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CASE REPORT

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A rare new presentation of pemphigus vulgaris

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Abstract

We report a new presentation for pemphigus vulgaris in a 51-year-old female patient that was complaining only from non-healing foot ulcer, but unfortunately pemphigus was not confirmed and the patient lost multiple nails. This new variation is reported to increase health workers' awareness especially in races pemphigus seems to be common.

KEYWORDS

mucocutaneous manifestations, pemphigus, pemphigus vulgaris

1 | INTRODUCTION

Pemphigus is a group of autoimmune bullous disorders that affects skin and mucosal membranes.[1] Pemphigus vulgaris is the most common type of pemphigus, and it is characterized by painful mucosal erosions and flaccid blisters that easily rupture.[1] The progression of PV usually begins with painful persistent mucosal erosions most commonly in the mouth with many patients developing also cutaneous manifestations.[2] The approximate time between the clinical presentations and diagnosis whether there is oral mucosal involvement or skin involvement or both is approximately 6.19 ± 3.82 months.[3] The diagnosis can be delayed especially in patients presenting only with oral mucosal involvement.[4]

Herein, we report a rare presentation of PV in 51-year-old female patient which presented with a 1 year foot ulcer and recently developed hand ulcer with oral erosions. Misdiagnosis leads to loss of some nails with recurrent bacterial infection. Early diagnosis for pemphigus tremendously important because till now and despite the use of adjuvant treatments, the mortality rate associated with disease is about 6%.[5]

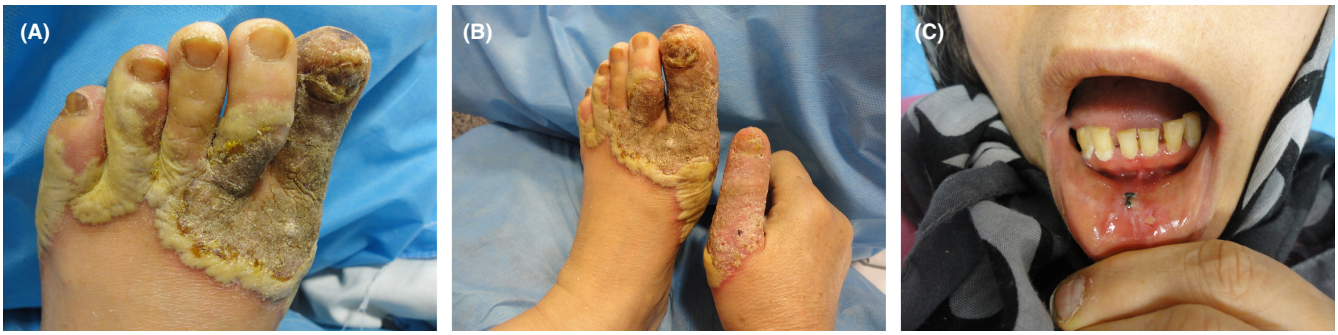

FIGURE 1 (A and B) Ulcerated plaques with elevated borders. (C) Erythematous erosion on the inner aspect of lower lip

2 | CASE PRESENTATION

A 51-year-old female patient with no significant past medical history presented to our clinic with ulcerated plaques on both left foot and right hand (Figure 1A,B). History goes back to 1 year before when she started complaining from progressive foot ulcer. Multiple skin biopsies were done to rule out malignancies, and all were negative. The patient was also evaluated for leishmaniasis and fungal infection, which were also negative. Only she had mixed bacterial colonization in her bacterial culture, and she was treated with multiple antibiotics that unfortunately were ineffective. The patient mentioned that her hand ulcer started about 1 month before and 2 weeks later was followed with oral erosions (Figure 1C).

Clinical examination showed left foot and right hand ulcers with elevated borders, in addition to loss of first toenail and first fingernail. Mucosal examination showed an ulcerative lesion on the lower labial mucosa. A skin biopsy was taken with differential diagnosis of pemphigus vulgaris, blastomycosis-like pyoderma, and pyodermatitis-pyostomatitis vegetans. The histopathological evaluation of the skin biopsy was compatible with pemphigus vulgaris. Also, direct immunofluorescence (DIF) from oral mucosa confirmed the diagnosis (Figure 2A,B).

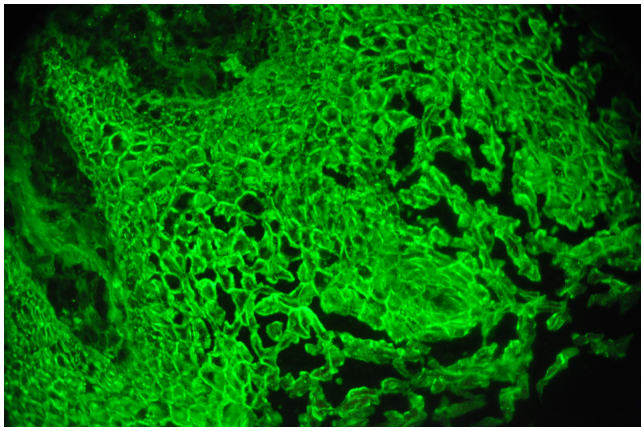

FIGURE 2 Intercellular deposits of IgG within the epidermis

The patient was admitted to dermatological ward, and she was treated with pulse methylprednisolone (1 mg/kg/day) in addition to oral azathioprine (100 mg/daily). Four days later, she was discharged home with oral prednisolone (60 mg/day) plus azathioprine. During the next 4 months, her medications were tapered gradually and she had complete clearance for her skin and oral lesions (Figure 3).

