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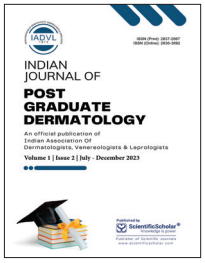

Case Report

Epidermolysis Bullosa Pruriginosa Treated with Acitretin

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ABSTRACT

Epidermolysis bullosa pruriginosa (EBP) is a rare inherited and distinctive clinical subtype of dystrophic epidermolysis bullosa. Here is a paediatric case of EBP with extremely pruritic lesions on extensor aspect of limbs and back, where the main modality of treatment given was topical retinoid (tazarotene) and oral retinoid (Acitretin), to which she had responded well clinically as well as symptomatically.

Keywords: Epidermolysis bullosa pruriginosa, Acitretin, Pediatric, Pruriginous papules

INTRODUCTION

Epidermolysis bullosa pruriginosa (EBP) is a rare inherited and distinctive clinical subtype of dystrophic epidermolysis bullosa (DEB). It is caused by COL7A1 gene mutation (collagen VII), in which patients present with intensely pruritic, lichenified or nodular prurigo-like lesions, occasional trauma induced blistering, excoriations, milia, and nail dystrophy.[1] The lesions are most evident on the limbs, particularly the shins. Most cases are sporadic, but some cases are reported with autosomal dominant or autosomal recessive pattern of inheritance.[2]

CASE REPORT

A 15-year-old female presented with gradually spreading multiple itchy skin lesions on the extensor aspect of forearms, legs, and scalp for 7 years. The patient was born of a consanguineous marriage, but there was no family history of similar complaints. Multiple skin coloured to erythematous scaly papules and plaques with atrophy and pigmentary changes over few lesions, and multiple nodular lesions with surrounding hyperpigmented rim were seen over the bilateral shins, ankles, and bilateral forearms [Figure 1]. Crusted papules were seen over the hairline and bilateral pinna. No nail involvement was noted. Differentials such as EBP, cutaneous amyloidosis, dermatitis artefacta, perforating disorder, hypertrophic lichen planus, and prurigo nodularis were considered. All routine blood and urine investigations were normal. Histopathological examination showed a sub-epidermal blister that contained plasma and fibrin. The floor of the blister showed sparse inflammatory infiltrate with dilatation of capillaries. The papillary dermis showed edema suggestive of EBP [Figure 2a and b]. Immunofluorescence-based mapping of the dermoepidermal junction and genetic diagnosis could not be done due to non-availability and financial constraint. Topical tazarotene cream (0.1%) once a day, Topical clobetasol propionate cream (0.05%) once a day and an emollient twice daily, along with tablet cetirizine 10 mg orally once a day were given for 2 months. The

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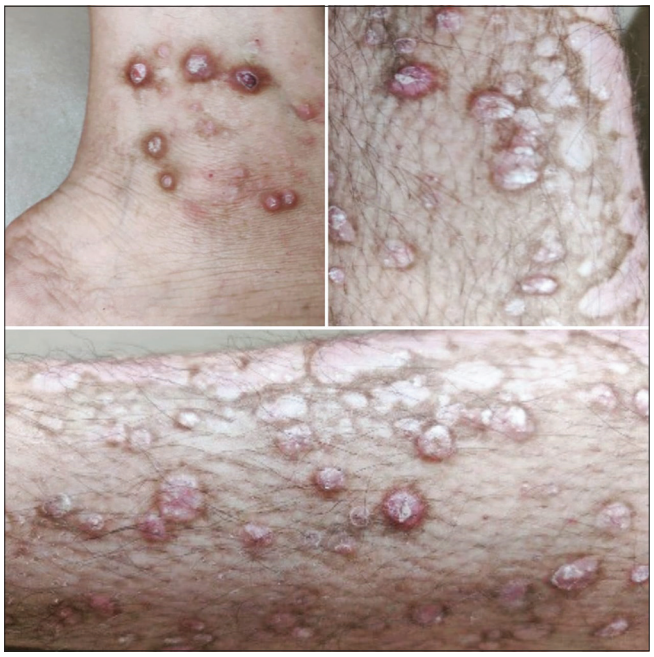

Figure 1: Pre-treatment clinical images of multiple skin coloured nodules and plaques with accentuated and hyperpigmented rim over bilateral shin and right ankle.

