reveals: hyperkeratosis, plugg follicular, discreet hyperchromia of the basal membrana and dilatation of the superficial dermis vases. The pillary follicles are enlarged with perianexial inflammatory infiltrate.^{1,3,4}

Treatment is not well defined but it should be avoided solarexposition as well as other sorces of ultraviolet light. Sunscreen is recommended¹. Various topic keratolytics are used including urea cream (10-20%), ammonium lactate 12%, tretinoin cream (0,05-0,1%) and combinations with hydroquinone 4%. Peelings with salicylic acid (30%) can be used. Vitamin C analogues are an option due to their regulating mechanism of keratinization⁸. In the most severe cases oral isotretinoin is used intermittently. Currently, the Pulsed Dyed Laser of 595nm is a therapeutic option to attenuate hyperpigmentation and erythema.⁹

Erythromelanosis follicularis faciei et colli (EFFC) is a rare disease for some authors but others consider it an underdiagnosed entity. Due to rarity or to underdiagnosis, less than 50 cases were described. Our case, apart from presenting clinical-pathological characteristics of EFFC, shows marked keratosis pilaris on the upper limbs and shoulders generating the discussion whether EFFC and keratosis pilaris rubra would be spectrum of the same disease. □

Abstract: Erythromelanosis follicularis faciei et colli is a rare disease of unknown etiology characterized by symmetric erythematous brownish hyperpigmentation on the frontal, malar and temporal areas associated with involvement of the pilary follicle. It is common to present pilar keratosis on the shoulders and neck. The three main clinical characteristics are: erythema (with or without telangiectasias), discrete follicular papules and brownish hyperpigmentation.

Keywords: Alopecia; Hyperpigmentation; Keratosis

Resumo: Eritromelanose folicular faciei et colli é uma doença rara, de origem desconhecida, caracterizada por hiperpigmentação eritêmato-acastanhada e simétrica nas regiões frontal, temporal e malar, associada com envolvimento do foliculo piloso. É comum apresentar ceratose pilar no pescoço e nos ombros. Sua característica clínica primária é composta pela tríade: eritema (com ou sem telangiectasias), pápulas foliculares discretas e hiperpigmentação acastanhada.

Palavras-chave: Alopecia; Ceratose; Hiperpigmentação

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