Case Report

Oral medullary plasmacytoma: Rare case reports

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Abstract

Medullary plasmacytoma (MP) or osseous or solitary bone plasmacytoma is a specializing hematopathology lesion typically present with local symptoms, such as pain, paresthesia and pathologic bone fractures as a result of proliferation of plasma cells. The most often involved sites are active hematopoietic long bones and the vertebrae. The clinical course of disease is identical to spectrum of other plasma cell dyscrasias. The diagnostic criteria include punched-out radiolucencies, monoclonal plasma cells and M protein. This lesion should be considered for the differential diagnosis of bone tumors. It is highly radiosensitive although combination modalities of radiation, surgery and chemotherapy have been used in the treatment. The long-term follow-up is essential. We report two rare cases of oral MP with unusual clinical presentation.

Keywords: Calcium levels, renal insufficiency, anemia and bone lesions' criteria, immunohistochemistry, monoclonal serum protein, positron emission tomography

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INTRODUCTION

Medullary plasmacytoma (MP) is defined as a localized tumor in the bone encompassing of a single clone of plasma cells. It is an infrequent variant of plasmacytoma, comprising of only 10% of plasma cell neoplasms. The disseminated form of plasma cell neoplasms is more common than the localized form. The lesion is a rare occurrence in jaws with a 4.4% predilection in the mandible. The most common sites involved the body, angle and ramus of the mandible. The tumors have a variable biological behavior ranging from periods of latency to sudden growth spurts and rapid dissemination.[1] The male-to-female ratio is 2:1. Clinical presentation of MP is pathologic bone fractures, jaw pain, paresthesia and mobility of the nearby teeth. Most patients may have severe pain or neurologic compromise.[2] MP also has various radiographic findings from "punched-out" appearance to ill-defined destructive radiolucencies and appearance of a multiloculated lesion.[3]

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CASE REPORTS

Case 1

A 60-year-old female patient came with complaints of pain and paresthesia of lower lip for 1 month. The patient had undergone extraction of healing socket-38 one week ago. Intraoral examination revealed the presence of a healing socket-38, and no other intraoral lesions were evident. Tenderness on palpation in relation to the left lower labial vestibule was elicited. An orthopantomograph revealed a well-defined unilocular radiolucent lesion involving the

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body of the mandible [Figure 1a]. Three-dimensional computed tomography (CT) scan revealed an osteolytic lesion measuring about 3 mm × 4 mm × 2 mm in the left posterior mandible [Figure 1b]. With provisional diagnosis of aneurysmal bone cyst, an incisional biopsy revealed solid proliferation of atypical plasmacytoid cells with eccentric nuclei and basophilic cytoplasm [Figure 1c_1 and c_2]. An Immunohistochemical analysis revealed membrane immunopositivity for kappa chain [Figure 1d_1] and immunonegative with faint background staining for lambda [Figure 1d_2]. Hematological examination revealed a red blood cell (RBC) count of 4.0 × 10^6/mm^3, hemoglobin (Hb) – 11.02 g/dl, white blood cell (WBC) count – 8800/mm^3, platelets – 3.15 × 10^5/mm^3, serum protein – 6.0 g/dl, serum calcium – 4.7 mg/dl and gamma globulin – 31.6, all of which were within normal limits, and other serum electrolytes were also found to be normal. A urine test, which was done for Bence Jones protein, was positive. An electrophoresis revealed abnormal free light chain ratio [Figure 1e].