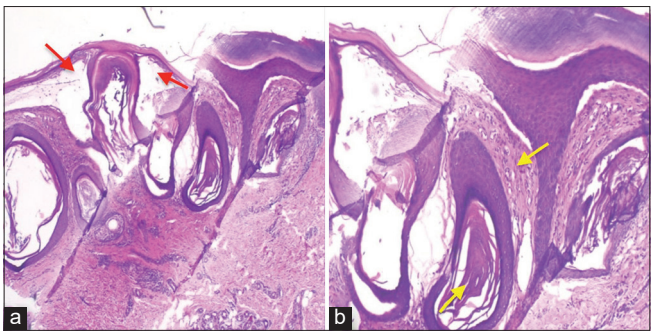

Figure 2: Hematoxylin and eosin staining of a tissue biopsy (4 mm) from left lower leg shows (a ×10) a subepidermal blister that contains plasma and fibrin (red arrows). (b ×40) The floor of the blister shows sparse inflammatory infiltrate with dilatation of capillaries (yellow arrows).

response was not satisfactory after 2 months; hence, oral acitretin 25 mg once at night was started for 3 months and alternately for the next three months. Topical tazarotene once a day and emollients were continued. After a duration of 6 months of oral acitretin, patient responded well clinically as well as symptomatically [Figure 3]. No significant adverse drug reactions to acitretin were reported.

DISCUSSION

EBP is a subtype of DEB, characterised by a combination of severe pruritus probably due to bradykinin cascade triggered by collagen VII and skin fragility that lead to scarring as well as hypertrophic, lichenified, and prurigo like nodules and plaques. Typically lesions mostly occur on the shins, and also on other parts of the legs, forearms, elbows, dorsal aspect of hands, shoulders, and lower back. The face and flexures are always spared, and nail dystrophy, albopapuloid lesions, blisters, and milia are other common but variable features.[3]

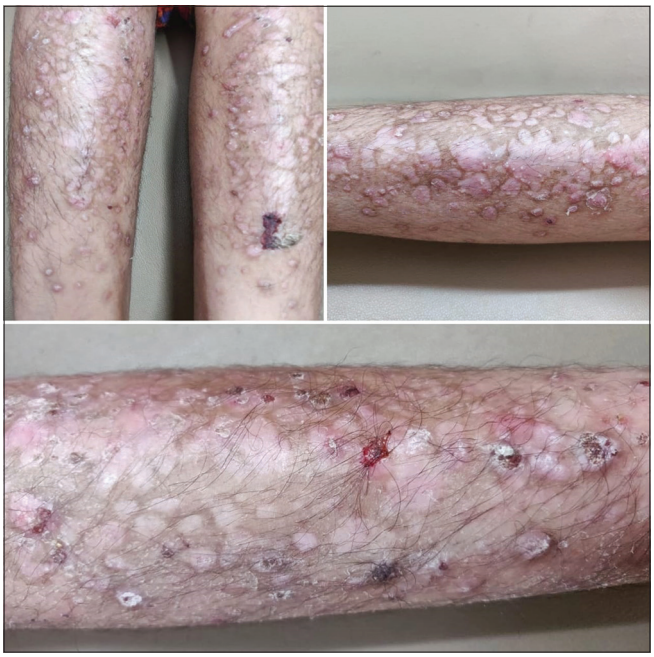

Figure 3: Six months post-therapy clinical images showing hypopigmented to depigmented plaques and patches with excoriation marks over bilateral shin.

Management is often difficult and usually unsatisfactory. Kim *et al.* have reported a case where significant improvement in reducing the hyper proliferative epidermal component was noted with a combination of daily oral isotretinoin 20 mg, intralesional triamcinolone acetonide injections and compression stockings.[4] Puri *et al.* gave symptomatic treatment for pruritus and tried treating with retinoids (etretinate) and thalidomide in a case of EBP, but the lesions recurred after stoppage of the therapy.[5] Inhibition of collagenase production by isotretinoin and all-trans-retinoic acid, respectively, in cultured synovial cells of patients with recessive DEB, has also been noted.[6] A study also points toward an immunomodulatory action of thalidomide that may suppress excessive production of Tumour necrosis factor-α,[7] while a study by Yamasaki *et al.* suggested successful outcome of EBP in a 10- year-old by oral cyclosporine therapy taken for 3 months with reduction in pruritus and no recurrence.[8] A study showed satisfactory results with oral dupilumab 600 mg first dose followed by 300 mg every 2 weeks for 6 weeks[9] while a case has also been reported where they successfully treated with topical

tacrolimus for 6 months.[10] Few cases reported satisfactory clinical outcome with good control over intense pruritus with oral baricitinib[11] and dapsone.[4]

In the present paediatric case, diagnosis was established on the basis of presence of lesions since childhood in a consanguineous marriage, intensely itchy lesions over extensors of upper limbs, lower limbs and back with presence of lichenified papules in a linear fashion with associated scarring and occasional blisters and normal hair and mucosa with the histopathology revealing a subepidermal cleft.

CONCLUSION

EBP is a missed-diagnosis. High index of suspicion, history and histopathology confirms the diagnosis.

EBP should be added to the differential diagnosis of chronically itchy and recurring papules on the shin, even though it's a rare condition.

Many treatment modalities were tried with varied responses. Present case responded satisfactorily to topical tazarotene with oral acitretin. Because of the potential for recurrence, acitretin therapy can be given in a tapered manner. In case of recurrence, it can be restarted with proper monitoring.

Declaration of patient consent

Patient's consent not required as patient's identity is not disclosed or compromised.

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Nil.

Conflicts of interest

There are no conflicts of interest.

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