**CASE REPORT**

**Multiple Myeloma: Report of a Very Rare Case and Review**

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**ABSTRACT**

Multiple myeloma consists of a clonal proliferation of plasma cells in the bone marrow, with varying degrees of differentiation. The disease is more frequently seen in men and the average age at diagnosis is about 60 years. The diagnosis is established by blood and urine examination and biopsy. Patients may present renal failure, bone pain, fatigue, recurrent infections and nervous system dysfunction. Oral manifestations may be the first sign of multiple myeloma, highlighting the importance of the dentist in the early diagnosis of the disease. This paper reports a case of multiple myeloma in a 72 years old male patient.

**Keywords:** Multiple myeloma, Plasma cells, Plasmacytoma, Clonal proliferation.


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**Conflict of interest:** None declared

**INTRODUCTION**

Multiple myeloma (MM) is a relatively rare malignant hematological disease, characterized by multicentric proliferation of plasma cells in the bone marrow. It accounts for about 1% of all types of malignancies and slightly more than 10% of hematologic malignancies. The term multiple myeloma was coined by von Rustizky in 1873.1,2 The common clinical manifestations are persistent pain in the bone, predominantly in the affected areas, a history of recurrent infection, fever, fatigue, hematological alterations, nephropathy and temporal arteritis. More than 30% of patients with MM develop jaw lesions and are more frequent in the posterior region of the mandible.3 This article reports two cases of multiple myeloma with initial presentation in the oral cavity.

**CASE REPORT**

A 72-year-old patient came to outpatient department with a chief complaint of bad breath for the past 2 months, which developed soon after getting up in the morning and in the evenings followed by a sensation of foul odor in his mouth throughout the day. The bad breath persisted even after brushing his teeth. The patient had supragingival calculus on the cervical aspect of the labial and lingual surfaces of all teeth, bleeding on probing and the marginal gingiva in relation to 41, 42, 31 and 32 appearing erythematous with gingival recession in relation to 41, 42, 31 and 32 (Fig. 1).

The medical history revealed pain in relation to both hips for 1 year, which was initially dull aching in nature, localized, intermittent and aggravated on lifting weights or standing, and relieved on medication. For the past 2 months patient gives history of pain over lower aspect of spine radiating to both lower limbs. Patient initially visited a nearby doctor and received treatment with analgesics. Patient now has headache along with pain in the hips and back pain for the past 1 month for which he is taking analgesics. Pain is dull aching in nature, localized, intermittent, aggravated on turning the head and relieved on medication.

Upon review of systems there is presence of headache, lower back pain in relation to lumbar spine L2 and pain present in relation to pelvis. Upon investigations, urine was positive for Bence Jones proteins with increase in the phosphate level in the urine. Blood investigations revealed raised ESR, raised serum urea and serum creatinine levels.
The rheumatology profile was positive for C-reactive protein, and urine chemistry showed raised levels of urine spot protein. Serum immunofixation electrophoresis showed the presence of lambda-free light chain disease.

Radiographic features of PA skull and lateral skull showed the presence of multiple punched-out radiolucencies of the entire skull with absence of any sclerotic border with size ranging from 2 to 7 mm throughout the entire skull (Figs 2A and B).

Lateral spine radiograph (Fig. 3) showed the presence of multiple radiolucencies of the spine with absence of any sclerotic border with size ranging from 2 to 5 mm throughout the entire spine.

PA of the pelvis (Fig. 4) showed the presence of multiple radiolucencies of the pelvis with absence of any sclerotic border with size ranging from 5 to 7 mm throughout the lower half of pelvis.

99m Tc-MDP whole body bone scintigraphy (Fig. 5) demonstrates diffusely intense, increased and inhomogeneous tracer concentration in both sternoclavicular joints and in the costochondral junction. The entire vertebral column has increased concentration, more in the lumbar spine. Both shoulders, elbow, wrist, sternoclavicular joint, knees, ankles, proximal and distal interproximal joints of hand and feet, show diffusely intense and symmetric tracer concentration.
Correlating the medical history, review of systems, blood, urine analysis, radiographic findings and bone scintigraphy, a final diagnosis of multiple myeloma was made.

