Multiple myeloma with primary manifestation in the mandible: a case report

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Abstract

Plasma cell neoplasms (multiple myeloma, solitary plasmocytoma of bone and extra medullar plasmocytoma) are characterized by a monoclonal neoplastic proliferation of plasma cells. Multiple myeloma is a monoclonal malignant proliferation of plasma cells that causes osteolytic lesions in the vertebrae, ribs, pelvic bone, skull and jaw. We report a case of a 69-year-old male patient who presented with a tumefaction in the body of mandible, which had evolved over the previous two months. In the radiographic examination, an extensive osteolytic lesion was observed in the body of mandible. An incisional biopsy was performed and histopathological study revealed a malignant hematopoietic neoplasm formed by plasmacytoid cells. Knowledge about the maxillofacial manifestations of multiple myeloma is important for the early diagnosis of the disease, since its primary form can manifest itself in the jaw.

Key words: Multiple myeloma, plasmacytoma, mandibular lesion.

Introduction

Multiple myeloma (MM) is a relatively rare malignant hematological disease, which is characterized by the multicentric proliferation of plasma cells in the bone marrow. It develops mainly in men aged 50 to 80 years, with a mean age of 60 years [(1)]. An intriguing feature of multiple myeloma is that the antibody-forming cells (ie, plasma cells) are malignant and, therefore, may cause unusual manifestations like recurrent infections, fever, fatigue, hematologic alterations, nephropathy and persistent bone pain. Its occurrence in the maxillaries is very common; however oral lesions rarely appear with primary manifestation of the disease. More than 30% of patients with MM develop osteolytic lesions in the jaw [(2)]. The maxillary lesions are more frequent in the posterior region of the mandible and the pain in the maxillaries may be the initial symptom of the disease. Manifestations such as gingival hemorrhage, odontalgia, paresthesias, dental mobility, ulcerations and increased volume may also be present [(3)]. The incidence is 4 cases per 100,000 population per year. Multiple myeloma is rare among the Asian population, with an incidence of 2 cases per 100,000. In this case report, we discuss a Multiple myeloma with primary manifestation in the mandible.

Case report

A 69 year old-male patient presented with complaint of painless swelling in the right lower face region of 2months duration. Initially he noticed a small swelling with a mobile tooth in the right posterior mandible region. The swelling was rapidly progressive and was not associated with other systemic symptoms like fever, night sweats, weight loss and paresthesia. The patient's previous medical history was non contributory. On general physical examination pallor was noticed and facial asymmetry was detected on the right side of the face due to well defined swelling involving right lower face region as shown in **Figure 1**. On palpation, there was hard consistency with no alteration in the local cutaneous tissue, and paresthesia . No significant lymphadenopathy was noticed.

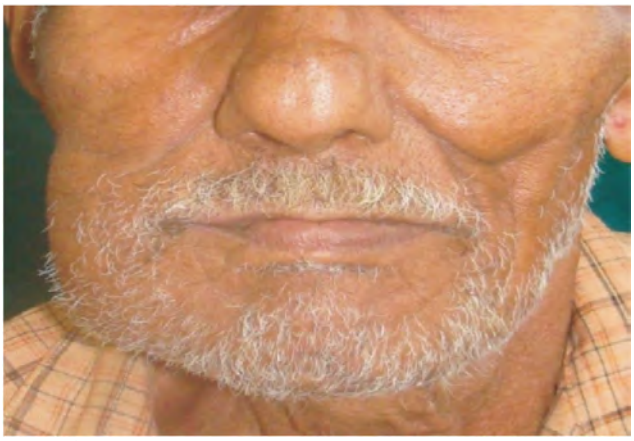

Figure 1 Extraoral photograph showed swelling on the right side of lower jaw.

On intraoral examination, a well defined expansile swelling involving the right side of the alveolus in the mandibular arch noticed measuring approximately 3x4 cm in size. Lingually causing expansion of lingual cortex with partial obliteration of floor of the mouth and buccally causing complete obliteration of the lower buccal vestibule due to expansion of buccal cortex **(Figure 2)**. The overlying mucosa appeared normal except at one area where ulceration was noticed probably due to impingement of the opposing teeth. The fine needle aspiration showed a minimal amount of blood.

