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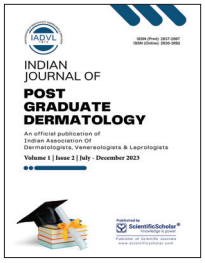

Case Report

Childhood Onset Erythrokeratoderma En Cocardes Treated with Oral Acitretin

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ABSTRACT

Erythrokeratoderma are genodermatose of keratinisation which usually presents in infancy. Erythrokeratoderma en cocardes is a rare variant of erythrokeratoderma which presents as polycyclic hyperkeratotic plaques with a peripheral desquamation, forming a concentric appearance termed 'en cocarde'. Here, we report a case of erythrokeratoderma en cocardes with onset in childhood showing excellent response after 2 months of acitretin therapy.

Keywords: Erythrokeratoderma variabilis, Erythrokeratoderma en cocardes, Acitretin

INTRODUCTION

Erythrokeratoderma en cocardes, first reported by Degos in 1947, is a rare subtype of erythrokeratoderma – a group of hereditary cornification disorders which usually present in the 1st year of life. Here, we report a case of erythrokeratoderma en cocardes with onset in childhood which was successfully treated with acitretin.

CASE REPORT

A 6-year-old boy born out of a non-consanguineous marriage presented with multiple hyperpigmented scaly lesions initially developing over the cubital and popliteal flexures which, then, progressed to involve the axillae, arms, gluteal region and thighs over the course of 8 months. Lesions were non-pruritic and though few lesions would resolve spontaneously; new lesions would occur over adjacent sites. There was a history of winter aggravation of lesions. He had been treated initially with both topical and systemic antifungals with no improvement and was only on emollients at the time of presentation. There was no history of atopy and other systemic symptoms. Developmental milestones were appropriate for age. There was no history of similar skin lesions in the family. On dermatological examination, we found multiple ill-defined scaly plaques with irregular, concentric and polycyclic borders over the lower chest, axillae, medial portion of the arms, abdomen and thighs [Figure 1a-c]. There were multiple ill-defined annular scaly plaques arranged as concentric rings involving the popliteal fossae as well [Figure 2a]. Palms and soles were spared. Hair, nails and teeth were normal. Hair microscopy did not show any hair shaft defects. Systemic examination revealed no abnormalities. Routine blood parameters were within normal limits. Potassium hydroxide (KOH)

