ERYTHROMELANOSIS FOLLICULARIS FACIEI

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ABSTRACT

Erythromelanosis follicularis faciei is not uncommon.

***Background**: Erythromelanosis follicularis is characterized by the triad of brown pigmentation, erythema and follicular papules on the face and neck. It is considered a rare, but actually underreported disease. Young adult males are principally involved. Only 11 females are yet reported.*

Methods: We observed 60 patients with this condition during a 2-year period from September 1994, in Razi Hospital and a private office, with special attention to the distribution of lesions, the age at onset, family history, sex, the presence of keratosis pilaris and the skin phenotype.

***Results :** 17 cases were female. The age range was from 4 to 37 years old and the age at onset of disease was in the second decade in the majority of patients. Two pairs of siblings were among our cases. Keratosis pilaris was a frequent finding.*

Conclusions : Erythromelanosis follicularis faciei, especially its milder forms, is not rare : It is rather missed or underreported by physicians. It seems this condition is ameliorated or cleared by increasing age, a suggestion that must be proved by further studies.

Introduction :

Erythromelanosis follicularis faciei et colli was first reported by kitamura et al, in 1960.

They described six cases in young Japanese men that consisted of a triad of hyperpigmentation, erythema, and follicular plugging. In all cases the face, neck, or both were involved bilaterally [1]. Although originally reported in Japanese adults, it occurs not infrequently in caucasian subjects [2]. Sodaifi et al reported 3 cases of erythromelanosis from Iran [3]. The disease frequently begins in the second decade, is more common in males [2] and only 11 female patients are yet reported [4,5].

Characteristically, it begins symmetrically on the preauricular cheeks and imperceptibly spreads onto the temples and lateral aspects of the neck [2]. When the neck is affected, the dermatosis is termed erythromelanosis faciei et colli. The eruption is usually bilateral but may be unilateral [4].

A background of reddish-brown pigmentation with telangiectasia studded with pale follicular papules [2]. This follicular papules produce a granular texture [4]. With diascopy, the erythema fades and the underlying pigmentation becomes more apparent [6]. Vellus hairs, and to a lesser degree, terminal hairs may be involved in affected areas [2], but there is no evidence of atrophy or scarring. The dermatosis is usually asymptomatic, although some patients describe a burning sensation. Keratosis pilaris of the upper extremities and trunk is a frequently associated finding [4]. The disease spreads slowly, is persistent and is not influenced by treatment [2].

Histopathologic features of EFF correlate well with the clinical findings. The individual hair follicles are enlarged from horny masses retained in the infundibular region.

Parakeratosis is noted at the follicular orifices. There is increased pigmentation of the basal layer inter follicularly.

The blood vessels in the papillary dermis are dilated with a mild lymphocytic infiltrate cuffing the vessels. Electron microscopy has revealed that many melanosomes appear abnormally large with a homogenous mass of pigment within the center of the melanosome [4].

The possible sexual predilection for male individuals, the relatively early age of onset, and the follicular papulation help to differentiate erythromelanosis follicularis from chloasma, Riehl's melanosis, poikiloderma of Civatte, and erythrose peribuccale pigmentaire of Brocq. The lack of seborrhea, comedones, pustules, and cysts helps to exclude the acneiform eruptions. The lack of atrophy and scarring helps to separate out ulerythema ophryongens and atrophoderma vermiculatum [7].

In recent years, we observed several cases of EFF in Razi Hospital (Tehran) and also a private office (M.M.S.) in some of them, EFF was an incidental finding and not the patient's complaint. The low number of reported cases in the world's literature (35 cases till 1995 [8]), urged us to study this disease in a 2-year period.

Materials and methods :

We studied patients with EFF who presented to Razi Hospital and the office of one of the authors (MMS) during a 2-year period from November, 1994.

The diagnosis of EFF was based on the presence of patches of erythema, pigmentation and follicular

papules of the cheeks and other areas of the face or neck, and approved by at least two dermatologists (fig 1,2,3,4).

Demographic and clinical data including age, sex, age at onset of lesions, family history, distribution of lesions, aggravating factors, the presence of keratosis pilaris, and the situation of vellus and terminal hairs were cited (table 1). Biopsy was performed in one patient (case 34).

