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Case Report

Suicidality in Epidermolysis Bullosa Pruriginosa: Unusual Presentation in a Rare Disorder

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ABSTRACT

Depression in association with dermatologic disease is not uncommon and increases the morbidity and mortality. Early recognition and treatment of depression associated with dermatological disorders can lead to improved therapeutic outcomes and avert disastrous consequences including suicide. We wish to report a case of a young man with Epidermolysis bullosa pruriginosa who attempted suicide due to secondary depression and improved significantly after management of depression.

Key words: Suicidality, depression, bullosa, epidermolysis, pruriginosa.

INTRODUCTION

Psychiatric disturbance is reported in approximately 30% of patients with dermatological diseases. [1] One can potentially encounter patients with suicidality in dermatology. A risk of suicide can be pre-existing, appear secondary to skin disorders or be triggered by medications. In dermatology suicide risk has been described in severe acne conglobata (especially men) and metastatic melanoma. [2] Patients at risk must be specifically asked about suicidal ideations and tendencies as suicide risk requires immediate crisis intervention.

This report describes the case of an educated young man, who developed depressive illness with suicidality secondary to Epidermolysis Bullosa Pruriginosa (EBP) a rare, inherited, distinctive clinical subtype of Dystrophic Epidermolysis Bullosa (DEB).

CASE REPORT

29 year old unmarried, educated and employed man presented with complaints of itchy, raised skin lesions over the limbs since four years of age. The paroxysms of itching used to disrupt daily activities and sleep. Trauma, scratching and sweating used to induce fluid filled lesions with summer aggravations. Patient was much distressed and he visited various health institutions, to get cured despite the prognosis being explained. Due to exacerbation of symptoms he was admitted in dermatology ward. On detailed exploration he revealed sadness of mood, decreased interest in previously enjoyable activities, low self-esteem, crying spells, decreased sleep and appetite. Patient expressed hopelessness regarding his further improvement. Recurrent troublesome itching and intermittent lesions with no hope for recovery lead to suicidal thoughts and attempt in form of jumping from height of about 200 feet sustaining multiple injuries. There was no history suggestive of systemic or other cutaneous cause for severe pruritus.

Muco-cutaneous examination revealed numerous excoriated and erythematous papules with crusted erosions and scaring symmetrically in a linear configuration over extensor aspects of forearms and shins and extending up to mid thighs. (Figure 1)

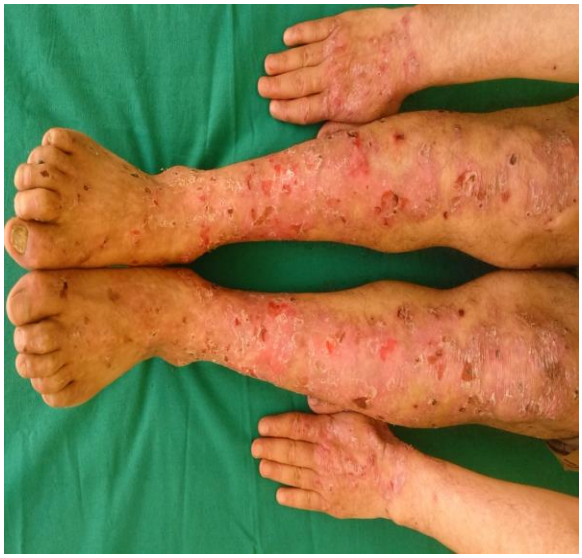

Figure 1: Numerous excoriated and erythematous papules with crusted erosions and scaring symmetrically in a linear configuration over extensor aspects of hands, forearms, shins and feet

Flaccid and clear fluid filled blisters were seen occasionally (Figure 2). Left toe nail revealed dystrophic changes (Figure3).

General physical and systemic examination was within normal limits. No abnormality was detected on routine investigations.

