

Cutaneous Plasmacytoma in Multiple Myeloma

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Abstract

Case Report

Skin involvement in multiple myeloma is rare. It is typically associated with aggressive progression and a poor prognosis. We report the case of a patient with aggressive skin involvement associated with extensive extramedullary dissemination, including orbital, peritoneal, and thoracoabdominal parietal sites, with a spectacular clinical and radiological response after intensive chemotherapy.

Keywords: Multiple myeloma, Extramedullary involvement, Cutaneous plasmacytoma, Orbital location, Intensive chemotherapy, Disease progression.

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INTRODUCTION

Multiple myeloma is a malignant blood disorder characterized by clonal proliferation of plasma cells in the bone marrow. Extramedullary involvement is a particularly aggressive form of the disease, defined by the ability of a tumor clone and/or subclone to proliferate independently of the medullary microenvironment. Its incidence is estimated to be between 0.5% and 4.8% in newly diagnosed patients, and between 3.4% and 14% in those with relapsed or refractory multiple myeloma [1].

Skin involvement in multiple myeloma is rare. It most often occurs at an advanced stage of the disease, although initial forms have been reported. The presence of cutaneous plasmacytoma is classically associated with aggressive progression and a poor prognosis [2].

We report the case of a patient with aggressive skin involvement associated with extensive extramedullary dissemination, including orbital, peritoneal, and thoracoabdominal parietal locations, with a spectacular clinical and radiological response after intensive chemotherapy.

OBSERVATION

This is a 59-year-old patient who was treated at our center for symptomatic IgG multiple myeloma and was initially treated with a VDT protocol combined with zoledronic acid. The evaluation after induction showed a very good partial response (VGPR). The patient was placed on maintenance therapy while awaiting an

autologous hematopoietic stem cell transplant. Six months later, the patient developed progressive rib tumor masses, associated with right exophthalmos and abdominal skin lesions.

Clinical examination revealed a large, polylobular, hard-consistency skin tumor mass measuring approximately 15 cm in length in the right hypochondrium, fixed to the deep plane, with an eroded surface covered with meliceric crusts and pigmented, indurated skin around the lesion. (Figure 1) In addition, there were multiple subcutaneous masses located in the left iliac fossa, axillary regions, and back. Ophthalmological examination revealed irreducible, non-pulsatile, non-inflammatory right exophthalmos associated with total ophthalmoplegia. (Figure 2) Light perception was preserved. Fundus examination revealed stage III papillary edema with exudative retinal detachment in the nasal area.

Thoracoabdominal-pelvic computed tomography showed a right intraorbital tissue mass causing grade III exophthalmos, as well as secondary peritoneal and thoracic and abdominal parietal locations measuring 60 × 47 mm, 25 × 23 mm, and 76 × 52 mm, respectively. Multiple bone lesions affected the axial skeleton, pelvic girdle, and scapular girdle. Brain MRI confirmed the presence of a large right intraorbital, intracone tissue process measuring 62 × 37 × 33 mm.

A skin biopsy was performed. Pathological examination showed plasma cell proliferation with

intense and diffuse expression of CD138 and light Kappa chain restriction, without Lambda expression. The findings were consistent with Kappa monotypic cutaneous plasmacytoma.

Serum protein electrophoresis revealed a monoclonal peak of 11.4 g/L in the gamma fraction. Weighted immunoglobulin measurement showed IgG at 23 g/L, with positive immunofixation for IgG Kappa. Blood count was normal, with no associated hypercalcemia or renal failure.

The patient underwent intensive VDT-PACE chemotherapy. Evaluation after three courses of treatment showed almost complete regression of the intraorbital process, complete disappearance of the peritoneal and thoracoabdominal parietal lesions, and stability of the osteolytic lesions. Serum protein electrophoresis no longer showed a monoclonal peak, with IgG levels reduced to 6 g/L. Unfortunately, the patient died after the fourth course of treatment due to severe infectious complications, with no documented signs of tumor progression.



Figure 1: A: Skin tumor mass

B: Response after treatment



Figure 2: A: Exophthalmos of the left eye

B: Response to treatment

DISCUSSION

Extramedullary involvement in multiple myeloma is a distinct clinical entity associated with aggressive biology and poor prognosis. It reflects the ability of plasma cell clones to proliferate independently of the medullary microenvironment, giving the disease systemic invasive potential [1]. Our observation illustrates this rare and severe form of extramedullary

dissemination, combining cutaneous, orbital, and peritoneal involvement, occurring in a patient being followed for early relapse of IgG multiple myeloma.

Cutaneous plasmacytoma corresponds to direct infiltration of the dermis and/or hypodermis by clonal plasma cells. It is the most specific but also the rarest form of cutaneous manifestation of multiple myeloma, with an estimated incidence of between 1 and 4% [3]. It

most often occurs at an advanced stage or in situations of relapse or therapeutic resistance and is generally associated with extensive extramedullary dissemination and limited survival.

The diagnostic approach must be rigorous, given the wide range of differential diagnoses. These include non-specific skin lesions associated with myeloma (leukocytoclastic vasculitis, opportunistic infections), manifestations related to plasma cell dyscrasias (AL amyloidosis, cryoglobulinemia, POEMS syndrome), as well as other malignant blood disorders infiltrating the skin, particularly cutaneous lymphomas and cutaneous leukemias [4]. Skin biopsy with histological and immunohistochemical examination is essential for diagnosis.

Management is based primarily on systemic treatment of the myeloma. Regimens combining proteasome inhibitors, immunomodulators, and monoclonal antibodies have improved tumor control, although the prognosis remains poor. Radiotherapy may be offered for symptomatic relief or local control.

In our observation, the coexistence of skin, orbital, and peritoneal involvement highlights the particularly aggressive nature of the disease. However, the spectacular clinical and radiological response obtained after intensive chemotherapy contrasts with the commonly reported prognosis, suggesting that intensive therapeutic strategies can achieve significant tumor control, even in the most disseminated forms [5].

CONCLUSION

Cutaneous plasmacytoma is a rare and aggressive manifestation of multiple myeloma. It most often occurs during the course of progressive disease. It is associated with a poor prognosis despite optimal management. Our observation nevertheless illustrates that intensive chemotherapy can achieve a major tumor response, highlighting the value of aggressive therapeutic management in selected patients.

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