

Solitary Plasmacytoma of the Mandible: An Uncommon Entity

Ezher Hamza Dayisoğlu,^{1*} Ozcan Ceneli,¹ and Esra Zeynep Coskunoglu¹

¹Department of Oral and Maxillofacial Surgery, Faculty of Dentistry, Baskent University, Bahcelievler, Ankara, Turkey

*Corresponding author: Ezher Hamza Dayisoğlu, Department of Oral and Maxillofacial Surgery, Faculty of Dentistry, Baskent University, Bahcelievler, Ankara, Turkey. Tel: +90-5326624778, Fax: +90-3322570637, E-mail: edayisoğlu@gmail.com

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Abstract

Introduction: Plasma cell dyscrasias are characterized by a monoclonal neoplastic proliferation of plasma cells. Solitary bone plasmacytoma (SBP) is a local form of the disease with the vertebrae and long bones being the most frequently encountered sites. Its prevalence in the maxillofacial area is extremely rare.

Case Presentation: A 70-year-old Caucasian male patient was referred for the extraction of his mobile premolar tooth with a poorly-defined radiolucent lesion. Histopathological analysis revealed an SBP and no distant lesion or serum M protein was noted on radiological and hematological examinations. The patient was under follow-up care with no recurrence at 2 years of follow up.

Conclusions: Diagnosis of an SBP is based on local radiological and neurological symptoms and similar systemic manifestations of multiple myeloma that are also distinctive for SBP. Skeletal radiological analysis including CT and PET-CT, bone marrow biopsy, and serum protein electrophoresis are essential for confirmation of the diagnosis. Although surgery, chemotherapy, and radiation, or a combination of these modalities, have been successfully used in the treatment of SBP, it should be managed in relation to its possible long-term evolution.

Keywords: Multiple Myeloma, Solitary Plasmacytoma, Mandible

1. Introduction

Plasma cell neoplasms are characterized by uncontrolled proliferation of plasma cells. Solitary bone plasmacytomas (SBPs) and extramedullary plasmacytomas (EPs) are the localized forms of the disease, whereas multiple myeloma (MM) is a systemic clonal proliferation of plasma cells based in the bone marrow (1). Fewer than 5% of patients with plasma cell dyscrasias present as SBP or EP without distinct evidence of systemic myeloma (2). The diagnosis of SBP is based on a biopsy of the specimen, skeletal radiological evaluation with a lack of any other distant lesions, and a negative result for monoclonal plasma cells on bone marrow aspirate to rule out MM. The majority of SBP lesions either progress to MM over a period of 2-3 years after diagnosis or local asymptomatic lesions tend to occur. Therefore, early diagnosis, proper treatment, and close follow up are crucial for survival (3). Clinical symptoms of SBP consist of jaw pain, paresthesia, and mobility of the surrounding teeth, or pathologic bone fractures. Radiological signs of SBP include ill defined unilocular or multilocular radiolucent lesions or a mass with cortical expansion (4). Although osteolytic jaw lesions commonly appear in patients with MM, the ileum, femur, humerus, and thoracic vertebrae are the most frequent locations for SBP and its incidence in the maxillofacial area is extremely rare. SBP

treatment consists of surgery, chemotherapy, or a combination of these modalities. There is also some controversy concerning management solely with surgery or radiotherapy alone (5). In this case report, the treatment of a 70-year-old Caucasian male patient with SBP of the mandible is presented.

2. Case Presentation

A 70-year-old Caucasian male patient was referred to Baskent University Department of Oral and Maxillofacial Surgery for the extraction of mobile tooth # 44. The patient's medical history was noncontributory, and the physical examination revealed no other abnormalities. An intraoral examination showed a mobile tooth with poor oral hygiene and pain on palpation. A radiological examination revealed a poorly-defined destructed radiolucent area around the mandibular premolar region (Figure 1). Numbness of the chin, expansion of the bone cortices, and other complaints were not observed. The mobile tooth was extracted and the lesion was enucleated. Following the curettage, an alveoplasty was performed under local anesthesia. The histopathological examination of the specimen showed diffuse stromal infiltration by plasma cells with eosinophilic cytoplasm and eccentric nuclei. The immunohistochemical examination showed positivity with CD138

and a monoclonal restriction for the Kappa chain. The patient was consulted to the hematology department and no monoclonal protein was observed with serum and urine immunofixation electrophoresis. The sedimentation rate was 5 mm/h and the beta 2 microglobulin level was 2.2 mg/L (reference range: 0.97 - 2.64 mg/L). Additionally, no renal dysfunction, hypercalcemia, or anemia were observed in age-related reference ranges. PET CT scans and cone beam CT (CBCT) evaluations revealed a lack of any local or distant lytic lesions except for an increased FDG uptake in the right hemimandible during PET-CT scan analysis. Therefore, we concluded that adequate surgical debridement had been achieved. The patient was followed up on a monthly basis for 6 months and no further lesions were observed at the 2-year follow-up examination.