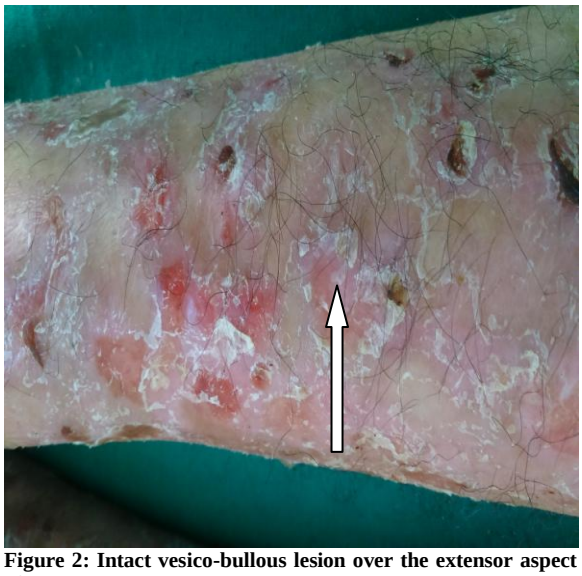

Figure 2: Intact vesico-bullous lesion over the extensor aspect of shin.

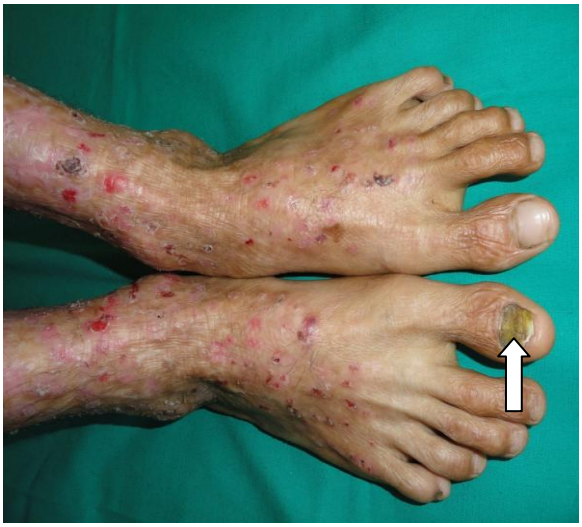

Figure 3: Cutaneous lesions with nail dystrophy (big toe nail).

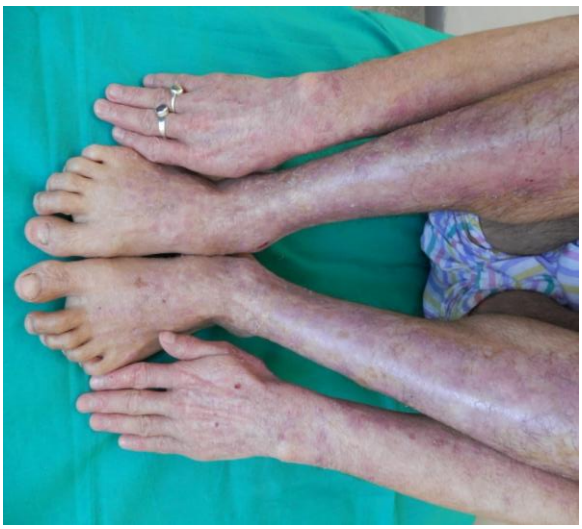

Figure 4: Post treatment resolution of lesions with mild scaring and hypo-pigmentation.

Histopathological examination of the fresh blister revealed evidence of hyperkeratosis, acanthosis and sub-epidermal bulla formation. Deeper tissue revealed perivascular and interstitial chronic inflammatory infiltrate composed of lymphocytes, histiocytes and eosinophils with capillary proliferation (Figure 5).

Direct immunofluorescence study was negative Electron microscopy and genetic analysis could not be done due to unavailability of these investigations.

Classical history, examination and histopathological findings suggested the diagnosis of epidermolysis bullosa pruriginosa (EBP). He was started on thalidomide, anti-histaminics and topical steroids. Minimal response was observed in itching after three weeks of treatment, so

short course of oral steroid (prednisolone 40 mg daily for two weeks) was added after which pruritus and lesions improved but he still had depressive features.

