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Case Report

A localized non-healing ulcer: an unusual clinical presentation of pemphigus vulgaris

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ABSTRACT

Pemphigus vulgaris is an autoimmune blistering disorder commonly involving oral mucosa and skin with characteristic acantholysis that results in formation of fluid filled blisters and painful erosions. Herein we report a case distinguished by its unusual clinical presentation. A twenty-five year old man, presented to our outpatient department of dermatology, complaining of ulcer on forehead in the last one year. Examination revealed single ulcer covered with crust on right side of forehead. Clinically lupus vulgaris, pyoderma gangrenosum, recurrent herpes simplex, leishmaniasis and pemphigus vulgaris were kept as differential diagnosis. Tzanck smear revealed multiple acantholytic cells and mild neutrophilic inflammatory infiltrate. Histological examination of biopsy sample showed suprabasal clefting, acantholytic cells and mild perivascular dermal lymphocytic infiltrate. Features were suggestive of pemphigus vulgaris. Significant improvement occurred after three months of treatment with oral prednisolone, azathioprine and topical steroid. Although oral and skin involvement is common, in the form of flaccid blisters and erosions, but non healing ulcer is unusual. This may represent a period of limited activity during this chronic disease. There is paucity of data in literature regarding this. So, this case report highlights a, perhaps unique, clinical presentation of this autoimmune disease.

Keywords: Non healing ulcer, Pemphigus vulgaris, Pyoderma gangrenosum

INTRODUCTION

Pemphigus is a group of chronic autoimmune blistering diseases characterized by presence of antibodies against desmosomal adhesion proteins. Pemphigus vulgaris is the most common form of pemphigus, accounting for over 80% of cases.[1] Nearly all the patients have mucosal lesions and pemphigus vulgaris presents with oral lesions in 50-70% of patients.[2-4] These may precede cutaneous lesions by months or can be the only manifestation of the disease. Flaccid blisters filled with clear fluid arise either on normal skin or an erythematous base. The contents may become turbid or the blisters may rupture, producing painful erosions which extend at the edges.

This case report describes the case of a patient complaining of ulcer over forehead in the last one year, which was diagnosed as pemphigus vulgaris.

CASE REPORT

A twenty-five years old man, presented to our outpatient department of dermatology, complaining of an ulcer on the forehead in the last one year. The lesion was started as small fluid filled blister over right side of forehead, not associated with any local symptom. Blister ruptured on its own within few days to form erosion which progressed to form an ulcer which developed crusts later on. Regarding this, he has taken treatment from various

local practitioners, the record of which was not available. But no improvement was seen.

There was no history of any mucosal involvement or any constitutional symptoms. There was no history of decreased appetite, weight loss and tuberculosis in past.

Examination revealed single well-defined ulcer of size 5×6 cm ×3 mm, covered with yellowish to brown colored crusts on right side of forehead. Surrounding skin was normal (Figure 1). Clinically lupus vulgaris, pyoderma gangrenosum, recurrent herpes simplex, leishmaniasis and pemphigus vulgaris were kept as differential diagnosis.

Complete blood count, Erythrocyte sedimentation rate and other routine investigations were within normal limit. Mantoux test was nonreactive. Chest X-ray was normal. Serology for herpes simplex virus 1 and 2 was non-reactive. Tzanck smear revealed multiple acantholytic cells and mild inflammatory infiltrate of neutrophils (Figure 2). Histological examination of biopsy sample taken from the lesion showed suprabasal clefting, acantholytic cells and mild perivascular lymphocytic infiltrate in the dermis (Figure 3).

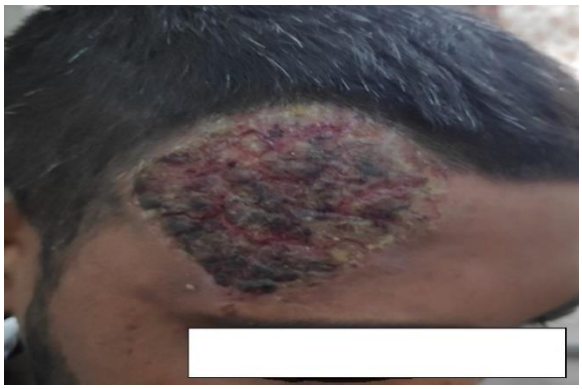

Figure 1: Single well-defined ulcer of size 5 cm × 6 cm × 3 mm, covered with yellow to brown colored crust over right side of forehead.