Figure 1. Note the Irregular Radiolucent Area Around Tooth # 44

3. Discussion

Plasma cell neoplasms are classified into three subgroups as multiple myeloma (MM), solitary bone plasmacytoma (SBP), and extramedullary plasmacytoma (EP). A plasmacytoma is a result of uncontrolled proliferation of a single clone of B cells with no distant spread that are able to evolve up to the stage of plasma cells (6). Whereas MM is the disseminated form of the plasma cell neoplasms, SBP and EP are separate entities that are observed as localized lesions in the bone or extramedullary soft tissue, respectively (4). Systemic alterations such as hypercalcemia, impaired renal function, anemia, bone lesions, leukopenia, thrombocytopenia, and proliferation of monoclonal plasma cells in the bone marrow are frequently observed in MM patients (7). Daghighi et al. noted that bone damage may also be responsible for alteration of blood calcium levels, especially in MM cases compared with SBP. Therefore, complete blood cell count (CBC), calcium, phosphorous, C-reactive protein (CRP), beta 2-microglobulin analyses, and skeletal bone surveys are highly recommended (8). On the other hand, SBP is a rare disease that constitutes approximately 3% - 10% of all plasma cell neoplasms and clinical

signs and symptoms of SBP may be really poor that differs from MM. Therefore, an early diagnosis of SBP may be challenging to perform (3, 9). Moreover, SBP commonly occurs in long bones and vertebrae, however, its involvement in the mandible is only 4.4% of the cases, which may also complicate its diagnosis (3). Pisano et al. noted SBP most frequently occurs at the posterior mandible, which is consistent with our case (10). SBP is frequently misdiagnosed as benign lesions, inflammatory diseases, and less frequently as malignant tumors. Clinical signs and symptoms indicated by patients include localized pain and paresthesia, but bone-impaired functions are also frequently reported (4). The most frequent clinical symptoms of SBP are referred pain in the jaws and teeth that may also be related to other jaw lesions, and less commonly seen are swelling, soft tissue masses, and pathologic fractures. The tumors are found as unilocular or multilocular radiolucent destructive lesions on radiological analysis that are also consistent with our case as a unilocular ill-defined lesion. Although no spontaneous jaw pain was observed in our patient except tooth mobility the OPG and CBCT revealed a lytic radiolucent area for SBP. Canger et al. (4) noted that patients are generally male with a 2:1 male to female ratio, and they are typically in their sixth or seventh decades of life; thus, our patient's profile was also consistent with the literature (11, 12).

Elevated serum or urine calcium levels, renal failure, anemia, bone lesions, and consequently pathological fractures are typically seen in MM patients. Alternatively, a lack of these findings increases the median survival rate in SBP patients. Furthermore, SBP may be an isolated disease or the first manifestation of a subsequent MM. Lesmes et al. claimed there is some controversy about SBP management solely with surgery or radiotherapy. On the other hand, there is often a need for a second surgical approach for biopsy for the eradication of the disease and the possible need for further radiotherapy (5). Daghighi et al. also noted CT and MRI findings are not specific for plasmacytoma and histopathological examinations are crucial for the diagnosis (8). Therefore, a surgery-first approach may be advantageous over solely using radiotherapy depending on the size and location of the lesion. In our case, following surgical debridement, no sign of local or distant lesions were noted and no radiotherapy was planned. Although radiation, chemotherapy, surgery, or a combination of these modalities provides good results with low recurrence rates, close follow up is essential (13). The prognosis of SBP could be worse if recurrence is present as in cases of evolution toward systemic diseases such as MM (13).

Footnote

Authors' Contribution: Ezher Hamza Dayisoylu: first visit of the patient, operation, and follow up of the patient and preparation of the manuscript; Ozcan Ceneli: diagnosis and follow up of the patient concerning a hematological viewpoint; Esra Zeynep Coskunoglu: pathological examination, diagnosis, and preparation of the paper.

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