Erythromelanosis Follicularis Faciei: A Case Report and Review of the Literature

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Key Words

Erythromelanosis follicularis faciei · Erythromelanosis follicularis faciei et colli · Keratosis pilaris

Abstract

Erythromelanosis follicularis faciei is a rare sporadic condition of unknown etiology characterized by reddish-brownish patches and follicular papules that appear commonly on the face and rarely on the neck. Herein, we report a 16-year-old male who had asymptomatic facial skin lesions since early childhood. His family history revealed a similar case in his younger brother. His parents are not consanguineous. Skin examination revealed diffuse nonscaly brownish patches with erythematous background and multiple skin-colored, hypopigmented follicular papules on both cheeks. A summary of previous reports of erythromelanosis follicularis faciei in the literature is presented in this report. © 2015 S. Karger AG, Basel

Introduction

Erythromelanosis follicularis faciei (EFF) is a rare sporadic condition of unknown etiology characterized by erythematous hyperpigmented patches and follicular papules on the face. It was first described in Japanese patients in 1960 by Kitamura and collaborators. When the neck is affected, the condition is called erythromelanosis follicularis faciei et colli (EFFC) [1]. The pathogenesis is unknown. However, a combination of vasodilation and hyperpigmentation has been found in the affected areas. Some authors consider EFF as part of the spectrum of keratosis pilaris atrophicans disorders [2]. EFF is characterized clinically by the

presence of red-brown patches on the lateral aspects of the cheeks, and rarely lateral aspects of the neck. Numerous pinhead-sized follicular papules are present within the involved areas that may sometimes appear relatively hypopigmented. Bilateral distribution is the main characteristic, but unilateral cases have been described [1–15]. EFF is usually asymptomatic. However, a burning sensation has been described in a few patients [16–21]. Keratosis pilaris elsewhere in the body is a common association with EFF. Histopathologically, there are hyperkeratosis, slight follicular hyperkeratosis (follicular plugging), increased basal layer pigmentation, dilatation of superficial dermal blood vessels, and periadnexal lymphocytic infiltrate [9].

Case Report

A 16-year-old male presented with asymptomatic facial skin lesions which he had since early childhood. There was no history of predisposing factors. He had been using topical treatment but without any help. Family history revealed a similar case in his younger brother. His parents are not consanguineous. Skin examination revealed diffuse nonscaly reddish-brownish patches and multiple skin-colored, hypopigmented follicular papules on both cheeks (fig. 1). On the basis of the above classical clinical findings, the diagnosis of EFF was made. The patient was reassured and put under periodic follow-up.

Discussion

Erythromelanosis follicularis faciei (EFF) is a pigmentary disease associated with erythema and follicular papules on the face. It affects all races. However, it shows a preponderance in the people of Asian ancestry [10–20]. The cause is unknown, but the hereditary component (autosomal recessive) seems to play a role in the pathogenesis [12, 19, 22, 24]. Table 1 summarizes the previous reports of erythromelanosis follicularis faciei (EFF) in the literature.

EFF primarily affects adolescents. However, it has been reported in children as young as 2 years old and in adults as old as 46 years old. Similarly, the onset of the disease shows a wide range, starting from birth to as old as 43 years old. The male:female ratio is 2:1 [1–24]. Differential diagnoses include keratosis pilaris rubra, poikiloderma of Civatte, Riehl's melanosis, and pigmented peribuccal erythrosis of Brocq. In skin type I patients, there may be only erythema, leading to a significant overlap with keratosis pilaris rubra, and it remains to be answered whether EFFC and keratosis pilaris rubra are two spectrums of the same condition [2, 9–18]. 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 [5–15]. Treatment is not well defined. Various modalities have been described. Topical agents have been used, including, ammonium lactate, retinoids, hydroquinone, vitamin C, salicylic acid peels (20–30%), glycolic acid peels, tacalcitol ointment, and metronidazole gel. The evidence for their use is anecdotal. Limited courses of isotretinoin (0.1–1 mg/kg/day) have been tried in severe cases. A combination of laser treatment (pulsed dye laser) for erythema and Q-switched Nd:YAG laser for hyperpigmentation have been tried but they require multiple sessions [1–24].

Statement of Ethics

Consent has been obtained from the parents of the patient for the purpose of using patient's photographs for print or online publication.

Disclosure Statement

The authors have no conflicts of interest that are directly relevant to the content of this paper. No sources of funding were used to assist in preparation of this paper.

