Hallopeau type of pemphigus vegetans, an unusual clinical presentation of an infrequent disease

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Abstract

Pemphigus vegetans is a rare variant of pemphigus vulgaris, characterized by vegetative lesions that develop mainly in flexural areas. Herein, we present a middle-aged woman who presented to the emergency department with a rare cutaneous finding of bilateral axillary pustules that progressed over months to vegetant and malodorous plaques associated with subsequent painful erosions.

Keywords: pemphigus vulgaris, vesicobullous disease, autoimmune disease

Introduction

Pemphigus vegetans is a rare and infrequent variant of pemphigus vulgaris, representing 1-2% of all pemphigus [1]. It is more common in women and the age at diagnosis is 40 to 60 years [2]. A triggering factor is identified in several cases, such as medications, infections, or malignancies [3]. We report the case of a woman in the fourth decade of life with a history of hypothyroidism, who presented with vegetative plaques of the axillae and recent appearance of painful erosions in the oral cavity. She was previously treated with antibiotics for presumed inflammatory and infectious disease with no improvement.

Case Synopsis

A 37-year-old woman with hypothyroidism presented to the emergency department with a 4-month history of a painful, itchy, furuncle-like lesion in the right axilla, with the subsequent appearance of

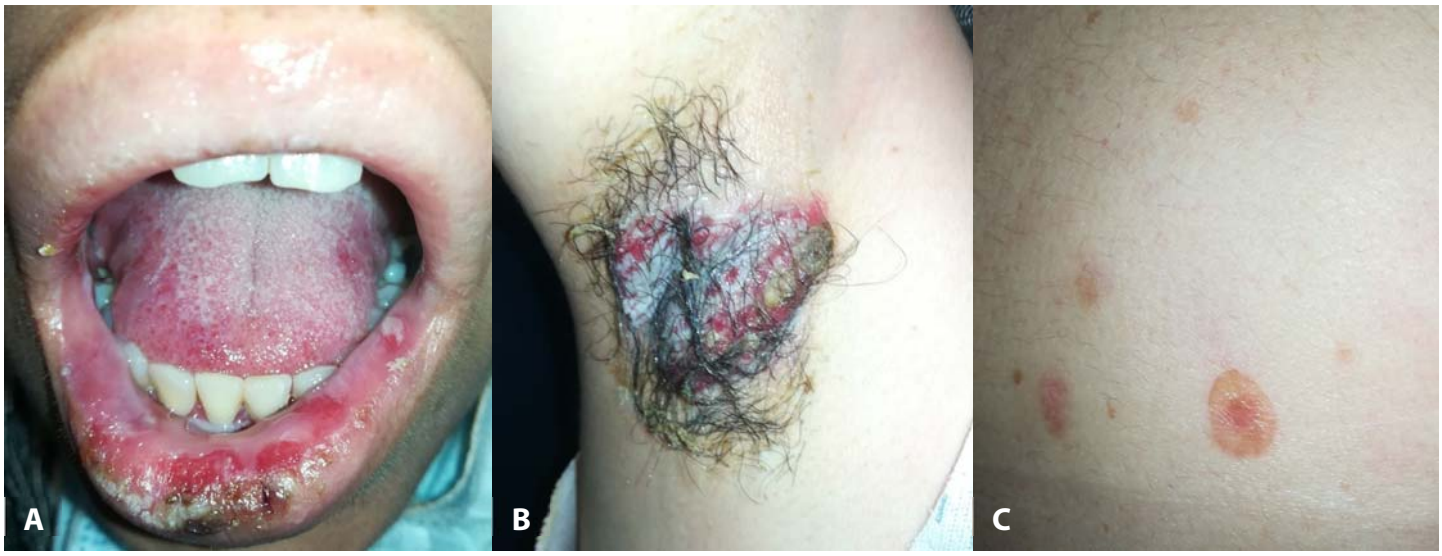

Figure 1. ***A)*** *Erosions and crusts of the oral mucosa.* ***B)*** *Erythematous, moist, vegetating plaques in the axillae with pustules on the surface.* ***C)*** *On the back, flaccid blisters with serous content and erythematous base.*

a similar lesion in the left axilla. She was previously prescribed topical and oral antibiotics on suspicion of hidradenitis suppurativa. In addition, she noted the recent appearance of painful lesions on the lips, tongue, and pharynx and eroded blisters on the chest.

On physical examination, multiple erosions were observed of the pharynx, tongue, cheeks, and lower lip, some of them covered with superficial bloody crust. The anterior thorax, abdomen, and back exhibited multiple erosions, and some flaccid blisters with positive Nikolsky sign. Armpits showed bilateral, vegetative, erythematous, eroded, painful, moist plaques that limited abduction (**Figure 1**). No lesions were observed in the genital mucosa or nail plate.