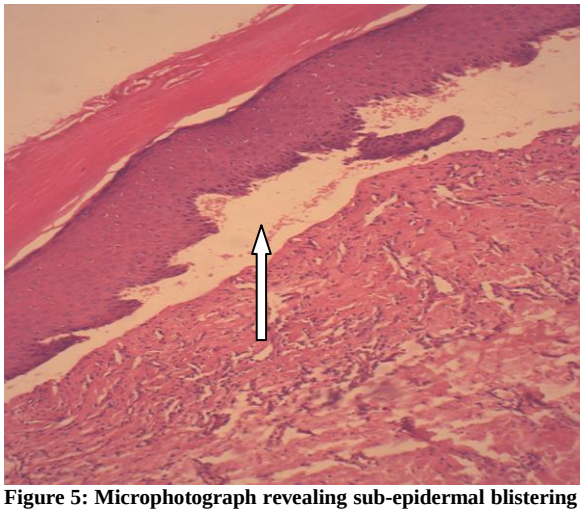

Figure 5: Microphotograph revealing sub-epidermal blistering (hematoxylin and eosin, 40X)

Psychiatric consultation was sought and patient was diagnosed to have severe depression with suicidality. He was managed with sertraline up to 100 mg, nortriptyline 25mg at bed time and risperidone 2mg daily along with supportive psychotherapy. Patient started showing mild improvement in depressive features after 2 weeks and in about 1month significant improvement was observed. Patient did not report suicidal ideations and expressed hope for recovery. Patient has been maintaining improvement both in cutaneous and psychiatric symptoms during subsequent monthly follow up for about one year.

DISCUSSION

Epidermolysis Bullosa pruriginosa (EBP) is a type of dystrophic epidermolysis bullosa (EB) coined by McGrath in 1994. [3] This is a rare disease with fewer than 100 patients reported in the literature. [4] It is caused by mutations in the *col7a1 (g2028r andg2028a)* gene which encodes the anchoring fibril protein, type VII collagen. [5] Various inheritance patterns have been described, including autosomal dominant, autosomal recessive and sporadic. [4] Age of onset for skin lesions in EBP is variable ranging from birth, infancy, childhood and even can be as late as 40 years. [6] Clinically it is characterized by severe pruritus, lichenified plaques or prurigo like lesions, violaceous linear scarring, and occasional trauma-induced blistering, excoriations, milia, and nail dystrophy in some cases. [7] Mostly lesions occur on legs predominantly over shins, but may affects forearms, elbows, dorsal aspect of hands, shoulders and even trunk symmetrically. [4]

Pruritus has been uniform features in all patients and is an important factor in the development of lesions and management of EBP. [8] Various proposed mechanisms for pruritus include development of immediate type hypersensitivity to environmental allergens, [4] exposure of type VII collagen activates bradykinin which possibly by interacting with other mediators results in severe pruritus. An aberrant wound repair process associated with increased protease-activated receptor (PAR-2) expression, which is an essential component of itch signaling and provide the link between the severe scarring and itching seen in EBP. [8]

Histologically, features of hyperkeratosis, acanthosis and split at dermal-epidermal-sublamina level are characteristic. Ultrastructurally, there is a sublamina densa level of blister formation and quantitative or qualitative changes in anchoring fibrils at the dermoepidermal junction. [9]

In the present case, diagnosis was established on the basis of onset of intensely itchy lesions since childhood. Typical distribution of cutaneous lesions in linear fashion over extensors of upper and lower limbs, with erythematous papules, erosions, crusting and occasional blisters with subsequent scaring was observed. Cutaneous histopathology was consistent with diagnosis of EBP. Clinical management of EBP is often difficult and unsatisfactory. Various treatment modalities include general measures for pruritus, oral anti-histaminic and topical steroid with or without occlusion have been tried. [10] Interventions such as topical tacrolimus, [11] systemic cyclosporine, [12] thalidomide [13]

etretinate, [3] cryotherapy [10] and dermabrasion or excision-grafting have been tried with variable success. [3] However, new lesions would usually continue to appear. Genetic counseling and gene therapy probably remain the most promising approaches.

CONCLUSION

Due to chronicity of disease and unsatisfactory treatment response patient may develop secondary depression and suicidality which complicates the course of skin disease. Depression can have varied presentation and needs early recognition and treatment for achieving favorable therapeutic response and avoid disastrous outcomes, including suicide. Our patient developed depression secondary to long standing symptoms starting at the age of four years and unsatisfactory treatment response. One should be vigilant to recognize depressive symptoms especially suicidality during the course of chronic dermatological conditions, to save life from a treatable condition as patient may not report these symptoms of their own.

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