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Case Report

Extramedullary Manifestation of Multiple Myeloma in the Oral Cavity

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Abstract

Background: Extramedullary plasmacytomas occurs in about 20% of multiple myeloma (MM) recurrences. Extramedullary disease seems to respond poorly to thalidomide and has adverse prognostic implication. When disease recurs in the oral cavity with soft tissue infiltration, some authors defend upfront surgical excision prior to radiotherapy with the aim of achieving better local control. We describe herein such an atypical case of recurrence from MM, with complete local response after 2 cycles of chemotherapy. Unfortunately, disease progressed later on, and the patient died after 9 months post-recurrence. This emphasizes the prognostic impact of extramedullary disease manifestation in MM.

Keywords: Multiple myeloma, Oral cavity, Recurrences

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Introduction

Multiple myeloma (MM) is a clonal B-cell malignancy, characterized by proliferation of plasma cells in the bone marrow although some patients develop extramedullary disease. In the past, the latter entity was rarely seen in daily medicine due to the short life span of affected patients, but recently its incidence appears to have risen steadily with prolonged survival achieved under novel drug treatments and autologous bone marrow transplantation.[1] It is estimated that extramedullary plasmacytomas occurs in about 20% of MM recurrences.[2] Oral cavity lesions may be the first sign of disease in MM and the sole manifestation of disease relapse or part of a group of symptoms of disease progression.[2,3]

Case Report

In the year 2015, a 61 year-old male was referred to the Oncology Department of A.C Camargo Cancer Center, for hematopoietic cell transplant evaluation. He had a past history of MM with diagnosis in June, 2010 and, by that time, after achieving an objective response under thalidomide plus dexamethasone therapy, he was submitted to an autologous stem cell transplant in February, 2011. He had a disease free interval of 54 months until August, 2015, when oncologic pain related to new sternal bone lesions developed and his disease was reappraised. According to the Durie-Salmon System, he had stage III disease with pronounced anemia, renal dysfunction and bone lesions. During the initial evaluation process, a gingival mass appeared and grew rapidly (Figure 1) and it was finally biopsied (Figure 2).

Pathology report indicated gingival infiltration by plasma cells with CD138 positivity and high mitotic rate, which was compatible with extramedullary multiple myeloma (EMM).

The patient was treated with a combination of cyclophosphamide, bortezomib and dexamethasone (CyBorD regimen) and he had complete local response in the oral cavity after only 2 cycles of chemotherapy (Figure 3).

Unfortunately, disease response was only partial systemically, and new bone lesions finally appeared in skull base after third cycle of CyBorD.

A radiotherapy course was applied to the skull base and a fourth CyBorD cycle was resumed.

This patient developed multiple complications under treatment: perforated diverticulitis after cycle 2 and H1N1 disease requiring hospitalization after cycle 4 are worth noting. Finally, he experienced disease progression with

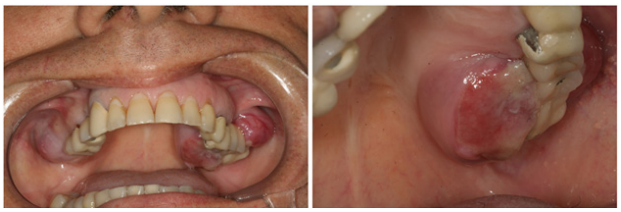

Figure 1. Photograph illustrates Ginvival Mass Both Sides of Maxilla.

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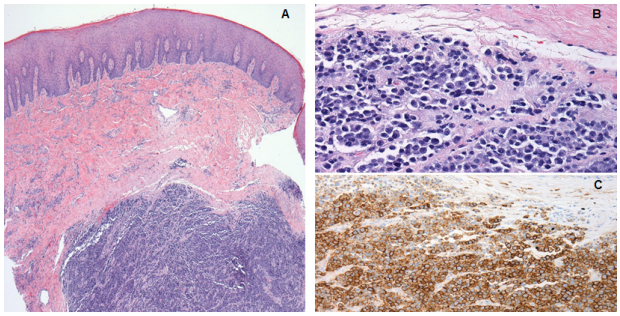

Figure 2. Gum Biopsy: H&E Stain Showing Neoplastic Infiltration of the Lamina Propria (**A**, x5). The morphology is consistent with a high grade plasmablastic neoplasm (**B**, x40). Immunohistochemical stain showing CD138 positivity (**C**, x20)