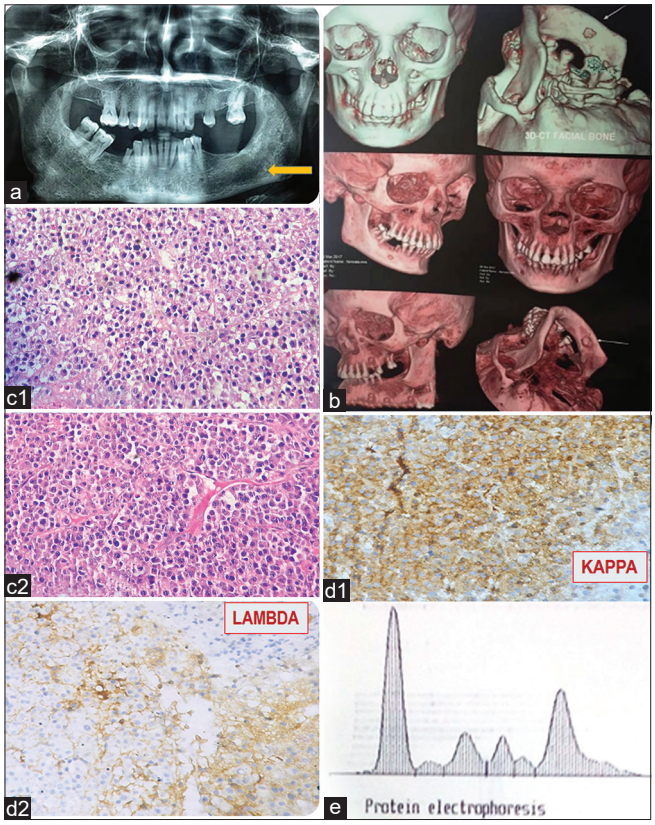

Figure 1: (a and b) Orthopantomograph shows osteolytic lesion (unilocular) in the posterior aspect of the left side of the mandible, and threedimensional computed tomography scan revealed an osteolytic lesion in the left posterior mandible. (c_1 and c_2) H&E staining ×20 shows sheets of closely packed cells resembling plasma cells and ×40 shows the presence of two nuclei within a single cell. (d_1 and d_2) immunohistochemistry staining (×40) shows tumor cells' strong Kappa positivity and negative lambda staining (×40) with faint background staining. (e) Electrophoresis shows the altered the serumfree light

Case 2

A 48-year-old male reported with complaint of a painful swelling in the left back jaw region over the past 2 months. The pain was dull and nonradiating in nature, and the swelling had gradually increased over the past 2 months. On extraoral examination, a firm mass measuring approximately 5 cm was noted, from the left preauricular region to the mandibular angle. Intraoral examination revealed a diffused mass on the retromolar pad area, covered with normal oral mucosa. Orthopantomograph revealed an ill-defined radiolucent mass in the left mandibular angle and ramus [Figure 2a]. CT scan showed a 4.8 cm × 3.7 cm × 3.7 cm destructive mass involving the body, angle and ramus of mandible [Figure 2b]. Based on the clinical and radiological findings, a provisional diagnosis of ameloblastoma or giant cell lesion was given. An incisional biopsy was done under local anesthesia. The microscopic examination showed monotonous sheets of

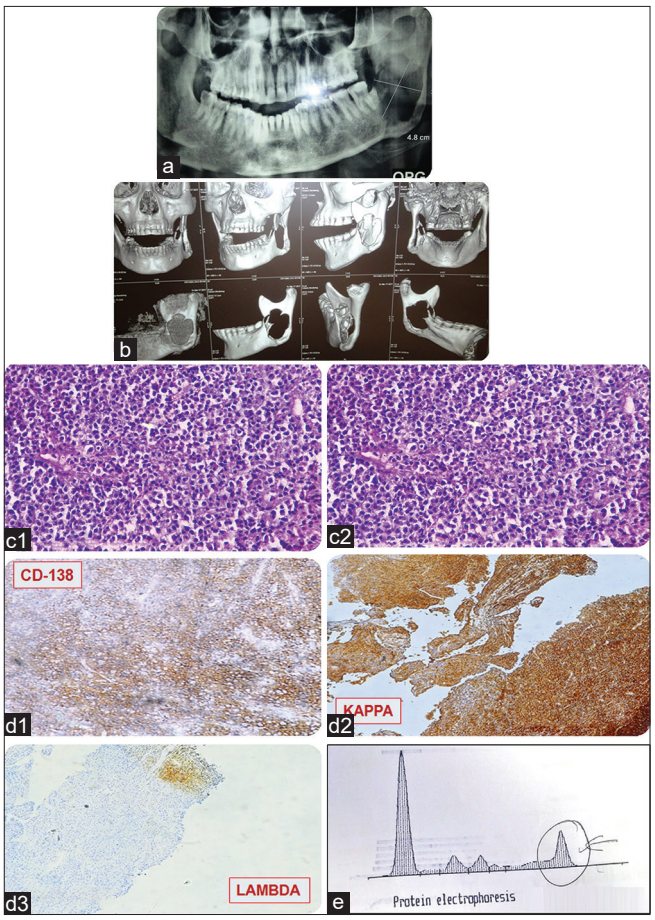

Figure 2: (a) OPG shows radiolucency involving the left ramus of the mandible. (b) Three-dimensional facial scan shows massive osteolytic lesion eroding ramus of mandible. (c_1 and c_2) H&E staining (×20) shows sheets of monotonous population of cells with eosinophilic cytoplasm and ×40 shows richly cellular areas with eccentric nucleus. (d_1-d_3) CD138, kappa chain immunostaining (×40) shows tumor cells showing strong positivity and immunonegative with faint background staining for lamda. (e) Serum protein assay shows altered the free light chain ratio

