

Plasmacytoma in the Oral Cavity: A Case Report

Plasmocitoma en la Cavidad Oral: Reporte de Caso

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ABSTRACT: The plasma cell neoplasms may present in soft tissue as extramedullary plasmacytoma (EMP), in bone as a solitary plasmacytoma of bone (SPB), or as part of the multifocal disseminated disease multiple myeloma (MM). The EMP is rare, comprising around 3% of all plasma cell neoplasm. The majority (80%) occurs in the head and neck region. In this study we report a case of a man, 70 years old, melanoderm, with a lesion of the oral cavity. Upon physical examination, a lesion was found that extended throughout the posterior upper alveolar ridge, as far as the maxillary tuber on the left side, extending towards the palate. Radiographic examination, complementary laboratory exams were performed. Based on the conclusive symptoms of plasmacytoma, the patient was referred to the hematology service for treatment with local radiotherapy. The patient responded satisfactorily to the treatment, and after 15 months, all clinical symptoms of the lesion in the oral cavity had disappeared.

KEY WORDS: plasma cell neoplasms, extramedullary plasmacytoma, bone marrow, oral cavity.

INTRODUCTION

The plasma cell neoplasms may present in soft tissue as extramedullary plasmacytoma (EMP), in bone as a solitary plasmacytoma of bone (SPB), or as part of the multifocal disseminated disease multiple myeloma (MM). These tumors are derived from bone marrow stem cells of B-Lymphocyte lineage, and are characterized by an expansion of a clone of immunoglobulin-secreting cells (Regezi *et al.*, 2003).

Multiple Mieloma (MM). The MM represents the most important and common plasma cell dyscrasia (Regezi *et al.*), involving large numbers of bone (Shafer *et al.*, 1974). Also known as generalized myeloma or myelomatosis, its etiology is unknown, it is characterized by the proliferation of cells that have the microscopic appearance of plasmacytes (Anil, 2007). It is considered a multifocal plasma cell cancer of the skeleton, but it may also affect extraskelatal sites during its progression (Dores *et al.*, 2009). The disease begins insidiously in bone marrow, and by the time it is diagnosed it is usually at an advanced stage. Extramedullary manifestations sometimes occur during the course of the disease in the form of a tumor mass

consisting predominantly of plasmacytes surrounded by a fine reticular network (Nasr Ben Ammar *et al.*, 2010).

Extramedullary Plasmacytoma (EMP). Shafer *et al.*, affirm that the MM and EMP presents similar histological features, however, the EMP is characterized by an extramedullary located mass infiltrated by monoclonal plasma cells without signs of MM elsewhere in the body (Dimopoulos *et al.*, 1999).

The EMP is rare, comprising around 3% of all plasma cell neoplasm (Dimopoulos *et al.*). The majority (80%) occurs in the head and neck region (Regezi *et al.*), in particular, in the nasopharynx, paranasal sinuses and tonsils (75%). The EMP occurs less commonly, in the oropharynx (12%) (Goel *et al.*, 2010). It accounts for around 4% of non-epithelial neoplasms of the nasal cavity, paranasal sinuses, and nasopharynx (Regezi *et al.*), occurring mainly in the fifth and sixth decades of life, with a higher prevalence in men (Gross *et al.*, 2004; Nasr Ben Ammar *et al.*). The incidence is higher among individuals of advanced

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age, with a mean age at diagnosis of 64 years, and two times higher among melanoderm individuals than caucasian (Nasr Ben Ammar *et al.*).

The signs and symptoms of plasmacytoma are pain, swelling, and tissue masses obstructing the respiratory tract or oral cavity that cause a local expansion by edema (Gross *et al.*). When it occurs in the nasopharynx, the main symptom is progressive nasal obstruction and intermittent epistaxis, and in the maxillary sinus, it presents enlarged volume and may affect and damage the adjacent teeth, due to destruction of the surrounding bone (Ersoy *et al.*, 2004). Other symptoms may appear depending on the location of the tumor. Edema is the most common complaint, and pain may also be present when the adjacent bone is affected. Discomfort or pain of the tooth can occur when there is involvement of the dental alveolus.

The most widely accepted treatment is chemotherapy with cyclophosphamide, melphalan, and steroid protocols. Radiotherapy may also be used since the tumor is radiosensitive (eradicable at a dose of 4.000 to 5.000 cGy). Another alternative is surgical resection (Romero *et al.*, 2009).

The aim of this study is to report a case of plasmacytoma with exteriorization to the oral cavity.

CASE REPORT

Male patient, 70 years old, melanoderm, was received at the Bucomaxillofacial Surgery and Traumatology Service of a public hospital located in São Paulo – Brazil, with a lesion in the oral cavity.

The patient reported rapid, painless growth of the lesion, and confirmed that he had been a smoker for 62 years. He denied alcohol consumption and did not know whether he had any systematic disease. The patient had been medicated with antibiotics for 14 days without regression of the lesion.

Upon physical examination, a lesion was found that extended throughout the posterior upper alveolar ridge, as far as the maxillary tuber on the left side, extending towards the palate (Fig. 1). The base was sessile, erythematous, and flaccid on palpation. On the radiographic examination, the maxillary sinus was foggy, with bone impairment. The complementary laboratory exams were normal.



Fig. 1. Lesion in the posterior upper alveolar ridge, as far as the maxillary tuber on the left side, extending towards the palate.

