Pemphigus vegetans with isolated involvement of the nose and chest: rare variant of pemphigus vulgaris*

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Abstract: Pemphigus vulgaris is chronic bullous disease that manifests as bullae and erosions of skin and mucosas, with intraepidermal suprabasal cleft formation seen in the histological examination. It has a rare variant called pemphigus vegetans, where vesicles and bullae are replaced by pustular, verrucous and hyperpigmented lesions, mainly in skin folds. The treatment is similar to that for classic pemphigus vulgaris. The authors present an exuberant case of pemphigus vegetans, covering the nose and chest exclusively, without oral or flexural lesions.

Keywords: Basement membrane; Pemphigus; Skin diseases, vesiculobullous; Treatment outcome

INTRODUCTION

Pemphigus vulgaris is a chronic bullous dermatosis, characterized by bullae and erosions of skin and mucosas, caused by IgG autoantibodies against desmosomal glycoproteins Desmoglein 1 (Dsg 1) and 3 (Dsg 3), present on the surface of basal keratinocytes, resulting in loss of cell adhesion and the formation of intraepidermal cleft.[1-3]

In PV, vesiculobullous lesions rupture easily, leaving erosions and ulcers, mainly in oral mucosa.[4] Pemphigus vegetans (PVeg) is a rare variant of PV, corresponding to 1–2% of cases, that is localized predominantly on flexural surfaces such as the axillae and groin, where pustules or vegetative plaques of verrucous and hyperpigmented aspect are visible.[5,6]

CASE REPORT

An 89-year-old male was referred from the basic health unit with suspected skin cancer. He had crusted vegetative lesions in the anterior nasal cavity and on the nasal tip, without oral lesions (Figure 1). There was an erythematous plaque with ulcerations, thick crusts, and a flaccid bulla with purulent content in its lower portion, with three months of evolution and no improvement after antibiotic therapy (Figure 2).

After biopsy of the two affected areas, the histopathological examination of the lesions showed epidermis with hyperkeratosis, foci of parakeratosis, suprabasal intraepidermal cleft, and acantholytic cells along the basal layer and the hair follicles; and in the dermis, mononuclear inflammatory infiltrate in between the

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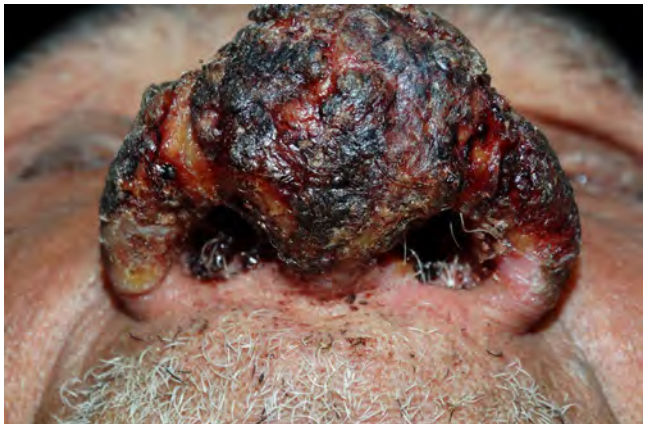

Figure 1: Thick crusts in anterior nasal cavity and on nasal tip with necrotic aspect

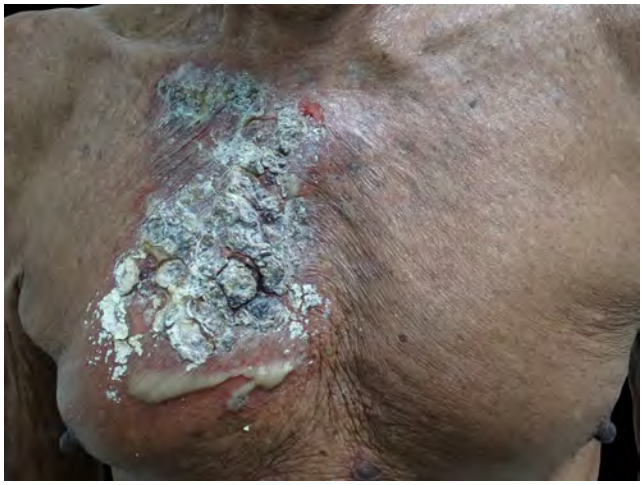

Figure 2: In chest, erythematous plaque, ulcerations, thick crusts, and flaccid bulla with purulent content in its lower portion

