

Solitary Plasmacytoma of the Mandible – A Rare Entity

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ABSTRACT

Plasma cell dyscrasias (multiple myeloma, solitary plasmacytoma of bone and extra medullary plasmacytoma) are characterized by a monoclonal neoplastic proliferation of plasma cells of which Solitary plasmacytoma of bone (SPB) is a localized form. SPB is most frequently seen in vertebrae and secondarily in long bones. Its presence in jaws is extremely rare. The malignant plasma cells express monotypic cytoplasmic immunoglobulins and plasma cell-associated antigens, with an absence of immature B-cell antigens. Here we report a unique case of plasmacytoma in the right side of mandible, a chronology for diagnosis of the lesion is also reviewed along with clinical, radiographic, histopathological and immunohistochemical evidence.

Key Words: Solitary Bone Plasmacytoma, Mandible, Multiple Myeloma.

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Background

Plasma cell neoplasms (Plasmacytomas) maybe either of a localized or a disseminated form, with both solitary plasmacytoma of bone (SPB) and extramedullary plasmacytoma (EMP) representing localized forms whereas Multiple myeloma (MM) representing the disseminated form of this disorder. Although both SPB and EMP initially are restricted to a single area, the former tends to disseminate and evolve into MM much more frequently than the latter. Thus, the two diseases are often considered as two distinct entities.

Solitary Bone Plasmacytoma (SBP) represents plasma cell dyscrasia with a single bone involvement (localized form) due to a malignant plasma cell infiltrate. It is a type of lymphoma which arises from bone –marrow based B- cells specifically those which have undergone terminal differentiation into plasma cells. These tumors are monoclonal and this can be assessed in tissue sections or biochemical evaluation of type of immunoglobulin secreted by the neoplastic cells. The incidence in males is three times that of females. The most affected age group is 50–70



Fig. 1 & 2 : Intraoral Clinical photograph

years. We here report a patient who had solitary plasmacytoma of the right mandible.

Case Report:

A 56-year-old male patient complained of loose and mobile teeth in the right- lower back region three months back, for which the patient visited a local dentist and got them extracted. However, few days post extraction patient noted a slow growing mass which subsequently reached the current size. No deformity of face was evident extraorally, but patient complained of pain and had difficulty in chewing food. Intra-orally, the right mandibular posterior region showed a soft,

proliferating growth, measuring 6cm x 5cm in dimension, extending antero-posteriorly from right lower canine to second molar region covering the edentulous ridge. The lesion was also observed extending into the buccal vestibule and impinging on to the tongue distally. The surface appeared ulcerated and showed indentations of the opposing dentition. (FIG-1, 2) . On palpation the mass felt firm and fixed. An Orthopantogram revealed a typical punched out radiolucency with ill-defined border with evidence of resorption of root of the adjacent



Fig. 3 : Orthopantogram.

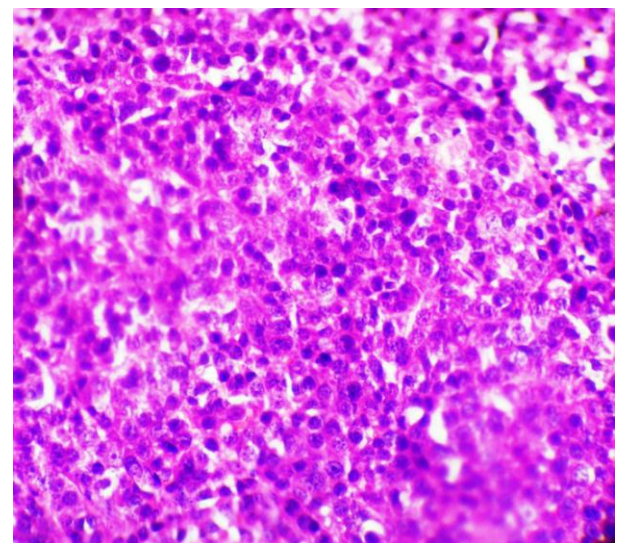


Fig. 4 : H & E picture at 40x showing plasmacytoid cells

teeth. (FIG-3).

