Clinicopathological and dermoscopic correlation of Erythromelanosis follicularis faciei et coli in skin of color

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Introduction

Erythromelanosis follicularis faciei et coli (EFFC) is a rare disorder of follicular keratinization of unknown etiology, which is first described in 1960 by Kitamura. The disease is characterized by the triad of hyper-pigmentation, erythema, and follicular papules[1]. As EFFC can be easily mistaken for other similar pigmentary, hyperkeratotic disorders, clinico pathological correlation with dermoscopic appearance are important in preventing diagnostic errors.

Herein we present correlation of clinicopathologic and dermoscopic appearance of EFFC in two Asian individuals.

Case report

First case

A 16-year-old boy presented with asymptomatic hyperpigmentation over bilateral cheeks and examination revealed Ill-defined erythematous to hyperpigmented patches studded with tiny follicular papules over bilateral cheeks and lateral neck (Figure 1). Hyperlinear palms were noted. However, keratosis pilaris was not evident. Rest of the cutaneous examination was normal.

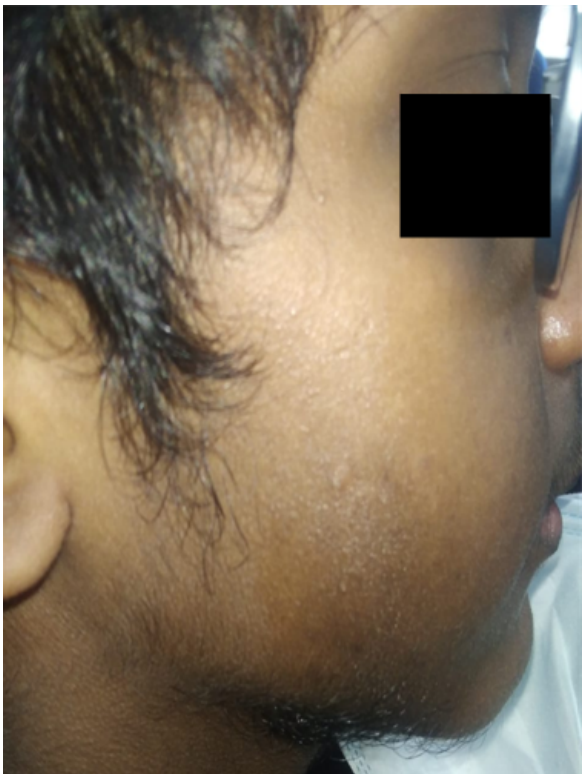

Figure 1.

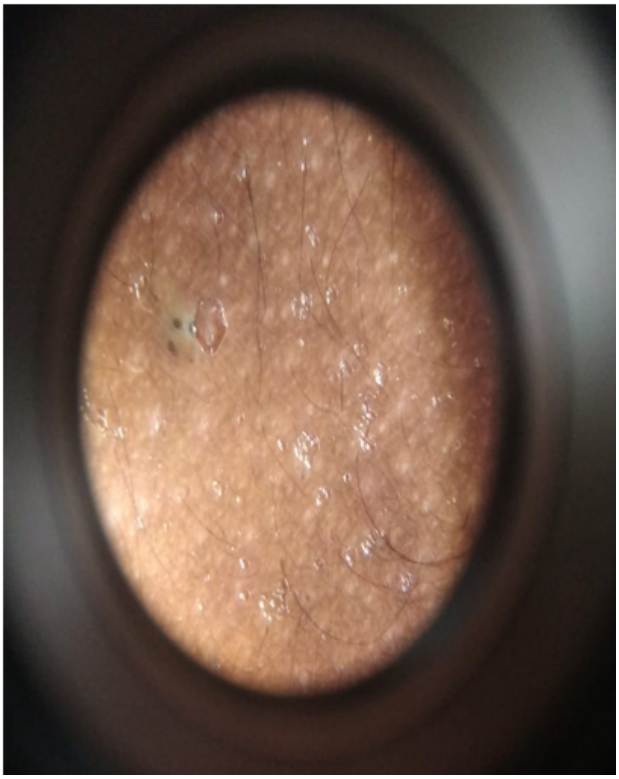

Figure 2.

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Dermoscopy of the lesion showed multiple whitish follicular keratotic plugs with perifollicular whitish scales over reddish brown background (Figure 2). However perifollicular or interfollicular blue grey peppering were not apparent.

Histology revealed orthokeratotic epidermis with a few keratin plugged follicles and dermal adnexa associated with these follicles appeared atrophied. Focal hyperpigmentation of the basal layer, perivascular and peri adnexal lymphocytic infiltrate were noted.

Clinicopathologic and dermoscopic evidence lead to the diagnosis of EFFC.

Second case

22-year-old young male presented with disfiguring asymptomatic hyper pigmented patches over bilateral cheeks, forehead, and neck of 3 years duration (Figure 3). Examination revealed Ill-defined hyperpigmented patches over either side of the face and neck with pigmented follicular papules. No pigmentation over rest of his body including the mucous membranes. hair and nail examination were normal no keratosis pilaris like lesions were noted. Differential diagnosis for the clinical appearance were EFFC and follicular lichen planus.

