Solitary Plasmacytoma of the Jaw- A Rare Case Report

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Abstract:
Plasma cell tumors are lymphoid neoplasms with an uncontrolled proliferation of B cells. These are divided into localized forms (solitary bone plasmacytoma -SBP and extramedullary plasmacytoma -EP) and disseminated forms (multiple myeloma–MM). SBP is most frequently seen in vertebrae and secondarily in long bones. Its presence in jaws is extremely rare. Solitary bone plasmacytoma is a rare condition affecting the jaws which manifests itself as a single osteolytic lesion without plasmacytosis of bone marrow and constitutes approximately 3% of all plasma containing tumors. It is different from multiple myeloma in terms of its clinical behavior and prognosis. The most common clinical presentation is local bone pain and lesion on alveolar ridge and affects mandible frequently than maxilla. We report a rare case of solitary plasmacytoma of mandible, diagnosed on the basis of distinct radiological and histopathological findings.

Keywords: B-cells, Plasmacytoma, Osteolytic lesion, Mandible

1. Introduction
Plasma cell neoplasms are a group of clinical disorders characterized by an uncontrolled monoclonal proliferation of plasma cells or plasma cells derived (stem cells and lymphoid cell line B) and, in the case of multiple myeloma, by monoclonal overproduction of immunoglobulins in blood and / or detecting light chains in the urine as Bence Jones protein.1

Plasma cell neoplasms (Plasmacytoma) maybe either of a localized or a disseminated form, with both solitary bone plasmacytoma (SBP) and extramedullary plasmacytoma (EMP) representing localized forms whereas Multiple myeloma (MM) representing the disseminated form of this disorder. Although both SBP and EMP initially are restricted to a single area, the former tends to disseminate and evolve into MM much more frequently than the latter. Thus, the two diseases are often considered as two distinct entities.2,1
Solitary Bone Plasmacytoma (SBP) is a type of lymphoma which arises from bone –marrow based B- cells specifically those which have undergone terminal differentiation into plasma cells. These tumors are monoclonal and this can be assessed in tissue sections or biochemical evaluation of type of immunoglobulin secreted by the neoplastic cells.

Plasma cells produce osteoclast-activating factors, which stimulate the growth of osteoclasts and therefore bone resorption occur. Plasmacytomas usually appear on radiographic images as radiolucent areas.

2. Case Report
A 49-year-old male patient complained of dull ache and mobility of tooth in the left lower back region. Clinical examination revealed a diffuse swelling was seen extra orally (Fig-1), which was starting anteriorly from corner of lip and extending posteriorly up to left pinna of the ear. The skin over the swelling appeared normal with no erythema. On Intraoral examination, a swelling at the left mandibular molar region was noticed. Tongue, lips, buccal and alveolar mucosa were normal in appearance and there was no history of swelling, discharge or any swelling in the body. On Palpation, the swelling was firm in consistency, mild tender and obliterated the buccal sulcus and was not attached to the overlying skin. The left submandibular lymph nodes were palpable measuring approximately 1 to 1.5 cm in diameter, firm in consistency, non-tender and mobile. An Orthopantomograph(fig-3) revealed an ill-defined multilocular, radiolucent lesion extending along the entire angle, ramus region without sclerotic border. The lesion extending along the left posterior border of the body of mandible, with marked expansion of buccal cortex. Based on the multilocular radiolucent lesion of the mandible on orthopantomograph, the differential diagnosis for this lesion included ameloblastoma, odontogenic myxoma, keratocystic odontogenic tumor; aneurysmal bone cyst; and metastatic carcinoma. Routine hematological and biochemical investigations (serum calcium, phosphorous, urea, uric acid, etc.) were within normal reference range. Biopsy was performed (Fig-2) and 38 was removed during biopsy and sample was sent for histopathological examination(Fig-5) which shows (Fig-4) there are many loose clusters and sheets of plasma cells. They contain moderate amount of amphophilic cytoplasm and peripherally placed round nucleus with dense chromatin pattern. Some of the large cells show prominent nucleolus. Based on the clinical presentation, radiological findings, and histopathology the diagnosis of solitary plasmacytoma was suggested.

