Solitary Bone Plasmacytoma of the maxilla- A rare case report

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Abstract

Aims and Objectives: Plasmacytoma is a discrete, uni-focal, monoclonal neoplastic proliferation of plasma cells. It can be either solitary or multiple and skeletal or extraskeletal. Solitary bone plasmacytoma accounts for 3% of all plasma cell neoplasms with 50% of cases transforming into multiple myeloma. Since plasmacytoma is a rare lesion in the oral cavity, this case presentation is an attempt to focus on the clinical, histopathological aspects of this lesion and the spectrum of plasma cell neoplasms.

Case description: We report a case of solitary bone plasmacytoma in a young patient, presenting as a painless ulcero-proliferative growth in the maxillary region. OPG showed radiolucency with respect to non-healing extracted tooth socket (16) with slight bony erosion. Microscopy revealed mature and immature plasma cells with eccentrically placed nucleus, suggestive of solitary plasmacytoma.

Conclusion: The purpose of this article is to report a rare case of solitary bone plasmacytoma with emphasis on diagnostic workup.

KEYWORDS: Plasmacytoma, plasma cell neoplasm, maxilla, myeloma

Introduction

Plasmacytoma is a lymphoid neoplastic proliferation of B cells which may occur alone in bone as solitary bone plasmacytoma (SBP), in soft tissue as extra medullary plasmacytoma (EMP) or in a multifocal disseminated form as multiple myeloma (MM)[1]. Solitary bone plasmacytoma originates as a clone of transformed malignant plasma cells in the bone marrow. It originates from the pathologic plasmablasts that dedifferentiate from primary and secondary B blasts to pathologic plasmablasts during maturation process. Cells may settle down in soft tissue or extracellular connective tissue with the assistance of adhesion molecules.[2]

50% SBP will transform into multiple myeloma and oral manifestations may be the first sign of multiple myeloma. In this regard, early diagnosis of the disease by the clinician is of utmost importance.

Case Report:

A 31-year-old male patient presented with an ulcerated swelling in the right maxillary tuberosity region of 3 months duration, following extraction of a tooth.

Intra-oral examination revealed a well defined ulcero-proliferative growth in the right maxillary posterior region in relation to 17, 15, and 14 without crossing the midline. (Fig 1) The ulcerated mass was irregular, covered with slough and was not associated with pain.

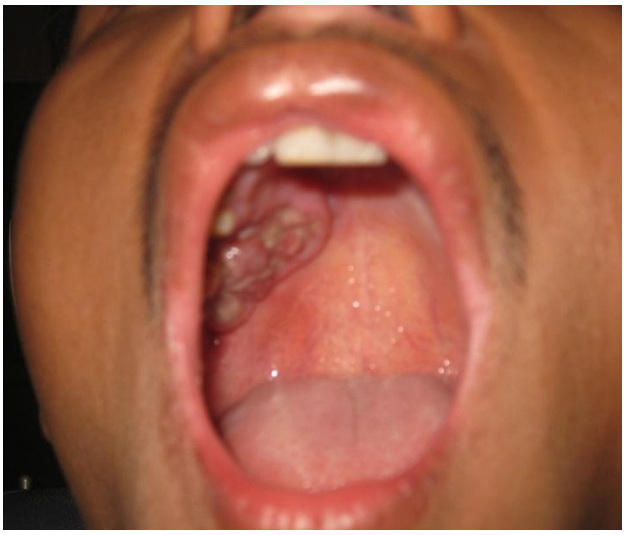

Figure 1: Intra-oral photograph showing lesion in the maxilla

OPG showed radiolucency with respect to non-healing extracted tooth socket (16) with slight bony erosion (Fig 2).