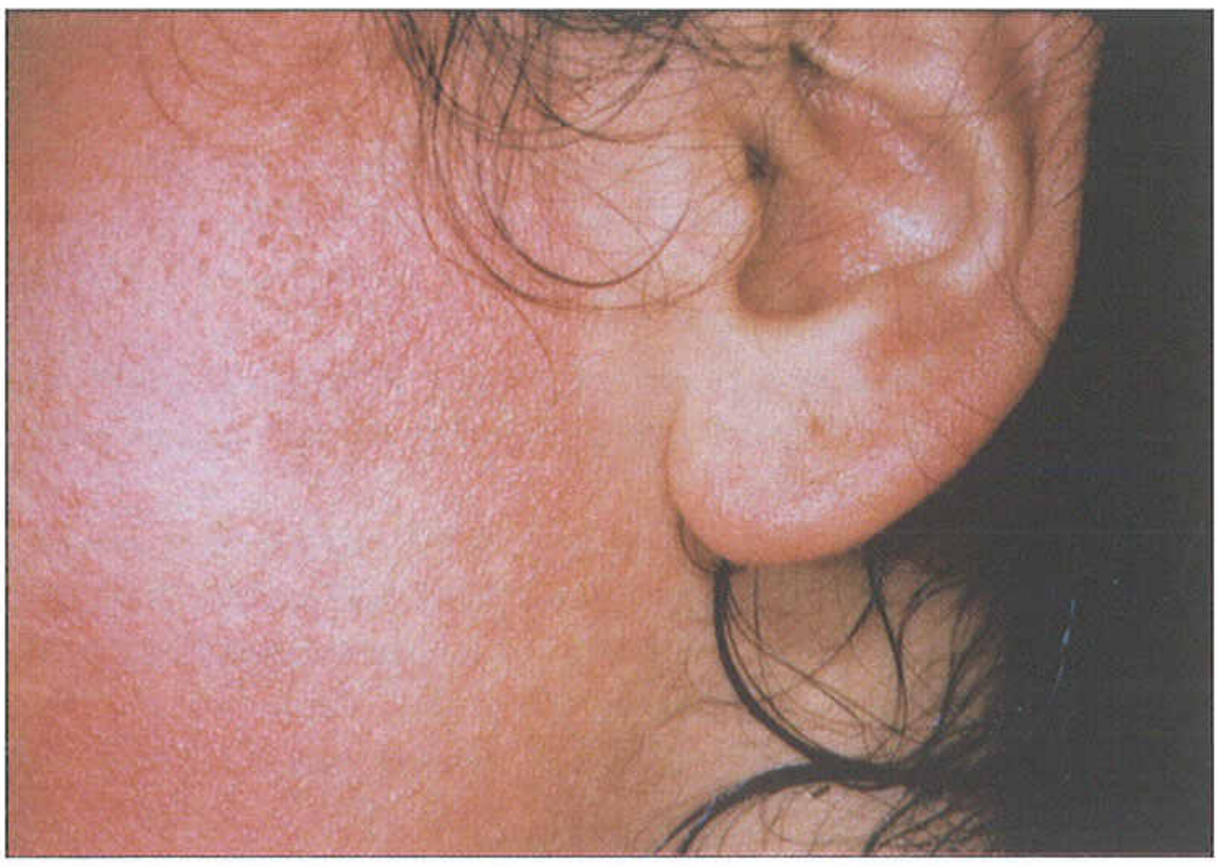

Fig. 2. Pigmentation, erythema and follicular micropapules on the face.