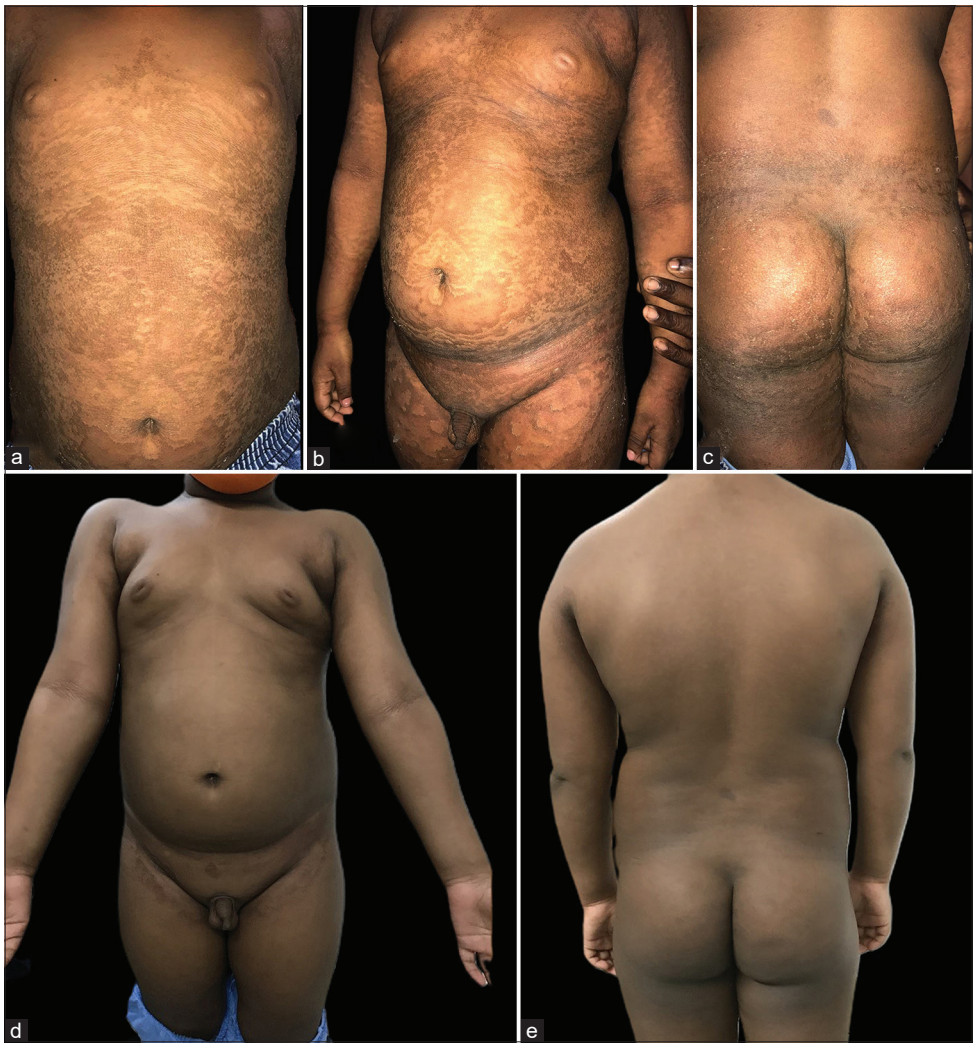

Figure 1: Concentric scaly plaques with irregular and polycyclic borders over (a) the lower chest and abdomen; (b) trunk, axillae, medial portion of the arms, abdomen and thighs; (c) the gluteal region and thighs; complete clearance of lesions post treatment – anterior view (d) and posterior view (e).

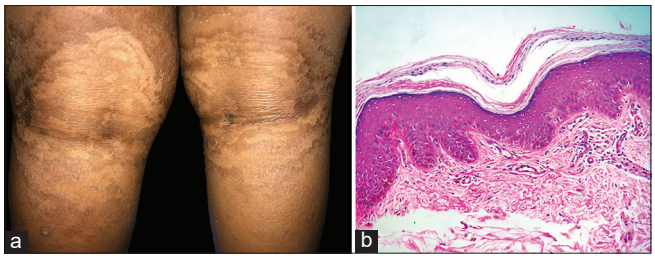

Figure 2: (a) Annular scaly plaques arranged in concentric rings in bilateral popliteal fossa; (b) Histopathological image showing parakeratosis, hypogranulosis and superficial dermal perivascular infiltrate (Haematoxylin-eosin stain, original magnification × 40).

mount of skin scrapings was negative for fungal elements. Histopathological examination showed focal hypogranulosis, alternating parakeratosis and perivascular lymphohistiocytic infiltrate in the upper dermis [Figure 2b].

Considering the distinct morphology of the lesions – annular scaly plaques with polycyclic borders, remitting and recurring nature of the lesions and the histopathological features, a diagnosis of erythrokeratoderma en cocardes was considered. Treatment options were explained to the family. After baseline blood workup, the patient was started on acitretin 10 mg tablets every alternate day. Lesions resolved almost completely with residual hyperpigmentation after 1 month of therapy [Figure 1d and e]. He is being maintained on the same for 3 months.

DISCUSSION

Erythrokeratoderma refers to a group of inherited heterogeneous cornification disorders. Earlier this group was thought to comprise of two distinct clinical entities – erythrokeratoderma variabilis (EKV) (Mendes de Costa

disease) presenting as transient migratory erythematous lesions and progressive symmetrical erythrokeratoderma (PSEK) presenting as fixed symmetrically distributed hyperkeratotic plaques. However, with the discovery of same genetic mutations in patients with PSEK and EKV; and observation of both the forms of erythrokeratoderma in the same family, PSEK and EKV is now considered to be different manifestations of a single disease. The term 'EKV et progressiva (EKVP)' is used to represent the diverse clinical phenotypes of this keratinisation disorder. EKVP is caused by mutations in the genes coding for the gap junction protein – connexin (GJB3, GJB4 or GJA1).[1] Rare variants of EKVP reported in the literature include erythrokeratoderma en cocardes (Degos syndrome), EKV with erythema gyratum repens-like lesions (EKV Cram Mevorah), EKV with erythema annulare centrifugum-like lesions and reticulate erythrokeratoderma.[2]

