

Mandibular Swelling as the First Manifestation of Multiple Myeloma

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Abstract

Multiple myeloma is a monoclonal malignant proliferation of plasma cells. The disease is more frequent in men, and the average age of patients at the time of diagnosis of the disease is about 60 years. Solitary myeloma may be the first manifestation of the disseminated form of multiple myeloma, which is characterized by multiple skeletal lesions, general metabolic alterations, impairment of renal function and eventually death.

The findings in regards to the present case suggest that oral manifestations may be the first sign of multiple myeloma. This might highlight the important role that a dentist can have in the early diagnosis of the disease.

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Keywords • Multiple myeloma • jaw neoplasm • oral manifestation

Introduction

Multiple myeloma is the most aggressive plasma cell neoplasm and the most common primary malignancy of bone.¹ It is primarily a disease of older adults, more common in men, and is usually diagnosed when the patients are around 60-years-old.^{1,2} Although the incidence is higher in populations exposed to radiation, pesticides and herbicides, the exact risk factors for the malignancy in most patients remain unclear.³ The monoclonal proliferation of plasma cells result in the production of a monoclonal immunoglobulin fragments. The accumulation of these products in bone marrow sets the patient at the risk of pathologic fracture, renal insufficiency anemia, infection and bleeding.³ The monoclonal proliferation of light- or heavy-chain immunoglobulin is remarkable in serum and/or urine in more than 99% of cases.⁴

The skeletal lesions are mainly confined to areas of red bone marrow, and are found in ribs, vertebrae, pelvis, skull, clavicle, sternum, femur and humerus.⁵ Clinically, multiple myeloma manifests as painful lytic bone lesions and fractures, and low blood counts.⁶ More than 30% of patients with multiple myeloma develop osteolytic lesions in the jaws. The maxillary lesions are more frequent in the posterior region of the jaw, and the pain in the maxillaries may be the initial symptom of the disease.⁷

Diagnosis can be established by the demonstration of malignant plasma cells on histology examination of affected bone with clonality usually confirmed by immunohistochemistry or flowcytometry for light chain restriction.³ Electrophoresis of urine and serum, and radiographic examination for multiple lytic bone lesions are also useful.²

The condition is almost always considered fatal, and the

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average survival time is thought to be 1.5-2 years after the diagnosis.⁵ Treatment involves mainly irradiation and chemotherapy, and prognosis is generally poor.⁶

The objective of this report is to present a case, which is the first manifestation of multiple myeloma in the jaw region.

Case Description

The case was a 58-year-old female referring to Department of Oral Medicine, Mashhad Dental School. Her medical history showed that she had not been prescribed medication, and had no medical allergies. There was no family history of cancer. The mandibular swelling had first been noted 2 months before patient's referral. Pain also appeared along with swelling. Initially her total denture was suspected as the cause of swelling. Clinical examination revealed a swelling with rubbery consistency extending from the right alveolar region and vestibule of first molar region to retromolar pad area with an ulcer in overlying mucosa adjacent to ascending ramus. No cervical lymphadenopathy was present (figure 1). No evidence of paresthesia or anesthesia of the face was seen. The primary diagnosis was a malignant mesenchymal tumor.



Figure 1: Intraoral examination of the tumor: swelling is realized in the right mandibular area

An orthopantograph revealed a paunch out radiolucencies in rarefied area extending from the first molar to the ascending ramus (figure 2, 3). The skull radiography was normal. Langerhans histiocytosis and metastatic malignant lesions should be included in radiographic differential diagnosis. Differential diagnosis of multiple myeloma from other metastatic bone lesions necessitate further clinical, histopathological and laboratory examination. Consequently, the patient was referred to a surgeon and an oncologist for further examination and treatment.



Figure 2: The panoramic view of the patient's face. It shows large ill-defined radiolucency in right mandibular side with multiple small round lesions that are paunch out.



Figure 3: Cropped and magnified view of right side of panoramic radiograph shows paunch out radiolucency

An incisional biopsy was obtained from the mass with the provisional diagnosis of malignant mesenchymal tumor. Histopathology examination revealed malignant plasma cells with

round, eccentric nuclear in a loose fibrous connective tissue stroma (figure 4). The histopathology diagnosis was plasmacytoma. Laboratory findings including complete blood counts (CBC), and serum concentrations of alkaline phosphatase (ALP), Calcium (Ca) and were within normal limits.