The provisional and differential diagnosis considered then were, Lymphoma, Peripheral Neuroectodermal tumor, Ewings sarcoma, Rhabdomyosarcoma and neuroblastoma. Subsequently an incisional biopsy was done and the tissue was sent for histopathological analysis. The H and E stained slides, revealed diffuse sheets of plasmacytoid cells with minimal fibrous stroma and few small capillaries. The cells showed eccentrically placed nucleus and peripheral chromatin beadings, resembling plasma cells. Few mitotic figures were noted and pleomorphism of cells was also evident. So we inferred a diagnosis of round cell malignant tumor [fig-4]. Accordingly, we carried out a panel of immunohistochemical markers for further confirmation of the source of the neoplasm. The IHC markers used were CD 138, CD117, EMA (epithelial membrane antigen), Kappa and Lambda light chain. The IHC reports revealed Kappa light chain and CD138, CD117, EMA membrane positivity of neoplastic cells [fig-5,6,7,8] . Also, the patient was screened for complete blood and radiographic analysis. Thus a final diagnosis of "Solitary plasmacytoma of mandible" was given.

Discussion:

Solitary plasmacytoma is a localized collection of monoclonal plasma cells. The median age of occurrence is 50–70 years with a male:female ratio of 3:1. Korolkowa et al reported that 40% occur in the nasal cavity and paranasal sinus, 20% in the nasopharynx, and 18% in the oropharynx¹⁻². The most common sites of SPB are long bones and vertebrae. It rarely involves jaws and when it is seen, only 4.4% of SPB occur in the mandible, most commonly in the bone marrow-rich areas of the body, angle and ramus of mandible. Most common clinical symptoms of SPB are pain in the jaws and teeth with paraesthesia/anesthesia,