Erythrokeratoderma en cocardes presents as polycyclic hyperkeratotic plaques, with a peripheral collarette of desquamation. These represent the classical targetoid or concentric appearance – 'en cocarde'. Although erythrokeratoderma tend to present at birth or during infancy, our patient developed lesions during childhood.[2] There are limited reports of EKV manifesting during adolescence and adulthood.[3]

The histopathological findings in erythrokeratoderma are usually nonspecific and are of little diagnostic significance. Common findings include hyperkeratosis, parakeratosis, acanthosis and perivascular lymphocytic infiltrate. Clinical presentation with genetic analysis remains the mainstay for diagnosis. In our patient, genetic analysis could not be done due to financial constraints.

Other differential diagnosis considered were tinea imbricata – which was ruled out due to negative KOH smears, lack of response to antifungal therapy, histopathological picture and lack of pruritus; there was no atopic history or hair shaft abnormalities as in ichthyosis linearis circumflexa.

There exist no definite treatment guidelines for EKV. Topical treatment modalities used with varying degrees of success include emollients, retinoids, keratolytics and corticosteroids. Systemic retinoids are used in treatment of various keratinisation disorders. Retinoids through retinoid receptors (retinoic acid receptors and retinoid X receptors) act on the transcription process and exhibit inhibitory effects on keratinocyte proliferation. Systemic therapy with retinoids such as acitretin and isotretinoin has been shown to cause remarkable improvement in EKV.[2,4] Dosing is usually 1 mg/kg for isotretinoin or 0.5 mg/kg for acitretin. However, recurrence of lesions post discontinuation of therapy has been reported. Retinoids have also been used in tandem with phototherapy – psoralen-ultraviolet A/ ultraviolet B (PUVA/UVB) for treatment of keratinisation disorders. Possible adverse events include hepatitis, hyperlipidemia, gastrointestinal like nausea, abdominal pain and skeletal abnormalities. Although premature epiphyseal closure and hyperostosis are potential complications in children, they have been reported with high dose treatments. Long-term follow-up of use of acitretin in children has failed to demonstrate significant side effects.[5]

Our patient was started on low-dose acitretin and showed dramatic improvement within 3 weeks of therapy and is planned for gradual tapering of acitretin with maintenance on topical retinoids and emollients. This case is reported for its characteristic clinical picture and the remarkable response to low-dose acitretin therapy.

CONCLUSION

In summary, we report a 6 year-old boy with characteristic finding of erythrokeratoderma en cocardes who showed an excellent response to low-dose oral acitretin, with remarkable improvement within 1 month of treatment. Low-dose acitretin therapy may be a viable treatment option for children with EKV.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent.

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Nil.

Conflicts of interest

There are no conflicts of interest.

REFERENCES

1. Ishida-Yamamoto A. Erythrokeratodermia variabilis et progressiva. J Dermatol 2016;43:280-5.
2. Srinivas SM, Dhar S. Erythrokeratodermia variabilis and erythrokeratoderma en cocardes: Case series with review of literature. Indian J Paediatr Dermatol 2016;17:202-5.
3. Rajagopalan B, Pulimood S, George S, Jacob M. Erythrokeratoderma en cocardes. Clin Exp Dermatol 1999;24:173-4.
4. Singh N, Thappa DM. Erythrokeratoderma variabilis responding to low-dose isotretinoin. Pediatr Dermatol 2010;27:111-3.
5. Lacour M, Mehta-Nikhar B, Atherton DJ, Harper JI. An appraisal of acitretin therapy in children with inherited disorders of keratinization. Br J Dermatol 1996;134:1023-9.

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