Case and Review

Erythromelanosus Follicularis Faciei et Colli: A Case Report in a Caucasian Male and Brief Review of the Literature

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Keywords

Erythromelanosus follicularis faciei et colli · Erythema · Hyperpigmentation · Follicular papules · Facial region

Abstract

Erythromelanosus follicularis faciei et colli, a rare condition of unknown etiology, was first described by Kitamura et al. from Japan in 1960. It is characterized by a triad consisting of well-demarcated erythema, hyperpigmentation, and follicular papules. We report the case of a 50-year-old Caucasian male, who had asymptomatic symmetrical facial lesions since the age of 42. His family history was unremarkable. Published erythromelanosus follicularis faciei et colli cases of the last 10 years are summarized in this report to demonstrate the variability and differences in the clinical presentation of this uncommon diagnosis.

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The uncommon diagnosis of erythromelanosus follicularis faciei et colli (EFFC) was first described in 1960 by Kitamura et al. in six Japanese males [1]. Clinical findings include well-demarcated erythema, hyperpigmentation, and follicular papules. The disease primarily affects darker-skinned young men and is seen less commonly in the Caucasian population. Until now, only few cases have been reported in Germany [2].

We report the case of a 50-year-old Caucasian male patient who presented to the Department of Dermatology, Venereology and Allergology, University Hospital Essen, Essen, Germany, for symmetrical erythematous lesions, which originated in the preauricular area and had spread to the patient's temples and forehead. The patient reported that the skin condition had been apparent for about 8 years. He had seen dermatologists before, and he had been treated with topical steroids and emollients, all without effect. The condition was static within the last years, although the patient had recently noticed some slight enlargement towards his forehead. Except for arterial hypertension, for which he is treated with ramipril, and an episode of Lyme disease, which was treated in 2018, his past medical history was unremarkable. He denied a history of atopy, photosensitivity, and pruritus. The family history for dermatologic conditions was unremarkable. The clinical examination revealed bilateral symmetrical erythema, partly with slight hyperpigmentation in the preauricular areas, the temples, the lateral forehead, and the posterior part of the submandibular areas (Fig. 1). There was no telangiectasia visible. Dermoscopy revealed round whitish areas with keratotic follicular plugs surrounded by gray spots in a reddish-brown background. Examination of the hair, nails, and mucous membranes did not show any alterations. The patient denied any changes of the condition with sunlight exposure or other factors. He did not mention any burning sensation, pain, or pruritus. A biopsy was not performed and the diagnosis of EFFC was made based on clinical findings. The patient was advised to the consequent usage of sunscreen and informed that topical steroids will not show any desired effect.

EFFC is a rare dermatosis of unknown etiopathogenesis, which is clinically characterized by the combination of mostly bilateral, symmetrical erythema and brownish pigmentation, associated with more or less evident follicular plugging. Keratosis pilaris was described as an associated condition in some cases [2]. Histopathologic findings include follicular plugging, hyperkeratosis, increased basal membrane pigmentation, and vascular dilation in the upper dermis. Development of the disease usually starts symmetrically in the preauricular area and spreads onto temples and lateral aspects of the neck; however, cases of unilateral involvement have also been reported. The cause of the disease is unknown, but a hereditary component seems to play a role at least in part of the cases. The onset of the disease is most commonly observed in childhood and adolescence [3]. However, our patient had no family history regarding dermatologic disorders including EFFC and his disease onset was at the age of 42 years. Thus, we hypothesize the presence of a non-familial form of EFFC in our patient.

An overview of available EFFC cases of the past 10 years is summarized in Table 1 for a better understanding of the clinical presentation and variations between patients.

The treatment of EFFC is not well defined. Topical treatments included metronidazole, ammonium lactate, tacalcitol ointments, retinoids, hydroquinone, and salicylic acid. Treatment with pulsed dye laser or Nd:YAG laser for erythema and hyperpigmentation have been reported [3].

Although EFFC is considered a rare disorder, it has been suggested that it actually may just be an underdiagnosed dermatosis [4]. The simultaneous presence of well-demarcated

erythema, hyperpigmentation, and follicular papules in the facial and neck region should make one think of EFFC.

Statement of Ethics

This research work was conducted in accordance with the Declaration of Helsinki. The subject of the case has given his written informed consent to publish this case.

Conflict of Interest Statement

Carl Maximilian Thielmann and Wiebke Sondermann have no conflicts of interests to disclose.

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Author Contributions

Carl Maximilian Thielmann – initial idea, writing the report, revising the report, treatment of the patient. Wiebke Sondermann – writing the report, revising the report, treatment of the patient.

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Fig. 1. Photograph of the patient's left (a) and right (b) side of the face and neck. Both sides exhibit symmetric areas of reddish, partly light-brown pigmentation, which extend from the preauricular areas to the temples and the forehead of the patient. Follicular papules are faintly visible.

Table 1. Summary of previous reports of EFFC in the literature of the past 10 years

Age of onset	Sex	Clinical findings	Year	Ref.
4 years	Male	Bilateral flat erythema of the cheeks with slight hyperpigmentation and follicular papules; lateral rarefaction of the eyebrows; keratosis pilaris	2014	2
Early childhood	Male	Diffuse non-scaly reddish-brownish patches and multiple skin-colored, hypopigmented follicular papules on both cheeks	2015	3
12 years	Male	Erythema with telangiectasia, mild hyperpigmentation in preauricular and sub-mandibular areas of the neck; keratosis pilaris	2010	5
7 years	Male	Bilateral, symmetrical patches of reddish-brown pigmentation with tiny papules on preauricular areas, cheeks, and submandibular areas; diffuse dark-brown reticulated pigmentation over both upper and lower limbs	2017	4
5 years	Female	Erythema over forehead, peribuccal area, malar region, and cheeks; follicular papules and hyperpigmentation; rarefaction of lateral eyebrows; keratosis pilaris	2016	6
27 years	Male	Asymptomatic reddish-brown pigmentation with roughness of the cheeks, temples, lateral aspects of the nose, and frontal area	2017	7
19 years	Male	Erythema and hyperpigmentation affecting cheeks, forehead, chin, preauricular region, and neck; follicular papules	2011	8
14 years	Male	Erythemato-pigmented patches; follicular papules on both cheeks	2019	9
7 years	Male	Erythema and brown-colored pigmentation over bilateral preauricular and malar areas; tiny whitish papules; keratosis pilaris	2016	10

The case reports reflect male predisposition and on average early onset of the disease with slight variation within the clinical appearance.