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Unusual Presentation of Solitary Plasmacytoma of Mandible with Review of Literature

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I. Introduction

Plasma cell myeloma is a monoclonal neoplastic proliferation of plasma cells of bone-marrow derivation, usually multicentric, that eventually infiltrates various organs but rarely produces plasma cell leukaemia[1]. It is characterized by osteolytic lesions, bone pain, hypercalcemia, a monoclonal gammopathy, and disorders due to depositon of abnormal immunoglobulin chains (amyloid) in various tissues, including kidney[1]. Plasmacytoma can present clinically as multiple myeloma (MM), solitary bone neoplasm (SBP) and extramedullary plasmacytoma (EMP). Localized plasmacytoma are less common than multiple myeloma and can occur as an extra osseous form and solitary bone neoplasm[2,3,4].The biological behaviour of these tumors is variable, from periods of clinical latency to rapid growth and progression from the localized forms (SBP and EMP) to MM. EMP has a better prognosis because 30% progress to MM compared with 70% in SBP[3,7]. Many authors have considered SBP as an early manifestation of MM. Solitary bone plasmacytoma is an immunoproliferative monoclonal disease, accounting for 3% of all plasma cell neoplasms[2]. The disease is more frequently seen in men, with a mean age of occurrence of 55 years[22] . Here we report a case of solitary plasmacytoma of mandible in a 59 year old female patient and reviewed the literature.

II. Case Report

A 59 year old female patient reported to the Department of oral pathology, Government dental college, kozhikode with the primary complaint of a painless swelling on right posterior mandible of 1 year duration. The swelling had gradually increased over the past 2 months. Extraorally, a firm non tender swelling on right posterior region of mandible measured 6cm x5cm which extended superiorly upto the Zygomatic arch inferiorly to the lower border of mandible, anteriorly 2cm from the commissure and posteriorly to the tragus (Fig1). Intraoral examination did not reveal any obvious features except an edentulous maxillary and resorbed mandibular ridge. No regional lymphadenopathy were noted.

Fig1: Extraoral photograph

Radiographic features

PAN view showed a multilocular lytic lesion involving the right ramus of mandible extending to condyle (Fig2). CT showed an expansile lytic lesion with central sclerosis, cortical discontinuity and scalloping involving the ramus of mandible on the right side (Fig3).Based on the clinical and radiographic features a provisional diagnosis of odontogenic tumour/cyst was considered.

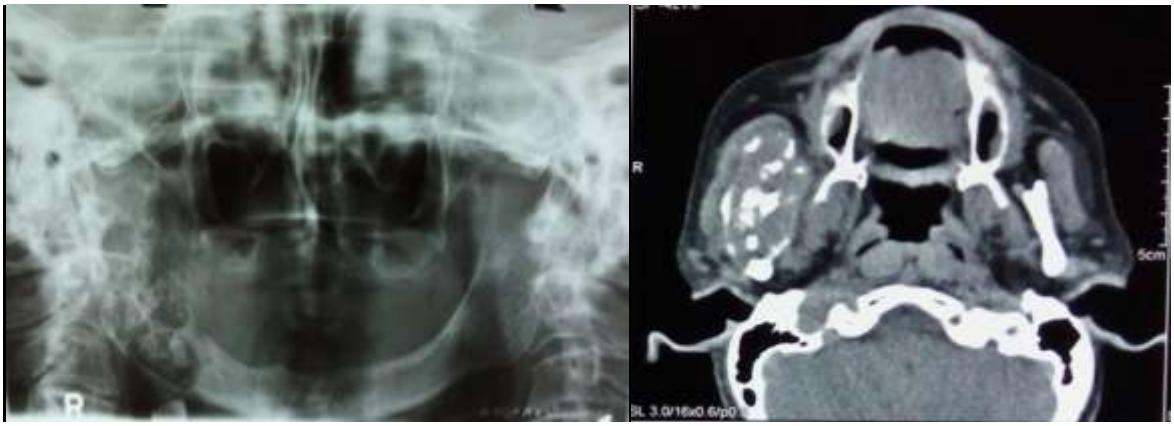

Fig2: Orthopantomogram showing multilocular lytic lesion on right posterior mandible
Fig3: CT shows expansile lytic lesion on right side of mandible

Routine hematological investigations were within normal limits and incision biopsy was performed. H & E stained sections showed sheets of round to ovoid cells with eccentrically placed nucleus and pale eosinophilic cytoplasm (Fig5) resembling plasma cells with occasional nuclear pleomorphism and prominent nucleoli. The intervening stroma showed pale homogenous eosinophilic material .

