Solitary Plasmacytoma of Mandible - Report of 2 Cases

B. Sekar, Dominic Augustine and S. Murali
Department of Oral & Maxillofacial Pathology, Vinayaka Missions Sankarachariyar Dental College
Ariyanoor, Tamil Nadu 636308, India

Abstract: A plasmacytoma is a discrete, solitary mass of neoplastic monoclonal plasma cells in either bone or soft tissue (extramedullary). Solitary plasmacytomas can be divided into 2 groups according to location, plasmacytoma of the skeletal system (SBP) and extramedullary plasmacytoma (EMP) respectively. Less than 5% of patients with a plasma cell dyscrasia present with a single bone (SBP) or extramedullary plasmacytoma (EMP). We present 2 rare cases of solitary plasmacytoma of bone affecting the mandible of 60 year old female and a 58 year old male.

Key words: Plasmacytoma, solitary, mandible.

Introduction

A plasmacytoma is a discrete, solitary mass of neoplastic monoclonal plasma cells in either bone or soft tissue extramedullary). Primary plasmacytoma, whether osseous or nonosseous, is distinguished from multiple myeloma by absence of hypercalcemia, renal insufficiency and anaemia, normal skeletal survey, absence of bone marrow plasmacytosis, and serum or urinary paraprotein < 2 g/dL.[1]

Epidemiological data continues to confirm a male:female ratio of 3:1 with median age of 60 years in patients with Solitary bone plasmacytoma [SBP].The axial skeleton especially the vertebrae remains the commonest site for SBP.[2] The mandible remains a rare site for occurrence of solitary bone plasmacytoma. Solitary bone plasmacytoma (SBP) develops into multiple myeloma in 50-60% of patients. Median overall survival time is 10 years.[3]

A solitary bone plasmacytoma (SBP) arises from the plasma cells located in the bone marrow, whereas extramedullary plasmacytoma (EMP) is thought to arise from plasma cells located in mucosal surfaces.[4] Both represent a different group of neoplasms in terms of location, tumor progression, and overall survival rate.[5] Some authors suggest a solitary bone plasmacytoma (SBP) represents marginal cell lymphomas with extensive plasmacytic differentiation.[6]

Case Report 1

A 60 year old female visited the Oral & maxillofacial pathology department (Vinayaka Missions Sankarachariyar Dental College and Hospital – Salem) with a chief complaint of swelling on the left side of the face and difficulty in eating for the past two months. Her medical history was non-contributory, extra oral examination showed a diffuse swelling over the ramus of the mandible. [Figure 1] It was non-tender and bony hard in consistency. No lymph nodes were palpable. Intraoral examination showed a swelling in the retromolar area. Patient had difficulty in opening her mouth. OPG revealed a lytic lesion without sclerotic border involving the angle of the mandible and the ascending ramus, impacted 38 was present. Pathologic fracture was present. [Figure 2] A provisional diagnosis of a bone malignancy was thought of.

Case Report 2

A 58 year old male presented with a fungating mass covering the mandibular edentulous alveolus, only teeth numbers 34, 35, 36, 37, 38 & 48 were present in the lower jaw. The maxillary posteriors were absent [Figure 3] It was tender to palpate and profuse bleed-
ing was observed while palpating. It was immobile and attached to the underlying bone. OPG revealed a large lytic lesion extending from 35 to 48 giving a moth eaten appearance. [Figure 4] A provisional diagnosis of primary intraosseous squamous cell carcinoma was made. An incisional biopsy was performed for both the cases under local anesthesia and the tissue was subjected to histopathological examination. Microscopically the tumour was composed of sheets of densely packed round and oval cells in a relatively sparse stroma. The neoplastic cells had large, single eccentric nucleus, resembling a typical plasma cell. Cells with double nuclei and abnormal mitotic figures were also seen. [Figure 5] Positivity for light chain kappa was found by immunohistochemistry [Figure 6] and negativity for lambda.

Table 1. International Myeloma Working Group diagnostic Criteria

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| Solitary plasmacytoma of bone | No M-protein in serum and/or urine*  
Single area of bone destruction due to clonal plasma cells  
Bone marrow not consistent with multiple myeloma (plasma cells <5%)  
Normal skeletal survey (and MRI of spine and pelvis if done)  
No related organ or tissue impairment |
| Extramedullary plasmacytoma | No M-protein in serum and/or urine*  
Extramedullary tumour of clonal plasma cells  
Normal bone marrow  
Normal skeletal survey  
No related organ or tissue impairment |
| Multiple solitary plasmacytomas (+/- recurrent) | No M-protein in serum and/or urine*  
More than one localised area of bone destruction or extramedullary tumour of clonal plasma cells which may be recurrent |

A final diagnosis of solitary plasmacytoma of bone was established for both the cases. Skeletal survey showed no lytic lesions. Urine was negative for Bence Jones Protein. Bone marrow study and serum protein electrophoresis continued the diagnosis as solitary plasmacytoma and multiple myeloma was ruled out. Both the patients are undergoing radiotherapy and are under follow up.

