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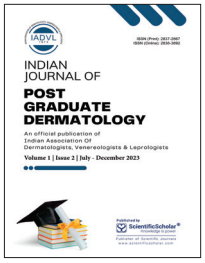

Case Report

Herpes Simplex Virus I Infection Complicating Pemphigus Foliaceus

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ABSTRACT

Pemphigus is a group of autoimmune blistering disease characterised by blisters affecting the skin and or mucosae. Herpes simplex virus (HSV) 1 is known to result in recalcitrant oral lesions of pemphigus. It is rarely described in association with pemphigus foliaceus. Here, we describe a middle aged man who had persistent erosions on the face despite adequate disease control in other body parts. Screening for secondary infection yielded HSV-1 by polymerase chain reaction.

Keywords: Pemphigus foliaceous, Herpes simplex virus 1, Autoimmune bullous disease

INTRODUCTION

Pemphigus is a collective term for a rare group of organ-specific autoimmune intraepidermal blistering diseases that affect the skin and the mucous membranes. Clinically, it manifests with vesicles and erosions affecting the mucous membranes and skin, often causing a prominent impairment of quality of life and increased morbidity and mortality.[1] Pemphigus vulgaris (PV) and pemphigus foliaceus (PF) are the two major forms of pemphigus accounting for more than 90% pemphigus in the clinical practice.[2] PF is a superficial type of pemphigus characterised by erosions, scaling and crusting affecting predominantly the seborrheic region of body. Histologically it is characterised by subcorneal acantholytic blister and immunopathologically by the presence of autoantibodies that target desmoglein 1 (Dsg1).[2] Herpes simplex virus (HSV) superinfection is known to influence the course of PV but rarely been reported in association with PF.[3-6] Here, we describe a case of PF with superimposed HSV infection.

CASE REPORT

A 55-year-old man presented with itchy and painful lesions over the body of 6-month duration. He developed fluid filled lesions initially over the back, which gradually progressed to involve face, scalp, chest and the upper limb. Cutaneous examination revealed few flaccid vesicles on the arm; scaly and crusted lesions were seen over the scalp, face and the trunk on the background of erythema [Figures 1a and b]. In addition, irregular and angulated erosions were seen over the face [Figure 1a]. A provisional diagnosis of PF was made. Histopathological examination from the intact vesicle showed subcorneal blister; direct immunofluorescence microscopy from the perilesional skin showed intercellular deposition of immunoglobulin G (IgG) and C3 in the epidermis. Indirect

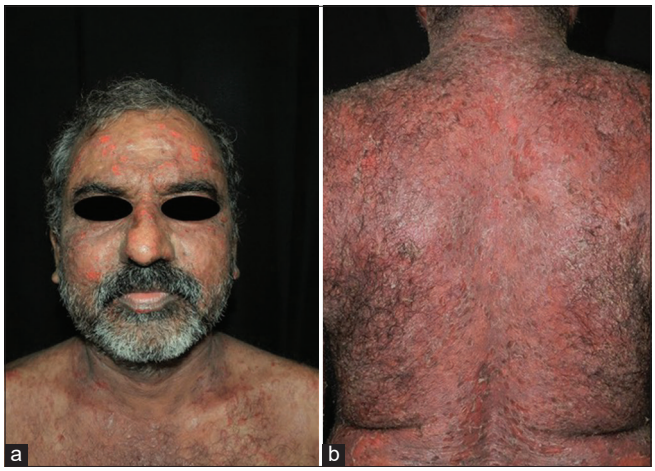

Figure 1: (a) Discrete angulated erosions on the face. (b) Scaly and crusted lesions on the back.

