

CASE REPORT

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Pemphigus vulgaris and koebner phenomenon

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Abstract

Pemphigus vulgaris is a rare disease characterized by loose bullae and erosions in the skin and mucous membranes. Koebner phenomenon is defined as the occurrence of new lesions that are characteristic of the disease at the site of trauma in normal-looking skin in other aspects. We report a 53-year-old woman who was followed for pemphigus. The patient's first complaints started 10 years ago, after tonsillectomy. The patient was operated 2 months ago for the application of spacer to infected hip prosthesis. One month after the operation, especially around the incision scar and in the mouth, common lesions emerged. Because the lesions around the incision scar appeared one month after the operation, it was interpreted as the Koebner phenomenon in this patient. In conclusion, it was aimed to draw attention to the fact that trauma may be a disease initiator factor or disease-triggering factor in remission, in patients with pemphigus vulgaris.

Keywords: Pemphigus vulgaris, Koebner phenomenon, trauma

Introduction

Pemphigus vulgaris is a rare autoimmune bullous disease characterized by loose bullae and erosions in the skin and mucous membranes [1]. The annual incidence of pemphigus in the world shows geographic differences and ranges from 0.5 to 16.1 per million [2]. Pemphigus has been reported to be triggered by various factors such as drugs, stressful events, radiation, burns, electrical damage and surgical intervention [1].

The isomorphic response or Koebner phenomenon is defined as the occurrence of new lesions that are characteristic of the disease at the site of trauma in healthy and normal-looking skin in other aspects [3]. The presentation of the case was found appropriate because of rare presence of the Koebner phenomenon in pemphigus disease.

Case

A 53-year-old female patient was followed with the diagnosis of pemphigus vulgaris. The patient's complaints started 10 years ago

as wounds that did not improve in the mouth after tonsillectomy operation. Approximately 4-5 months later, she had a diagnosis of pemphigus vulgaris after performing biopsy and direct immune fluorescence from the lesions on the skin and scalp. She had been on methylprednisolone and azathiopurine treatments for the first 5 years of her disease. Six years ago, she had a prosthetic replacement of the left hip joint due to aseptic necrosis of the femoral head. She had been receiving methylprednisolone and azathiopurine treatments at the time of the prosthesis application and she had no recent lesions. She received intravenous immunoglobulin (IVIG) treatment lasting 2 years. After IVIG treatment, remission was achieved for approximately 2.5 years.

The patient was operated 2 months ago for the application of spacer to the infected hip prosthesis. One month after the operation, especially around the incision scar and in the mouth, common lesions emerged. The distribution of the lesions of the patient with pemphigus vulgaris is given in Figure 1. She was admitted to our clinic 1 month after the onset of her symptoms and she was started on IVIG 2 g/kg/day and azathiopurine 2x50 mg / day. No biopsy was performed because the lesions were typical. The Nikolsky's sign was positive. Her laboratory values were within normal ranges. The patient's anti-desmoglein 3 value was 1000 U / mL and the anti-desmoglein 1 value was 36 U / mL, both above the cut-off value (> 20 U / mL).

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Figure 1. Distribution of lesions of the patient with pemphigus vulgaris
A. Erosions on tongue and right buccal mucosa
B. Placed erode plaques scattered on the back
C. Loose bullae and erode plaque in the right inguinal area
D. Wide plaque surrounding the incision scar on the outer face of the left thigh

Discussion

Although the pathogenesis of the Koebner phenomenon in pemphigus vulgaris is not known exactly, genetic factors, environmental triggers, local and systemic factors affecting the immune response and wound healing are blamed [4]. Dermatological diseases such as psoriasis, vitiligo and lichen planus are known to be Koebner positive, but the condition in pemphigus is controversial. In this disease, the Koebner phenomenon must be distinguished from Nikolsky's sign. In Nikolsky's sign, blisters appear in normal-appearing skin immediately after lateral pressure is applied to the skin. In order to be able to say 'Koebner phenomenon', there should be a period of time between trauma and bulla formation [5]. Because the lesions around the incision scar appeared one month after the operation, it was interpreted as the Koebner phenomenon in this patient.

In a study, the time between trauma and the onset of pemphigus was ranged from 7 days to 3 years and it was reported to be about 2 months [1]. Pemphigus is characterized by intraepithelial acantholysis caused by immunoglobulin G (IgG) autoantibodies against desmoglein 1 and 3, which are desmosomal transmembrane glycoproteins in keratinocytes. Increased expression of epidermal antigens and increased antigen presentation in genetically predisposed patients due to injuries that disrupt the dermoepidermal composition of the skin or oral mucosa is one of the theories that explain the Koebner phenomenon [6]. Anti-desmoglein antibody levels are associated with disease severity and disease activity [7]. Similar to the literature, desmoglein 3 and 1 levels were found to

be high during the active period in which the lesions recurred.

After ionized radiation, surgical interventions, burns, chemical peeling, periodontal surgery, tuberculin test and hair transplantation, new pemphigus lesions have been reported in the literature [1,5,8]. In a study evaluating 36 pemphigus patients induced by trauma in Iran, the hijama (cupping therapy) and laser treatment were highlighted as triggering factors [1]. In this case, pemphigus disease started with non-healing wounds after tonsillectomy. In a case report, it was reported that the lesions were not limited to the trauma area in the newly diagnosed pemphigus patient and then became generalized [4].

In a study by Daneshpazhooh et al. [1], some of the patients had lesions in the limited area of the trauma area. In another part, new lesions have been reported outside the trauma area. In this case, in a period when the patient was in remission without treatment, she had new lesions around the scar, around the body and in the mouth after the surgical intervention. The patient did not have a history of a new lesion after surgery in the period of using azothiopurine and methylprednisolone. This condition has been associated with the preservation of medical treatment. In conclusion, it was aimed to draw attention to the fact that trauma may be a disease initiator factor or disease-triggering factor in remission, in patients with pemphigus vulgaris.

Competing interests

The authors declare that they have no competing interest

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