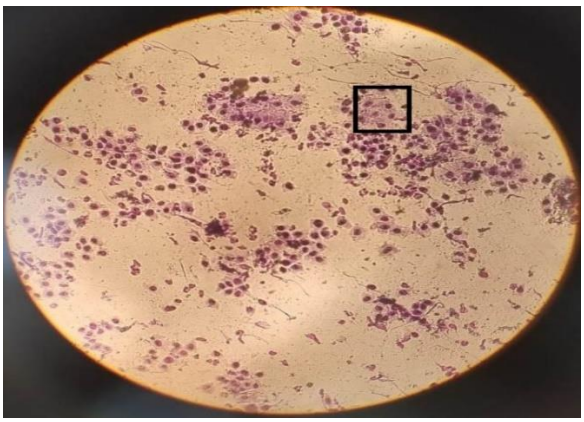

Figure 2: Tzanck smear showing multiple acantholytic cells.

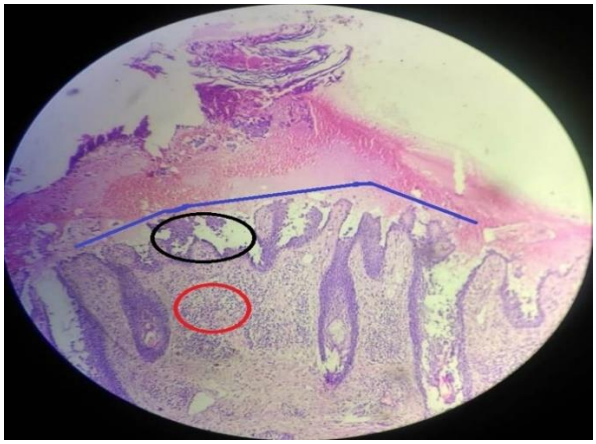

Figure 3: Histopathology- blue line indicates suprabasal clefting, black circle indicates acantholytic cells in blister cavity, red circle indicates lymphocytic inflammatory infiltrate in dermis

Features were suggestive of pemphigus vulgaris. Patient was given treatment in the form of oral prednisolone, azathioprine and topical steroid. Significant improvement occurred after three months of treatment.

DISCUSSION

Pemphigus is a group of chronic autoimmune blistering diseases characterized by presence of antibodies against desmosomal adhesion proteins. In most cases, the first sign of disease appears on the oral mucosa. While the lesions can be present anywhere within the oral cavity, they are most commonly found in areas subjected to frictional trauma such as the buccal mucosa, pharynx, larynx, esophagus, genital mucosa as well as the skin.[5] The underlying mechanism responsible for causing the intraepithelial lesions of pemphigus vulgaris is the binding of Immunoglobulin G autoantibodies to desmoglein 3 and 1.

Pemphigus vulgaris can be differentiated from other similar conditions by tzanck smear, biopsy and direct immunofluorescence. Supra basilar split seen in pemphigus vulgaris helps distinguish this condition from sub-epithelial blistering diseases. The direct immunofluorescence (DIF) shows characteristic deposition of IgG and other C3 antibodies that bind to cell surface of perilesional skin or mucosa.[6,7] ELISA has been developed which can detect desmoglein 1 and 3 in serum sample of patients with pemphigus vulgaris.[7] Indirect immunofluorescence is useful in monitoring the disease activity.

An important aspect of patient management is early diagnosis and institution of early treatment, which can prevent serious involvement of other body sites and various fatal complications. Systemic corticosteroid treatment remains the mainstay of therapy, generally in combination with a steroid-sparing immunosuppressant.

Rituximab, intravenous immunoglobulin, plasmapheresis and immunoadsorption are other treatment modalities.

In this case, patient must have taken oral steroids in small doses prior to visiting our department because of which the disease got localized to one site in the form of non healing ulcer without developing generalized blisters.

CONCLUSION

The case we report, illustrates a special situation in which pemphigus vulgaris was revealed by an unusual skin lesion. Although oral and skin involvement is very common, in the form of flaccid blisters and erosions, but non healing ulcer is unusual presentation. Localized non healing ulcer may represent a period of limited activity during this chronic disease. There is paucity of data in the literature regarding this. Atypical presentations can lead to a delay in diagnosis and initiation of the appropriate treatment. Pemphigus vulgaris may have various unusual manifestations, such as nail dystrophy, paronychia or granulation tissue-like lesions, but this case of nonhealing forehead ulceration highlights a further, perhaps unique, clinical presentation of this autoimmune disease. So, pemphigus vulgaris has to be kept in mind as one of the differential diagnosis for non healing ulcer.

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