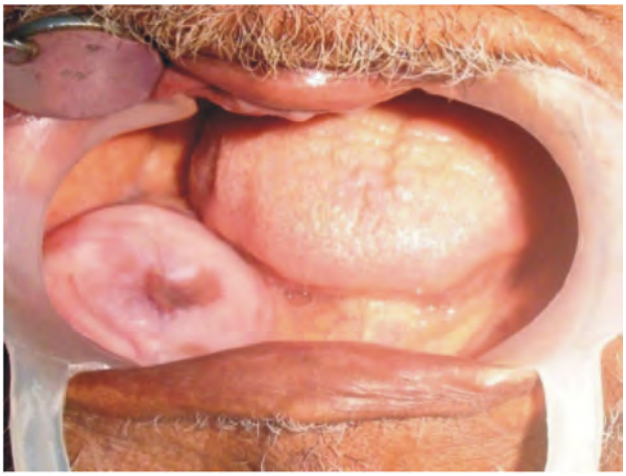

Figure 2 Intraoral photograph showed a well defined swelling of right posterior region of the mandible with bicortical expansion

A provisional diagnosis of odontogenic tumor of the body of the mandible was made depending upon the clinical picture. On radiographic examination, the panoramic view showed a solitary radiolucent lesion involving the right side of the body of the mandible measuring roughly 5x4 cm in size. The Interior of the radiolucency showed haziness and presence of trabeculae in various directions. The superior border of the body appeared discontinuous and the inferior cortex of the mandible appeared continuous without any thinning or breach but it showed few fine radiating, radiopaque lines suggestive of periosteal reaction.**(Figure 3)** Expansion of bone in the bucco-lingual direction, cortical bone destruction and periosteal reaction were also realized from the occlusal radiograph.**(Figure 4)** The lateral skull view showed multiple, well defined, round, radiolucent lesions measuring involving the calveria.**(Figure 5)**

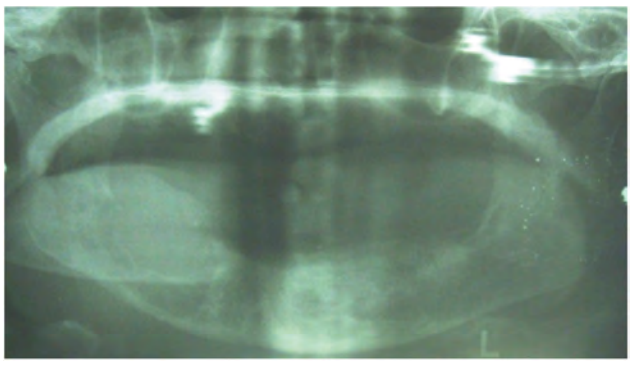

Figure 3 Panoramic radiograph showed a solitary radiolucent lesion involving right side body of the mandible with few fine radiopaque lines at the inferior border of mandible suggestive of periosteal reaction.

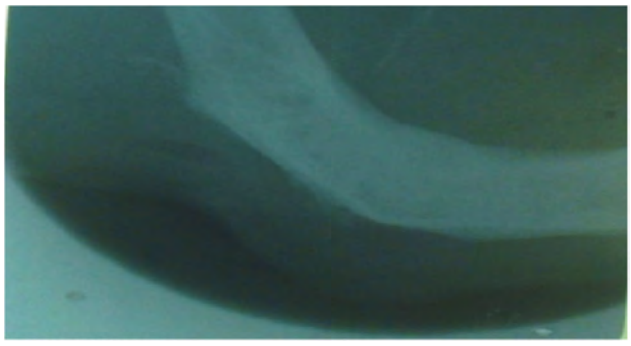

Figure 4 Mandibular occlusal view showed bicortical expansion with perforation of buccal and lingual cortex.

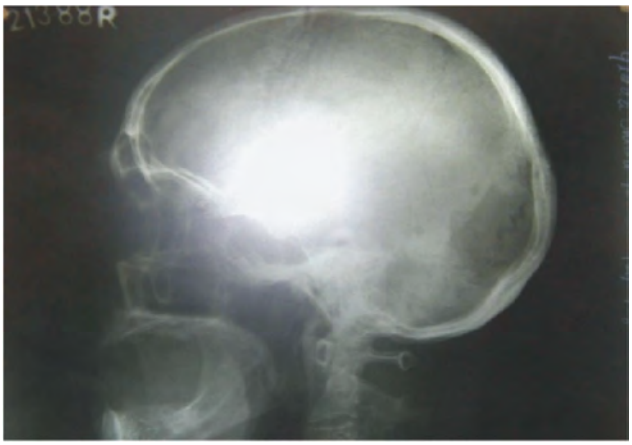

Figure 5 Lateral skull radiograph showed multiple, well defined, round, radiolucent lesions involving the calveria.

In the hemogram, a decrease in the number of erythrocytes, concentration of hemoglobin, hematocrit and average corpuscular volume was observed, which is indicative of normochromic microcytic anemia. The erythrocyte sedimentation rate (ESR) was also raised.

An incisional biopsy was performed. Histological analysis revealed sheets of plasmacytoid cells, with round, eccentric nuclei with fine granular chromatin and evident nucleolus and intervening fibrous tissue, characteristics of a solid malignant hematopoietic neoplasm **(Figure 6)**. The diagnosis was of plasmocytoma.