Dermoscopic examination revealed whitish follicular plugs with perifollicular blue grey peppering and follicular whitish scaling which supporting the diagnosis of EFFC (Figure 4).

Histology revealed mildly orthokeratotic epidermis, follicular hyperkeratosis, increased basal layer pigmentation and peri adnexal lymphocytic infiltrate, compatible with the clinical diagnosis of EFFC.

Discussion

EFFC is an underdiagnosed follicular hyperkeratotic disorder of unknown etiology which typically involves lateral cheeks and lateral aspects of the neck. Clinical diagnosis is based on the presence of triad of erythema, hyperpigmentation, and follicular papules[1]. However in skin type 1 patients there may be only erythema and it is more commonly seen in patients with Asian ancestry.

EFFC can have an overlap with keratosis pilaris rubra, keratosis pilaris and erythrosis pigmentosa peribuccalis[2].

Diagnosis of EFFC can be often get delayed due to confusion with other facial pigmentary disorders like lichen planus pigmentosus, poikiloderma of Cuvette, keratosis pilaris rubra atrophicans, follicular lichen planus and demodiconiosis etc.

This shows the importance of clinicopathologic and dermoscopic correlation in early diagnosis of this underdiagnosed facial pigmentary disorder.

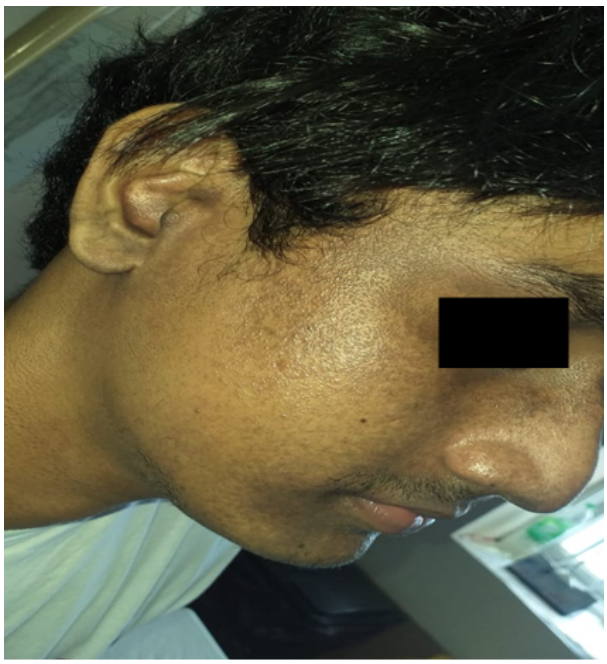

Figure 3.

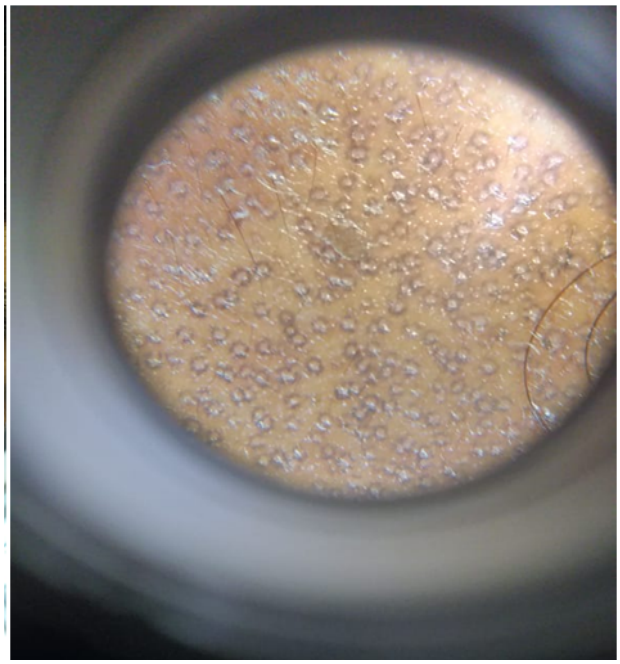

Figure 4.

Histology of EFFC shows follicular hyperkeratosis, follicular plugging, increased pigmentation in basal layer, perivascular and peri adnexal inflammatory infiltrate, follicular dilatation, and dilatation of blood vessels in the upper dermis[1,2].

Dermoscopic examination of EFFC shows follicular plugging, white scaling, perifollicular, interfollicular blue grey peppering on the reddish-brown background[2,3].

Dermoscopic findings seen in our patients can be related to peculiar histologic features of the condition. E.g. follicular plugging to the hyperkeratosis of the hair follicle, scaling to the orthokeratosis, blue grey peppering to the pigment incontinence and dermal melanophages and reddish brown pigmentation to the hyperpigmentation of the basal layer and vasodilation of the superficial basal vessels etc.

As there is strong correlation between clinicopathologic and dermoscopic examination we can use dermoscopic examination as a noninvasive bed site tool of investigation in diagnosing this mimicker.

References

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