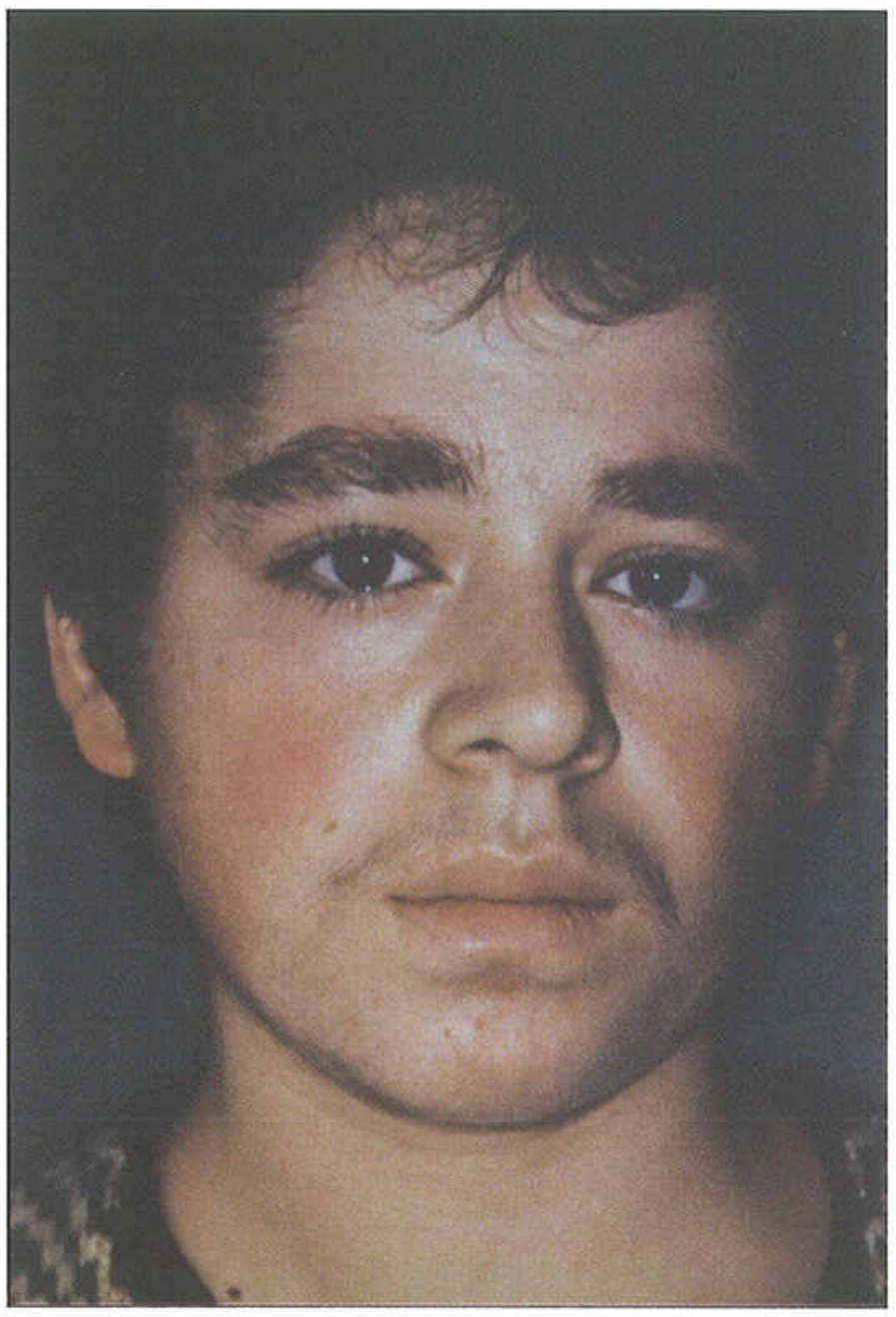

Fig 1 - Erythromelanosis on the cheeks, eyebrows and chin

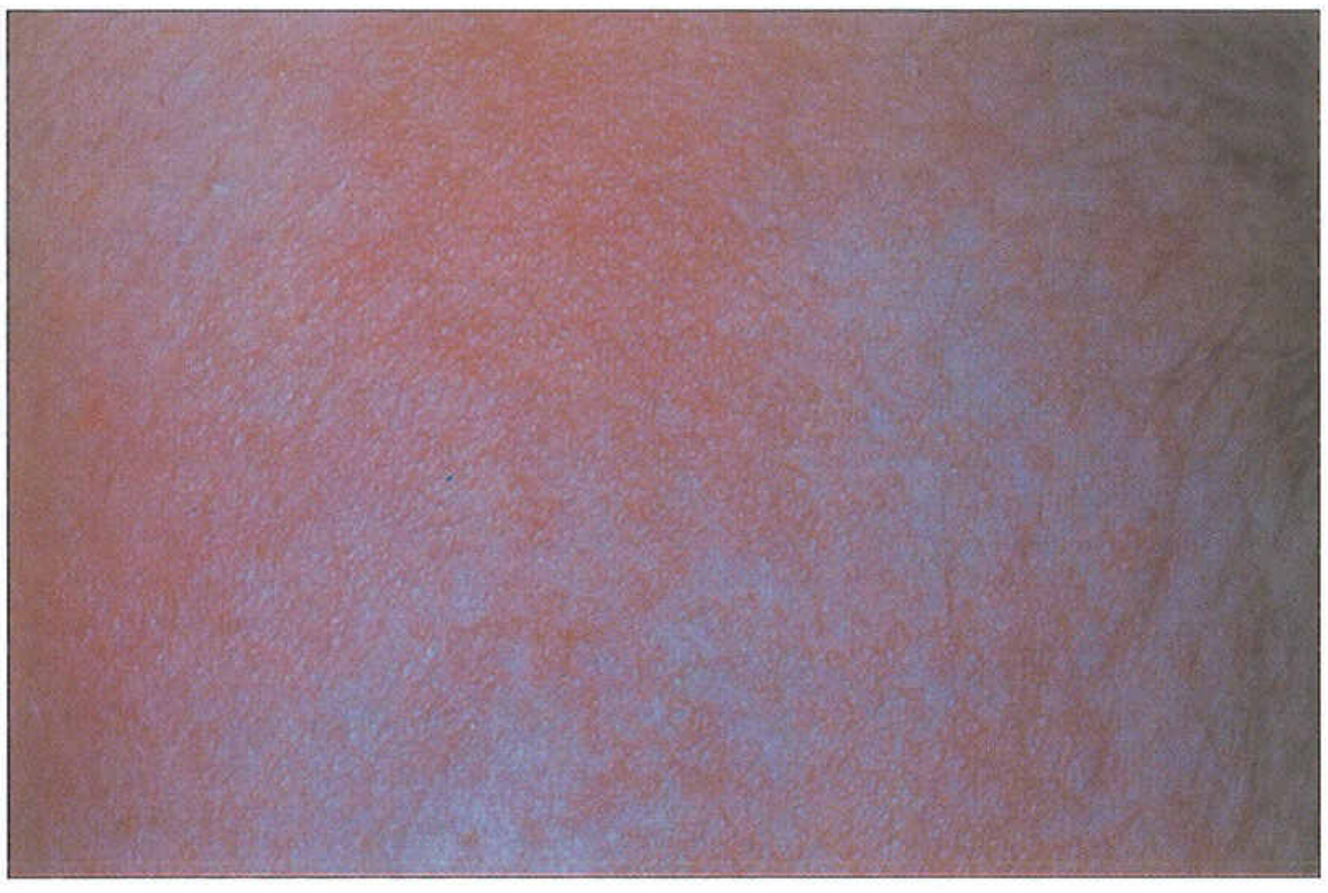

Fig. 3. Close-up of erythromelanosis follicularis

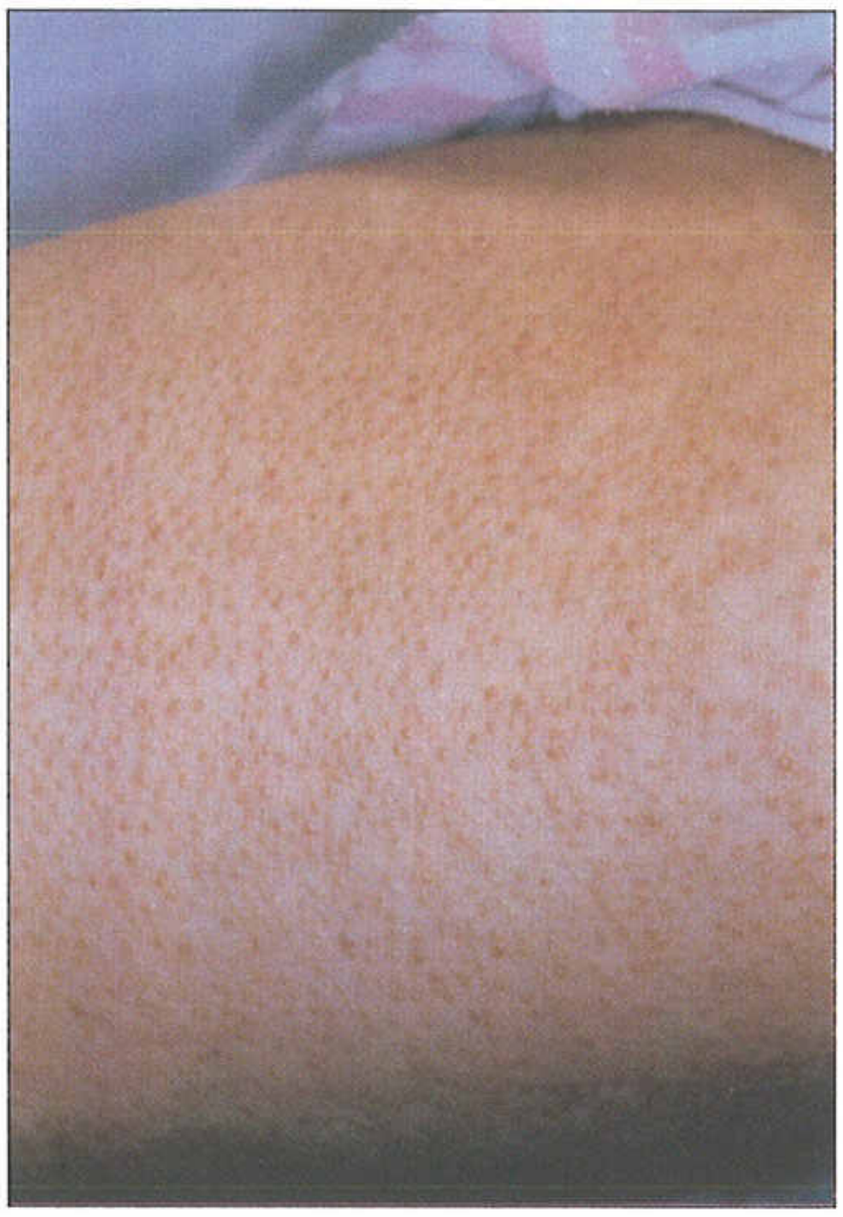

Fig. 4. Keratosis pilaris of the arm.

Results

From 60 patients who have EFF, 17 were female and 43 male. The peak age of patients was in the 2nd decade (49 cases) and the age range was from 4 to 39 years. The age range at onset of the disease was claimed from birth to the 4th decade, but the peak age was in the second decade (33 cases). Two sisters (cases 2 and 3), and a brother and sister (cases 34 and 35) were among our patients. In four other cases a positive family history was mentioned. A history of keratosis pilaris was present in the family members of 6 cases. 41 cases were not fair-skinned (phenotype 3) and 15 patients have a fair skin (phenotypes 1and 2).

The situation of hairs in involved areas was as follows : vellus hair loss in 14 cases, reduced terminal hair in 14 patients and reduction of both hair types in 5 cases. In all the 4 patients