Co-existent Erythromelanosis Follicularis Faciei et Colli and Erythroses Pigmentosa Mediofacialis in a Patient of Generalized Keratosis Pilaris - A Rare Report in a Young Female

Sir,
Erythromelanosis Follicularis Faciei et Colli (EFFC) and Erythroses Pigmentosa Mediofacialis (EPM) are considered to be the disorders of abnormal follicular keratinization characterized by a triad of hyperpigmentation, erythema, and keratotic follicular papules. Herein, we report a young female with a features of both in association with generalized keratosis pilaris which is a rare co-occurrence, diagnosis of which was confirmed by dermoscopy and histopathology.

A 16-year-old female reported with multiple asymptomatic elevated lesions associated with darkening of skin which appeared first over the sides of cheeks, upper part of ear and gradually extended over the neck since 4 years. Similar lesions were also seen over central part of face since 2 years. Patient denied history of topical application and photosensitivity. Her past, personal, and family history was insignificant. Cutaneous examination revealed bilaterally symmetrical follicular micropapules associated with well-demarcated erythema and hyperpigmentation over the pre-auricular region, pinnae of ear and neck [Figure 1a and b]. Forehead, perinasal, peribuccal region, and chin also showed lesions of similar morphology [Figure 2]. In addition, patient had keratotic papules over bilateral extremities, back, chest, abdomen, and thighs suggestive of generalized keratosis pilaris [Figure 3a-d]. On diascopy of facial lesion erythema disappeared but pigmentation persisted. Dermoscopy using 3 Gen Dermlite DL4 (CA, USA) 10 × polarized mode, from cheeks revealed multiple small whitish rounds with keratotic follicular plugs surrounded by gray spots in a background of brownish discoloration suggestive of EFFC [Figure 4a]. Forehead lesion on dermoscopy showed numerous dilated follicles with keratotic plugging, perifollicular erythema, and scaling suggestive of EPM [Figure 4b]. Dermoscopy of the lesion from upper arm showed coiled hair, keratotic plugs, perifollicular erythema, and scaling suggestive of keratosis pilaris [Figure 4c]. Skin biopsy (one from cheek and another from forehead) with H and E staining revealed follicular hyperkeratosis with acanthosis with increased pigmentation in the basal layer. Dermis showed superficial perivascular lymphocytic infiltrate [Figure 5a and b]. Based on clinical, dermoscopy and histopathology a final diagnosis of coexistent EFFC and EPM in a patient of generalized keratosis pilaris was made. The patient was counselled and started on topical tretinoin 0.025% cream and sunscreen for face and an emollient for the body. After 8 weeks of treatment patient showed only mild improvement and is under regular follow-up.

EFFC is an unfamiliar syndrome described by Kitamura, in 1960, with well-demarcated erythema, telangiectasia, hyperpigmentation, and follicular papules primarily on lateral aspect of the face, with an extension on to the lateral half of the neck, thus the name et colli (in Latin “et” = “and”, “colli” = “neck”). It affects patients of all ethnic group but majorly seen in Asian or Middle Eastern men.[1] The male to female ratio is observed to be 2:1.[2] However, it is increasingly reported in female patients as noticed in our case.[1] EPM as described by Brocq, is erythematous to hyperpigmented, rough skin over midline of the face. It is synonymously