dense proliferation of neoplastic cells with highly vascular stroma and inflammatory cells. The cell population varied from small, well-differentiated cells with an eccentric nucleus and basophilic cytoplasm to less differentiated atypical cells resembling immunoblast [Figure 2c_1 and c_2]. An immunohistochemical analysis revealed positivity with Vimentin, CD45, CD138, kappa chain and negative expression for lambda chain [Figure 2d_1-d_3]. Hematological examination revealed a RBC count of 4.73 × 10^{6}/mm^3, Hb – 14.9 g/dl, WBC count – 8600/mm^3, platelets – 1.5 × 10^5/mm^3, total serum protein – 7.8 g/dl, globulin – 2.2 g/dl and serum calcium – 9.7 mg/dl and gamma globulin – 23.4, all of which were within normal limits, and other serum electrolytes were also found to be normal. A urine test performed for Bence Jones protein was negative. The free light chain ratio was altered [Figure 2e].

DISCUSSION

Plasma cell neoplasm usually affects the bone and bone marrow in the older age groups. The most common clinical presentation is bone pain, pathologic fractures, hypercalcemia and neurological symptoms. The roentgenographic findings varies from well-defined radiolucency or punched-out appearance to ill-defined destructive radiolucencies with ragged border. Clinical and radiologic variants of plasmacytoma can be identified by genetic loss and genetic gains in chromosome – 13, 1p, 14q, 19p, 9q and 1q. Many authors have considered solitary plasmacytoma as a primary manifestation of further devastating disease – multiple myeloma (MM). MP progressed to MM due to high-grade angiogenesis.[4] A few authors proposed that MP could be a middle plane between monoclonal gammopathy and MM.[5]

Microscopically, the lesions are composed of sheets or aggregates of atypical plasma cells and eccentrically placed nucleus with pink cytoplasm and perinuclear halo. Amyloid deposition may be seen in some cases. It is indeed a challenging task to the pathologists to disentangle potential differences in disease pattern of MP and MM. Some authors stated that role of chemokine receptors and CAM in the tumor microenvironment contributes to the disease presentation. A well-differentiated plasmacytoma is difficult to distinguish from reactive proliferation. Differential diagnosis of moderate and poorly differentiated plasmacytoma is metastatic melanoma and malignant lymphoma.[6,7] A complete blood picture, biochemical analysis of serum protein and marrow aspiration/biopsy are required for the diagnosis of this disease. A positron emission tomography (PET) scan or CT is required for evaluation of metastasis.[8] The diagnostic CRAB criteria are calcium levels, renal insufficiency, anemia and bone lesions. Serum electrophoresis and immunohistochemistry attributed to rule out its disseminated form.[9] The final diagnosis of the present cases were done based on radiological, histopathological findings, immunohistochemical and supportive investigative modalities.

The primary treatment for patients with solitary lesion is localized radiation therapy. Surgical therapy is recommended for structural unstable bony lesions.

Bisphosphonates is advised for patients with osteopenia. Two concurrent distinct plasmacytomas with no bone marrow involvement, radiation to both sites followed by systemic therapy is recommended. A follow-up is essential after completion of radiotherapy, complete laboratory tests every 4–6 months for 1 year and annually thereafter.[10]

CONCLUSION

MP of the oral cavity is a distinct plasma cell neoplasm that requires clinical, radiological, and histopathological and immunohistochemical correlation to arrive at the right diagnosis. New modalities such as PET, electrophoresis will have a prognostic value of disease status and the survival analyses, especially in the highrisk category of patients.

Declaration of patient consent

The authors certify that they have obtained all appropriate patient consent forms. In the form the patient(s) has/have given his/her/their consent for his/her/their images and other clinical information to be reported in the journal. The patients understand that their names and initials will not be published and due efforts will be made to conceal their identity, but anonymity cannot be guaranteed.

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Conflicts of interest

There are no conflicts of interest.

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