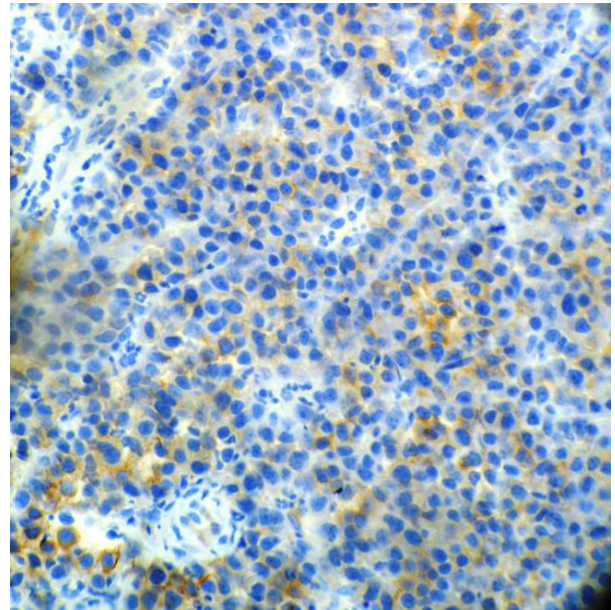


Fig. 5 : Membrane Immunopositivity Picture for EMA

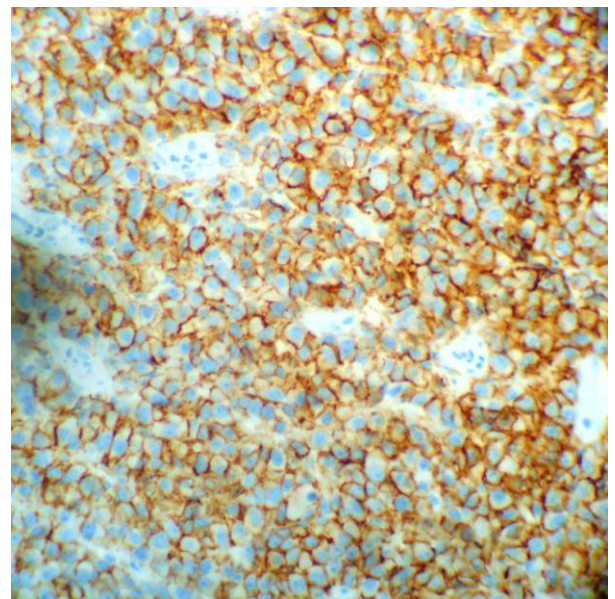


Fig. 6 : Membrane Immunopositivity Picture for CD117

mobility and migration of the teeth, hemorrhage, swelling in hard and soft tissues and pathological fractures. The clinical presentation in the present case was in harmony with literature right up to the site of occurrence.

Clinical and microscopic features may not be sufficient for distinguishing plasmacytoma from other malignancies commonly arising in the oral cavity, such as poorly differentiated carcinoma, and other types of Lymphoproliferative diseases.

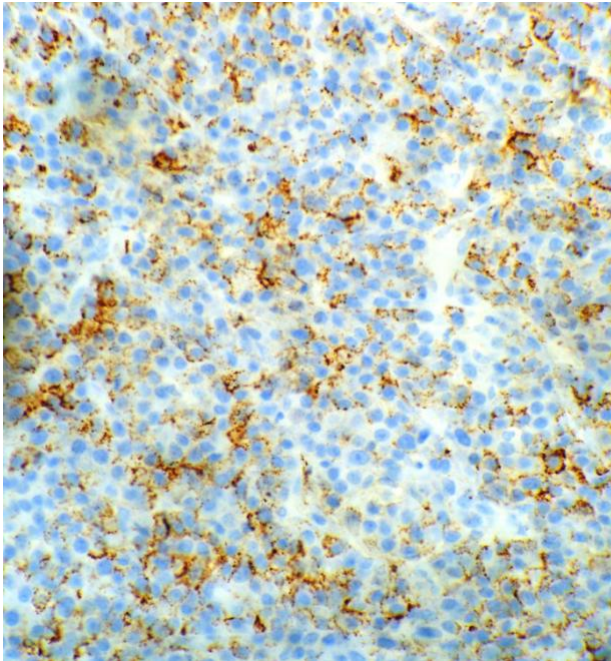


Fig. 7 : Membrane Immunopositivity Picture for CD138

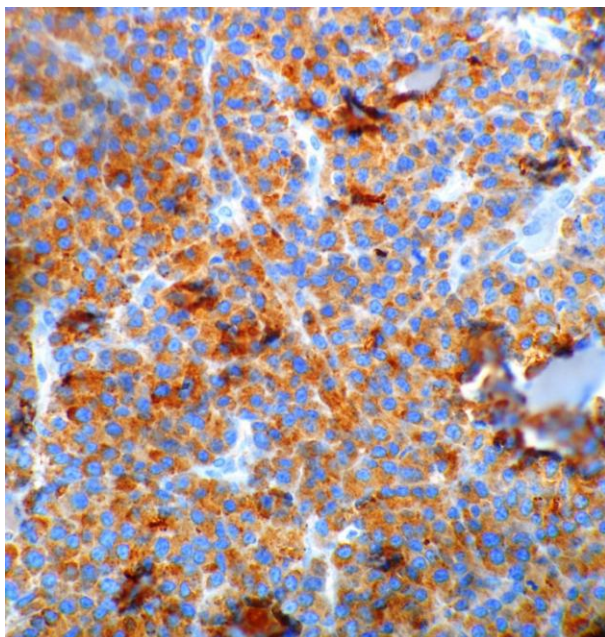


Fig. 8 : Immunopositivity picture for Kappa light chain.