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called as erythrose pigmentaire faciale, dermatose pigmentée mé diofaciale, erythrosis pigmentosa peribuccalis, erythrosis pigmentata faciei, erythromelanosis follicularis faciei and erythrosis pigmentosa faciei colli.\(^4\) The etiology of EFFC is multifactorial and includes genetic (autosomal recessive) and hereditary components.\(^5\) Previous data suggest spontaneous mutation in EFFC.\(^6\) Tuzun and Wolf et al., postulated that EFFC to be a chromosomal instability syndrome.\(^7\) It is hypothesized that various environmental factors such as cosmetics, ultraviolet radiation, and
topical formulations can cause hyperpigmentation over the period of years. Dermoscopy of EFFC shows multiple round whitish areas with follicular plugs, some shows central hair, surrounded by blue gray spots or peppering in a reddish-brown background. Dermoscopy of EPM includes follicular dilatation with yellowish keratotic plugs, perifollicular erythema, and scaling surrounded by slate-gray globules. Histology of EFFC and EPM is similar and shows follicular hyperkeratosis, increased basal layer pigmentation, and incontinence of melanin pigment with dermal melanophages. Dermis shows perivascular and periadnexal lymphocytic infiltrate with vasodilatation. EFFC has been found to be associated with keratosis pilaris of different body parts which postulated that EFFC is a variant of keratosis pilaris. Juhlin et al. has reported a single case of EFFC and EPM occurring in a 34-year-old female postulating that the two conditions are same. Our case is unique since she had concurrent EFFC, EPM, and generalized KP. It can be stated that both EFFC and EPM are etiologically same conditions manifesting at different sites. Dermoscopy plays an important beside tool to identify these entities. Differential diagnosis of EFFC and EPM includes ulerythema ophryogenes, atrophoderma vermiculatum, Riehl’s melanosi, poikiloderma of Civatte, tricostasis spinulosa, lichen spinulosus. Treatment of EFFC and EPM is same but unsatisfactory. Various topical keratolytics such as tretinoin, glycolic acid, salicylic acid (20–30%), urea, ammonium lactate, vitamin C can be...
Table 1: Contd...

| Diagnosis | Site | Clinical Features | Histopathology | Dermoscopy |
|-----------|------|------------------|----------------|-----------|
| 5) Poikiloderma of Civate<sup>1<sup>8</sup>,<sup>19</sup> | Lateral aspect neck | Triad of reticulate hyper-pigmentation, atrophy, and telangiectasia. Telangiectasias are interfollicular with sparing of thin rim of skin around each hair follicle | Epidermal, papillary dermal atrophy with loss of papillae, hyaline masses at the dermoepidermal junction and disappearance of the elastic network in the upper dermis. In fully developed lesions cavities in the connective tissue filled with epidermal cells, pigment and lymphocytes may be seen. | Dotted/globular vessels and linear irregular vessels, giving rise to a “spaghetti and meatballs” appearance along with perifollicular whitish areas. Additional findings include follicular keratotic plugs and delicate reticular or structureless brownish areas. |
| 6) Lichen spinulosus<sup>17</sup> | Extensor aspects of extremities, neck, abdomen, thigh, buttocks, popliteal fossa | Multiple grouped follicular papules with projecting keratin spine without erythema | Follicular hyperkeratosis and keratotic plugging of infundibulum with perifollicular and perivascular mononuclear infiltrate | Follicular papules with translucent spines |
| 7) Riehl’s melanosis<sup>20</sup> | Cheeks, temporal area, forehead | Patches of hyperpigmentation over these areas, sometimes associated with pruritus, prolong use of cosmeceuticals can be elicited | Dilated infundibulum with inflammatory cell infiltrate, liquefaction of basal cells, pigment incontinence, dilated vessels with perivascular inflammatory infiltration. | Sparse scaling, pseudonetwork, grey dots/granules, follicular keratotic plugs, perifollicular whitish halo, telangiectatic vessels |
| 8) Trichostasis spinulosa<sup>17</sup> | Nose, abdomen, back, rarely diffuse | Multiple tiny black dots on face especially over tips of nose, black dots represent vellus hair, without erythema | Follicular hyperkeratosis, Dilated infundibulum with numerous pigmented vellus hairs and keratin material | Tuft of short vellus hairs emerging from the same follicular opening. Keratotic plugs within dilated follicle |
| 9) Comedonal acne<sup>21</sup> | Cheeks, forehead, neck, chest, back | Tiny papules sometimes black (open comedons) containing cellular debris and sometimes white (closed comedons) with intact overlying epithelium | Epidermis shows follicular dilatation with keratinous material, mild perivascular lymphocytic infiltrate in dermis | Numerous homogenous areas, light & dark-brown sometimes black in color depending on the type of acne open or closed comedones, predominantly circular and situated superficially. |

Conflicts of interest

There are no conflicts of interest.

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