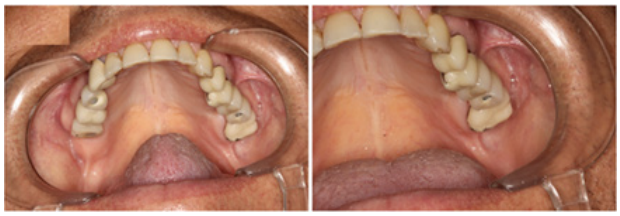

Figure 3. Photograph Shows Resolution of Soft Tissue Infiltration on Maxilla After 2 Cycles of Chemotherapy.

an extramedullary paravertebral mass causing spinal cord compression.

Radiotherapy to thoracic spine – including the paravertebral mass – was used and an alternative chemotherapy protocol was applied. It consisted of bortezomib-thalidomide-dexamethasone (VTD) and cisplatin-adriamycin-cyclophosphamide-etoposide (PACE). On the eighth day of the first VTD-PACE cycle, the patient developed neutropenic sepsis and was transferred to the intensive care unit (ICU), but his clinical condition deteriorated with septic shock, multiple organ failure, and death.

Discussion

We describe herein an atypical case of MM recurrence, with extramedullary disease manifestation in the oral cavity. For soft tissue plasmacytoma at this location, some authors defend upfront surgical excision prior to radiotherapy with the aim of achieving better local control.[4] It is general rule to consider extramedullary disease relapse as poor predictor of survival and some case series describe median overall survival of only 5–7 months, even in the era of novel drug treatments and hematopoietic cell transplantation.[5,6] A possible explanation for that resides in the fact that tumor cells in EMM lack expression of adhesion molecules such as CD56, have a high mitotic rate and tend to disseminate early in the course of disease.[1]

In the case reported herein, there was an excellent local response in the oral cavity (Figure 3) on systemic treatment with the CyBord regimen. Unfortunately, patient developed new bone lesions and spinal cord compression ensued as result of another soft tissue plasmacytoma (this time not in the oral cavity but in the paravertebral location instead). Systemic therapy was modified to VTD-PACE regimen and the patient died with septic complications soon after beginning of such intensive treatment. Overall survival – from time of disease recurrence to death – was only 9 months.

Authors' Contribution

CdAPH assessed the patient, wrote de manuscript and reviewed it. JSF, MdMN, FAA, TN and FDC assessed the patient and reviewed the manuscript.

Conflict of Interest Disclosures

The authors have no conflicts of interest.

Ethical Statement

The authors declare no conflict of interest. Written consent was obtained.

References

1. Blade J, Fernandez de Larrea C, Rosinol L, Cibeira MT, Jimenez R, Powles R. Soft-tissue plasmacytomas in multiple myeloma: incidence, mechanisms of extramedullary spread, and treatment approach. J Clin Oncol. 2011;29(28):3805-12. doi: 10.1200/jco.2011.34.9290.
2. Cardoso RC, Gerngross PJ, Hofstede TM, Weber DM, Chambers MS. The multiple oral presentations of multiple myeloma. Support Care Cancer. 2014;22(1):259-67. doi: 10.1007/s00520-013-1960-y.
3. Lee SH, Huang JJ, Pan WL, Chan CP. Gingival mass as the primary manifestation of multiple myeloma: report of two cases. Oral Surg Oral Med Oral Pathol Oral Radiol Endod. 1996;82(1):75-9.
4. Tesei F, Caliceti U, Sorrenti G, Canciullo A, Sabbatini E, Pileri S, et al. [Extramedullary plasmocytoma (EMP) of the head and neck: a series of 22 cases]. Acta Otorhinolaryngol Ital. 1995;15(6):437-42.
5. Rasche L, Bernard C, Topp MS, Kapp M, Duell J, Wesemeier C, et al. Features of extramedullary myeloma relapse: high proliferation, minimal marrow involvement, adverse cytogenetics: a retrospective single-center study of 24 cases. Ann Hematol. 2012;91(7):1031-7. doi: 10.1007/s00277-012-1414-5.
6. Pour L, Sevcikova S, Greslikova H, Kupska R, Majkova P, Zahradova L, et al. Soft-tissue extramedullary multiple myeloma prognosis is significantly worse in comparison to bone-related extramedullary relapse. Haematologica. 2014;99(2):360-4. doi: 10.3324/haematol.2013.094409.