

Exceptional Localization of Multiple Myeloma: Cutaneous Involvement, a Case Report

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Abstract: Multiple myeloma is a malignant hemopathy characterized by the proliferation of a clone of plasma cells invading the hematopoietic marrow. Bone manifestations (pain, pathological fractures) frequently dominate the clinical presentation. The disease can also be revealed by complications, including anemia or kidney damage. Skin localization is extremely rare. Primitives it precedes the appearance of multiple myeloma. Secondary it appears during multiple myeloma already well established. The presence of this skin disorders reflects a severe form with a high mortality rate. We report an observation with cutaneous involvement in a woman suffering from IgG lambda multiple myeloma.

Keywords: Multiple myeloma; Plasmacytoma; Cutaneous involvement.

1. INTRODUCTION

Multiple myelomas with dermatological manifestations in the form of cutaneous plasmocytomas are extremely rare. They represent less than 2% of cases [1]. These plasmocytomas are usually contiguous to bone lesions. Exceptionally these plasmocytomas develop at a distance from bone lesions. The presence of these skin disorders is associated with a pejorative prognosis [2]. This observation reports a case of cutaneous plasmocytomas in a 74-year-old patient treated for 14 months for IgG lambda multiple myeloma.

2. CASE REPORT

A 74-year-old woman with IgG lambda multiple myeloma:

- a monoclonal peak,
- anemia
- 23% presence of dystrophic plasma cells within the bone marrow. These plasmocytes have in 83% of cases a trisomy of the CKS1B locus in 1q21 and the deletion of the CDKN2C locus (P18) in 1p32. There

was no translocation t (4; 14) nor deletion 17p.

There was no damage to bone.

The disease progressed under 3 first lines of treatment:

- 1st line: Bortezomib - Melphalan - Prednisone
- 2nd line: Bortezomib - Lénalidomide - Dexamethasone
- 3rd line: Bortezomib - Cyclophosphamide - Dexamethasone

A partial response was finally obtained after the 4th therapeutic line: Dexamethasone - Cyclophosphamide - Etoposide - Cisplatin.

In view of the general condition of the patient considered satisfactory, it was then decided to carry out intensive chemotherapy followed by an autograft of hematopoietic stem cells. But after the collection of peripheral stem cells, cutaneous and subcutaneous nodules appeared. They were purplish, not painful, measuring 1 to 4 cm and localized on the abdomen, chest, back and limbs.



Figures 1 and 2. Multiple purplish papules located in the right leg (image on the left) and the supero-external quadrant of the right breast (image on the right)

A cutaneous biopsy was performed, revealing an infiltration of the subcutaneous fat by diffuse tumor proliferation made up of rather abundant cytoplasmic cells and nuclei of variable size more or less nucleolated.

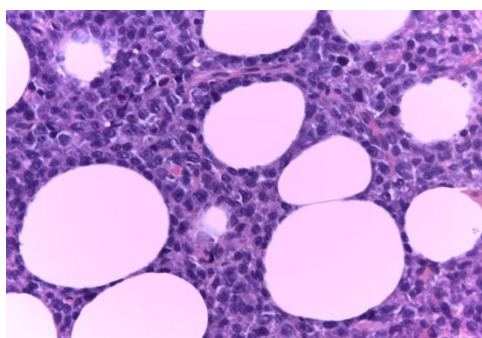


Figure 3. Diffuse tumor proliferation consisting of cells with plasmocytoid differentiation (HES x 400)

In immunohistochemistry the tumor cells were CD138 +

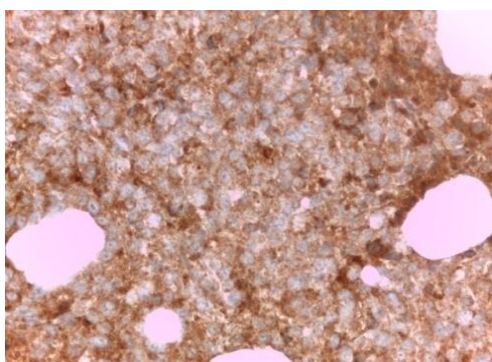


Figure 4. The tumor cells are CD138 + in immunohistochemistry (x400).