3. Discussion
Neoplastic proliferation of plasma cells was discussed by Dalrymple and Bence Jones in 1846. These are described as a group of clinical disorders characterized by uncontrolled clonal proliferation of plasma cells. Plasma cell tumors are B-cell lymphoid neoplasm classified in three types by the site and no. of the lesion: Multiple myeloma (MM), Solitary bone plasmacytoma (SBP) and extramedullary plasmacytoma (EMP), Multiple myeloma is the most common type of plasma cell malignancy. Other types include extra-medullary plasmacytoma and solitary plasmacytoma. Solitary bone plasmacytoma is rare. Plasmacytomas are localized proliferations of plasma cells giving rise to a localized tumor mass which can be classified into two groups depending upon the site where they appear:
Solitary plasmacytoma is characterized by the collection of monoclonal plasma cells in the bones. It is seen generally in long bones but rarely occurred in the jaws and only 4.4% of SBP seen in the mandible. Solitary bone plasmacytoma is a localized malignant monoclonal plasma cells, which constitutes approximately 3-10% of all plasma cell tumors. The peak incidence is in the sixth decade of life, being more prevalent in male than females with a ratio 2:1. It rarely involves jaws and when it is seen, only 4.4% of SBP occur in the mandible, most commonly in the bone marrow-rich areas of the body, angle and ramus of mandible. Clinical signs and symptoms may be really poor; therefore, it may be very difficult to perform an early diagnosis of solitary bone plasmacytoma. Specific symptoms to suspect a diagnosis of solitary bone plasmacytoma localized in a unique bone segment are: pain, impairment of bone function and movements, presence of bone swelling with or without local erosion, involvement of local mucosa or tissues. Radiographically SBP is seen as a well-defined uni or multilocular lytic lesion without periosteal reaction, or as a protruding mass with cortical expansion. CT, MRI and more recently PET, are useful to establish the characteristics of the lesion and tissues involved and to rule out other affected areas.

Review of the literature over the last 20 years showed a total of 25 reported cases in the mandible and two cases in the maxilla with the most common site being the premolar-molar region. Involvement of the anterior region is rare with only one case being reported. Two cases of solitary plasmacytoma has been reported in the maxilla. Plasmacytomas are clinical entities where it is very difficult to confirm the diagnosis without radiological, histopathological, immunohistochemical and other supportive investigative modalities. In our case along with radiological, histopathological findings, support of Immunohistochemistry (IHC) positivity for CD138 was also done for establishing the final diagnosis.

Solitary bone plasmacytomas are highly radiosensitive lesions. Radiation therapy, radical extensive surgery or a combination of both is recommended as primary treatment. Radical radiotherapy comprising of 40-50Gy has shown 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 deficit or in cases where pathological fracture is anticipated (to prevent that fracture and stabilize the fracture mandible).

4. Conclusion

Solitary bone plasmacytoma is a localized form of plasma cell tumours. Maxillofacial area it is rarely affected, most commonly the mandible. This entity requires a meticulous overview of the patient by the specialist and overall the control of any signs or symptoms of systemic diseases, a fact that would mark a dramatic change in the treatment and prognosis of the patient. The course of SBP is relatively benign and survival is 50-80% at 10 years. The prognosis is worse if recurrence is present as in cases of evolution toward systemic disease.

References


**Figures**

**Figure 1:** Clinical picture showing a diffuse swelling appreciated on left lower side of face.

**Figure 2:** Clinical picture showing the sutures placed on extraction socket of 38.
**Figure 3:** OPG shows an ill-defined multilocular, radiolucent lesion in the left mandibular ramus and 3rd molar region.

**Figure 4:** Surgical specimen showing the extracted 3rd molar and the Biopsy sample

**Figure 5:** Histopathological picture showing loose clusters of plasma cells (H & E stain, 40x)