

Solitary Plasmacytoma of Mandible – A Rare Dyscrasia- Case Report and Review of Literature.

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ABSTRACT

Introduction: The plasma cell neoplasms may present in soft tissue as extramedullary plasmacytomas, bone as a solitary plasmacytoma of bone, or as part of the multifocal disseminated disease multiple myeloma.

Aim of study: The study aims to report solitary plasmacytoma in the gnathic bone oral cavity, which is also mimicking as malignant neoplasm of bone, seen in a female patient.

Case Report: A 38-year-old female patient reported to the outpatient department of our hospital complaining of pain and swelling over the left lower one-third region of the face for one month CBCT analysis shows a hypodense area involving 35 regions extending towards ascending rami of the mandible.

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 the remission of the lesion

Keywords: Plasma cell neoplasms, Multiple myeloma, Solitary Plasmacytoma

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INTRODUCTION

Dalrymple first described the plasma cell myeloma in 1848, and in the same year Bence Jones discovered the urinary protein that was named after him (quoted in Dominok and Knoch 1977).¹ The term "multiple myeloma" was introduced in 1873 (von Rustitzky). In 1889 Kahler published a complete description of the disease. It was not until 1920 that the lesion was recognized as being a plasma cell tumor (Wallgren). The term plasmacytoma was introduced in 1940 (Apitz).²

Solitary plasmacytoma of bone (SPB) and extramedullary plasmacytomas (EMP) are rare plasma cell proliferative disorders. Their diagnosis is based on histologic confirmation of monoclonal plasma cell infiltration of a single disease site and the exclusion of systemic myeloma.³ Patients with Solitary plasmacytoma of bone SPB are more likely to progress to multiple myeloma (MM), which adversely affects their survival compared with those with EMP.^{4,5,8}

The Solitary plasmacytoma of bone SPB usually occurs in the vertebra and skull and is more common than EMP that almost always arise in the head and neck and may spread to regional lymph nodes.^{1,2} Prognosis is relatively good and is better for patients with EMP compared with those presenting with SPB Solitary plasmacytoma of bone. Localized pain, swelling, a raised red lesion on the alveolar ridge are the most clinical signs for plasmacytoma.^{3,4} This disease includes patients with more than one lesion, for it is elevated levels of myeloma protein, and excludes patients whose disease progressed within two years or whose abnormal protein persisted after radiotherapy.⁶ Absence of anemia, hypercalcemia, or renal impairment

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attributable to myeloma low, if present, concentrations of serum or urine monoclonal protein preserved levels of uninvolved immunoglobulins. The study aims to report large expansile plasmacytoma in the mandible of the oral cavity, which also mimicking malignant bone neoplasms in a female patient.

CASE REPORT

A 38-year-old female patient reported to the outpatient department of our hospital complaining of pain and swelling over the left lower one-third region of the face for one month. Past dental history revealed extraction of 38 one month back, after which she noticed a painful swelling which was initially peanut in size and gradually increased to attain the present size. Past medical history was non-contributory, and general physical

examination revealed no abnormality.

On extraoral examination, facial asymmetry on the left side of lower one-third of face is presented, with a hard non-tender swelling measuring 2x2cm in size, which was covered by skin (Fig.1).

Intraoral examination (Figure 2) revealed a well-defined solitary

erythematous growth in the left retromolar region. The lesion was extending posteriorly from the distal aspect of 36 to the retromolar pad, measuring approximately 3 cm × 3 cm. All the inspeactory findings were confirmed on palpation, and growth was soft to firm in consistency with smooth surface and well-defined margins.



Fig. 1: Extra Oral Findings: a hard non-tender swelling measuring 2x2cm in size was observed on the lower one-third of the posterior mandible, which was covered by normal-appearing skin



Fig. 2: Intra Oral Findings: A well-defined solitary erythematous growth in the left retromolar region. The lesion was extending posteriorly from the distal aspect of 36 to the retromolar pad, measuring approximately 3 cm × 3 cm.



Fig. 3: Orthopantomograph: a well-defined radiolucency involving 35 to ascending ramus of the mandible. Evidence of root resorption is seen in relation to 36.

