


## CASE REPORT

# Erythromelanosis follicularis faciei et colli with reticulated hyperpigmentation of the extremities

Fahad Mohammad Al-Saif<sup>1</sup> , Abdullah Ali Baqays<sup>2</sup>, Hadeel Fahad AlSaif<sup>3</sup> & Ahmed Abdullah Alhumidi<sup>4</sup>

<sup>1</sup>Department of Dermatology, College of Medicine, King Saud University, Riyadh, Saudi Arabia

<sup>2</sup>Dermatology Resident, Saudi Commission for Health Specialties (SCFHS), Riyadh, Saudi Arabia

<sup>3</sup>Pediatrics Resident, Saudi Commission for Health Specialties (SCFHS), Riyadh, Saudi Arabia

<sup>4</sup>Pathology Department, College of Medicine, King Saud University, Riyadh, Saudi Arabia

### Correspondence

Fahad Mohammad Al-Saif, Department of Dermatology, College of Medicine, King Saud University, P.O. Box 7805, Riyadh 11472, Saudi Arabia. Tel: +966-11-4691426; Fax: +966-11-4691432; E-mails: fsaif1000@hotmail.com and falsaif1@ksu.edu.sa

### Funding Information

No sources of funding were declared for this study.

Received: 31 March 2017; Revised: 4 June 2017; Accepted: 21 June 2017

*Clinical Case Reports* 2017; 5(10): 1576–1579

doi: 10.1002/ccr3.1095

## Introduction

Erythromelanosis follicularis faciei et colli (EFFC) is a rare disease that was originally described by Kitamura et al. [1]. The etiology of this condition is unknown, but an autosomal recessive mode of inheritance, familial cases, and spontaneous mutation have been reported [2, 3]. EFFC was initially reported in young male patients, but a recent study showed nearly equal incidence between males and females [4]. This disease is characterized by the following triad of symptoms: well-demarcated erythema of the face and neck, hyperpigmentation, and follicular plugging. We report the case of a patient with EFFC that was associated with unusual acral reticulated hyperpigmentation.

## Case Report

A 12-year-old boy, born of nonconsanguineous parents after a normal, full-term vaginal delivery, presented with

### Key Clinical Message

Erythromelanosis follicularis faciei et colli (EFFC) is a distinct, rare, and underdiagnosed condition of unknown etiology that is characterized by well-demarcated erythema, hyperpigmentation, and follicular papules. The pigmentation is usually on both maxillary, preauricular regions, and the cheeks. We report a case of a 12-year-old boy with EFFC that was associated with reticulated pigmentation of the extremities.

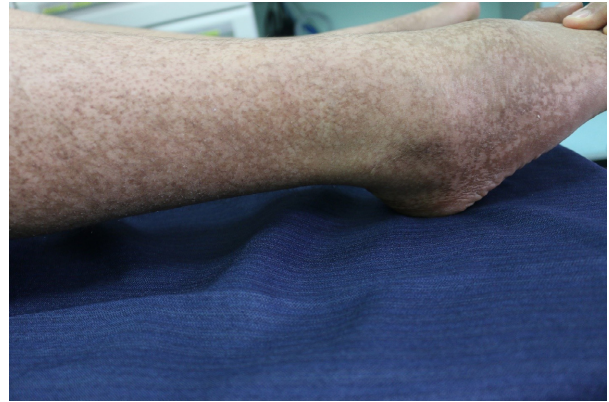
### Keywords

Erythromelanosis, faciei, follicularis, hyperpigmentation, reticulated.

a 5-year history of progressive bilateral and symmetrical patches of reddish-brown pigmentation with tiny papules that begin on preauricular areas and cheeks and gradually spread to the submandibular areas. The child also concurrently developed reticulated pigmentation over the acral part of upper and lower limbs. On examination, there were symmetrical well-demarcated erythematous patches without telangiectasia, and they were studded with tiny follicular papules over his cheeks. Over both upper and lower limbs, there was diffuse dark-brown reticulated pigmentation (Figs 1–4). No palmer pits or hypopigmentations were present. The skin texture of the trunk and limbs was rough with many pale follicular papules. The patient's hair, mucous membranes, and nails were normal, and the results of systemic examination were normal. His family history was negative for EFFC, and the histologic features, although nondiagnostic, correlated well with the clinical features. The microscopic examination of a biopsy taken from the leg skin showed orthokeratosis,



