Mandibular involvement as a first sign in Multiple Myeloma: a case report

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CASE REPORT

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Envolvimento mandibular como primeiro sinal clínico em mieloma múltiplo: relato de caso

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RESUMO

Objetivo: Relatar um caso de mieloma múltiplo (MM) na mandíbula com parestesia. Material e Métodos: O relato de caso descreve a ocorrência de um inchaço na região mandibular posterior em uma paciente de 70 anos e chama a atenção para o primeiro sinal clínico de uma doença sistêmica disseminada que se manifesta na região da cabeça e pescoço. Resultados: O diagnóstico presuntivo inicial foi invalidado pelo exame histopatológico e outros testes diagnósticos que confirmaram as características do MM. Conclusão: Sendo uma doença rara, o MM deve ser considerado como uma primeira alternativa de diagnóstico diferencial de lesões mandibulares em pacientes geriátricos com parestesia.

PALAVRAS-CHAVE

Mieloma múltiplo; Mandíbula; Parestesia.

INTRODUCTION

Multiple myeloma (MM) is a relatively rare malignant hematological disease accounting for 1% of all cancers and slightly over 10% of all hematologic malignancies [1]. It is characterized by the multicentric proliferation of plasma cells in the bone marrow, an excessive amount of abnormal monoclonal immunoglobulins and CRAB symptoms which includes hypercalcemia, renal insufficiency, anemia, and bone lytic lesions [2,3]. The median age of presentation of this disease is 66 years and is seldom diagnosed before the age of 40 years [4]. Clinically, the presentation of MM in the oral cavity may vary from small gingival mass to intra-osseous involvement causing facial asymmetry [5,6]. This present paper highlights jaw involvement as the first clinical sign in a 70-year-old female patient diagnosed with MM.

CASE REPORT

A 70-year-old woman presented with a chief complaint of gradually increasing swelling in left mandibular posterior region for 4 months. Extra-oral examination revealed a diffuse painless swelling in the left mandibular posterior region which was bony hard in consistency, non-fluctuant, showing paraesthesia. Regional lymphadenopathy and
Mandibular involvement as a first sign in Multiple Myeloma: a case report

Bansal S et al.

Braz Dent Sci 2018 Oct/Dec;21(4)

Evidence of any discharge was not observed. Intra-oral examination showed bony hard swelling extending from permanent left mandibular canine to permanent left mandibular third molar region with obliteration of buccal vestibule and cortical expansion. Mobility of teeth and any intra-oral draining sinus was not observed. (Figure 1A). The patient’s medical and family history was non-contributory. Oral hygiene was fair. Local factors such as plaque and calculus were minimum without any carious tooth on the affected side.

A panoramic radiograph revealed ill-defined radiolucency extending beyond left permanent mandibular second premolar region involving body of ramus and condylar region. External root resorption was seen w.r.t left permanent mandibular second and third molars. (Figure 1B). Based on the size of the lesion attained with presence of paraesthesia, provisional diagnosis of malignancy was considered. Differential diagnosis of ameloblastoma, giant cell lesion and osteomyelitis were also taken into consideration.

The histological examination of incisional biopsy under local anaesthesia revealed small, monotonous undifferentiated round cells with increased nuclear-cytoplasmic ratio and basophilic cytoplasm infiltrating the fibrovascular component suggestive of blue round cell tumour. (Figures 1C and 1D). The term round cell tumour describes a group of highly aggressive malignant tumors which includes langerhans cell histiocytosis, Ewing’s sarcoma, plasmacytoma, rhabdomyosarcoma, primitive neuroectodermal tumour, lymphoma and leukemic infiltrate. Considering patient’s age and histological presentation, provisional diagnosis of plasma cell neoplasm was made, and immunohistochemistry was performed with results showing positivity for CD138, MUM1 with dual expression of kappa and lambda and negative for Cd1a, CD99, desmin, synaptophysin, CD3, CD20. (Figures 2A, 2B, 2C and 2D). Further radiographic examinations and a skeletal survey revealed multiple scattered osteolytic lesions involving the calvarium showing characteristic rain drop skull appearance on a lateral cephalogram suggestive of plasmacytoma. (Figure 2E). Cone Beam Computed Tomography revealed a fairly expansile osteolytic lesion in the left body of the mandible with expansion and thinning of buccal cortical plate. (Figure 2F).

Figure 1 - A: Intra-oral swelling extending from distal of permanent left mandibular second premolar posteriorly to permanent left mandibular third molar region with buccal cortical plate expansion. B: Ill-defined multilocular radiolucency extending extending beyond left permanent mandibular second premolar region involving body of ramus and condylar region. C and D: Photomicrograph illustrates H and E staining in sheets of atypical plasma cells: A(x 10), B(x 40)
Mandibular involvement as a first sign in Multiple Myeloma: a case report

Bansal S et al.