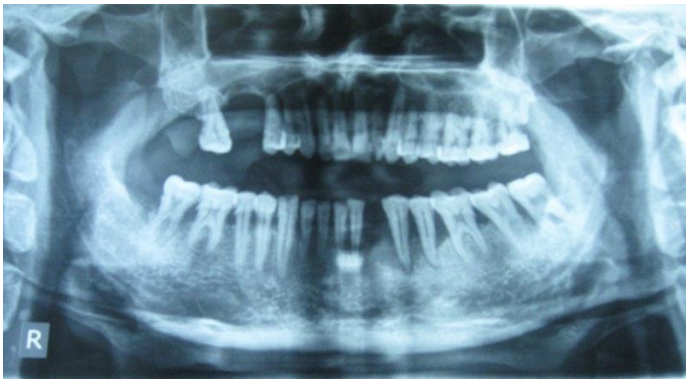

Figure 2: OPG showing radiolucency with slight bony erosion

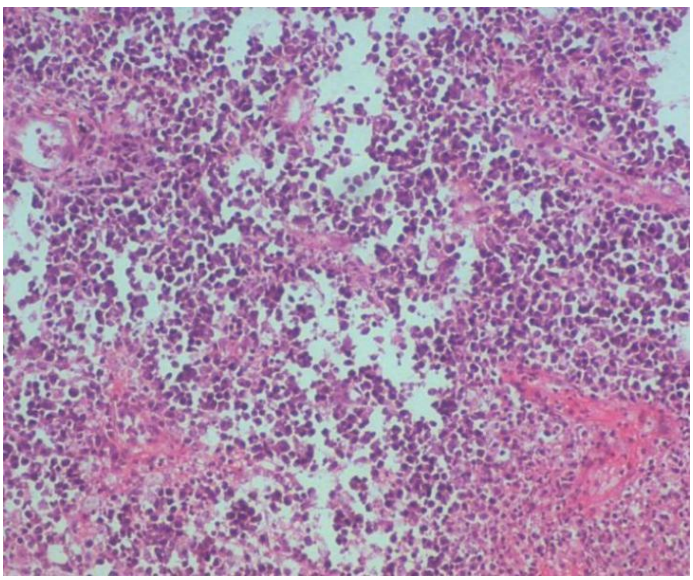

Figure 3: H&E (20x) sheets of mature and immature plasma cells

Incisional biopsy was done and histopathological examination revealed sheets of round to ovoid cells in a scanty stroma. Cells with eccentrically placed nucleus and abundant cytoplasm and few cells with centrally placed enlarged nucleus with prominent nucleoli and dispersed chromatin were present representing mature and immature plasma cells. (Fig 3) Perinuclear halo was evident. Mild cytological atypia and mitosis was noted. Focal areas showed Russell bodies. Giemsa stain was done for the assessment of cytological details to demonstrate the perinuclear halo in the cytoplasm of the plasma cells. (Fig 4) The microscopic features were in favour of Plasmacytoma.

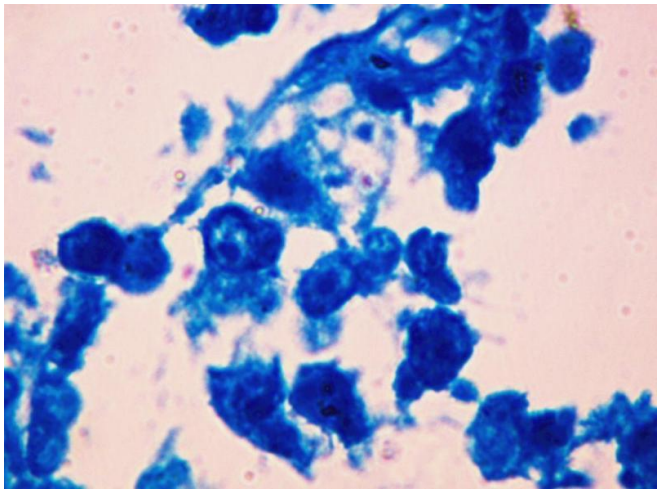

Figure 4: Giemsa stain (100x) demonstrating the perinuclear halo

Routine blood investigations were within normal limits with no evidence of hypercalcemia and urinary Bence-Jones proteins. Patient tested negative for HIV. Complete body skeletal survey showed no other bony lesions. Bone marrow biopsy was normal. Immunohistochemistry showed positivity for κ light chain. (Fig 5)

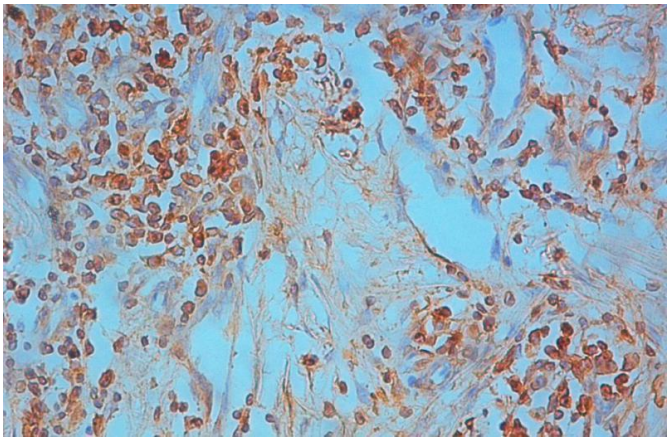

Figure 5: IHC (40x) immunopositivity for κ light chain.

