

Solitary Bone Plasmacytoma of Posterior Mandible: A Case Report

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Abstract

Background: Plasmacytoma is a monoclonal, neoplastic proliferation of plasma cells that usually arises within bone marrow or soft tissue sites. It can involve either a single bone (solitary) or multiple bones. Solitary plasmacytoma has a predisposition for the red marrow-containing axial skeleton and is most frequently seen in the thoracic vertebrae, followed by the ribs, sternum, clavicle, or scapula. Its presence in the jaws is extremely

rare. In the absence of typical clinical presentation, diagnosis may not be suspected clinico-radiologically.

Case presentation: We present a case of solitary plasmacytoma in a 70-year-old male with a painful swelling in the posterior region of mandible. An osteolytic lesion with periosteal reaction was evident on panoramic radiographs as well as CBCT images. An incisional biopsy confirmed it to be a case of Plasmacytoma.

Conclusions: Solitary plasmacytoma of the jaw bone is a rare clinical condition. But if it gets diagnosed on histological and immunohistochemistry studies, special precaution should be taken to rule out multiple myeloma.

Keywords: Mandible, myeloma, plasma cell, solitary plasmacytoma.

Introduction

Plasma cell dyscrasias are neoplastic proliferation of B-cells, and classified as multiple myeloma (MM, generalized medullary type) and plasmacytoma (localized extramedullary type). When the plasmacytoma occurs only in bone, it is known as solitary bone plasmacytoma (SBP) and when involving the soft tissue, it is named as extramedullary plasmacytoma (EMP); however, both the lesions are characterized by absence of systemic involvement attributing to myeloma. SBP arises from plasma cells of bone marrow (BM), whereas EMP arises from those in the mucosal surfaces. SBP is infrequent, representing 3-7% of all plasmacytoma. The mean age for SBP is 55 years and involves bones of the axial skeleton (vertebra, skull); involvement of jaw is rare. SBP has a significantly high risk of progression to MM (65-84% in 10 years).¹The jaws are rarely involved, with more predispositions toward the mandible. The most common symptom is pain at the site of the skeletal lesion due to bone destruction by the infiltrating plasma cells. Pathologic migration of teeth associated with swelling and paresthesia are observed in the case of larger lesions infiltrating into the neurovascular bundle. Fatigue and fever are the most common systemic symptoms.² Asymptomatic solitary bone plasmacytoma of the jaw is very rare but such a clinical form without pain has been described previously.³ In this case report, we present a case of 70 year old male with an ill defined radiolucent lesion in the left body region of the mandible later diagnosed as plasmacytoma.

Case Report

A 70-year-old male patient reported to our OPD with the chief complaint of pain and growth in the left lower back tooth region over the past 1 week. The pain was dull and nonradiating in nature and the growth had suddenly increased over the past 1 week. There was history of mobility of teeth in that region. On general examination patient was poorly built and nourished. General systemic examination was within normal limits. Extraoral examination revealed a firm, poorly circumscribed and mildly tender swelling measuring approximately 2x2 cm over the left lower cheek region [Figure 1a, 1b]. There was no evidence of palpable cervical lymphadenopathy. On intraoral examination, a solitary dome shaped soft tissue swelling of about 2.5x 1.5 cm appeared to be protruding out of the alveolus from the clinically missing 36 and 37 regions. The swelling was firm, mildly tender, nonfluctuant, with a slight, bluish discoloration and ulceration of the overlying mucosa [Figure 2]. 34 and 38 showed grade III mobility. He had generalized periodontitis and oral hygiene was poor. Panoramic imaging revealed an ill defined radiolucency involving the residual alveolus of 35, 36 and 37 38 simulating a floating tooth appearance. Root stumps of 14, 44 and 45 with periapical radiolucencies, generalized severe horizontal bone loss and multiple carious teeth were also noted on panoramic radiograph [Figure 3]. Cone Beam Computed tomography (CBCT) scan revealed an ill defined osteolytic lesion with bicortical expansion and perforation of the both the cortical plates along with periosteal reaction [Figure 4]. On the basis of clinical and radiological findings, a provisional diagnosis of primary intraosseous carcinoma was made. Incisional biopsy of the lesion and extraction of 38 were done. The tissue was sent for histopathological examination. The histopathological study showed sheets of mature plasma

cells along with immature and nucleolated cells permeating the bone, suggestive of plasmacytoma/myeloma [Figure 5]. In Complete Blood Count test, white blood cell, red blood cell, hemoglobin and hematocrit were low but Mean Corpuscular Volume (MCV) and platelet were in the normal range. In biochemistry test, urea and creatinine were normal and hypercalcaemia was not found so renal insufficiency was ruled out. Serum immunoelectrophoresis showed an increase in M-protein [immunoglobulin (IgG) κ type] and also a decrease in albumin of plasma and albumin/globulin ratio was lower than normal. In immunohistochemical staining, CD138, vimentin, Ki67, and EMA were positive. A complete clinical, hematological, biochemical and radiological work-up was done to exclude MM. All the above investigations were conclusive of solitary plasmacytoma of the mandible. The patient was referred to the radiation oncology unit for adjuvant radiotherapy. Initially he responded well, but did not turn up for further follow-ups due to postradiation complications.



Figure 2: Bluish red coloured dome shaped soft tissue swelling with superficial mucosal ulceration in relation to residual alveolus of 35 36 & 37 region.