Braz Dent Sci 2018 Oct/Dec;21(4)

Figure 1 - A,B,C,D) Photomicrograph illustrates positive immunohistochemistry membranous stain in plasma cells for CD138 (x 40), kappa (x 40), lambda (x 40) and nuclear staining pattern for MUM1 (x 40). E) Lateral cephalogram shows characteristic rain drop skull appearance. F) Cone Beam Computed Tomography revealed a fairly expansile osteolytic lesion in the left body of the mandible with expansion and thinning of buccal cortical plate.

A comprehensive metabolic panel including complete blood counts revealed decreased haemoglobin levels of 8.9mg/dl (Biological reference range: 12gm/dL to 15gm/dL) and raised erythrocyte sedimentation rate of 152 (Biological reference: Males 0-10; Females 0-20). Serum assays revealed normal calcium level of 8.7mg/dl (Biological reference range: 8.6 to 10mg/dL), increased serum proteins of 11.2g/dL (Biological reference range: 6.6 to 8.3 gm/dL) and serum beta-2 microglobulin of 6.75mg/dL (Biological reference range: less than 60yrs: 0.8 to 2.4; more than 60 years: less than 3.0). Serum protein electrophoresis (SPEP) revealed monoclonal immunoglobulin IgG exhibiting an abnormal dense M band with concentration of 800g/dL. Nephelometry revealed raised serum IgG levels of 7140mg/dL (Biological Reference range: 751 to 1560mg/dL) along with increased lambda light chain up to 3760mg/dL (Biological Reference Range: 313 to 723mg/dL). Immunofixation revealed monoclonal band of IgG lambda type. Surprisingly, urine analysis showed absence of Bence Jones Proteins.

Bone Marrow aspiration report exhibited hypercellular marrow with increase in interstitial sheets of plasma cells suggestive of plasma cell neoplasm that exhibited positivity for CD38, CD117, CD56, lambda with 98.8% of abnormal plasma cells. DNA hyperdiploidy with DNA index of 1.41 was found.

A diagnosis of MM ISS III was attained, and the patient was referred to cancer speciality centre where she received systemic chemotherapy with bortezomib-1mg/m2 Cyclophosphamide-200 mg/m2, Dexamethasone-20 mg on days 1,8,15,22 with every cycle in 28 days along with palliative radiation. The treatment was complicated by worsening respiratory conditions and patient expired three months later, despite treatment.

DISCUSSION

Plasma cell neoplasms are B-cell lymphoid neoplasms classified as solitary
bone plasmocytoma, multiple myeloma and extramedullary plasmacytoma. The etiology remains unknown, but exposure to certain chemicals, overdose irradiation, viruses and genetic factors have been suggested as etiologic factors [7]. Epstein et al reviewed 783 patients in literature and found oral manifestations in 14% of patients. Oral manifestations of MM are seldom the first sign of disease and only 30% of patients with MM develop osteolytic lesions in the jaw which occurs secondary to plasma cell infiltration of the jaw and are more frequent in advanced disease with extensive skeletal involvement [8-11]. Clinically, tumefaction and pain are the most common features followed by paresthesia, tooth mobility, hemorrhages and pathological fractures [12]. The present case presented with a painless swelling in the mandibular body with paraesthesia. The radiographic appearance may vary from widespread involvement to rounded, well-defined radiolucencies, with or without osteosclerotic changes [13,14]. Mandibular lesions are never an isolated radiographic finding, but only observed in patients with involvement of the skull and other bones [15]. Accordingly, the present case showed ill-defined radiolucency of the left side involving body of ramus extending to the condylar region along with the presence of multiple scattered osteolytic lesions involving the calvarium.

The diagnosis of MM requires demonstration of malignant plasma cells on histologic examination of bone marrow or a soft tissue plasmacytoma, with clonality usually established by immunohistochemistry or flow cytometry for light chain restriction [16]. The most common laboratory finding associated with MM is the presence of monoclonal protein in serum or urine [17]. In the present case, serum protein electrophoresis (SPEP) revealed monoclonal immunoglobulin IgG exhibiting abnormal dense M band. Immunophenotyping of the bone marrow revealed plasma cells positive for CD38, CD117, CD56 and lambda antibodies. Present case fulfilled diagnostic criteria of MM ISS III confirmed by clinical, radiographic, laboratory examination and immunophenotyping with presence of 98.8% of abnormal plasma cells along with monoclonal immunoglobulin IgG in serum [2].

A suspected diagnosis of multiple myeloma warrants a multidisciplinary approach including a full blood evaluation, biochemical assessment, serum protein electrophoresis, immuno electrophoresis, bone marrow biopsy along with aspiration, urine analysis which includes immunofixation electrophoresis (IFE) and a radiographic skeleton survey. Treatment involves irradiation, chemotherapy, autologous stem cell transplantation and other supportive measures [8].

CONCLUSION
Knowledge of the maxillofacial manifestations of multiple myeloma is important for early diagnosis of the disease, especially when it occurs as primary solitary lesion in the jawbones. Being a rare disease, MM should be kept on forefront of differential diagnosis for jaw lesions in a geriatric patient with paraesthesia.

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Mandibular involvement as a first sign in Multiple Myeloma: a case report

Bansal S et al.

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