Case for diagnosis*

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CASE REPORT

A 72-year-old man in complete remission of an immunoglobulin G kappa multiple myeloma. Ten months later he presented firm painful erythemato-violaceous or flesh color nodules and tumors on the trunk, axillae and face (Figures 1 and 2). The patient referred concomitantly asthenia and fever for 3 days. An incisional biopsy showed a dense infiltrate of plasmablasts in the dermis and hypodermis (Figure 3). The immunohistochemistry evidenced a dense infiltrate of monoclonal plasma cells expressing kappa light chains, strongly positive for CD138 (Figure 3). Serum immunoelectrophoresis identified monoclonal IgG 2152 mg/dL (700-1600 mg/dL) and κ chains 702 mg/dL (170-370 mg/dL). Urine immunoelectrophoresis showed monoclonal κ chains: 0,925 mg/24h (0-0,7 mg). Bone marrow biopsy evidenced plasmocytosis with cellularity of almost 100%.

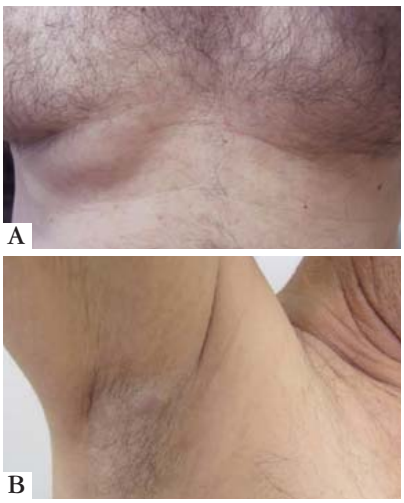

FIGURE 2: A - Clinical images with more details of the cutaneous lesions on the right inframammary area and **B -** on the right axilla

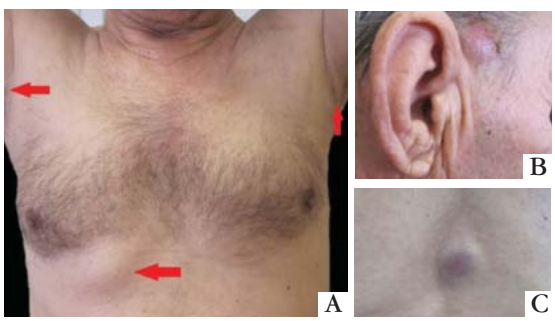

FIGURE 1: A - Clinical images of the axillary lesions and of the right inframammary tumor, **B -** Images of the cutaneous lesions on the face and **C-** on the back

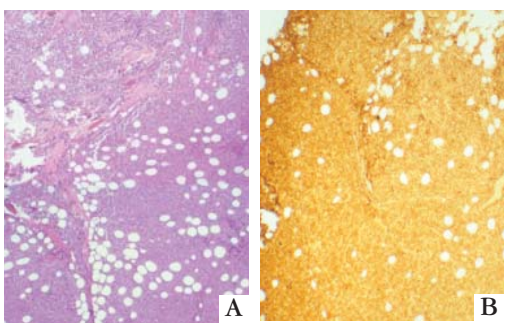

FIGURE 3: A - Histopathologic features of the dense infiltrate of plasma cells in the dermis and hypodermis (Hematoxylin-eosin stain, original magnification X 400). **B -** Immunohistochemical staining of neoplastic plasma cells with CD138 antibody

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DISCUSSION

Cutaneous involvement associated to multiple myeloma varies from 5 to 10% of multiple myeloma.[1] The cutaneous plasma cell tumors may arise from hematogenic spread or from direct extension of bone lesions. Metastatic skin lesions without adjacent bone involvement are rare.[2,3,4,5]

Cutaneous metastatic plasmocytomas are clinically erythemato-violaceous cutaneous or subcutaneous papules, plaques and/or nodules, with a smooth-surface, hard consistency, ranging from 1 to 5 cm in diameter, solitary or multiple. Any area of the skin can be involved, but it has been reported most frequently on the trunk and abdomen, followed by the scalp, face, neck, lower extremities and upper extremities.

No lytic lesion of bone should be evidenced directly below by x-ray.

These specific lesions are mainly associated with Ig G (56%) but in any of the others myeloma proteins may be involved. It is currently apparent that the risk of cutaneous involvement is independent of the immunoglobulin class type. Histopathology reveals the typical pattern of a dense monomorphic dermal plasmacytic infiltrate. Immunohistochemical study demonstrates monoclonality of plasma cells with strong immunoexpression for CD 79a and CD 138. Several entities can have a similar appearance, namely cutaneous sarcomas or cutaneous B-cell lymphomas.

Therapy of cutaneous plasmocytomas in the setting of multiple myeloma includes chemotherapy (melphalan and prednisolone) and local radiotherapy. Surgical excision has a role in lesions resistant to radiotherapy. Our patient was treated with melphalan, prednisolone, radiotherapy, bortezomib, dexamethasone and chemotherapy due to new extra-cutaneous plasmocytomas (pelvic, pulmonary, ganglionary and ocular).

The specific cutaneous lesions are a sign of poor prognosis, leading to death within 12 months after diagnosis.[2,5] Less than 20% remain progression-free at 5 years. Despite cutaneous metastasis in multiple myeloma usually indicating aggressive behavior, longer survivals are possible.[2,3] For example, after 2 years of follow-up, no cutaneous metastasis recurrences were observed in our patient.

The cutaneous metastatic plasmocytomas can be the first sign of the progression of Multiple Myeloma or signal a deteriorating clinical course in a preexisting disease. ❑

Abstract: Cutaneous involvement associated to multiple myeloma varies from 5 to 10% of cases and is infrequently recognized. Cutaneous metastatic plasmacitomas are rare. We present the case of a 72-year-old man with multiple myeloma in complete remission since 2 years ago with cutaneous tumors on the trunk and face. A cutaneous biopsy was consistent with plasmacytoma. The patient was treated with melphalan, prednisolone and radiotherapy. Despite optimal therapeutic response of the lesions, the disease progressed, with the appearance of new extra-cutaneous plasmocytomas. The cutaneous metastatic plasmocytomas were the first sign of progression of the disease.

Keywords: Multiple myeloma; Plasmacytoma; Skin manifestations

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