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Erythromelanosus follicularis faciei et colli: Report of a case and literature review

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Abstract

Erythromelanosus follicularis faciei et colli (EFFC), characterized by hyperpigmentation, erythema and follicular papules on face and neck, is a rare disease and not previously reported from Nepal. We report a case of 13 years old boy who presented with classical triad of EFFC and present a literature review on this condition.

Case report

A 13 years old boy presented with redness, pigmentation and raised rough lesions on the face since early childhood. He recognized an increase in the erythema and burning sensation on exposure to light. He denied any similar history in his family. Physical examination revealed follicular papules, erythema and hyperpigmentation present on the malar area, which was bilaterally symmetrical extending to the pre-auricular area, ear lobules and neck as shown in **Figure 1** and **Figure 2**. There was no atrophy, scarring or alopecia. Systemic examination failed to reveal any significant abnormality. Patient was put on topical tretinoin 0.025% cream, emollients and sunscreen. Patient denied for skin biopsy. On follow up over 1 month his symptoms remain static.



Fig 1: follicular papules, erythema and hyperpigmentation seen in malar area, preauricular and ear lobules.



Fig 2: Extension of the follicular papules, erythema and hyperpigmentation on the neck

Discussion

Erythromelanosus follicularis faciei et colli (EFFC) is a disorder of unknown etiology which classically presents with a triad of hyperpigmentation, erythema (with or without telangiectasia) and follicular papules on the pre-auricular and cheek areas. Our case is also a classic presentation of EFFC. Initially coined by Kitamura in 1960, EFFC is a rare disease and only 55 cases are reported in the literature. Common age of presentation is adolescence and preferably seen in males [1,2,3]. EFFC also affects

children and young adults. There have been many reports of EFF in women since last two decades [4,5]. Bilateral distribution is the main characteristic but unilateral cases were described [1]. Clinical presentations of most reported cases are similar. However, there are variations in symptoms and seasonal influences [4,6]. Our case also had exaggeration of symptoms on photo exposure. EFFC emerge sporadically, however, there are reports of cases people from the same family. The disease may have an autosomal recessive mode of inheritance as reported by Yanez et al [7,8]. EFFC has recently been considered to be a poly-etiological disorder with the possibility of a chromosomal instability syndrome [4]. Histopathology though not diagnostic, shows follicular plugging, hyperkeratosis, increase pigmentation in the basal membrane, perivascular and periadnexal inflammatory infiltrate and follicular dilatation [1]. Differential diagnoses include athrophoderma vermiculatum, ulerythema ophryogenes and poikiloderma of Civatte. Keratosis pilaris is known to be associated with EFFC [4,9]. Treatment so far is unsatisfactory. Topicals like ammonium lactate 12%, tretinoin cream (0,050,1%), salicylic acid 2%, metronidazole and combinations with hydroquinone 4% has been tried, so is chemical Peel with salicylic acid (30%) [1,5,10]. In severe cases oral isotretinoin is used intermittently. Pulsed Dyed Laser of 595nm is a newer therapeutic option to attenuate hyperpigmentation and erythema [1].

Conclusion

Erythromelanosis follicularis faciei et colli (EFFC), is a disorder of unknown etiology classically present with a triad of hyperpigmentation, erythema (with or without telangiectasia) and follicular papules on face and neck. Treatment till date is not satisfactory. Rarity of this condition demands more case description to characterize the disease. We report this classic case for the first time in Nepal.

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