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Case Report

Pemphigus Vulgaris

Riofrío Ivonne[1]*, Suárez Efraín[1], Vaca Wilma[2], Rosero Carla[2], Bedón Ricardo[2], Vergara Eduardo[1], Manteca Angeles[1], Cueva Andrea[1], Bastidas Jacqueline[1], Ramírez Lisseth[1] and Echeverria Julian[1]

[1]Dermatology Resident, Universidad Tecnológica Equinoccial. Quito, Ecuador

[2]Department of Dermatology, Hospital General de Calderón, Quito, Ecuador

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***Corresponding author:** Ivonne Alejandra Riofrío Costa, Universidad Tecnológica Equinoccial, Federico Páez e14 214 u Guanguiltagua (Quito-Ecuador), Tel: +593- 96014645, email: ivo.aleriofrioc1008@gmail.com

ABSTRACT

Pemphigus is an unusual skin disease characterized by blistering of the skin and mucous membranes. It has a predilection for people of middle or advanced age; clinically it manifests with the appearance of blisters in the oral and genital mucosa, face, and extremities which are usually painful but do not cause itching. It can reach a mortality of up to 75% without treatment.

Keywords: Pemphigus vulgaris; Blister; Desmoglein

Abbreviations: DSG 3: Desmoglein 3; IL4: Interleukin 4; DSG1: Desmoglein 1

Introduction

Pemphigus, a term from Greek meaning blister, is a rare, chronic, autoimmune vesiculobullous disease that attacks the mucous membranes and skin. There are two main types; pemphigus vulgaris and foliaceous. Other classifications of pemphigus include paraneoplastic, erythematous, vegetative, and IgA subtypes. The diagnosis depends on clinical suspicion and confirmation by biopsy which demonstrates intraepithelial vesicle formation, acantholysis[1]. A Tzanck test which can be performed bedside demonstrates the presence of acantholytic cells.

In pemphigus vulgaris there are autoantibodies directed against Dsg1, mainly located in the superficial layers of the epidermis and Dsg 3, which tends to the basal layers. The formation of these autoantibodies is associated with the presence of CD4+ Th2 lymphocytes, which mainly secrete IL-4[2]; this induces a humoral immune response that promotes the differentiation of B lymphocytes into IgG4-secreting cells present in patients with pemphigus vulgaris and pemphigus foliaceus.

Case Presentation

A 36-year-old female patient, with history of left ovarian cancer, gastritis, duodenitis and hypothyroidism presented tense, painful blisters beginning in the oral mucosa which generalized to the upper limbs, then chest, then full body surface over the course of 3 months **(Figure 1 and Figure 2)**.

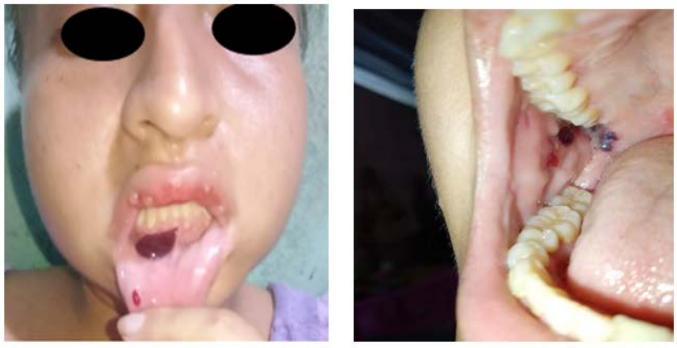

Figure 1: Dermatosis located at the level of the oral mucosa characterized by blisters, with serohematic content.

She was treated in a primary care unit where she received treatment with prednisone 60 mg/day. As her conditioned worsened, she was admitted to Hospital Eugenio Espejo, a tertiary care center, in Quito. She presented tense blisters at the level of the left upper limb and multiple post-inflammatory spots, as well as gastroesophageal reflux. At the time of admission, she had been without treatment for one month, so intravenous methylprednisolone at 1gram daily for three doses was instituted, resulting in a significant remission **(Figure 3)**.

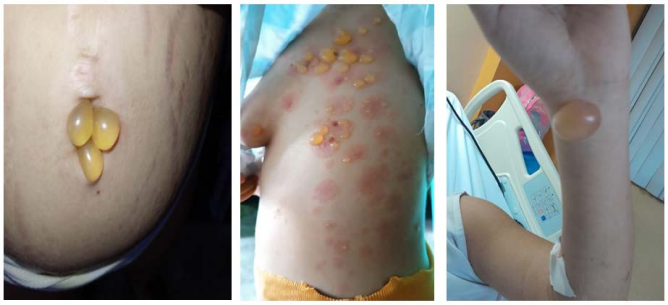

Figure 2: Dermatosis located at the level of the left rib cage, periumbilical area and left forearm characterized by tense blisters with serous content, on an erythematous base.

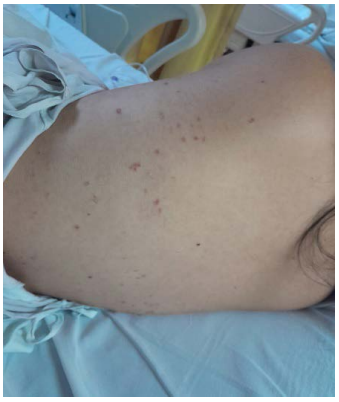

Figure 3: Post-inflammatory spots.

Discussion

Pemphigus vulgaris is a rare chronic blistering disease mediated by antibodies against adhesion molecules on keratinocyte cell surface. Factors such as increased age, comorbidities and larger affected body surface define the evolution and prognosis of the pathology[3].

The prevalence is 0.5 to 3.2 cases per 100,000 inhabitants and the incidence is 0.1 to 0.5 per 1000,000 inhabitants. Clinically, 75% of pemphigus vulgaris begin in the oral mucosa, initially being small blistering lesions that break easily, causing painful and bleeding erosions.

In the majority of patients, the cutaneous phase begins three months after the appearance of lesions in the oral mucosa. On the skin they appear in the same way as on the mucous membranes, on normal or slightly erythematous skin, located anywhere on the body, most frequently on the face, trunk and scalp. The diagnosis is made based on clinical suspicion and confirmed with biopsy[4].

Thanks to the introduction of corticosteroid therapy, mortality has decreased, the main cause of mortality being associated with infections and hydroelectrolyte imbalance.

Conclusions

Although it is a rare, autoimmune, rare and chronic disease, early diagnosis is important since its outcome can be fatal. Early diagnosis occurs through the detection of lesions in the oral mucosa, since the majority of cases begin at this anatomical site, relating and evaluating the accompanying comorbidities and the level of extension in order to provide adequate management.

Conflict of Interest: The authors declare no conflicts of interest.

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