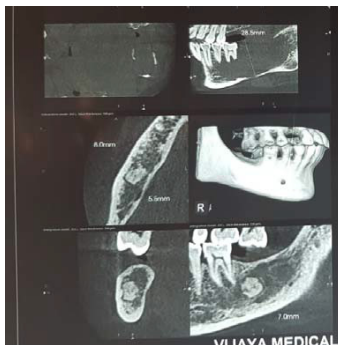


Fig. 4: CBCT- a hypodense area involving 35 regions extending towards ascending rami of the mandible



Fig. 5: Gross specimen -nine bits of soft tissue and one bit of hard tissue

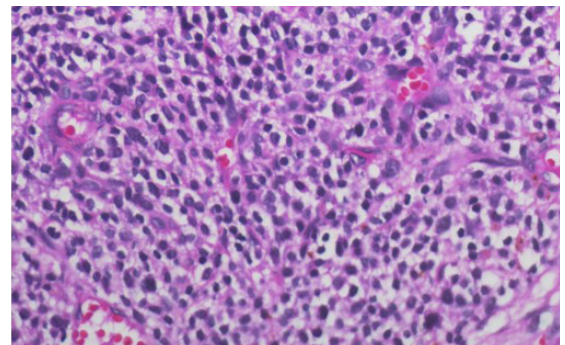


Fig. 6: Monotonous arrangement of plasma cells were seen (Hand E)

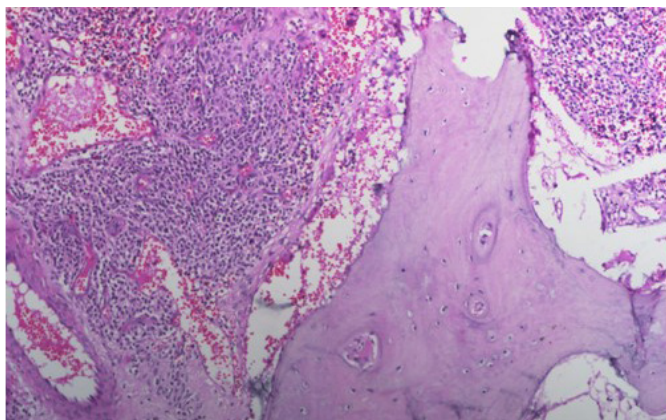


Fig. 7: Bone is fibroblastic and adjacent to bone tumor infiltration is evident as shown (H and E)

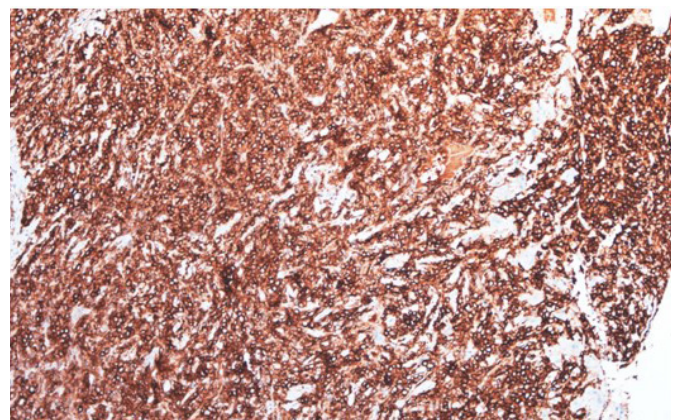


Fig. 8: CD138, shows diffusely positive (IHC).

On orthopantomogram analysis (Fig. 3), a well-defined radiolucency involving 35 to ascending ramus of the mandible is seen. Evidence of root resorption is seen in 36.

CBCT analysis shows (Fig. 4) a hypodense area involving 35 regions extending towards ascending rami of the mandible.

On blood analysis no changes were evident. Full body analysis was done on gross examination (Fig. 5) shows nine bits of soft tissue and one bit of hard tissue—the soft tissues were brownish-red and soft to firm inconsistency. A large bit was taken for processing.

On histopathological examination, the hematoxylin and eosin-stained sections exhibit epithelium and underlying connective tissue stroma. Bone is fibroblastic and adjacent to bone tumor infiltration is evident as shown in (Fig. 6). On immunohistochemistry analysis, CD138 (Fig. 7), shows positive.

Based on the clinical, radiographical histopathological and immune histochemical analysis final diagnosis is suggestive of Plasmacytoma and surgical excision was done.

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., 1983¹¹; 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.)⁷, 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 seems 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). ⁸Dimopoulos et al. affirm 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. ⁷Mock et al. (1987) suggested that the cases with lambda light chain may be more immature and more likely to progress to Multiple myeloma.¹⁰ In another study, Susnerwala et

al. observed that the cases that progressed to Multiple myeloma showed kappa light chain restriction.

Holland et al.¹⁰ affirm 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 Multiple myeloma.

Susnerwala et al.¹¹ affirm 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.).

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 the remission of the lesion.

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