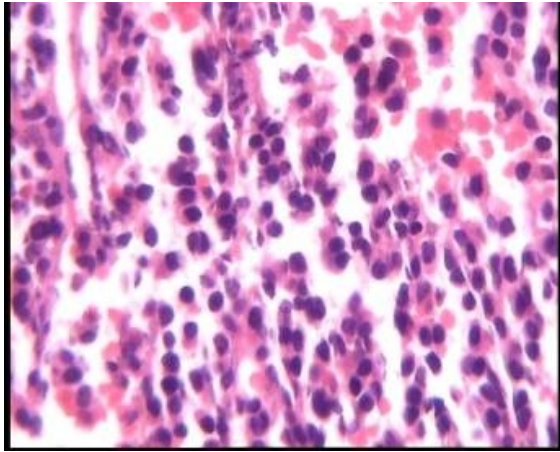


Figure 4: Histopathology of the biopsy from the tumor: malignant neoplastic cells with plasmacytoid pattern can be seen.

After referring the patient to an oncologist, treatment plan consisted of performing bone marrow aspiration and complete laboratory work up. In bone marrow aspiration, over %30 of cellular elements were plasma cells, many of which were atypical, binuclear or immature. Diagnosis of the bone marrow aspiration was multiple myeloma. Serum electrophoresis evaluation revealed no gammopathy (IgG, IgM and IgA were within normal range). However, urine protein electrophoresis showed high monoclonal Kappa light chain. In urine immunofixation, kappa was positive and lambda was negative. Immunohistochemistry of tumor cells also showed a positive Kappa marker.

Based on the clinical, laboratory and radiographic findings, the definitive diagnosis of multiple myeloma was made. The patient was referred to the Imam Reza Hospital for treatment. The patient underwent chemotherapy and radiotherapy. The patient was under medical care after two year of follow up.

Discussion

The present report describes a 58-year-old female patient who developed unilateral swelling of mandible as the first manifestation of multiple myeloma. She experienced no other signs and symptoms characteristic for multiple myeloma.

The patient was in the 6th decade of her life, which has been described as an age range in which multiple myeloma occurs frequently.

Multiple myeloma is a hematologic malignancy that is characterized by monoclonal proliferation of an abnormal plasma cell. Diagnostic techniques used to evaluate patients with suspected multiple myeloma include radiographic evaluation, blood test, and urine analysis and bone marrow biopsy.³

Osteolytic lesions are seen in 79% of multiple myeloma cases. The most often involved sites are the vertebrae, skull, pelvic bones, ribs, humerus and femurs. Jaws may be involved in 30% of the cases.⁶ Although jaw involvement is frequent, the primary manifestation of the disease in the head and neck is uncommon, as detected in our case. The radiographic appearance of the lesions is generally paunch out osteolytic lesions (60%).^{6,8} In the present case, a typical paunch out lesion was found in a rare area.

Laboratorial tests for multiple myeloma usually reveal anemia (73%).² Hypocalcaemia is also a regular feature, which was not detected in the present case. Serum electrophoresis shows M-protein in about 93% of the patients. Approximately 70% of myeloma secretes IgG, and kappa light chains are common as well.² However, in the present case serum protein electrophoresis showed no monoclonal spike.

Immunoglobulin can be detected within the cytoplasm of malignant plasma cells.² Immunohistochemistry revealed that immunoglobulin was present in urine and biopsy sample of the present case.

Bone marrow examination has shown that a level of 10% or more plasmacytosis is observed in 96% of patients with multiple myeloma.² Medullar examination of the present patient showed that more than 30% plasma cells were present, however after all the final diagnosis of multiple myeloma the patient was without renal failure.

The treatment of multiple myeloma consists of chemotherapy.⁶ Allopurinol, Melphalan, Pamidronate and prednisone were prescribed for this case. Although there was no evidence of paresthesia or anesthesia of face at the first visit, the patient complained of bone pain, fatigue and paresthesia of lower lip several days after treatment. Such manifestations might suggest that multiple myeloma needs to be included in the differential diagnosis of cranial neuropathies. They also might highlight the important role that a dentist can play in the early diagnosis of the disease.

When a plasmocytoma lesion is encountered, multiple myeloma should always be excluded.¹ Therefore, the need for a detailed investigation has been emphasized not only to distinguish plasmocytoma and multiple myeloma, but also to detect complications which may necessitate adjunctive treatment.⁴

Conclusion

A dentist's knowledge about the maxillofacial manifestations and diagnostic techniques of multiple myeloma is important for the early diagnosis of the disease, since, as was seen in the present case, the primary form of the disease can manifest in the orofacial region.

Conflict of Interest: None declared

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