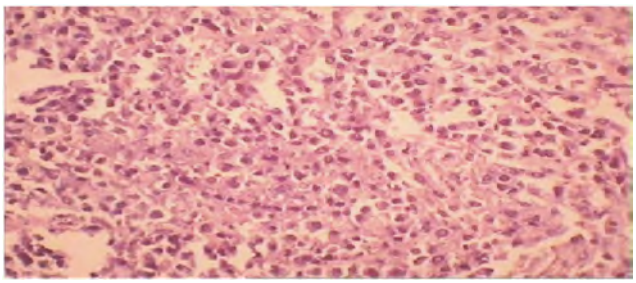

Figure 6 Photomicrograph of intraoral lesion revealed sheets of plasmacytoid cells, with round, eccentric nuclei and evident nucleolus and intervening fibrous tissue.

Later, chest x-ray was taken which showed no significant changes. Urine protein electrophoresis which was performed to identify the presence of the Bence Jones protein in urine was negative. Serum protein electrophoresis showed raised concentration of M-band proteins. Chemical screening, including serum calcium and creatinine, alkaline phosphatase level, and albumin showed no significant changes. Based on the clinical laboratory finding and radiodiagnosis, the definitive diagnosis of Multiple Myeloma was established. The patient was referred to the Oncological Hospital for treatment

Discussion

Plasma cell myeloma (plasmacytoma) may present as one of three distinct clinical entities: multiple myeloma, solitary plasmacytoma of bone, and extramedullary plasmacytoma [(1)]. Multiple myeloma is the multifocal disseminated form and is a malignant neoplastic condition characterized by the proliferation of a single clone of abnormal plasma cells [(4)]. Secondary invasion of the skeletal tissue is one of the most important characteristics of the disease [(5)]. Myelomatous infiltrates commonly involve the calvaria and mandible, pelvis, sternum, clavicle and proximal portions of the humerus and femur[(6)].

Multiple myeloma is most common in patients older than 40 years of age with a peak incidence rate at 60-70 years. The incidence of the oral lesions in multiple myeloma varies from less than 2% to 70% [7]. Epstein et al[4] reviewed 783 patients in the literature and indicated that approximately 14% of patients had oral manifestations. Its occurrence in the jaw is common and frequently occurs in the advanced stage of the disease but it rarely occur as the first sign of the disease with an incidence varying from 8% to15% [7] . In the present case, the lesions damaged the mandible and calvaria. Owing to its dimension and the bone damage caused by the mandibular lesion, we believe that it was a case of primary mandibular lesion. In the mandible , the posterior body and ramus are favoured which is compatible with this case. Orally, patient may complain of dental pain, swelling, paresthesia, dental mobility, hemorrhages and pathological fractures or there may have no complaints [4]. In the present case there was increased volume in the mandibular body with no painful symptomatology and paresthesia.

The most frequent radiographic characteristics in MM are osteolytic lesions with a "punched out" appearance. However, many appeared ragged and even infiltrative or may give multilocular appearance in case of aggressive lesions. Occasionally islands of residual bone yet unaffected by tumor, give the appearance of the presence of new trabecular bone within the mass. Periosteal reaction is uncommon, but if present, it takes the form of a single radioopaque line or more rarely a sunray appearance[8]. In the present case, a solitary osteolytic lesion was noticed involving body of mandible with ragged borders, interior of the lesion shows residual bone and periosteal reaction at the inferior border giving sunray appearance. A lateral skull radiograph showed osteolytic lesions with "punched out" appearance and contributed to establishing the definitive diagnosis.

The neoplastic aspect of the plasmacytoid cells was compatible with that described in the literature. Knowledge of the maxillofacial manifestations of multiple myeloma on the part of the dentist is important for early diagnosis of the disease, especially when it occurs in its primary form in the maxillary bones.

Although multiple myeloma (MM) remains incurable, several drug therapies are valuable in the treatment of patients with MM. A combination of melphalan and prednisone remains the standard chemotherapy. As monotherapy or in combination, interferon alfa-2b and prednisone modestly prolong the disease-free interval. Early evidence suggests that bisphosphonates may be effective in treating bone pain and in decreasing the likelihood of lesion recurrence.

Combination chemotherapies may be appropriate. Vincristine, Adriamycin, and dexamethasone (VAD) chemotherapy is also used in MM treatment. It is the best standard-dose treatment for patients in whom relapse occurs.

For MM therapy, the MP regimen is no longer the criterion standard, as it is suitable for less than 50% of patients. Alternative approaches, such as VAD-based regimens and high-dose chemotherapies with stem-cell support are preferred for most patients.

Conclusion

This case of multiple myeloma is a rarity and illustrates the wide variation in presentation. Knowledge of the maxillofacial manifestations of multiple myeloma on the part of the dentist is important for early diagnosis of the disease, especially when it occurs in its primary form in the maxillary bones.

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