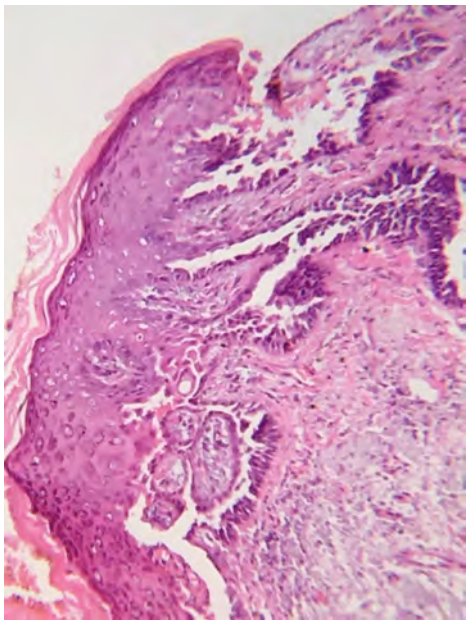

Figure 3: Histopathology examination showing hyperkeratosis, epidermal acanthosis, suprabasal cleft with acantholytic cells along the basal layer. (Hematoxylin & eosin, x400)

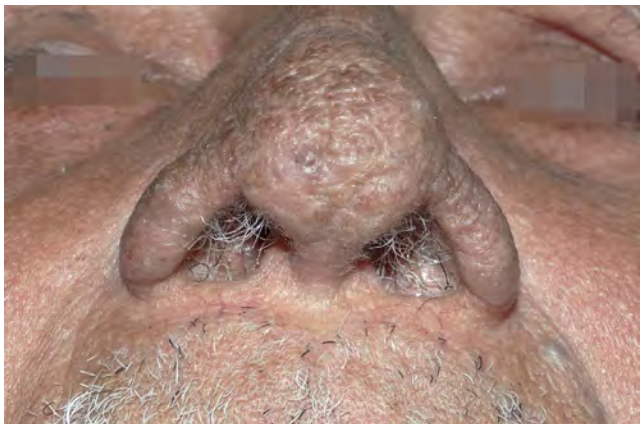

Figure 4: Clinical aspect of nose after treatment with prednisone

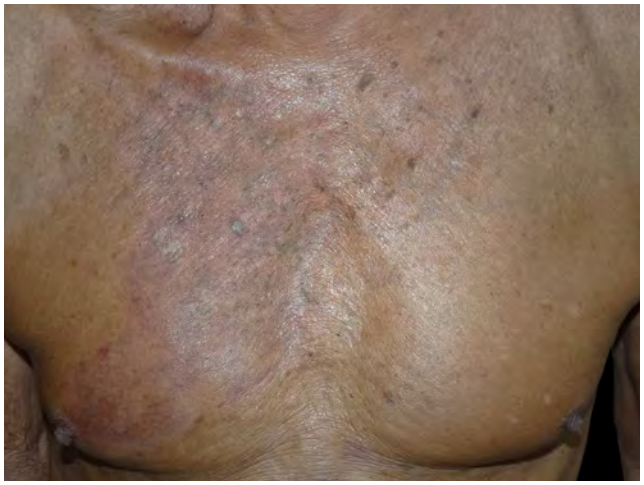

Figure 5: Chest without lesions or scars after treatment

basophilic degeneration of collagen (Figure 3). These findings were compatible with pemphigus vulgaris. In view of the clinical aspect and histopathological findings, the diagnosis of pemphigus vegetans was confirmed.

The patient was treated with prednisone at a dose of 1 mg/kg/day, with excellent response to treatment, evolving to complete remission of the condition (Figures 4 and 5).

DISCUSSION

Originally described by Neumann in 1876, PVeg is a rare variant of PV, with two known subtypes: Neumann and Hallopeau.[2,7] The lesions of this type of pemphigus are primarily flexural but can affect other sites. Isolated involvement of the lips, face and foot has been described.[2,5,6] The clinical course is variable and may resemble PV, with periods of improvement and exacerbation, often requiring immunosuppressive drugs, or evolving to remission and control with low doses of corticosteroids.[5,6]

In addition to autoantibodies against Dsg 1 and Dsg 3, as in PV, there are reports of the presence of antibodies against des-

mocollin 1, desmocollin 2 and periplakin in PVeg cases.[7] Captopril and Enalapril, angiotensin-converting enzyme inhibitors, have been associated as inducers of this rare form of pemphigus.[5,7]

The histopathological findings that can be seen in PVeg are suprabasal acantholysis (equal to classic PV), epidermal hyperplasia, papillomatosis and eosinophilic infiltrate (intraepidermal eosinophilic microabscesses).[5,6,7]

In the reported case, PVeg presented with predominance of crusts and verrucous surface affecting only the nose and chest, without lesions of the oral mucosa or flexural areas. The clinical diagnosis was confirmed by the presence of suprabasal intraepidermal cleft with acantholytic keratinocytes in histopathological examination. Despite the exuberance of the lesions, the disease presented a benign course, evolving to total regression after systemic corticosteroid therapy.❑

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