with involvement of eyebrows, hairs were reduced. The beard was involved in the remaining cases of reduced terminal hair and in the age range of 17 to 24 years.

Aggravating factors were mentioned by 47 cases such as Sun (26 cases), heat (26 cases) and cold (23 cases). Exercise, excitement and bathing were among other factors.

Keratosis pilaris was present in 53 cases and from a mild form to diffuse keratosis rubrum. Fine milia were seen in 7 patients.

In light microscopic examination of the biopsy obtained from case 34, mild hyperkeratosis without parakeratosis, mild acanthosis and hyperpigmentation of basal layer were seen in the epidermis. Dilation of hair follicles and keratotic plugs with cyst, capillary dilation and chronic inflammatory cells with some macrophages around vessels, with other findings.

Discussion :

The cases of EFF in our study were more than the total cases reported in the literature. Of course, the latter point is not due to the low frequency of the disease in other countries or its high frequency in Iran. It seems the disease is underdiagnosed or underreported by dermatologists, a point emphasized in different articles. In fact, mild forms of the disease are frequently missed. The peak age at onset of the disease in our case series was in the second decade as other reports [2]. Our youngest patient was 4 years old, while the child reported by Anderson was a 12 year-old boy [8]. The disease is considered rare in women: Warren and Davis reported the 10th and 11th female cases [4,5]. The presence of 17 females in our case series is significant in this respect. The presence of two pairs of siblings (two sisters, a sister and a brother) among our cases, is another interesting point. Only a few familial cases are reported in the literature.

We observed keratosis pilaris in association with EFF in the patients and their families as seen in other reports. It's worth mentioning that some authors consider EFF and keratosis rubra pilaris as variants of each other [7]. In some articles, aggravating factors such as heat and cold were cited [3]. Many of our cases, too, complained of exacerbation of EFF following sun exposure, heat and cold.

