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A Case of Pemphigus Vulgaris in a Patient with Human Immunodeficiency Virus

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ABSTRACT

Pemphigus vulgaris is the most common variant of autoimmune chronic blistering skin diseases. Its appearance in patients infected with acquired immunodeficiency virus (HIV) is rare. A 40-year-old woman is reported, who presented skin lesions in 90% of the body surface, characterized by blisters of 3 to 4 cm that, when broken, left denuded areas, excoriations and pruritic meliceric crusts. A skin biopsy was performed that showed a characteristic pattern of pemphigus vulgaris. Elisa for HIV positive. She evolved with septic shock and died 72 hours later. Understanding the relationship between HIV and autoimmune blistering disorders can help guide the therapeutic and prognostic management of these patients. Proposed mechanisms include molecular mimicry, polyclonal B-cell stimulation, and highly active antiretroviral therapy possibly associated with immune restoration. Clinically, these patients may present limited mucosal symptoms or develop multiple erosions that may spread, merge, and progress to exfoliative erythroderma. Glucocorticoids serve as the first line of management; however, they can have important adverse effects such as infectious complications that pose a therapeutic challenge for the doctor and could influence mortality.

ARTICLE DETAILS

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BACKGROUND

Pemphigus encompasses a heterogeneous group of chronic autoimmune blistering skin diseases that affect both the mucosa and the skin. It can be divided into three main forms: pemphigus vulgaris (PV), pemphigus foliaceus (PF), and paraneoplastic pemphigus (PNP). PV is the most common clinical variant in approximately 70% of cases. The annual incidence rate has been reported between 0.76 and 16.1 per million inhabitants, depending on the geographic area and ethnicity[1]. The mechanism of autoantibody formation is complex, it results from the breakdown in immunological tolerance in genetically predisposed subjects, the strongest evidence for the risk alleles are HLA DRB1*04:02 and DQB1*05:03 before triggers such as drugs (thiols, angiotensin converting enzyme inhibitors, non-steroidal antiinflammatory drugs, aspirin, rifampin, etc)[2]. As for paraneoplastic pemphigus, it is related to non-Hodgkin's lymphoma, chronic lymphocytic leukemia, Castleman's disease, thymoma, and other mixed connective tissue tumors[3]. The pathophysiology focuses on the underlying loss of epidermal cell adhesion caused by IgG autoantibodies against the desmosomal adhesion proteins, desmoglein 3 (Dsg3) and/or Dsg1, in epidermal keratinocytes, however, the pathological mechanisms underlying acantholysis of the pemphigus are complex and not completely clear[4,5]. Clinically, it can manifest as a dominant mucocutaneous type or, less frequently, exclusively cutaneous[6].

The appearance of pemphigus vulgaris in HIV-infected patients is rare and although some cases have been reported, there are no definitive statistics and the course of the disease is not known[7,8]. A case treated at the third level of care is reported.

CLINICAL CASE

A 40-year-old woman, previously healthy, presented skin lesions of 2 months duration that affected 90% of the body surface (Fig. 1A) characterized by blisters of 3 to 4 cm that, when broken, left denuded areas, excoriations, and meliceric crusts. pruritic with serohematic secretion: in gingival mucosa, palate, oropharynx, trunk and extremities. A skin biopsy was performed (Fig. 1B) which showed a characteristic pattern of pemphigus vulgaris. With a report

during Elisa's hospitalization for HIV which was positive. Respiratory acidosis was added, for which a chest X-ray was performed (Fig. 1D) which showed increased hila, with right basal pneumonia, the patient evolved with septic shock, and she died 72 hours later.

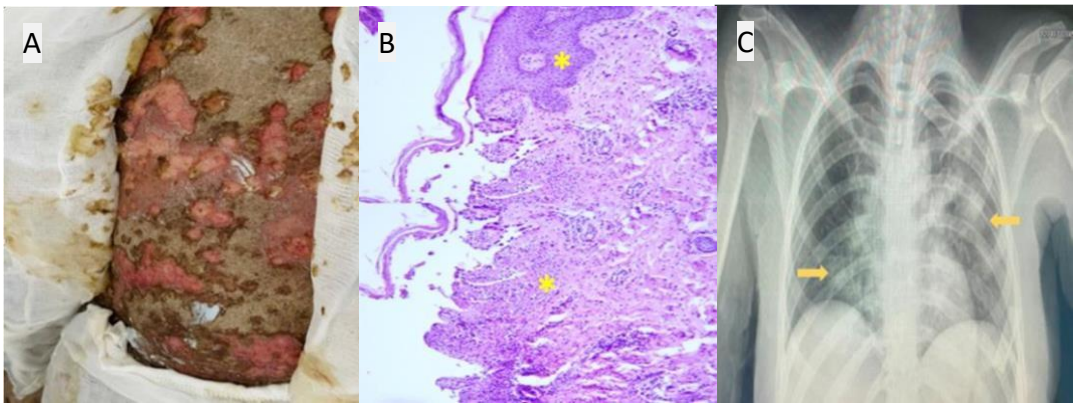

Figure 1. (A) Confluent bare areas with erythroderma in the chest and abdomen (B) Intraepidermal acanthosis in the lower portions of the epidermis and spongiosis with exocytosis of neutrophils, perivascular infiltrate with eosinophils in the dermis [asterisks]. (C) Increased pulmonary hila and right basal consolidation [arrows].

DISCUSSION

Understanding the relationship between HIV and autoimmune blistering disorders can help guide the therapeutic and prognostic management of these patients. According to Etzioni A99, molecular mimicry of the HIV virion is a proposed mechanism in the development of autoimmune diseases. A small study conducted in the pre-antiretroviral therapy (ART) era looking for auto antibodies indicative of autoimmune blistering skin disease found that autoantibodies directed against the dermal-epidermal basement membrane proteins BP180 and BP230 were present in 38% of patients. cases of HIV-infected patients and were more frequent in the later stages of infection; On the other hand, more recent studies indicate that in the early stage of infection, the cytokines that are released from HIV-infected macrophages stimulate non-specific polyclonal B cells10. In addition, an increase in prevalence has been documented since the introduction of highly active antiretroviral therapy, possibly associated with immune restoration[7,10].

Clinically, these patients may present with limited mucosal symptoms or progress to multiple pruritic, scaling, and crusted erosions with circumscribed scaly patches in mainly seborrheic areas that may spread, merge, and progress to exfoliative erythroderma (erythema and scaling that covers >90% of the skin area). body surface); Blisters are rarely seen due to their superficial location and consecutive rupture[6,10].

Effective treatments must be implemented promptly. Glucocorticoids serve as a first-line approach due to their rapid onset of therapeutic effects and remission of the acute phase.

However, applications can have significant adverse effects that outweigh the benefits[11].

CONCLUSIONS

PV in the presence of immunocompromised states such as HIV has an uncertain course, direct infectious complications on skin lesions and the system in general pose a therapeutic challenge for the physician and could influence high mortality.

AUTHOR CONTRIBUTIONS

All authors have contributed equally.

CONFLICTS OF INTEREST

None reported.

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