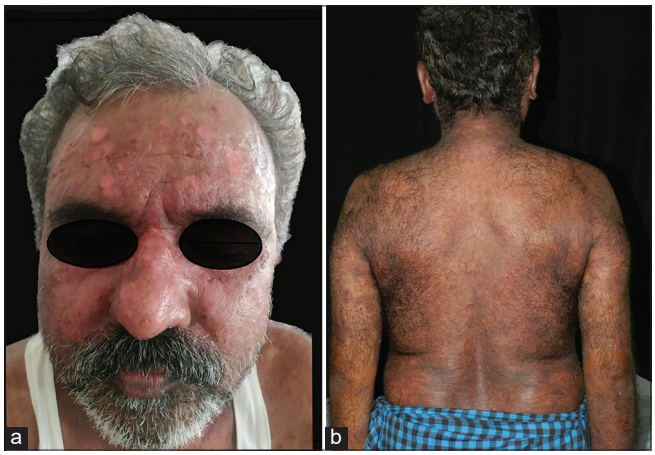

Figure 2: (a) Facial lesions after treatment with prednisolone, azathioprine and valacyclovir. (b) Back lesions after treatment with prednisolone, azathioprine and valacyclovir.

immunofluorescence microscopy on normal human skin revealed high titre of circulation antibodies (titre of 1:200) while enzyme linked immunosorbent assay detected IgG anti-Dsg1 antibodies (titre >200). His baseline haematological and biochemical tests were within the normal range for his age. He was commenced on prednisolone at a dose of 1 mg/kg/body weight along with azathioprine 50 mg/day. Although most of the lesions responded well to this regimen after 2 weeks of starting medications, he had persistent painful erosions on the face. He was investigated further; skin swab from the erosions revealed the presence of HSV-1 by polymerase chain reaction (PCR). Valacyclovir 3000 mg in three equal divided doses was added to his treatment regimen. After 3 days of starting Valacyclovir, his facial lesions started showing signs of healing and at the end of 1 week complete resolution was seen [Figures 2a and b]. Antivirals were stopped at the end of 1 week.

DISCUSSION

Impaired skin barrier and attenuated immune response are the underlying factors that favour the occurrence of HSV superinfection in pemphigus. Use of topical and systemic immunosuppressive agents in the treatment of these conditions may further facilitate the occurrence of infection. The sources of HSV in the pemphigus lesions can be either from endogenous or exogenous source.[3] HSV superinfection may result in severe, persistent and recalcitrant pemphigus lesions and also with exacerbations and relapse of pemphigus.[3] It can either be localised to the erosions or disseminated with systemic involvement that is, eczema herpeticum (EH) or Kaposi's varicelliform eruption. The latter has been reported in the setting of diverse pre-existing dermatoses such as atopic dermatitis, seborrheic dermatitis, impetigo, scabies, ichthyosiform erythroderma, Darier's disease, benign familial pemphigus, bullous pemphigoid, ichthyosis vulgaris, mycosis fungoides, Sezary syndrome and other dermatoses. (IJD 1996). EH is characterised by presence of disseminated vesicles which become pustular and markedly umbilicated.[5,7] We did not consider the possibility of EH in our patient as the erosions were localised to the face and there were no umbilicated lesions. It is challenging to diagnose HSV superinfection in cases of pemphigus, as both present with flaccid blisters which rupture to form painful erosions on the skin as well as mucosae. The presence of virus in the lesion may lead to excessive production of interleukin (IL)-4 and IL-10, which cause a shift from T helper cell type 1 to T helper cell type 2 response, increasing antibody production. They can also directly infect B and T lymphocytes, contributing to the production of autoreactive B lymphocytes and autoimmune antibodies.[8] Among various methods available for the detection of HSV in the skin lesions, PCR is highly sensitive technique;[9] biopsy from the lesion to demonstrate the cytopathic effect would have ruled out asymptomatic colonisation. However, our patient denied repeat biopsy; nevertheless, he responded well after the initiation of oral antiviral therapy.

It would be prudent for clinicians involved in the management of pemphigus to elicit history of cold sore or genital blisters; our patient denied history of such episodes in the past. Clinician should have low threshold for screening for HSV infection especially in patients who have refractory or resistant disease.[5] Timely recognition of herpetic superinfection helps to avoid unnecessary changes of immunosuppressive treatment and to start an effective antiviral treatment without delay.[4]

CONCLUSION

Dermatologists should carefully look for secondary cutaneous HSV infection in cases of Pemphigus foliaceus, when the standard therapy fails to respond.

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.

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