Based on the morphological and immunophenotypical findings, final diagnosis of Solitary Bone Plasmacytoma of maxilla was considered.

Table 1: **Comparison of plasma cell neoplasms**

Solitary bone plasmacytoma (SBP)	Extra medullary plasmacytoma(EMP)	Multiple Myeloma (MM)
• Occurs in the bone • In the oral cavity, jaw bones are involved with predilection for ramus, angle of the mandible, maxillary sinus [2] • Osteolytic lesion	• Occurs in the soft tissue • In the oral cavity, it usually involves tongue, palate, gingiva & parotid gland[3].	• Involves multiple bones • Anaemia, thrombocytopenia, Hypercalcemia [2].

Table 2: Characteristic features of plasma cell inclusion bodies

Features	Russell bodies	Dutcher bodies
1. Synonyms	• Mott cell/ Morula cell/ Russell cell[6]	• Intranuclear Russell body
2. Site	• Cytoplasmic globules, rarely in the nucleus	• Intranuclear inclusion body
3. Nature	• Dilated ER cisternae containing condensed immunoglobulins	• Crystalline in nature, consisting of monoclonal Ig G[3]
4. Morphology	• Round	• Uniform square/ rectangular
5. Physiologic role	• It helps in the mechanism that facilitates the segregation of insoluble aggregates which might otherwise disrupt the secretory pathway.[6]	• Unknown with respect to tumor progression, clinical course & • prognosis
6. Special stain	• Red in Masson's trichrome[6] • MGG Stain • PAS	----------

Discussion:

Solitary bone plasmacytoma is an immunoproliferative monoclonal disease, accounting for 3-7% of all cases of plasmacytoma[3]. The diagnosis of the SBP depends on the biopsy evidence of plasma cell proliferation and absence of evidence of involvement of other bones. Plasma cell neoplasms mainly comprise of Solitary Bone Neoplasm (SBP), Extra Medullary Plasmacytoma (EMP) & Multiple Myeloma (MM). (Table -1)

SBP is more prevalent in men with the M: F of 2:1[1]. The oral manifestations of Plasmacytoma include jaw pain, tooth pain, parasthesia, swelling, pathologic fracture, mobility and migration of teeth [3]. Though the etiology remains uncertain, certain etiologic agents like radiation, exposure to chemicals, viruses, and genetic factors have been implicated[3]. Plasmacytoma has been reported in HIV positive patients. Cytogenetic studies in Plasmacytoma reveal loss in chromosome 13, 1p, 14q, gain in 19p, 9q, 1q and IL-6 is considered as the principal growth factor in the pathogenesis[5]. Radiologically, lesions are osteolytic with unilocular / multilocular radiolucency. Osteoclastic activation by cytokines increases the osteoclast numbers in areas invaded by malignant plasma cells, resulting in an osteolytic lesion[3.]

Microscopically, the plasma cells show varying degrees of differentiation with sparse stroma. Plasmacytomas can be graded as lesions of minimal dysplasia and severe dysplasia[1].

Nucleus can be binucleated. The spherical nuclei are set eccentrically and show regular or irregular margination of chromatin often showing a cart-wheel pattern[6]. Sometimes the chromatin may be coarsely clumped showing a clock-face pattern. The cells show paranuclear globular, pale-staining, cytoplasmic space called hof [7.] Pseudo- angiomatous areas, giant cell formation, amyloid deposition and myxoid change can be seen in some cases[8]. The plasma cells may contain intracytoplasmic (Russell body) & nuclear inclusions called Dutcher bodies (Table 2). The immature plasma cells have larger or more irregular nuclei, less condensed chromatin and occasional nuclei. The immature cells are larger and more irregular with ample slightly basophilic cytoplasm[7].

Plasmacytoma has to be differentiated from reactive inflammatory lesions & lymphomas. Monoclonal proliferation is suggestive of a neoplasm over an inflammatory lesion and Presence of nonplasmacytic neoplastic component and Ig M expression favors Lymphomas[8].

The clinical behaviour of Solitary Bone Plasmacytoma is relatively benign. The prognosis is poorer than EMP with approximately 70 % of lesions transforming into multiple myeloma.[1]

Conclusion:

Solitary bone plasmacytoma manifests itself as a single osteolytic lesion without plasmacytosis of bone marrow and constitutes approximately 3% of plasma cell neoplasms. As SBP is considered to be an early MM by some authors, it reinforces for the meticulous diagnostic work up .Radiation is the treatment of choice, as plasma cell neoplasms are highly radiosensitive.

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