**Figure 1.** Erythematous patches over preauricular area and cheek.



**Figure 4.** Reticulate hyperpigmented macules present over feet.

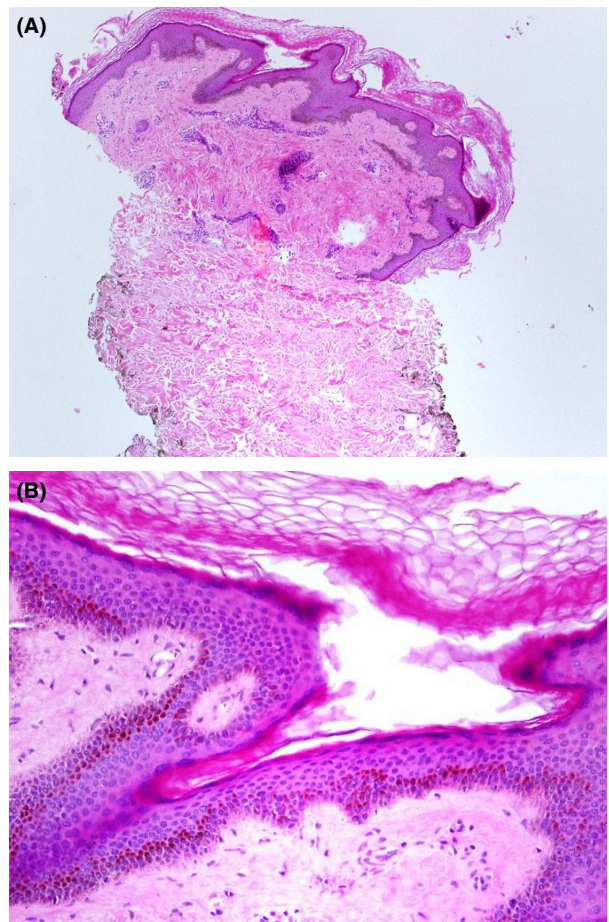


**Figure 2.** Bilateral reticulated hyperpigmentation over hands.



**Figure 3.** Reticulate hyperpigmented macules present over hands.

follicular keratin plugging, and basal layer hyperpigmentation (Fig. 5A and B). Another skin punch biopsy from the face revealed mild hyperkeratosis, basal layer hyperpigmentation with perifolliculitis, and scattered necrotic keratinocytes in the hair follicle epithelium (Fig. 6A and B).

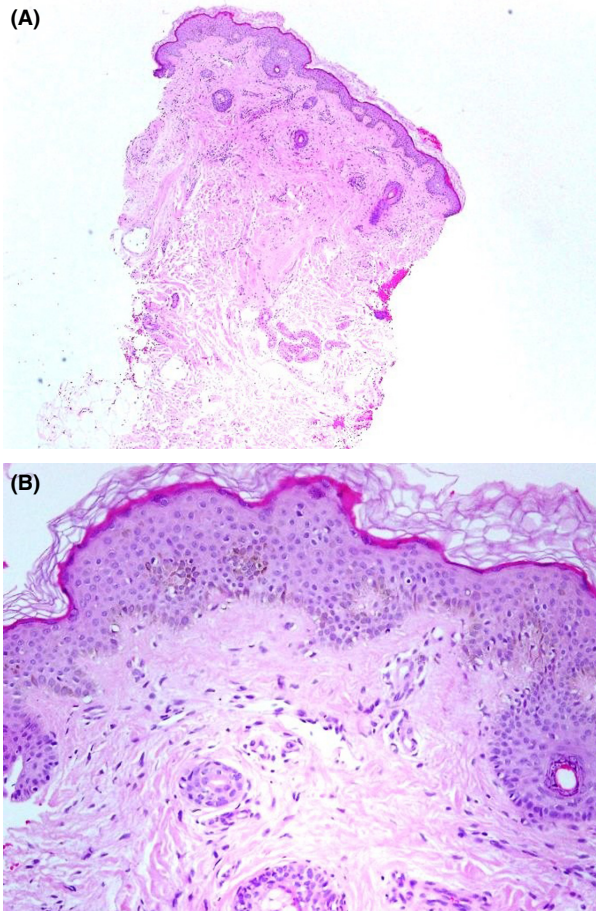


**Figure 5.** (A) Punch skin biopsy from leg (H/E stain X100). (B) High power (H/E stain X400).

## Discussion

Erythromelanosus follicularis faciei et colli is a rare disease of unknown etiology, but an autosomal recessive pattern of inheritance and familial cases [2–5], and spontaneous





**Figure 6.** (A) Punch skin biopsy from face (H/E stain X100). (B) High power (H/E stain X400).

mutation and chromosomal instability have been reported in some cases [5, 6]. This condition was initially reported in young male patients, but a recent study showed nearly equal incidence between males and females [4]. The mean age of onset for the disease was 12.1 years [7].

Erythromelanosis follicularis faciei et colli is characterized by well-demarcated reddish-brown patches in the preauricular and maxillary areas, and it is associated with keratosis pilaris either over affected areas or over classical sites [3, 4]. Telangiectasia can be observed in some patients especially over a sun-exposed area [8]. Pigmentation in EFFC usually occurs in association with erythema and as a result of exposure to sunlight in the predisposed skin [3]. Our patient has diffuse dark-brown reticulated pigmentation over acral areas, and this type of pigmentation has not been previously reported.

