Solitary Plasmacytoma of The Jaws: Therapeutically Considerations and Prognosis: A Case Report

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ABSTRACT

The plasma cell neoplasm is a rare malignant neoplasm of plasma cells that may present as Extramedullary Plasmacytoma (EMP) in soft tissues especially in the upper respiratory tract, in the bone as a Solitary Plasmacytoma of bone (SPB), or as a part of the multifocal disseminated disease as Multiple Myeloma (MM). The majority of 80% occurs in the head and neck region. In our case, a 23-year-old female patient presented with a non-tender swelling of the gingiva in the upper and lower jaws with the mobility of all teeth. The radiological examination showed severe destruction of bone around all teeth.

Keywords
Solitary Plasmacytoma, Radiotherapy, Multiple Myeloma.

Introduction

Solitary plasmacytoma (SP) is a rare malignant neoplasm of plasma cells with no evidence of systemic spread [1] It is considered one of the three variations of plasma cell neoplasms, the further two are being multiple myeloma (MM) that is associated with the involvement of several skeletal sites and extramedullary plasmacytoma (EMP), a localized plasma cell neoplasm that occurs in tissues other than bone [1,2].

SP presents a prevalence of less than 5% of all neoplasms [3]. EMP is a relatively rare lesion, constituting 3% of all plasma cell neoplasms [4]. About 1% of head and neck tumors are EMPs. It is found most commonly in the head and neck region, with 80% of cases occurring in the upper respiratory tract the nasopharynx, paranasal sinuses, and tonsils. EMPs occur less commonly in the gingiva [5].

The definite etiology of SP is unknown, however, there is a predisposing factor that can contribute to the progress of the lesion such as chronic stimulation, viral infections, and radiation overdose [6]. SP of the jaws can affect persons of any age, nevertheless, it is common between the 6th and the 7th decades of life with a predilection for males than female patients, in overall, present a primary complaint of swelling accompanying with slight pain [7,8]. The present case report discusses a rarely described SP of the maxillary and mandibular jaws.

Case Presentation

A 23-year-old female patient reported to the Periodontology Department faculty of dentistry Tanta university with the chief complaint of swelling of the gingiva in the upper and lower jaws. The patient complained of dull aching pain and mobility of her teeth, difficulty in practicing oral hygiene, and a poor aesthetic appearance. The gingival mass is associated with bleeding and from 6 months’ period. The patient's medical history was non-contributory.
On intra-oral examination, the gingival mass was oval, with a lobulated appearance. The lesion was reddish, sessile, firm, and non-tender, involved the labial and lingual gingiva and alveolar mucosa, and extended around all teeth in both arches, the surface was smooth with no ulceration or pus discharge and associated with tooth mobility (Figure 1). A panoramic radiograph and cone-beam CT radiographs have shown generalized advanced bone loss (Figure 2). Routine blood investigations were within normal limits except for microcytic hypochromic anemia, fasting and postprandial blood glucose levels were normal, liver and kidney function tests were all normal.

**Diagnosis**

An incisional biopsy from lower labial gingiva was done and the tissue was referred for histopathological examination (Figure 3). The section displayed epidermis lined with stratified squamous epithelia. The subepithelial stroma presented diffuse sheets of plasma cells. The plasma cells were mainly mature and variable in size, with an eccentric nucleus and perinuclear halo surrounded by abundant eosinophilic cytoplasm. Quite a few binucleate and multinucleate plasma cells were also seen (Figures 4). To exclude the possibility of plasmacytoma. Immunohistochemistry for CD 138 was done which showed diffuse membranous positivity.

![Figure 1](image1.png)  ![Figure 2](image2.png)  ![Figure 3](image3.png)

**Figure 1:** Clinical presentation of the case.

**Figure 2:** A panoramic radiograph, and cone beam CT radiographs have showed generalized advanced bone loss.

**Figure 3:** An incisional biopsy was done and the tissue was referred for histopathological examination.
To exclude other types of plasmacytoma (MM & EMP) Bone Marrow aspiration from the sternal was done which showed Normocellular bone marrow. Megakaryocytes are normal on screening, Myeloid series are normal in maturation and morphology with myeloid hyperplasia (M/E ratio 8.4/1), Erythroid series are normoblasts and normal in maturation and morphology but relatively reduced, no infiltration by abnormal cells could be detected. Serum electrophoresis shows an increase in the gamma globulin fraction suggesting polyclonal gammopathy for clinical correlation No monoclonal band could be detected. Therefore, MM was excluded and according to clinic histopathological examination, a confirmatory diagnosis of plasmacytoma was done.

Discussion
Solitary plasmacytoma of the jaw usually has a manifestation that includes localized pain in the jaws and teeth, paresthesia, swelling, soft tissue masses, mobility and migration of teeth, hemorrhage, and pathological fracture. Fatigue and fever are the most common systemic symptoms [6,9]. Our patient just suffered from mobility and migration of her teeth that probably due to aggressive periodontitis. She did not report a history of pain or paresthesia in the jaws and teeth. Asymptomatic solitary bone plasmacytoma of the jaw is very rare but such a clinical form without pain has been described previously [9].

Diagnosis is based on the presence of malignant proliferation of plasma cells in the biopsy. Histological features of solitary plasmacytoma are identical to MM, Sheets, or clusters of atypical monoclonal plasma cells with various types of differentiation [9,10]. Bone marrow biopsies are performed to ensure that the disease is localized. In solitary and extramedullary plasmacytoma, there will not be an increase of monoclonal plasma cells in bone marrow [11]. In immunohistochemical painting, CD138 was positive. The results of the present case are in accordance with some studies which reported that these atypical cells react positively for CD138 [9-11].

Solitary plasmacytomas are extremely radiosensitive lesions. Radiation therapy, radical extensive surgery, or a combination of both is recommended as primary treatment. Radical radiotherapy comprising of 40-50Gy has revealed 80% of local disease control. Surgical treatment is recommended to those cases where the whole tumor is to be removed to minimize cosmetic or functional discrepancy or in cases where the pathological fracture is anticipated (to prevent that fracture and stabilize the fractured mandible). Research is being gone on for the role of angiogenesis inhibitors, thalidomide, protease inhibitors or inhibitors of vascular endothelial growth factors in plasma cell neoplasm's as an alternate mode of treatment [3,9,10]. All patients with plasmacytomas require follow-up for at least the first five years after treatment has been accomplished. The rate of progression of EMP to MM is 15-

(Figure 5) favoring the diagnosis as plasmacytoma (plasma cell myeloma).
20%. Harwood et al. reported the rate of conversion to MM if EMP involved the underlying bone is high [12].

**Conclusion**
Solitary bone plasmacytomas rarely occur in maxillofacial areas affecting the maxilla and mandible. In this case, the patient was referred to a cancer institute for complete treatment which includes radiation therapy, radical extensive surgery, or a combination of both. Chemotherapy should be reserved for those cases progressing to multiple myeloma.

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**References**

**Supplemental data**

Video Graphic 1. See video, Supplemental Digital Content, which displays a demonstration of Clinical presentation of the case with incisional biopsy harvesting from lower facial gingiva and the tissue was referred for histopathological examination. available at https://youtu.be/ttAGfzMHsg4.