

Plasma cell mucositis with exclusive involvement of the perianal region

Abstract

We present the case of a woman who has had perianal ulcers for several years. Since no improvement was achieved with topical treatments, a biopsy was performed, showing a dense lymphoplasmocellular infiltrate. After ruling out other differentials, a diagnosis of plasma cell mucositis was made and intralesional betametasone was attempted, with almost complete resolution of the lesions in a few days and no recurrence in one-year follow-up.

Keywords: plasma cell mucositis, idiopathic lymphoplasmocellular mucositis-dermatitis, zoon mucositis

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Clinical findings

A 67-year-old woman presented with a 4-year history of painful cutaneous lesions on her perianal region, which had been treated with high-potency topical corticosteroids, antibiotics, calcineurine inhibitors and antifungals without improvement. She referred slow but progressive growth of the ulcers, with no periods of remission. She denied gastrointestinal symptoms, fever or weight loss. She also denied manipulation of the area or application of irritating products. Her personal and family history were unremarkable apart from hypothyroidism treated with levothyroxine.

Physical examination revealed 2cm, well defined erosions with a slightly raised, erythematous border, with no exudation, symmetrical, on the lower region of her intergluteal fold (Figure 1a). On the upper aspect of this fold a smaller ulcer could be observed (Figure 1b). Perilesional skin showed a shiny, whitish discoloration. Routine laboratory tests including quantiferon test, C-reactive protein, erythrocyte sedimentation rate, serologies for lues, hepatitis B and C and human immunodeficiency virus, and chest X-ray were normal. Biopsies from both lesions were taken.

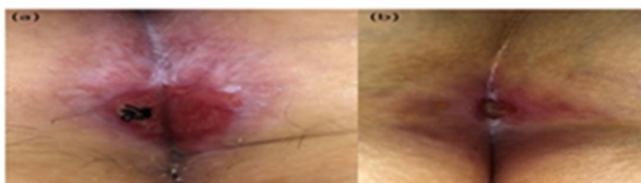


Figure 1 (a) Erythematous, erosive plaque surrounded by whitish perilesional skin on the lower aspect of the intergluteal cleft. (b) Smaller ulcer with erythematous perilesional skin on the upper aspect of the intergluteal cleft.

Histological findings

Histopathological findings of both lesions were similar and showed hyperkeratosis and hypergranulosis with no interface dermatitis and a dense lymphoplasmocitary infiltrate (Figure 2). No granulomas or

microorganisms were observed. Immunohistochemistry techniques were performed, showing positivity for IgG4 lower than 1%, no light-chain restriction and negativity for anti-treponema antibody. These findings supported the diagnosis of plasma-cell mucositis.

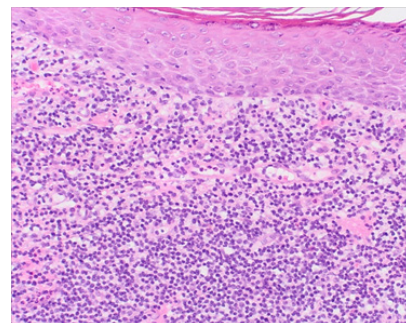


Figure 2 Histopathological features of the largest ulcer, showing a dense infiltrate compound of lymphocytes and plasma cells.

Discussion

Plasma-cell mucositis was first described by Zoon¹ in 1952, for which has also received the name of Zoon mucositis¹. This article described lesions on the penis and, since then, cases affecting the vulva, glans, oral mucosa, conjunctiva and vagina have been reported.^{2,3} In 2014 Mitkov et al.,² reported the first case of plasmocellular mucositis affecting perianal region. Plasma-cell mucositis is more common in postmenopausal women and in males on the fifth and sixth decade of life. These lesions have received many names, some of them making reference to the anatomic location and others to the clinical aspect. To avoid confusion some authors have proposed the term “idiopathic lymphoplasmocellular mucositis-dermatitis” to include this type of affection.⁴

Lesions consist of erythematous plaques, sometimes ulcerated or eroded, with well-demarcated borders and frequently involving genital region (vulva, glans), and that can be asymptomatic or produce itching,

pain or bleeding. Etiology is unknown, although cases secondary to trauma and autoimmune diseases have been reported.⁵ Although considered a benign condition, some authors think that, when affecting the glans, it could be considered as a premalignant condition, due to the maintained, chronic inflammation.⁶ Histopathology examination reveals a non-neoplastic, dense, rich in plasma cells infiltrate with epidermal atrophy, spongiosis and dilated capillaries. Differential diagnosis includes extra mammary Paget's disease, erosive lichen planus, squamous cell carcinoma, cutaneous tuberculosis and sexually transmitted diseases. In our case, immunohistochemistry stains (CD3, kappa and lambda chains, IgG4, anti-treponema antibody) confirmed the non-neoplastic nature of the cells and the absence of spirochaete. IgG4 stained positive for less than 1% of the cells, ruling out the possibility of an IgG4-related disease.

There is no standard treatment, and different responses to both topical and intralesional corticosteroids, topical antibiotics, tacrolimus and imiquimod have been reported.² Since our patient had already used several topical treatments without improvement, we decided to infiltrate betametasone achieving a fast healing of the lesions with no recurrence after one year follow-up (Figure 3a&3b).

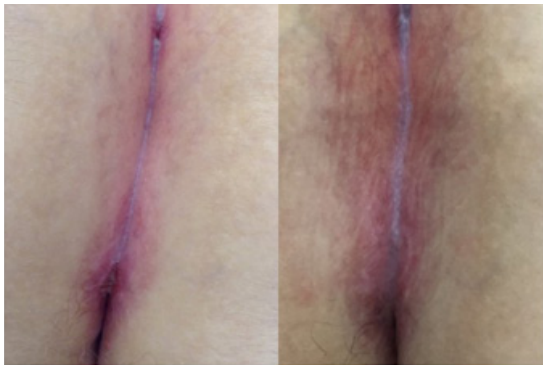


Figure 3 (a) Evolution of the lesions five days after one infiltration with betametasone. **(b)** one year later

We consider this case of interest for dermatologists, since differential diagnosis of perianal plaques is wide, and only one case of this entity with exclusive affection of the perianal region had been reported before; besides, we would like to highlight the fast and sustained response with intralesional corticosteroids, suggesting this treatment if topical therapy fails.

Conflicts of interest

The author declare that there is no conflicts of interests.

Acknowledgments

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