Histopathology of EFFC usually reveals follicular plugging, hyperkeratosis, increased pigmentation in the basal membrane, perivascular and periadnexal inflammatory infiltrate, follicular dilatation, and dilated blood vessels in

the upper dermis. However, some studies found a correlation between disease severity and basal layer pigmentation in which our case has. [4, 9].

Differential diagnosis includes keratosis pilaris rubra, poikiloderma of Civatte, and Atrophoderma vermiculatum. EFFC has been considered to be a variant of keratosis pilaris in terms of follicular keratin plugging although in skin type I patients keratosis pilaris rubra presents with erythema of the cheeks, but EFFC is associated with well-defined patches of erythema and pigmentation. Poikiloderma of Civatte is seen in sides of the neck in middle-aged women. Atrophoderma vermiculatum has honeycombed atrophy that affecting the cheeks, and no erythema or pigmentations are usually noticed [10, 11].

There is no satisfactory treatment for EFFC, and most of the lesions usually recur shortly after treatment is discontinued. Emollients, keratolytic, tacalcitol ointment, metronidazole, hydroquinone 4%, chemical peel, and laser systems have been used with variable results [2, 7, 12–14].

## Conclusion

We report this case for its rarity and to highlight the association of EFFC with reticulated pigmentation of the extremities.

## Conflict of Interest

None declared.

## Authorship

FMA: conceived the work and reviewed the manuscript. AAB: wrote the manuscript. HFA: wrote the manuscript. AAA: wrote the histopathology section of the manuscript.

## References

1. Kitamura, K., H. Kato, Y. Mishima, and S. Sonoda. 1960. Erythromelanosis follicularis faciei. *Hautarzt* 11:391–393.
2. Yanez, S., J. A. Velasco, and M. P. Gonzalez. 1993. Familial erythromelanosis follicularis et colli—an autosomal recessive mode of inheritance. *Clin. Exp. Dermatol.* 18:283–285.
3. Augustine, M., and E. Jayaseelan. 2008. Erythromelanosis follicularis faciei et colli: relationship with keratosis pilaris. *Indian J. Dermatol. Venereol. Leprol.* 74:47–49.
4. Rather, S., A. Yaseen, and M. Mukhija. 2016. Erythromelanosis follicularis faciei et colli - A cross-sectional, descriptive study. *Indian J. Dermatol.* 61:308–313.
5. Tüzün, Y., R. Wolf, B. Tüzün, M. Ozdemir, C. Demirkesen, A. Deviren, and A. Kotogyan. Familial erythromelanosis follicularis and chromosomal instability. *J Eur Acad Dermatol Venereol.* 15:150–152.

6. Acay, M. C. 1993. Erythromelanosus follicularis faciei et colli: a genetic disorder? *Int. J. Dermatol.* 32:542.
7. Jang, H. S., C. W. Lee, J. H. Cha, S. J. Kim, C. K. Oh, and K. S. Kwon. 2001. Clinicopathological study of 12 cases of erythromelanosus follicularis faciei et colli. *Korean J. Dermatol.* 39:574–580.
8. Warren, F. M., and L. S. Davis. 1995. Erythromelanosus follicularis faciei et colli in women. *J. Am. Acad. Dermatol.* 32(5Pt2):863–866.
9. Kim, M. G., S. J. Hong, S. J. Son, H. J. Song, I. H. Kim, C. H. Ch, and J. S. Moon, 2001. Quantitative histopathologic findings of erythromelanosus follicularis faciei et colli. *J. Cutan. Pathol.* 28:160–164.
10. Augustine, M., and E. Jayaseelan. 2008. Erythromelanosus follicularis faciei et colli: Relationship with keratosis pilaris. *Indian J Dermatol Venereol Leprol* 74:47–49.
11. Al Hawsawi, K., O. Aljuhani, G. Niaz, H. Fallatah, and A. Alhawsawi. 2015. Erythromelanosus follicularis faciei: A case report and review of the literature. *Case Rep Dermatol* 7:335–339.
12. Kim, W. J., M. Song, H. C. Ko, B. S. Kim, and M. B. Kim. 2012. Topical tacalcitol ointment can be a good therapeutic choice in erythromelanosus follicularis faciei et colli. *J Am Acad Dermatol.* 67:320–321.
13. Li, Y. H., X. Zhu, J. Z. Chen, Y. Wu, H. C. Wei, X. H. Gao, and H. D. Chen. 2010. Treatment of erythromelanosus follicularis faciei et colli using a dual-wavelength laser system: a split-face treatment. *Dermatol Surg.* 36:1344–1347.
14. Kurita, M., A. Momosawa, M. Ozaki, I. Ban, and K. Harii. 2006. Long-pulsed dye laser for the treatment of erythromelanosus follicularis faciei: report of two clinical cases. *Dermatol Surg.* 32:1414–1417.