

Erythromelanosus Follicularis Faciei et Colli: Case Reports of Bilateral Lesions in 2 Females

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Erythromelanosus follicularis faciei et colli (EFFC) is a rare disorder of unknown etiology characterized by the clinical triad of well-demarcated erythema, hyperpigmentation, and follicular plugging on the face. For many years, it was thought to be a disease occurring in males with ethnical cutaneous pigmentation. There are less than 50 cases reported in the literature, most of which are males. We describe bilateral EFFC in 2 females.

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Case Reports

Patient 1—A 24-year-old woman presented with hyperpigmented plaques on both cheeks and both sides of the neck (Figure 1). The lesions were well-demarcated and had a rough surface on palpation due

to the presence of multiple follicular papules. The patient reported that the exanthem presented one year prior and was asymptomatic. She also reported no previous dermatologic diseases, and no relatives had similar lesions. Histologic evaluation revealed marked hyperkeratosis with follicular plugging and slight dilatation of the pilosebaceous follicles.



Figure 1. Hyperpigmented plaques on the cheeks and both sides of the neck (A). The lesions were well-demarcated with a rough surface on palpation due to the presence of multiple follicular papules (B).

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The authors report no conflict of interest.

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Figure 2. Well-demarcated erythematous plaques on both cheeks and the jaw (A). The lesions consisted of numerous follicular micropapules (B).

Patient 2—A 14-year-old adolescent girl presented with well-demarcated erythematous plaques on both cheeks and the jaw that consisted of numerous follicular micropapules (Figure 2). The first lesions appeared 4 years prior. Other than aesthetic discomfort, she reported no pruritus or other symptoms. She had no medical or family history.

Both Patients—Histologic findings were the same in both patients. The main feature was the presence of follicular plugging, sparse lymphocytic perifollicular infiltration, and melanophages in the upper dermis (Figure 3). Both patients were treated with topical retinoids. After 3 months of treatment, there was a slight improvement in the roughness of the follicular papules; however, the hyperpigmentation was persistent.

Comment

EFFC was first described in Japanese youths by Kitamura et al² in 1960, and for many years, it was thought to be a disease occurring in males with ethnical cutaneous pigmentation. The first case of unilateral distribution in a white girl was reported in 1991.^{3,4}

The main clinical features of EFFC are the presence of reddish brown, irregularly marginated pigmentation with fine telangiectatic vessels and pale follicular papules symmetrically distributed on the preauricular cheeks. The lesions spread to the temples and lateral aspects of the neck. Patchy alopecia of vellus hair also may be present.^{5,6} Similar lesions rarely can occur on the auricles and eyebrows.⁷ The disease is asymptomatic.

The histologic features, although nondiagnostic, correlate well with the clinical features. Acanthosis, hyperkeratosis with follicular plugging, a slight lymphocytic infiltrate, and dilatation of the dermal vessels have been reported.⁸ The possible relationship to

keratosis pilaris rouge is discussed⁶ because keratosis pilaris of the arms has been described.⁹

The differential diagnosis in bilateral forms of EFFC includes keratosis pilaris atrophicans faciei and its variants (ie, ulerythema ophryogenes, atrophoderma vermiculatum), lichen spinulosus, and poikiloderma of Civatte.¹⁰ Keratosis pilaris atrophicans faciei and its disorders are associated with scarring or distinctive ice pick depressions. In lichen spinulosus, the face usually is spared and the typical lesions are 2- to 6-cm patches composed of grouped skin-colored keratotic follicular papules with a central keratotic spine. Patches may be surrounded by faint erythema. Poikiloderma of Civatte is a phototoxic reaction that occurs in adults. It is characterized by areas of erythema as a reflection of multiple interfollicular telangiectases that spare the thin rim of skin around each follicle. Unilateral forms of EFFC must be differentiated from fixed pigmented erythema and berloque dermatitis and rarely from Ota nevus and Becker nevus.¹⁰

Erythrosis pigmentosa peribuccalis (also known as Brocq or erythrosis pigmentosa mediofacialis) is a disorder of the mediofacial area and is common in women.¹¹ The disorder shows histologic signs of abnormal follicular keratinization with telangiectasis and round cell infiltrate. An increase in the level of melanin has been seen in some patients.¹¹ Because these findings also are present in EFFC, Juhlin and Alkemade¹¹ suggested that the 2 conditions are the same disease and proposed the neutral term *erythrosis pigmentosa faciei et colli*.

Electron microscopic examination results reveal abnormalities consisting of a homogeneous mass of pigment in the center of the melanosome and a stippled appearance to the periphery of the organelle. Many of the melanosomes appear to be abnormally large.¹²