The diagnosis of SBP requires a solitary bone lesion, with both confirmatory histo-pathological and immunohistochemical (IHC) analysis with a definite support of haematological investigation. Poorly differentiated carcinoma may be differentiated from SBP based on its consistent immunoreactivity for cytokeratins. Phenotypic

studies positive for CD138, CD117, EMA and monoclonal cytoplasmic light chain expression of malignant plasma cells obtained by biopsy or fine needle aspiration of the solitary lesion differentiates it from other round cell tumors³⁻⁴. Plasmacytic infiltrates are also common in various odontogenic infections, and hence need to be differentiated from (SBP). In most inflammatory conditions with characterizing high plasma cell infiltrate also exhibit other leukocytes and mainly collagenous stroma whereas in plasmacytoma the cell population is homogenous as was evident in the histopathology evaluation of the present case. Moreover, plasma cells in inflammatory conditions are polyclonal and hence will express both kappa or lambda immunoglobulin light

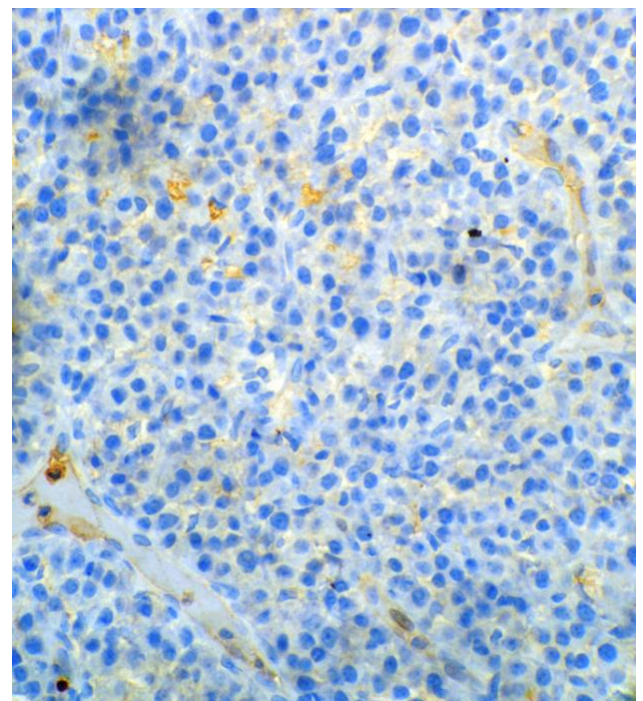


Fig. 9 : Immunonegative picture for lambda light chain

chain using immunohistochemistry, in contrast to SBP which is monoclonal which will express either kappa or lambda light chain⁵. In the present case the monoclonal IHC cytoplasmic reactivity was positive only for the kappa light chain thus distinguishing this lesion from an inflammatory condition. This monoclonal reactivity along with the other IHC panel of antigens (CD138, CD117,

and EMA) that stained the neoplastic cells positively helped differentiate the round cell tumors as plasmacytic.

Evidence from a thorough clinical and radiographic evaluation helped rule out multiple lesions, along with the confirmatory histopathology and IHC analysis helped in arriving at the definitive diagnosis of this rare Solitary Bone Plasmacytoma of the mandible. SPB requires a meticulous overview of the patient by the specialist and control of any signs or symptoms of systemic diseases, a fact that would mark a dramatic change in the treatment and prognosis of the patient. Definitive local radiotherapy is the treatment of choice as decided by the surgeon for SBP as these lesions are highly radiosensitive⁶.

Conclusion:

SPB have been characterized along with EMP as solitary variants of Plasmacytomas with former considered to be more aggressive in nature. However SPB has less credential to be characterized as a localized form as literature indicates them to having a stronger propensity to transform into MM. It appears; however, that this apparent stronger propensity is actually due to the great number of cases that are have been misdiagnosed for a occult MM. SPB requires a

meticulous overview of the patient by the specialist as distinguishing it from the other variants is critical for treatment and survival of the patient.

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