# Case Report

## Solitary ulcerated plaque on the face – an unusual presentation of cutaneous plasmacytosis?*

Ana Marta António[1]
Ricardo Coelho[2]
João Vitor Alves[1]
Elvira Bártolo[1]

DOI: http://dx.doi.org/10.1590/abd1806-4841.20175355

**Abstract:** Cutaneous and systemic plasmacytosis is a rare disorder characterized by cutaneous polyclonal plasma cell infiltration frequently associated with polyclonal hypergammaglobulinemia and lymphadenopathy. We report a case of a 67-year-old woman with an inflammatory ulcerated plaque in the left masseter region. A skin biopsy showed dense perivascular infiltrate of mature plasma cells in the dermis without atypia and immunoglobulin light chain restriction. After physical examination and further investigation, we ruled out systemic disease. Our patient was successfully treated only with hydrocortisone cream application. Few cases of isolated benign primary cutaneous plasmacytosis have been described, particularly in children. After excluding the diagnosis of a reactive process to an infection, which is unlikely in this case, we suspected of a rare manifestation of primary cutaneous plasmacytosis in adults with distinct presentation and clinical course.
**Keywords:** Immunoglobulin light chains; Hypergammaglobulinemia; Plasmacytoma

### INTRODUCTION

Plasma cells may be a prominent component of inflammatory infiltrates in a variety of dermatoses and neoplastic disorders. They can also be found in lesions of the forehead and scalp previously submitted to cryotherapy.[1] Cutaneous and systemic plasmacytosis is a rare disorder typically described in Asian patients, characterized by a cutaneous polyclonal plasma cell infiltrate frequently associated with polyclonal hypergammaglobulinemia.[2,3] It usually manifests as multiple red to dark brown patches located on the trunk. It may be accompanied by extracutaneous involvement with superficial lymphadenopathy. Infiltration of lung, liver, spleen and kidney occurs less frequently.[3] The diagnosis of primary cutaneous plasmacytosis is assumed based on histologic findings, presence of a polyclonal population of plasma cells and negative results from a diagnostic workup for systemic disease.[4] It is very important to differentiate cutaneous polyclonal plasmacytosis from malignant plasmacytoma, which is characterized by the proliferation of monoclonal plasma cells that could also appear in the skin, usually as metastatic lesions of multiple myeloma and more rarely without evidence of systemic disease in primary involvement of the skin. Clinical lesions could be identical to those of cutaneous and systemic plasmacytosis as reported in this publication of multiple primary cutaneous plasmacytoma .[5]

### CASE REPORT

We report a 67-year-old woman presented with a 3-month history of a large asymptomatic ulcerated lesion on her face. The patient was otherwise well and denied fever, malaise, night sweats, weight loss, or other constitutional symptoms. Physical examination revealed an ulcerated erythematous plaque with 7 cm in longest axis, located in the left masseter region (Figure 1). We also observed multiple actinic keratosis over the zigomatic and nasal regions, but no actinic keratosis was observed adjacent to the ulcerated plaque. The patient denied previous history of treatments like local cryotherapy. There was no lymphadenopathy or hepatosplenomegaly. Based on clinical features, squamous cell carcinoma or other ulcerated skin neoplasm were the most probable diagnoses. We performed two punch biopsies and histopathological examination showed a dense perivascular infiltrate of mature plasma cells without evident atypia in the superficial and deep dermis (Figure 2). Immunohistochemical study revealed that a great number of the infiltrating cells were CD138-positive. Plasma cells expressed both kappa and lambda light chains. We noted no evidence of clonal immunoglobulin gene rearrangements or human herpesvirus type 8 infection. Routine bacteriology, mycobacteriology, and mycology cultures were negative. Further investigation excluded systemic disease. Laboratory tests revealed no abnormalities in hemogram, lactate dehy-

Received on 10.11.2015
Approved by the Advisory Board and accepted for publication on 21.02.2016
* Work performed at the Dermatovenereology Service, Hospital Garcia de Orta – Almada, Portugal.
Financial support: none.
Conflict of interest: none.

[1] Dermatovenereology Service, Hospital Garcia de Orta – Almada, Portugal.
[2] Dermatology Service, Centro Hospitalar Conde de São Januário – Macau, China.