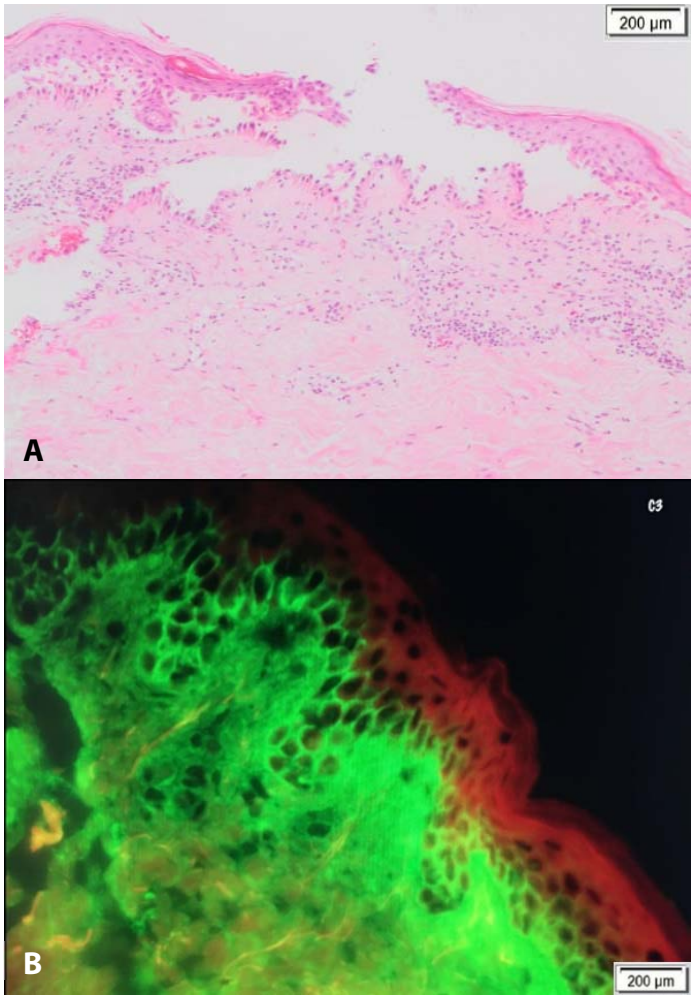

Figure 2. ***A)*** *Eroded zone with detached necrotic epithelium and a suprabasal blister with acantholytic cells, H&E. In the dermis, edema and perivascular and interstitial inflammatory infiltrate constituted by lymphocytes, neutrophils, and eosinophils.* ***B)*** *Intercellular C3 complement binding.*

With these findings, a diagnostic impression of pemphigus vegetans was made. A biopsy of a lesion on the back was performed, which showed typical findings of pemphigus vulgaris. Direct immunofluorescence identified intercellular epidermal deposits of IgG and C3 (**Figure 2**). Extensive evaluation for a hidden neoplasm or other associated autoimmune phenomenon was performed and revealed neither.

Prednisone was initiated at 1mg/kg/day, and clobetasol propionate applied in an oral analgesic base. Once oral intake was resumed she was discharged to follow up as an outpatient. Disease control was achieved after three months of prednisone treatment. She remains lesion free with a 10mg prednisone daily dose, plus calcium and vitamin D3 supplementation.

Case Discussion

Pemphigus vegetans is a rare variant of pemphigus vulgaris that presents with flaccid blisters that erode and form vegetating papules that coalesce in erythematous, moist, malodorous plaques that can secrete purulent material, predominantly in intertriginous areas. Oral involvement is frequent (60-80%) and may be present since the beginning of the disease in more than half of patients; pain may limit food intake [1]. Two variants have been described: Hallopeau and Neumann. The Hallopeau subtype is characterized by starting with grouped pustules that erode, forming painful vegetative lesions. This type usually has an adequate response to treatment. The Neumann variant starts with the appearance of blisters (similar to pemphigus vulgaris) that subsequently form exudative vegetative lesions; this form often shows a poorer response to management [2]. Our patient's skin findings and their evolution were consistent with the rare pemphigus vulgaris variant Hallopeau type, owing to the characteristic development of pustules in the axillae, which coalesced, eroded, and progressed to vegetative, moist erythematous plaques.

The histopathology of the vegetative lesions shows suprabasal intraepidermal clefts, acantholytic cells, pseudoepitheliomatous hyperplasia, and intraepi-

dermal microabscesses composed of eosinophils and neutrophils. Direct immunofluorescence shows deposition of IgG and C3 in intercellular junctions [1].

The exact pathophysiology is unknown but it has been associated with HIV infection, herpes simplex, deep vein thrombosis, use of intranasal cocaine, solid organ neoplasms, and medications [1,3-5].

The first line treatment is with systemic corticosteroids at a dose of 1mg/kg/day, associated with immunosuppressants such as azathioprine, cyclophosphamide, cyclosporine, mycophenolate mofetil, and methotrexate. For localized and limited presentations, the use of topical or intralesional corticosteroids can be considered. In addition, dapsone, tetracyclines, rituximab, and others have been described [2,6-7]. Our patient responded rapidly to first line treatment, which supports the diagnosis of Hallopeau type pemphigus vegetans.

Conclusion

Pemphigus vegetans is a rare variant of pemphigus vulgaris that should be considered in patients with vegetative lesions predominantly in folds. The diagnosis can be confirmed by histopathology and immunofluorescence, the findings of which vary according to the type of biopsied lesion.

It was determined that the patient had pemphigus vegetans type Hallopeau, of primary origin, which contributes to a small number of cases of this variant in the Latin American population. With this report, we seek to emphasize the importance of knowing the clinical characteristics of this entity and of taking a skin biopsy to confirm the diagnosis.

Potential conflicts of interest

The authors declare no conflicts of interests.

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