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Table 1. Summary of previous reports of EFF in the literature

First author [Ref.]	Cases, n	Patient age, years	Gender	Patient age at disease onset	EFFC cases in the family, n	Site of lesion	Keratosis pilaris	Other associated conditions	Treatment
Shahshahani [1]	60	range: 4–39 average: 22	M = 43 F = 17	at birth, 3 cases 1st decade, 16 cases 2nd decade, 33 cases 4th decade, 1 case	8	cheeks and neck	trunk, arms, thigh	milium in 4 cases	not available
Volks [2]	3	15	not available	4 years	no	cheeks	trunk, limbs	no	no
		2	F	1 year	no	cheeks	limbs	no	
		4	F	birth	1 (mother)	cheeks, forehead	arms, limbs	no	
Silva [3]	1	11	M	3 years	no	face, shoulder, arms	face, neck	no	no
Augustine [4]	3	19	F	15 years	2 (sister and brother)	cheeks, neck, ears	upper back, shoulders, arms	no	no
		10	F	6 years	no	cheeks, chin	upper back, shoulders, arms	no	
		13	M	8 years	no	cheeks	shoulders	no	
Ertam [5]	2	17	F	12 years	no	face, neck, arms	arms, upper back, thigh	no	topical salicylic acid 2% and retinoic acid 0.01%
		19	F	15 years	no	face, neck	arms, upper back, thigh	no	
Aljabre [6]	1	17	M	11 years	no	cheeks, lower lip and auricles	shoulder areas	no	no
Karakatsanis [7]	2	24	F	23 years	no	cheeks and neck	not available	no	topical retinoids
		14	F	10 years	no	cheeks and jaw	not available	no	
Kurita [8]	2	26	M	not available	no	cheeks and neck	not available	no	pulsed dye laser
		22	M	not available	no	cheeks	not available	no	
Kim [9]	10	range: 12–46 average: 22	M = 8 F = 2	range: 8–43 years average: 16.5 years	no	cheeks and neck	1 patient with keratosis pilaris on arms	no	not available
Ermertcan [10]	1	18	M	childhood	1 (brother)	maxilla, cheeks, neck	arm	no	oral isotretinoin
Whittaker [11]	1	15	M	13 years	no	preauricular regions	arms	no	topical retinoic acid
Tuzun [12]	1	17	M	10 years	2 (sister and father)	cheeks	arms and trunk	diabetes mellitus and congenital leukokeratosis	no
Lee [13]	1	18	M	13 years	no	cheeks, neck	no	no	no
Warren [14]	2	35	F	25 years	no	cheeks	arms	no	topical ammonium lactate 12% or metronidazole gel
		43	F	24 1/2 years	no			no	
McGillis [15]	1	13	F	8 years	no	face and neck	upper arms	no	topical tretinoin 0.05%, ammonium lactate and hydroquinone
Watt [16]	1	15	M	several months	no	temples, cheeks and neck	upper arms	no	no
Wang [17]	12	not available	F = 7 M = 5	not available	not available	cheeks	not available	not available	dual wavelength laser (pulsed dye laser + Q-switched Nd:YAG laser)
Kim [18]	11	14.4 ± 7.7	M = 6 F = 5	not available	not available	preauricular and maxillary regions	not available	not available	topical tacalcitol ointment
Lalit [19]	1	21	M	19 years	no	cheeks forehead chin	arms, shoulders, back	no	no
Li [20]	1	20	M	12 years	no	preauricular area, neck	shoulders, lateral arms, thighs	no	dual-wavelength laser system (pulsed dye laser + Q-switched Nd:YAG laser)
Sodaify [21]	3	15, 18, 28	M	childhood	no	cheeks	arms and legs	no	tretinoin and hydroquinone cream
Acay [22]	2	17	M	9 years	1 (sister)	cheeks and neck	upper arms	no	not available
		17	M	childhood	2 (mother and grandmother)	cheeks	face, neck	no	
Sardana [23]	5	19	M	13 years	no	temples, cheeks, neck	arms, shoulders, back	not available	oral isotretinoin
		13	M	not available	no			not available	topical retinoic acid cream
		18	M	12 years	no			not available	tretinoin and hydroquinone cream
		11	M	5 years	no			not available	
		13	F	not available	no			not available	
Yanez [24]	2	15	M	10 years	1 (sister)	cheeks, forehead and neck	arms	no	topical retinoic acid cream 0.05%
		18	F	childhood	1 (brother)	cheeks	upper limbs	no	
Al Hawsawi, this study	1	16	M	childhood	1 (brother)	cheeks	no	no	no

Fig. 1. Diffuse nonscaly reddish-brownish patches with multiple skin-colored, hypopigmented follicular papules on both cheeks.