The presence of fine milia on the involved areas in 7 cases was interesting. Light microscopy of EFF is compatible with this clinical finding [3]. The reduction of vellus hairs in 14 cases was predictable, but reduced terminal hair of eyebrows without scar or atrophy in 4 cases with the more severe form of the disease, is interesting. Spare beard hair is considered normal regarding the age range of the patients. The helix was involved in many of our cases, a finding noted by Sodaifi et al in one of their patients [3]. EFF appeared rare after the 4th decade. this finding could be due to a change or amelioration of the clinical aspects of the disease with increasing age. So, a prospective study to follow the course of EFF is invaluable.

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The authors wish to thank Zahra Safai Naraghi, M.D., the chief of the department of Pathology, for her cooperation.

Table 1- Characteristics of patients with erythromelanosis follicularis

Age(y)/No/Sex	Family history	Phenotype	Site	Age at onset (y)	Aggravating factors	Reduced hair	Keratosis pilaris	Associations
1/M/15	-	not fair	cheek,ear,neck, forehead	<1	-	vellus	Rubrum (limbs,trunk	milia
2/F/14	+(sister) (K.p.father)	not fair	Forehead,cheek, chin, upper lip, ear	7-8	sun	vellus, terminal eyebrow	Rubrum,(arm,thigh, trunk)	
3/F/12	+ (sister) (K.p.father)	not fair	Chin, upper lip,cheek, ear, eyebrow (slight)	11	cold	vellus ±	--	
4/M/21	(K.p.sister)	not fair	Cheek,ear	14	cold,heat,sun	terminal	Rubrum(upper	
5/M/16	-	Not fair	Cheek,ear	13	sun	-	-	
6/F/14	-	fair	Cheek,ear	6-7	sun,heat	-	Rubrum(diffuse	
7/F/16	-	not fair	Cheek,ear	14	heat,cold,sun	-	Rubrum (buttock, arm)	
8/M/14	(K.p.sister)	not fair	Cheek,ear	8	sun,heat	vellus	Rubrum (thigh,trunk, arm)	
9/M/24	-	not fair	Cheek,ear,temple eyebrow,nose		sun,exercise	-	Rubrum (arm), keratosis pilaris (thigh)	
10/M/13	+	not fair	Cheek,nose	8-9	Exercise	vellus	-	milia
11/M/14	-	not fair	Cheek,ear	11	summer	vellus ±	Diffuse	
12/M/14	-(K.p.mother)	not fair	Cheek	12			Forearm	
13/F/15	-	not fair	Chin,ear,under the jaw cheek,forehead	13	sun	vellus ±	Rubrum (limbs)	
14/M/24	-	fair	Cheek,ear,chin,forehead	school age	Sun,cold,heat	terminal	Arm	

Table 1- Characteristics of patients with erythromelanosis follicularis (continued)

Age(y)/No/Sex	Family history	Phenotype	Site	Age at onset (y)	Aggravating factors	Reduced hair	Keratosis pilaris	Associations
15/M/21	-	not fair	Cheek		Sun,heat	terminal	Arm, thigh	
16/M/18	-	not fair	Cheek	15	Sun,cold, heat	terminal	Arm., thigh	
17/M/17	+(Father and uncles	fair	Cheek	early childhood	Exercise	terminal and vellus	Rubrum (Arm,thigh	
18/M/15	-	not fair	Cheek		Sun	-	Arm	
19/M/15	-	not fair	Cheek,nose,ear,forehead upper lip,eyebrow,chin	12	Sun,dust		Arm,thigh	
20/M/17	-	not fair	Cheek,ear		-	Vellus ±	-	
21/M/17	+(Brother)	not fair	Cheek,nose	15	Exercise,heat	-	Rubrum (limbs,trunk)	
22/M/12	-	fair	Cheek,ear,eyebrow	3-4	Cold	vellus	Arm, thigh	
23/M/4	-	not fair	Cheek,upper lip, forehead,ear	1	-	-	limbs, trunk	milia
24/M/15	-	not fair	Cheek, temples, neck, chin	12	Sun,cold	-	limbs, trunk	
25/M/17	-	not fair	Ear, cheek		sun, cold	terminal and vellus	-	
26/M/17	-	fair	cheek,ear	15	sun,heat,cold		Rubrum,(arm,chest)	
27/M/21	-	not fair	Cheek,ear	17	sun,heat,cold	-	Rubrum,	
28/M/17	-	not fair	cheek	17				
29/F/14	-	not fair	Cheek	12	Heat,sun			
30/M/16	-	not fair	Cheek,ear	13	-		rubrum (Arm)	milia
31/M/18	-		Cheek	15		vellus +	limbs,trunk	

