

Multiple primary cutaneous plasmacytoma: first reported case in Brazil *

Plasmocitoma cutâneo primário múltiplo: primeiro caso relatado no Brasil

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Abstract: Primary cutaneous plasmacytoma is a rare disease characterized by monoclonal proliferation of plasma cells in the skin, in the absence of bone or systemic disease. It can be solitary or multiple, the latter being even more rare and presenting a higher mortality rate. We describe the clinical, histopathological and immunohistochemical aspects as well as the evolution of an 87-year-old female patient, diagnosed as having multiple primary cutaneous plasmacytomas.

Keywords: Brazil; Lymphoma; Plasmacytoma

Resumo: O plasmocitoma cutâneo primário é uma entidade rara, caracterizada pela proliferação monoclonal de plasmócitos na pele, na ausência de doença óssea ou sistêmica. Pode ser único ou múltiplo, sendo esta última forma ainda mais rara e de pior prognóstico. Descreveremos os aspectos clínicos, histopatológicos, imunohistoquímicos e a evolução de um caso de paciente do sexo feminino, 87 anos, com diagnóstico de plasmocitoma cutâneo primário com múltiplas lesões.

Palavras-chave: Brasil; Linfoma; Plasmocitoma

INTRODUCTION

A plasmacytoma is a malignant tumor characterized by proliferation of monoclonal plasma cells. The primary involvement of the skin without evidence of systemic or bone disease is extremely rare.^{1,2,3} According to the classification of skin lymphomas, approved in 2005 by the World Health Organization-European Organization for Research and Treatment of Cancer (WHO-EORTC), extramedullary plasmacytomas of the skin are nowadays included within the

group of primary cutaneous marginal-zone B-cell lymphoma.⁴ We describe the first reported case of multiple primary cutaneous plasmacytoma in Brazil.

CASE REPORT

An 87-year-old female presented with a six-month history of painless, disseminated erythematous-violaceous papules and plaques, well circumscribed, measuring 0.5 to 2.5 cm on the chest,

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FIGURE 1: Disseminated erythematous-violaceous papules and plaques

abdomen, upper and lower extremities (Figure 1). Her past medical history included classic Kaposi's sarcoma (CD 34 and Human Herpes Virus 8 were positive in tumor cells on immunohistochemical analysis), which was treated with radiotherapy in 1993, and Alzheimer's disease since 1999.

Skin biopsy demonstrated diffuse proliferation of plasma cells in superficial and deep dermis (Figure 2). On immunohistochemical analysis for kappa and lambda, light chain restriction was demonstrated with a ratio of kappa/lambda 7:1 confirming the monoclonal nature of the plasma cells, which were also positive for CD138 (Figure 3).

The patient underwent screening laboratory tests (blood cell count, creatinine, calcium, immunoglobulins, urine), skeletal exams and bone marrow biopsy. All data collected excluded the possibility of multiple myeloma and supported the diagnosis of multiple primary cutaneous plasmacytoma.

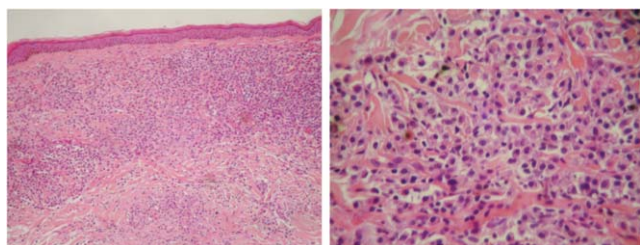


FIGURE 2: Light microscopy with diffuse proliferation of plasma cells in superficial and deep dermis (HE100x/HE400x)

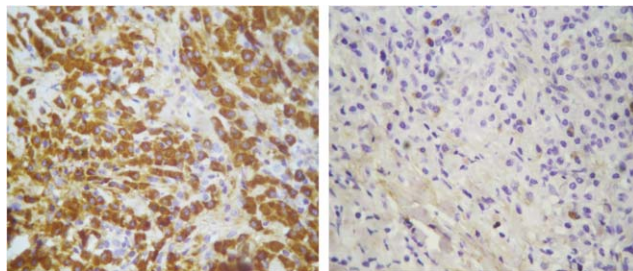


FIGURE 3: Immunohistochemical analysis for kappa and lambda, light chain restriction was demonstrated with a ratio of kappa/lambda 7:1 (400x)

Considering the patient's age, comorbidities and lack of change in quality of life, the option of delaying chemotherapy was preferred. After 12 months of rigorous follow-up the patient showed no signs of systemic disease.

DISCUSSION

Cutaneous plasmacytoma can either present as a primary or a secondary tumor. The most common in primary presentation is the solitary plasmacytoma of the bone, originally from the bone marrow. The primary extramedullary forms occur in the upper respiratory tract, gastrointestinal tract, lymphoreticular system and skin, in absence of systemic disease. The secondary forms result from hematogenic systemic disease, the multiple myeloma, or extending directly from the bone lesions.^{1,5,6,7} The mean age of the patients is 59.5 years, with male predominance.^{2,7}

Clinically, the lesions are erythematous-violaceous cutaneous or subcutaneous papules, plaques and/or nodules, with a smooth-surface, ranging from 1 to 5 cm in diameter. Ulceration of the lesions is rarely observed. The tumor can be solitary or multiple, most commonly found on the face, trunk and extremities.^{1,4,5,7}

Histologically, a dense monomorphic plasmacytic infiltrate is found in the superficial dermis, possibly involving the reticular dermis and subcutaneous tissue. Atypia, binucleation and increase in the mitotic activity can be present in plasma cells.^{1,5,7} The epidermis is generally spared. Immunohistochemistry allows to distinguish reactive plasma-cell proliferation by infections, inflammatory processes or other neoplasms (plasmacytosis) from monoclonal plasma cell proliferation (plasmacytoma).^{5,8} A positive CD138, CD79a and CD38 supports plasma cell lineage. CD20 is typically negative. The plasmacytoma displays clonal kappa or lambda light chains, as a marker of malignancy.^{1,5,9}

The preferred mode of therapy in the case of solitary lesions is surgical excision, local radiotherapy or the combination of both. Systemic chemotherapy is

indicated in patients with multiple lesions.^{1,2,5,9}

Very few cases were described in the literature, therefore it is challenging to evaluate the risks of the transformation to a systemic disease, although it is known that the risks are higher in patients with exten-

sive and multiple lesions, explaining the worse prognosis and higher mortality rate described in this group.^{2,3,7,9,10} Follow up in these cases must be rigorous with appropriate and efficient treatment in case the disease evolves into a multiple myeloma.¹ □

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