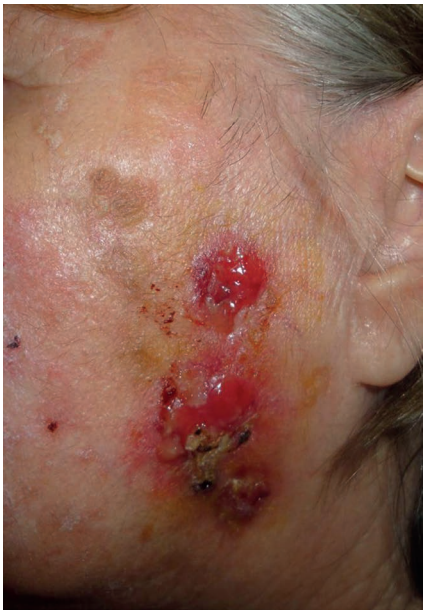

**Figure 1:** Well-defined, ulcerated and erythematous plaque, covered by crust in the left masseter region

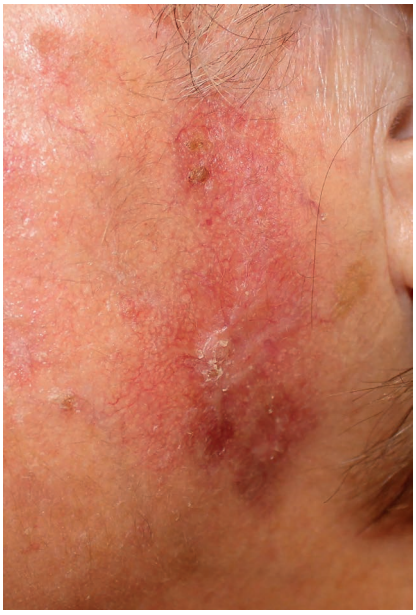

**Figure 3:** Clinical lesion after three weeks of therapy – erythema and crusting

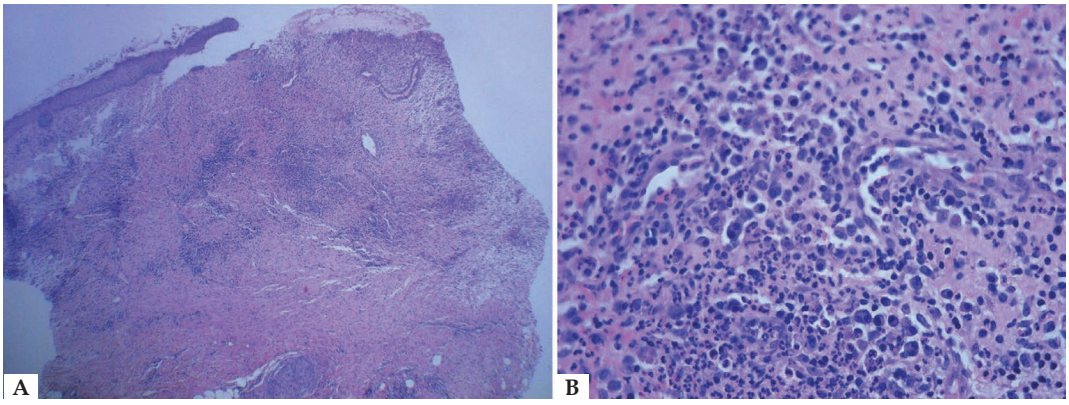


**Figure 2:**
**a)** Dense perivascular inflammatory infiltrate in the superficial and deep dermis (Hematoxylin & eosin 40x); **b)** Inflammatory infiltrate with mature plasma cells without evident atypia in the deep dermis (Hematoxylin & eosin 400x)