3 | DISCUSSION

Pemphigus vulgaris is an IgG autoantibody-mediated blistering dermatosis against desmogleins 1 and 3 occurring usually between 40 and 60 years of age.[6] It usually starts with oral blisters and erosions, which can be followed later with involvement of other mucosa like genitalia, nose and eyes, and flaccid blisters on the skin.[6] In PV, there are formation immune complex deposits consisting of IgG antibodies that bind to desmosomal transmembrane proteins of keratinocytes desmoglein 1 (Dsg1) and Dsg3 leading to loss of cell adhesion between keratinocytes.[7]

Clinical variants of PV include cutaneous-only disease, mucosal-only disease, or more commonly mucocutaneous variant.[8] PV starts usually with superficial oral blister that ruptures rapidly leading to slow healing painful erosions and ulcers.[8] Development of skin lesions is usually from weeks to months after the initial oral presentation.[8]

In our case, there was an unusual presentation for PV which presented with cutaneous ulcer for 1 year and the patient had not complained from any mucosal lesions. What makes our case distinct is the limited skin involvement which seems very rare in pemphigus vulgaris presentations.

Routine histopathological examination for PV shows intraepidermal bulla formation in the suprabasal layer of epidermis associated with loss of cell-to-cell adhesion called acantholysis.[9] The floor of the bulla is formed from basal cells, which remains attached to the basement membrane giving the appearance of row of tomb

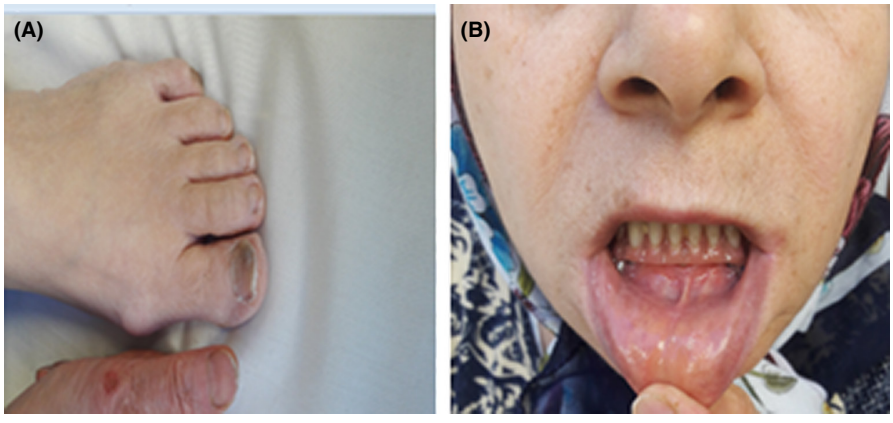

FIGURE 3 Complete resolution of skin and oral lesions 4 months after starting therapy

stones.[9] Demonstration of the IgG autoantibodies by DIF, IIF, ELISA, or immunoblotting is the gold standard tool for confirming the diagnosis of pemphigus.[4] In our case, skin biopsies were done multiple times, and the last time was confirmed with direct immunofluorescence. Biopsy site selection is very important in order to get an accurate pathological information.[10] Regarding blisters or bullae, the most appropriate site for biopsy is the edge of the lesion including the perilesional skin, whereas the best site for biopsy in ulcerated and necrotic lesions is the edge of the ulcer in addition to the adjacent skin.[10]

The cornerstone for treatment of PV was systemic corticosteroids, which led to dramatic decrease in mortality among pemphigus patients.[11] Controlling the disease requires several weeks to months depending on the severity of the disease, and complete remission can be achieved.[11] Because of the long-term side effects of corticosteroid, new corticosteroid-sparing adjuvant therapies including azathioprine, high-dose intravenous immunoglobulins, immunoadsorption, and rituximab have been established as adjunctive successful therapy choices.[11]

With such new presentation, PV can be easily missed by healthcare professionals and dermatologists, hence delaying the diagnosis; thus, awareness for these presentation is essential. As dermatologists, we must be familiar with these new cutaneous manifestations and variations of pemphigus vulgaris in order to start treatment as soon as possible and to avoid undesirable complications. We recommend performing direct immunofluorescence for all non-healing bullous lesion to avoid delay in diagnosis for these diseases.

AUTHOR CONTRIBUTIONS

NM was involved in the diagnosis and management of the patients and has been responsible for the clinical part of the manuscript. RG and NM did literature review and drafted the manuscript. NM was responsible for final editing of the manuscript and coordinated the study. All authors have read and approved the final manuscript.

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We would like to thank the patient for granting us permission to publish this article.

CONFLICT OF INTEREST

The authors have no conflicts of interest to declare.

DATA AVAILABILITY STATEMENT

The data that support the findings of this study are available from the corresponding author upon reasonable request.

CONSENT

Written informed consent was obtained from the patient for the publication of this case report and accompanying images.

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