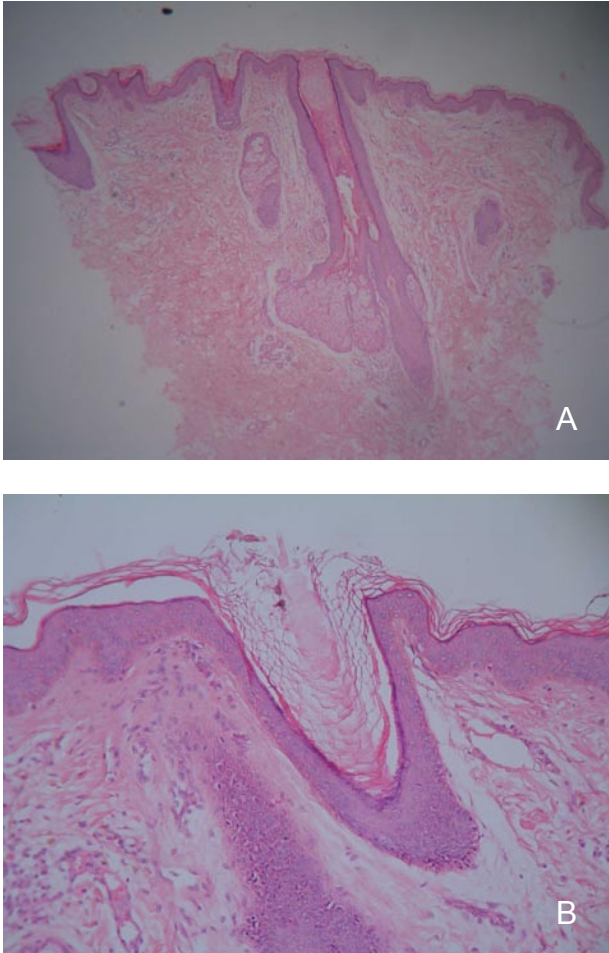


Figure 3. Follicular plugging, sparse lymphocytic perifollicular infiltration, and melanophages in the upper dermis (H&E, original magnification $\times 40$)(A). Follicular plugging (H&E, original magnification $\times 100$)(B).

In a quantitative clinicopathologic study by Kim et al,¹³ the correlation between the histologic findings and clinical grading in EFFC was assessed. Although follicular plugging is a characteristic clinical feature of EFFC, enlargement of hair follicles and hair shafts were not characteristic pathologic changes of EFFC in this study. The pigmentation of the basal layer and the percentage area of the inner spaces of the superficial dermal blood vessels reflected the clinical severity more than the other parameters.¹³

There are many theories about the pathogenesis of EFFC. In 1993, Yanez et al¹⁴ proposed an autosomal recessive mode of inheritance. Several years later, Tuzun et al¹⁵ thought that EFFC might be a polyetiologic disorder (eg, familial, environmental) or that it might be considered one of the chromosomal instability syndromes.

Various keratolytic agents containing urea, ammonium lactate, or isotretinoin, as well as glycolic or salicylic acid peels, have been topically used

in the treatment of EFFC. Limited courses of oral isotretinoin can be tried in severe cases.

Although EFFC was reported as a rare disorder for decades, we now believe it is not so rare. Clearly, more data are needed to define the etiology and epidemiology of EFFC and to help find more effective therapeutic modalities.

REFERENCES

1. Ermertcan AT, Ozturkcan S, Sahin MT, et al. Erythromelanosis follicularis faciei et colli associated with keratosis pilaris in two brothers. *Pediatr Dermatol.* 2006;23:31-34.
2. Kitamura K, Kato H, Mishima Y, et al. Erythromelanosis follicularis faciei [in German]. *Hautarzt.* 1960;11:391-393.
3. McGillis ST, Tuthill RJ, Ratz JL, et al. Unilateral erythromelanosis follicularis faciei et colli in a young girl. *J Am Acad Dermatol.* 1991;25(2 pt 2):430-432.
4. Warren FM, Davis LS. Erythromelanosis follicularis faciei in women. *J Am Acad Dermatol.* 1995;32(5 pt 2):863-866.
5. Andersen BL. Erythromelanosis follicularis faciei et colli. *Br J Dermatol.* 1980;102:323-325.
6. Watt TL, Kaiser JS. Erythromelanosis follicularis faciei et colli. a case report. *J Am Acad Dermatol.* 1981;5:533-534.
7. Seki T, Takahashi S, Morohashi M. A case of erythromelanosis follicularis faciei with a unique distribution. *J Dermatol.* 1991;18:167-170.
8. Mishima Y, Rudner E. Erythromelanosis follicularis faciei et colli. *Dermatologica.* 1966;132:269-287.
9. Sodaify M, Baghestani S, Handjani F, et al. Erythromelanosis follicularis faciei et colli. *Int J Dermatol.* 1994;33:643-644.
10. Stephan F, Ayoub N, Klein-Tomb L, et al. Erythromelanosis follicularis faciei et colli. *Ann Dermatol Venereol.* 2002;129:63-65.
11. Juhlin L, Alkemade H. Erythrosis pigmentosa mediofacialis (Brocq) and erythromelanosis follicularis faciei et colli in the same patient. *Acta Derm Venereol.* 1999;79:65-66.
12. Borkovic SP, Schwartz RA, McNutt NS. Unilateral erythromelanosis follicularis faciei et colli. *Cutis.* 1984;33:163-170.
13. Kim MG, Hong SJ, Son SJ, et al. Quantitative histopathologic findings of erythromelanosis follicularis faciei et colli. *J Cutan Pathol.* 2001;28:160-164.
14. Yanez S, Velasco JA, Gonzalez MP. Familial erythromelanosis follicularis faciei et colli—an autosomal recessive mode of inheritance. *Clin Exp Dermatol.* 1993;18:283-285.
15. Tuzun Y, Wolf R, Tuzun B, et al. Familial erythromelanosis follicularis and chromosomal instability. *J Eur Acad Dermatol Venereol.* 2000;15:150-152.