**DISCUSSION**

Plasma cell myeloma (plasmacytoma) may occur as one of three distinct clinical variants: Multiple myeloma, solitary plasmacytoma of bone and extramedullary plasmacytoma. The first well-documented case of multiple myeloma was described by Samuel Solley in 1844.4 Multiple myeloma occurs in the disseminated form, affecting several bones. It is most common in patients older than 40 years of age with a mean age of 66 years and males are more affected than females. The common systemic manifestations include bone pain and pathologic fracture, renal failure, hypercalcemia, weight loss, fatigue, weakness, shortening of the spine, fever, thrombocytopenia, neutropenia, diarrhea, orthostatic hypotension and infections. Kyle et al reported in his review of 1027 patients that bone pain, especially back pain (58%) and fatigue related to anemia (32%) were the most common symptoms.

In a review of 783 patients by Epstein et al, approximately 14% of the patients had oral presentations.4 Clinical manifestations in the jaws include swelling, pain, gingival hemorrhage, odontalgia, paresthesia, tooth mobility and migration, amyloid deposition in the tongue and other oral tissues and pathological fractures. Secondary invasion of the skeletal tissue is one of the principal characteristics of the disease. Earlier, bone manifestations were ascribed directly to tumor cell infiltration and replacement of bone substance. Although, it is now reported that ‘osteoclastic activating factor’, lymphokine, is responsible for the changes.5 Myelomatous infiltrates frequently involve the calvaria and mandible, pelvis, sternum, clavicle and proximal portions of the humerus and femur.6

The most common radiographic findings are punched-out radiolucencies without sclerotic borders. Typical punched-out lesions are radiolucent and their borders are distinct but without a definitive cortical margin. In a retrospective study of 77 patients, Witt et al stated that skull manifestations were present in 46.7% and jaw manifestations in 15.6% of the cases.7

Diagnostic criteria include end-organ damage, presence of at least 10% plasma cells on examination of the bone marrow, and monoclonal protein in serum or urine.8 A multidisciplinary approach toward the diagnosis should always be followed when multiple myeloma is suspected. Hematological examination with differential and total blood counts, biochemical assessment of renal function, calcium status, serum protein electrophoresis, quantification of immunoglobulins, immunoelectrophoresis, bone marrow biopsy and aspiration, urinalysis which includes immunofixation electrophoresis (IFE) and a radiographic skeleton survey should be obtained.

![Fig. 5: 99m Tc MDP whole body bone scintigraphy demonstrates diffusely intense, increased and inhomogeneous tracer concentration in both sternoclavicular joints and in the costochondral junction](image)
Urine electrophoresis may show M-protein in about 60% of the patients, but myeloma protein was not detected in our patient. The differential diagnosis of small, multiple, separate, well-defined radiolucencies include multiple myeloma, multiple metastatic lesions and Langerhans’ cell disease. Langerhans’ cell disease and multiple metastatic lesions are the most probable diagnoses for multiple punched-out bony lesions in children. In adults, multiple myeloma and metastatic carcinoma are likely diagnosis in cases of multiple bone involvement. In the recent years, high-dose chemotherapy with hematopoietic stem cell transplantation has become the preferred treatment for patients under the age of 65 years. The patients receive an initial course of induction chemotherapy prior to stem-cell transplantation. The most common induction regimens currently followed are thalidomide-dexamethasone, bortezomib-based regimens and lenalidomide-dexamethasone.9

Autologous stem cell transplantation after chemotherapy is the most frequent type of stem cell transplantation for multiple myeloma. Though not curative, it prolongs overall survival. Radiation therapy is useful only as a palliative treatment for painful bone lesions. Bisphosphonates reduce the risk of myeloma-related fracture, but bisphosphonate related osteonecrosis of the jaws occurs in a small percentage of the patients.10 The reported case died within a year after the diagnosis, probably because of the degree of dissemination of the disease.

CONCLUSION
Early diagnosis of multiple myeloma is essential for the better prognosis in myeloma patients. So every clinician should be aware of the maxillofacial manifestations of multiple myeloma, especially when it occurs in its primary form in the maxilla and mandible.

REFERENCES

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