drogenase, erythrocyte sedimentation, or serum proteins. Tests for syphilis, hepatitis B and C, and HIV infection were negative. Serum immunoelectrophoresis and urinary electrophoresis were normal. Antinuclear antibody titer was negative. Bone marrow study was not remarkable. Flow cytometry on peripheral blood revealed normal B and T cells and no evidence of malignant lymphoproliferative disorder. Thoracoabdominal computed tomography was normal, excluding lymphadenopathies and organomegaly. Plasma level of interleukin-6 was < 2,0 pg/mL (normal value < 17,0 pg/mL). This cytokine induces the final differentiation of B cells into plasma cells and is elevated in some cases of plasmacytosis, particularly when systemic involvement is present.[3] Assuming the diagnosis of a reactive inflammatory process or a rare manifestation of cutaneous plasmacytosis, we treated the patient with hydrocortisone cream applied at night for 1 month. We observed an excellent response to therapy with complete healing of the lesion in 4 weeks (Figure 3). After 1 year of follow-up, there was no evidence of relapse.

## DISCUSSION

The etiopathogenesis of cutaneous and systemic plasmacytosis is uncertain. The prevailing hypothesis advocates that it represents a reactive plasmacytic disorder, an overreaction to unknown stimuli, which includes trauma, infections or malignancies.[3,4,6,7] A few cases described in literature point to the existence of a new entity within the spectrum of primary cutaneous and systemic plasmacytosis manifestations – an isolated benign primary cutaneous plasmacytosis, without any sign of systemic involvement occurring particularly in children.[4,8] The present case is peculiar due to its clinical and histopathological features. The plasmacytic infiltrate found in biopsy could be correlated to an immunological reaction to infection of a tumor and/or trauma occurred on the face. However, no features of these were found in the clinical and histological examination or history. Possibly, it can also represent a rare manifestation of cutaneous plasmacytosis. If so, this case supports the existence of a primary cutaneous plasmacytosis characterized by mature plasma cell infiltration without systemic findings. Moreover, the age of

onset, localization of the lesion, absence of hypergammaglobulinaemia, lymphadenopathy and hepatosplenomegaly, presence of an infiltration of mature polyclonal plasma cells restricted to the skin, and the excellent response to therapy were different from those described so far. Despite the good response to therapy and the benign course of this case, a careful monitoring of such patients is recommended. ❑

**REFERENCES**


1. Weedon D. Plasma cell infiltrates. In: Weedon D. Weedon's Skin Pathology. London: Churchill Livingstone; 2010. p. 943.
2. Honda R, Cerroni L, Tanikawa A, Ebihara T, Amagai M, Ishiko A. Cutaneous plasmacytosis: report of 6 cases with or without systemic involvement. J Am Acad Dermatol. 2013;68:978-85.
3. Leonard AL, Meehan SA, Ramsey D, Brown L, Sen F. Cutaneous and systemic plasmacytosis. J Am Acad Dermatol. 2007;56:S38-40.
4. Gilliam AC, Mullen RH, Oviedo G, Bhatnagar R, Smith MK, Patton DF, et al. Isolated benign primary cutaneous plasmacytosis in children: two illustrative cases. Arch Dermatol. 2009;145:299-302.
5. Saback TL, Botelho LF, Enokihara MM, Michalany NS, Floriano MC. Multiple primary cutaneous plasmacytoma: first reported case in Brazil. An Bras Dermatol. 2012;87:629-31.
6. Martín JM, Calduch L, Monteagudo C, Molina I, Ramón D, Alonso V, et al. Cutaneous plasmacytosis associated with lung and anal carcinomas. J Eur Acad Dermatol Venereol. 2006;20:428-31.
7. Cannistraci C, Lesnoni La Parola I, Donati P, Amantea A, Cordone I, Berardesca E. Case of benign cutaneous plasmacytosis: immunohistochemical and flow cytometry study. J Eur Acad Dermatol Venereol. 2010;24:111-2.
8. Shiba Y, Satoh T. Isolated benign primary cutaneous plasmacytosis. Int J Dermatol. 2014;53:e397-8.



*MAILING ADDRESS:*
*Ana Marta Salvado Ferreira António Daniel*
*Avenida Torrado da Silva,*
*2801-951, Almada, Portugal.*
*E-mail: anamartaantonio@gmail.com*


**How to cite this article:** António AM, Alves JV, Coelho R, Bártolo E. Solitary ulcerated plaque on the face - an unusual presentation of cutaneous plasmacytosis? An Bras Dermatol. 2017;92(3):410-2.