They showed lambda monotype and were negative for the kappa light chain. This aspect confirmed the diagnosis of cutaneous plasmocytomas. Despite two new therapeutic lines (Carfilzomib - Lénalidomide - Dexamethasone and Daratumumab - Pomalidomide - Dexamethasone), the disease has progressed rapidly. At the appearance of

new cutaneous nodules, some of which were flesh-colored, progressive left exophthalmia, left jugular tumefaction and anesthesia of the chin tassel were progressively associated. The analysis of the cerebrospinal fluid did not find any tumor cells. F18-FDG PET-CT

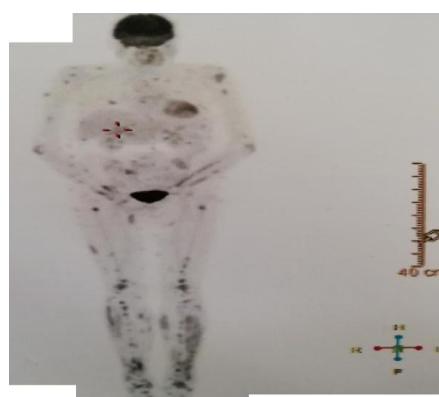


Figure 5. F18-FDG PET-CT showing multiple hypermetabolic diffuse cutaneous-mucous, muscular, skeletal and abdominal foci in multiple myeloma.

Was performed, demonstrating bone, ophthalmologic, ENT, hepatic, splenic, diaphragmatic, retro peritoneal fascia of the kidneys, muscular and diffuse mucocutaneous (SUVmax 8 in the right leg). Many skin nodules, including abdominal and mammary, were located at a distance from bone lesions. Intensive chemotherapy with autologous hematopoietic stem cell transplantation was performed 10 months after onset of cutaneous involvement. It was well tolerated and the patient is waiting for consolidation treatment.

3. DISCUSSION

Specific skin lesions in multiple myeloma are rare, the literature being limited to a few isolated cases or small series of cases [1-3]. Two types are usually individualized. Primary cutaneous plasmocytomas that occur outside myeloma and may precede its appearance or remain isolated.

Secondary cutaneous plasmacytomas, of which we report an observation, appear during the course of a known multiple myeloma. They are usually contiguous to bone involvement but can exceptionally, as with our patient, appear at a distance. In the latter case they are called metastatic cutaneous plasmacytomas [4-6]. The typical appearance is that of purplish or flesh-colored cutaneous or subcutaneous nodules, preferentially localized on the trunk and the extremities. More rarely, it realizes a papular or urticarial rash. Diagnosis is based on histological analysis and immunostaining [1,5,7,8]. The median time to onset of lesions is 2 years after the diagnosis of multiple myeloma. Cutaneous involvement is often associated with other soft tissue disorders. The management of secondary cutaneous plasmacytomas is usually based on chemotherapy with or without radiotherapy. The prognosis of these patients is pejorative. The median overall survival is 8.5 months from the onset of cutaneous involvement despite aggressive treatments. These treatments combine intensive chemotherapy and autologous hematopoietic stem cell transplantation [1-3]. Cutaneous involvement in multiple myeloma is evidence of advanced and aggressive disease.

REFERENCES

- [1] Woo YR, Kim JS, Lim JH, Hwang S, Kim M, Bae JM and al. Prevalence and clinicopathologic characteristics of multiple myeloma with cutaneous involvement: A case series from Korea. *J Am Acad Dermatol.* 2018; 78 (3):471-78.
- [2] Jurczynszyn A, Olszewska-Szopa M, Hungria V, Crusoe E, Pika T, Delforge M and al. Cutaneous involvement in multiple myeloma: a multi-institutional retrospective study of 53 patients. *Leuk Lymphoma.* 2016; 57(9):2071-6.
- [3] Malysz J, Talamo G, Zhu J, Clarke LE, Bayer MG, Ali L and al. Cutaneous involvement in multiple myeloma (MM): A case series with clinicopathologic correlation. *J Am Acad Dermatol.* 2016;74(5):878-84.
- [4] Comfere NI, Gonzalez Santiago TM, Peters MS, Knudson RA, Ketterling RP, Gibson LE. Cutaneous extramedullary plasmacytoma: clinical, prognostic, and interphase cytogenetic analysis. *Am J Dermatopathol.* 2013; 35 (3): 357-63.
- [5] Collet E, Dalac S, Lorcerie B et al. Plasmocytomes cutanés: revue de la littérature à propos d'une observation. *Rev Med Interne.* 1991; 12(3): 183-6.
- [6] Kato N, Kimura K, Yasukawa K, Aikawa K. Metastatic cutaneous plasmacytoma: a case report associated with IgA lambda multiple myeloma and a review of the literature of metastatic cutaneous plasmacytomas associated with multiple myeloma and primary cutaneous plasmacytomas. *J Dermatol.* 1999;26(9):587-94.
- [7] Kubica AW, Wetter DA, Lehman JS. Cutaneous involvement by multiple myeloma. *Am J Hematol.* 2014;89(10):1014-5.
- [8] Santos G, Sousa L, Fernandes T, João A. Case for diagnosis. Cutaneous involvement associated to multiple myeloma. *An Bras Dermatol.* 2014;89 (1):173-4.

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