Pemphigus vulgaris and lichen planus pigmentosus in an HIV-positive patient

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Abstract

Pemphigus vulgaris is a rare autoimmune blistering disorder which can occur with other disorder with autoimmune etiology like lichen planus pigmentosus. The concurrence of pemphigus vulgaris and HIV infection has been rarely reported in literature. Here we report a 31 year old patient who came with oral and skin erosions suggestive of pemphigus vulgaris and later developed HIV infection with lichen planus pigmentosus.

Key words: Acquired immune deficiency syndrome, human immunodeficiency virus, lichen planus, pemphigus

Introduction

Pemphigus vulgaris is an acquired acantholytic disorder that has high mortality and morbidity rate without treatment. Although symptoms of autoimmune conditions such as pemphigus vulgaris have been reported to improve in HIV infection, therapy of autoimmune blistering disorders in such case scenario poses a clinical challenge.[1] Here, we discuss a patient who presented with pemphigus vulgaris and periorbital lichen planus pigmentosus with HIV infection.

Case Report

A 31-year-old female came with a history of fluid-filled lesions in the oral cavity that ruptured to form erosions for a duration of 3 weeks associated with burning sensation. The baseline blood investigations such as complete blood count, liver and renal function test, urine routine, and chest X-ray including serology for HIV and hepatitis B were normal. Tzanck smear showed acantholytic cells and indirect immunofluorescence showed intercellular staining of the epithelium with IgG in 1:200 titer in oral mucosa and 1:10 titer in human skin suggestive of pemphigus vulgaris. She was given intravenous pulse therapy with dexamethasone 100 mg in 500 ml normal saline for 8 cycles and once the disease was under remission changed to daily oral prednisolone therapy which was started at the dose of 1 mg/kg/day (50 mg/day). The routine investigations including serology for HIV were done after 1 year as azathioprine was considered a steroid sparing agent for maintenance of

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Figure 1: The periorbital violaceous pigmentation

remission. The patient became positive for HIV serology and was started on antiretroviral therapy (ART) therapy with the regimen tenofovir 300 mg, lamivudine 300 mg, and efavirenz 600 mg. The patient was on irregular follow-up, and recently, she presented with periorbital hyperpigmentation in violaceous hue with photosensitivity for the past 2 months and flaccid vesicles over the bilateral forearm with positive Nikolsky's sign [Figures 1 and 2]. The CD4 count was 250 cells/mm^3 and histopathology of the facial pigmentation showed interface changes with significant melanin incontinence suggestive of lichen planus pigmentosus which could have been induced by the ART drugs. The patient was restarted oral prednisolone (1 mg/kg/day) to control the disease activity of pemphigus along with photoprotection with physical sunscreen.

Discussion

A review article published in PubMed in 2018 revealed that only six cases of pemphigus vulgaris and HIV were reported.[2] A PubMed search using the term "pemphigus" and "HIV" showed 15 results in the last 5 years and only one was a clinical report about a study in Botswana about autoimmune skin disease including pemphigus which had stated that 19.8% of the study population were HIV positive.[3] In the case reports mentioned above, four patients had HIV preceding their pemphigus vulgaris and only two had a concurrent presentation of PV and HIV.

The pathological mechanism of HIV preceding pemphigus has been explained by nonspecific polyclonal B-cell stimulation, leading to persistent antigenic stimulation and highly active antiretroviral therapy (HAART)-induced pemphigus lesions.[4] Similarly, the development of HIV in pemphigus patients could be due to treatment with immunosuppressive drugs. The loss of the epidermal barrier due to genital erosions of pemphigus could also predispose to acquiring HIV infection. While the clinical features and response to therapy of pemphigus vulgaris are reported not to be modified by HIV, the impact of HIV on pemphigus remains obscure.[5]

The treatment of autoimmune bullous disorders (AIBDs) which primarily involves immunosuppressants becomes a conundrum in an already immunosuppressed HIV-positive individual. Topical corticosteroids are the drug of choice in limited AIBD, whereas systemic corticosteroids are considered a first-line therapy in patients with extensive BP and PV.[2] Cyclosporine, azathioprine, cyclophosphamide, mycophenolate mofetil, rituximab, and thalidomide in a dose similar to HIV-negative individuals are the other treatment options that have been documented in literature.[6] Periorbital lichen planus pigmentosus is a rare condition that has been reported in association with pemphigus vulgaris.[7] The occurence could be due to the established

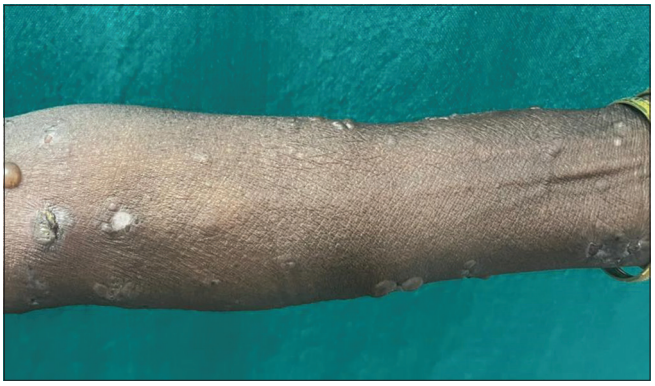

Figure 2: The flaccid vesicles with erosion on the ventral and dorsal aspect forearm

association of pemphigus vulgaris with other autoimmune diseases or lichen planus pigmentosus could have been induced by the drugs of ART regimen.

In conclusion, patients with pemphigus vulgaris who are treated with immunosuppressive therapy should be monitored by serological tests (Hepatitis B or C, tuberculosis, HIV) during follow-up visits. It is important to remember that reconstitution of immunity in patients on HAART can lead to flare-up of the disease activity of pemphigus, and periodic CD4 count monitoring is essential in such patients.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form, the patient has given her consent for her images and other clinical information to be reported in the journal. The patient understands that her name and initials will not be published and due efforts will be made to conceal identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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