Figure 3: Ill defined osteolytic lesion involving the residual alveolus of 35, 36 and 37



Figure 1a, 1b: A roughly circumscribed swelling over the lower left cheek region on extra oral examination

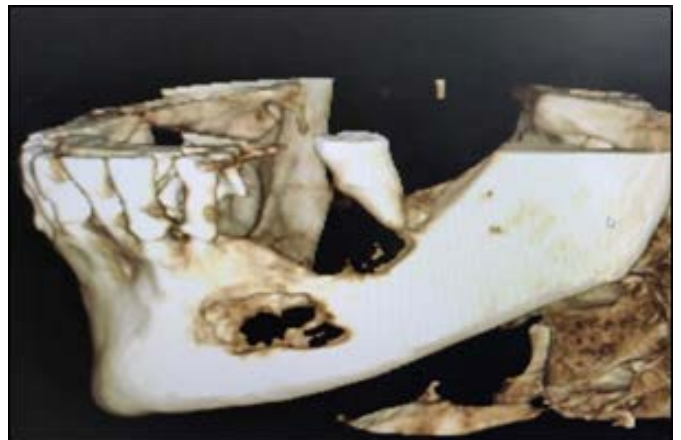


Figure 4a: CBCT 3D View



Figure 4b: Axial and coronal CBCT sections: Osteolytic lesion with bicortical perforation and periosteal reaction involving body of mandible on left side with adjacent soft tissue thickening

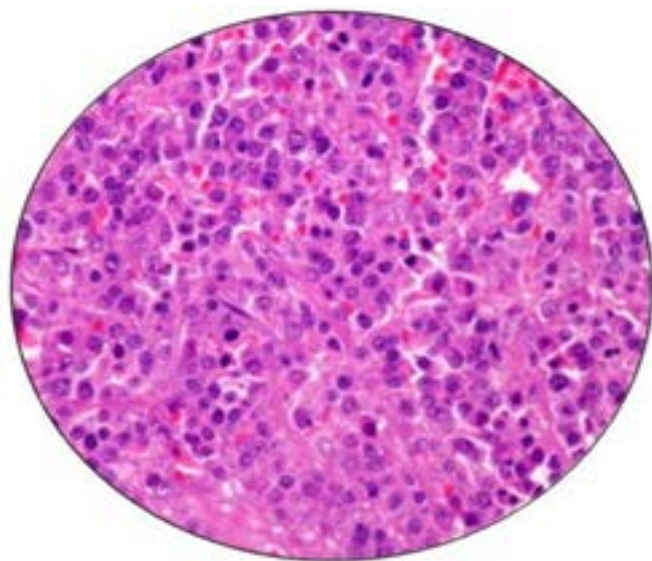


Figure 5: Photomicrograph revealing sheets of mature plasma cells along with immature and nucleated cells

permeating the bone, suggestive of plasmacytoma / myeloma.

Discussion

Solitary bone plasmacytoma is a rare immunoproliferative monoclonal disease with localized proliferation of plasma cells. It occurs more commonly in men than in women (M:F-2:1) and presents a decade younger than MM. It mostly involves the axial skeleton, that is, vertebra and skull; involvement of the jaw bone is rare. EMP generally involves the head and neck region with most common sites being nasal cavity and nasopharynx. In contrast to MM, both SBP and EMP show absence of CRAB (increased serum calcium, renal insufficiency, anemia, and multiple bone lesions). The diagnostic criteria for SBP includes a solitary bone lesion confirmed by skeletal survey, biopsy proven clonal plasma cell infiltration, lack of myeloma-related organ damage, plasma cell constituting <10% of BM nucleated cells and absence of urine/serum “M” protein. EMP, on the other hand, will be characterized by absence of any osteolytic bone lesion with monoclonal plasma cells on tissue biopsy⁴. Durie and Salmon⁵ have graded SBP as stage-I myeloma. Solitary plasmacytoma of the mandible also has various radiographic findings: from well-defined, unilocular radiolucency or “punched-out” appearance similar to multiple myeloma (MM) to ill-defined destructive radiolucencies with ragged borders but without periosteal reaction. A complete skeletal survey is beneficial in detecting an osteoblastic response to bone destruction. Histopathological analyses alone are not sufficient for making a diagnosis; all the above investigations are mandatory to exclude MM. To the best of our search, there are only few cases were published literature describing SBP of the jaw. However, recent advancements have improved the precision of diagnosis. Flow cytometry and molecular detection of heavy and light chain

rearrangements may reveal monoclonality of plasma cells⁶. The current criteria to make a diagnosis of solitary plasmacytoma are the following: Isolated area of bone destruction due to clonal plasma cells, bone marrow plasma cell infiltration not exceeding 5% of all nucleated cells, absence of further osteolytic bone lesions or other tissue involvement (i.e., no evidence of systemic plasmacytoma); absence of anemia, hypercalcemia, or renal impairment attributable to myeloma; low concentrations of serum or urine monoclonal protein (i.e., myeloma protein); or preserved levels of immunoglobulins⁷. Solitary plasmacytomas are highly radiosensitive lesions. Radiation therapy, radical extensive surgery, or a combination of both is recommended as primary treatment and surgical approach gave good outcomes^{8,9}. SBP has a benign clinical behavior but has a poor prognosis in comparison to EMP.

Conclusion

Solitary plasmacytoma of the jaw bone mostly in the mandible is a rare clinical condition. It is uncommon to suspect an ill defined radiolucency of the mandible as plasmacytoma. But if it gets diagnosed on histological and IHC studies, it should be taken into account to rule out its disseminated form, i.e., multiple myeloma. In spite of curative treatment, the median time of progression to MM is 2-3 years and the rate is 65-84% in 10 years¹⁰. Therefore an early diagnosis of solitary plasmacytoma of bone is essential for a better survival of the patient.

Declaration of patient consent: Written informed consent was obtained from the patient for publication of this case report and any accompanying images.

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