Table 1- Characteristics of patients with erythromelanosis follicularis (continued)

Age(y)/No/Sex	Family history	Phenotype	Site	Age at onset (y)	Aggravating factors	Reduced hair	Keratosis pilaris	Associations
32/F/13	-	not fair	Cheek	12	Heat	-	Arm, buttock,thigh	
33/F/12	-	fair	Cheek,ear	<10	Sun, heat,cold wind	-	Rubrum (Limbs)	
34/F/16	+(brother, sister)	fair	Cheek,ear,eyebrow	childhood	sun,emotion, heat	vellus,terminal eye-brow	+	
35/M/22	+ (2 sister)	fair	Cheek,ear,eyebrow	childhood		vellus,terminal eye-brow	+	
36/M/17	-		Cheek,,ear, eyebrow,	9-10			+	
37/M/16	-		Cheek,ear				Arm	
38/M/17	-	not fair	Cheek,ear	sun,heat, emotion,cold	-		Arm,trunk	
39/M/19	-	not fair	Cheek	15	sun,heat	terminal	Rubrum (arm)	
40/M/18	-	not fair	Cheek	13-14	Sun,wind,cold certain foods	vellus +	Rubrum (arm,trunk)	
41/M/19	-(K.P.father	not fair	Cheek,ear, eyebrow		-	vellus,terminal +	Rubrum (arm,thigh)	
42/M/22	-	not fair	Cheek,forehead,nose	17	delayed bathing,shaving	terminal	Rubrum (arm,thigh)	
43/F/14	-	not fair	Cheek,forehead, periauricular	12	sun	-	arm,thigh	
44/M/17	-	not fair	Cheeks	16	-	vellus +	Rubrum (arm)	
45/M/18	-	not fair	Cheeks	15-16	cold	-	Arm	Milia
46/F/17	-	fair	Cheek,ear	9	Heat,sun, cold,wind	-	Arm	
47/M/20	-	not fair	Cheek,ear	16	Emotion, heat, sun	terminal,vellus	Arm,thigh,trunk	

Table 1- Characteristics of patients with erythromelanosis follicularis (continued)

Age(y)/No/Sex	Family history	Phenotype	Site	Age at onset (y)	Aggravating factors	Reduced hair	Keratosis pilaris	Associations
48/F/8	-	fair	Cheek	birth ?	Cold,Heat			
49/F/13	-	fair	Cheek,	12	Sun, heat,wind	-	Trunk,lower limb	
50/M/11	-	fair	Cheek,preauricular	4-5	Exercise,bath, cold,heat, emotion	vellus	Rubrum (arm,trunk, thigh)	
51/M/19	-	not fair	Cheek,eyebrow,ear	childhood	Cold,heat, emotion	-	Rubrum (arm,trunk, thigh)	
52/F/13	-	not fair	Cheek,,ear,	birth ?	Mense,hunger	-	Arm	Milia
53/F/39	-	fair	Cheek,forehead, eyebrow, neck	37	-	terminal eyebrow	Rubrum (limbs)	
54/F/13	-	fair	Cheek,helix,arm	13	Heat	-	Rubrum (arm)	
55/F/14	-		Cheek,ear	12	Bathing, summer	vellus	Thigh, arm	
56/M/17	-	not fair	Cheek,preauricular, temple,neck	14	Bathing	-	Rubrum (arm)	
57/M/16	-	not fair	Cheek,ear,neck	12-13	Exercise, cold	Arm, thigh	Milia	
58/M/17	+ (Brother)	not fair	Cheek,temple, ear	9-10	Exercise, heat,emotion	terminal,vellus ±	Rubrum (arm,thigh)	
59/M/16	+ (2 cousins)	fair	Cheek,ear	12-13	Cold,heat, emotion		Rubrum (arm,thigh)	
60/M/17	-	not fair	Cheek,temple	13-14	Cold,heat exercise	vellus	Arm	