Erythromelanosis follicularis faciei et colli with reticulated hyperpigmentation of the extremities

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Abstract

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 the case of a 12-year-old boy with EFFC that was associated with reticulated pigmentation of the extremities.

Introduction

Erythromelanosis follicularis faciei et colli (EFFC) is a rare disease that was originally described by Kitamura et al. [1]. EFFC was initially reported in young male patients. 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 non-consanguineous parents after a normal, full-term vaginal delivery, presented with 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 reticulatedpigmentation 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 checks. Over both upper and lower limbs, there was diffuse dark-brown reticulated pigmentation (Figures 1, 2, 3,4).

The microscopic examination of a biopsy taken from the leg skin showed orthokeratosis, follicular keratin plugging, and basal layer hyperpigmentation (Figure 5A,B). Another skin punch biopsy from the face revealed mild hyperkeratosis, basal layer hyperpigmentation with perifolliculaitis, and scattered necrotic keratinocytes in the hair follicle epithelium (Figure 6A,B).

Discussion

EFFC is a rare disease of unknown etiology, but an autosomal recessive pattern of inheritance and familial cases [2], and spontaneous mutation and chromosomal instability have been reported in some cases [3]. EFFC 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 Pigmentation in EFFC usually occurs in association with erythema and as a result of exposure to sunlight in the predisposed skin [4]. Our patient has diffuse dark-brown reticulated pigmentation over acral areas and this type of pigmentation not been previously reported.

Conclusion

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

Figures



Figure 1



Figure 2

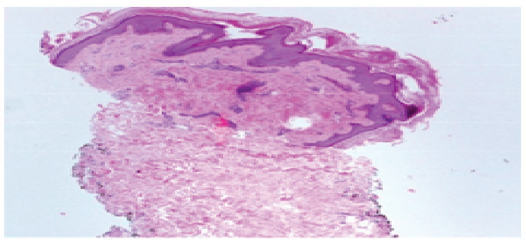


Figure 3



Figure 4

Figure 5(A,B) Leg



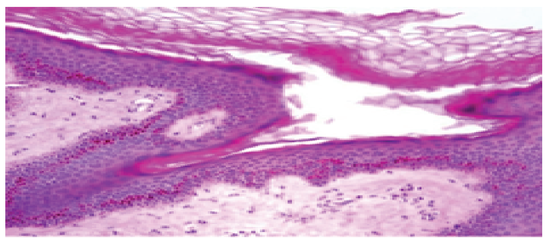
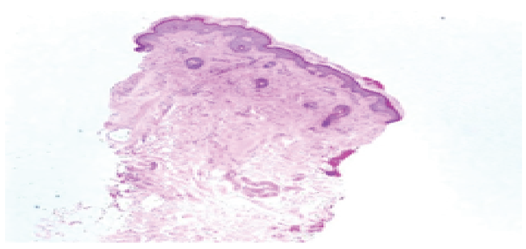
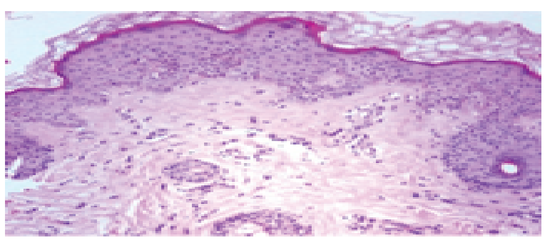


Figure 6 (A,B) Face





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