Figure 1. A large diffuse swelling over the left ramus of the mandible.

Figure 2. Orthopantomogram (OPG): A lytic lesion without sclerotic border involving the angle of the mandible and the ascending ramus with pathologic fracture.

Figure 3. A fungating mass covering the mandibular edentulous alveolus.
Discussion

Plasma cell tumours include the disseminated form, multiple myeloma, and the medullary (solitary plasmacytoma of the bone) and the extramedullary localized form. Some patients with plasma cell myeloma present with a single painful bone lesion due to a monoclonal plasma cell infiltrate, and further studies show no evidence of myeloma elsewhere. In other cases, solitary bone plasmacytoma (SBP) may be discovered during roentgenographic studies for another condition or the patient presents with a painless swelling of the sternum, rib, or other bone. SBP affects less than 5% of patients with plasma cell myeloma. [7]

Solitary bone plasmacytoma (SBP) may involve any bone, but it has a predilection for the red marrow-containing axial skeleton. Spinal disease is observed in 34-72% of cases. The thoracic vertebrae are most commonly involved, followed by lumbar, sacral, and cervical vertebrae. The rib, sternum, clavicle, or scapula is involved in 20% of cases. Involvement of mandible is rare as present in two of our cases. Physical findings are related to the site of involvement, presenting as a painful mass, pathologic fracture, or root or spinal cord compression syndrome. [8] Occasionally, patients with solitary bone plasmacytoma (SBP) may present with peripheral polyneuropathy or with features that are consistent with POEMS syndrome (polyneuropathy, organomegaly, endocrinopathy, M protein, and skin changes). [9]

The majority of patients with apparent SBP continue to develop myeloma. Up to 75% of patients at diagnosis may have a monoclonal protein in blood or urine, this is usually <10g/l. The rare cancer network published data on 206 patients with SBP, the largest series to date, in 2006. Despite treatment, 104 of 206 (50.4%) patients developed myeloma with a median time to development of 21 months. [10] Both of our patients haven’t developed myeloma yet and are under regular follow up.

In 2003 the International Myeloma Working Group (IMWG) published criteria for the classification of monoclonal gammopathies, multiple myeloma and related disorders recognising solitary plasmacytoma of bone, extramedullary plasmacytoma and multiple solitary plasmacytomas (+/-recurrent) as distinct entities. [11] The diagnostic criteria are listed in Table 1.

Diagnosis requires a biopsy-proven monoclonal plasma cell infiltration of a single lytic bone lesion; no additional lesions on bone survey; absence of clonal plasma cells on a random marrow sample; and no evidence of systemic myeloma (normocalcemia, absence of anemia or renal disease attributable to myeloma). These findings were consistent with our cases. Additionally, plasma cells with neoplastic phenotype demonstrable by flow cytometry at bone marrow sites distant to solitary plasmacytoma would also appear to predict for progression to myeloma.

Cytogenetic studies show recurrent losses in chromosome 13, chromosome arm 1p, and chromosome arm 14q, as well as gains in chromosome arms 19p, 9q, and 1q. Interleukin 6 (IL-6) is still considered the principal growth factor in the progression of plasma cell disorders. [12]

Radical radiotherapy remains the treatment of choice for SBP. Knobel et al [13] confirmed excellent local dis-
ease control with radiotherapy alone in their review of 206 patients with SBP. Local relapse occurred in 21 (14%) out of 148 patients who received radiotherapy alone compared with 4(80%) out of 5 patients who were treated with surgery +/- chemotherapy. Surgery (radiotherapy versus partial or complete resection and radiotherapy) did not influence the 10-year probability of local control. Recently, free light chain assays have provided a measurable parameter to follow in approximately 65% of patients previously diagnosed with “nonsecretory” multiple myeloma by standard electrophoretic studies. It is likely that these assays will also be useful in a similar percentage of patients with SBP. Like MRI, this would improve the precision of monitoring SBP and provide more sensitive identification of patients who achieve complete disappearance of paraprotein after radiation therapy and are most likely to be cured. 

Kyle [15] described 3 patterns of treatment failure: development of multiple myeloma (54%), local recurrence (11%), and development of new bone lesions in the absence of multiple myeloma (2%). Both our patients are undergoing radiotherapy and are showing signs of improvement.

**Conclusion**

While the majority of patients with solitary plasmacytoma of bone develop myeloma after a median of 2-3 years, the overall median survival of 7-12 years is longer than for patients in early phases of symptomatic myeloma. Approximately 15%-45% of patients remain disease free at 10 years, and although the majority of these appear to be cured, rare late recurrences have been reported. We have reported 2 rare cases of plasmacytoma occurring in the mandible.

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**References**