The patient was referred to the surgical center for incisional biopsy of the lesion, and the fragment was sent for anatomopathological examination. The exam results showed fibroadipose tissue with infiltration of small atypical cells, requiring complementary immunohistochemical exam. In this latter exam, histological aspects of a plasmacytoma were observed, with evidence of cell differentiation into kappa chains. Tumor cells were positive for Ki-67, Mib-1; CD45; Bcl-2; and positive diffused for CD-138.

Based on the conclusive symptoms of plasmacytoma, the patient was referred to the hematology service for treatment with local radiotherapy (at a dose of 4000 to 5000 cGy).

The patient responded satisfactorily to the treatment, and after 15 months, all clinical symptoms of the lesion in the oral cavity had disappeared (Fig. 2). He is still in follow-up care.



Fig. 2. Oral cavity of the patient showing good results for remission of the lesion.

DISCUSSION

The majority of patients with extramedullary plasmacytoma are male (63%-86%). This neoplasm is most common in the sixth to eighth decade, and the head and the neck are the regions most commonly affected, 56% to 90% was disease free at 10 years, and the median survival was 8 to 20 years (Knowling *et al.*, 1983; Shih *et al.*, 1995; Bolek *et al.*, 1996; Liebross *et al.*, 1999; Galieni *et al.*, 2000). In other studies the median survival rate was 5.66 years (Susnerwala *et al.*, 1997).

Plasmacytoma is a clinical finding that is difficult to diagnose. Only the anatomopathological exam, preferably accompanied by an immunohistochemical study (Romero *et al.*), can confirm the diagnosis through positive plasma cells that express CD38 with concomitant cytoplasmic expression of kappa or lambda light chains (Dimopoulos *et al.*). In our case, incisional biopsy was carried out, with confirmation for kappa chains, CD45, CD138 and negative confirmation for lambda light chains.

Some authors affirm that approximately 10-30% of treated patients distant failure seem to occur (Corwin & Lindberg, 1979; Holland *et al.*, 1992; Susnerwala *et al.*). This takes place on average within 2 to 3 years after diagnosis (Knowling *et al.*; Liebross *et al.*; Galieni *et al.*; Straetmans & Stokroos, 2008). Dimopoulos *et al.* affirms that the prognosis of patients with solitary extramedullary plasmacytoma (SEP) appears to be better than for patients with SBP because approximately 70% of patients with SEP remain disease-free at 10 years. Mock *et al.* (1987) suggested that the cases with lambda light chain may be more immature and more likely to progress to MM. In other study, Susnerwala *et al.* observed that the cases that progressed to MM showed kappa light

chain restriction. In our study the patient presents the kappa light chain, however there was no evidence of progression to MM. Holland *et al.* affirms that the lesion size, total serum protein levels, and the presence of a monoclonal spike on serum electrophoresis may be of prognostic significance in identifying those solitary lesions that ultimately will convert to MM.

Susnerwala *et al.* affirms that the higher grade tumors were generally associated with more bulky disease. There is every indication that plasmacytomas that emerge from the soft tissues of the nasopharynx, oral cavity, or larynx, and do not involve adjacent bone, have a better prognosis than those located in areas such as the maxillary, mandible, alveolus and others (Romero *et al.*).

As EMP are highly radiosensitive, almost all patients obtained successful treatment with local control (Weber, 2005). Radiation therapy is the most effective form of local treatment in the management of solitary plasmacytomas. The recommended dose of radiation is 40 to 60 Gy (Corwin & Lindberg; Holland *et al.*; Romero *et al.*; Nasr Ben Ammar *et al.*). Weber and Soutar *et al.* (2004) recommend that for EMP <5 cm a radiotherapy dose of 40 Gy in 20 fractions, and recommend for bulky EMP of >5cm, a higher dose of up to 50Gy in 25 fractions. The appropriate therapy results in long-term stability and potential cure in approximately 67% of patients (Weber).

In conclusion, plasmacytoma, despite being a lesion with slow, asymptomatic growth, can assume large volumes, making proper treatment difficult. When there is no bone involvement and it is diagnosed early, the success of treatment is generally higher. The treatment of choice is radiotherapy, with good results for remission of the lesion, as we have demonstrated in this case study.

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RESUMEN: La neoplasia de células plasmáticas se presenta en los tejidos blandos como plasmocitoma extramedular (EMP), en el hueso como plasmocitoma solitario (SPB), o como parte de una enfermedad diseminada, el mieloma múltiple (MM). El EMP es poco frecuente, manifestándose en alrededor de un 3%. Se presentan generalmente (80%) en las regiones de cabeza y cuello. En este reporte de caso, un hombre de 70 años, melanoderma, presentaba una lesión en la cavidad oral. En el examen físico se observó la lesión a lo largo de la parte posterior del reborde alveolar superior, en la tuberosidad maxilar en el lado izquierdo hacia el paladar. Se realizaron exámenes radiográficos y de laboratorio. Con base en los signos de plasmocitoma, el paciente fue referido a tratamiento con radioterapia. El tratamiento fue muy exitoso y después de 15 meses se observó que el paciente no presentaba los signos clínicos característicos de la lesión neoplásica.

PALABRAS CLAVE: neoplasia de células plasmáticas, plasmocitoma extramedular, médula ósea, cavidad oral.

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