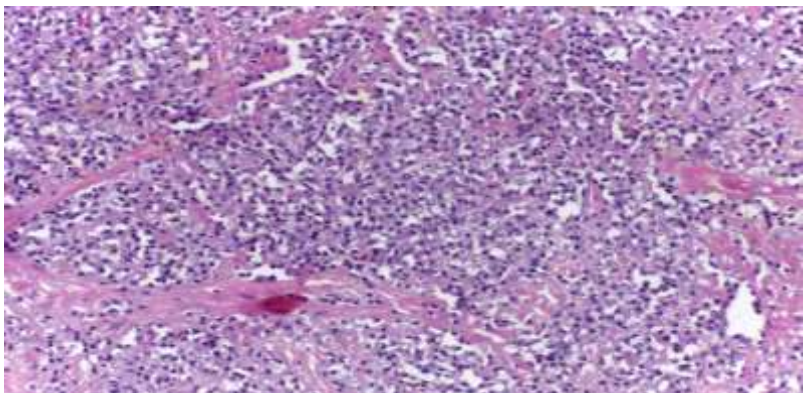

Fig4: H & E picture showing plasmacytoid cells (10x)

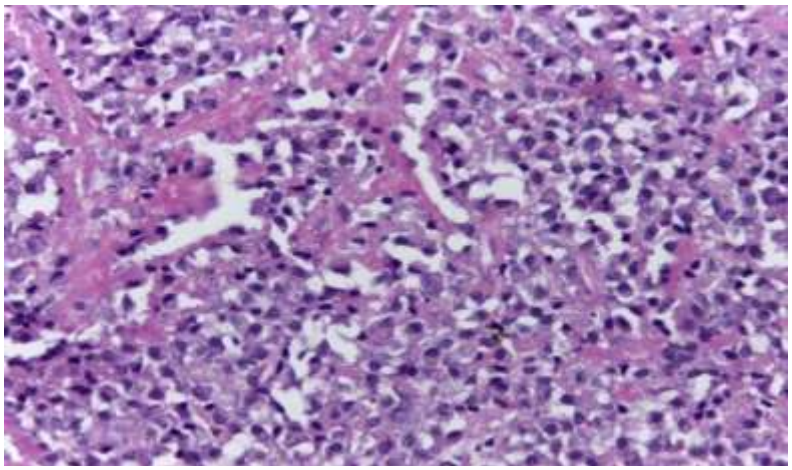

Fig5: H & E picture showing atypical plasma cells with eccentrically placed nucleus (40x)

Based on the histologic findings diagnosis of reactive plasmacytosis was made. To confirm the source of the neoplasm immunohistochemistry was done. The IHC markers used were CD 138, Kappa and Lambda light chain and reports revealed lambda light chain and CD138, membrane positivity of neoplastic cells (Fig6,8).

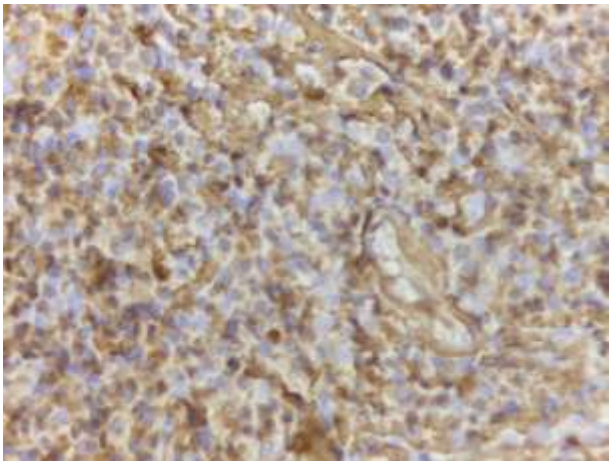

Fig6: Tumor cells were positive for CD138 which is a reliable marker for identifying and quantifying normal and tumoural plasma cells in paraffin sections.

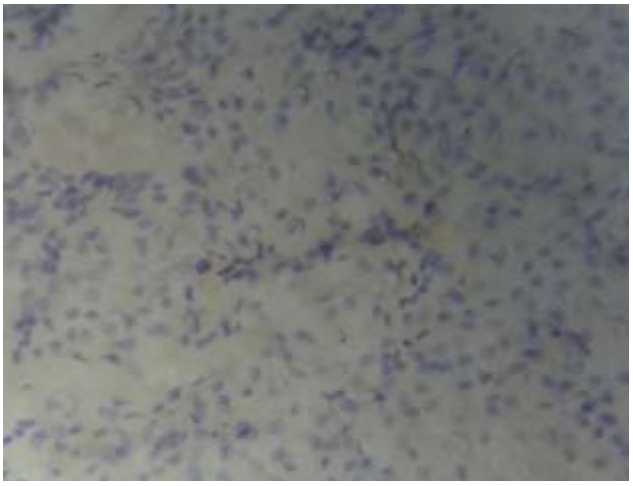

Fig7: Immunonegative picture for Kappa light chain

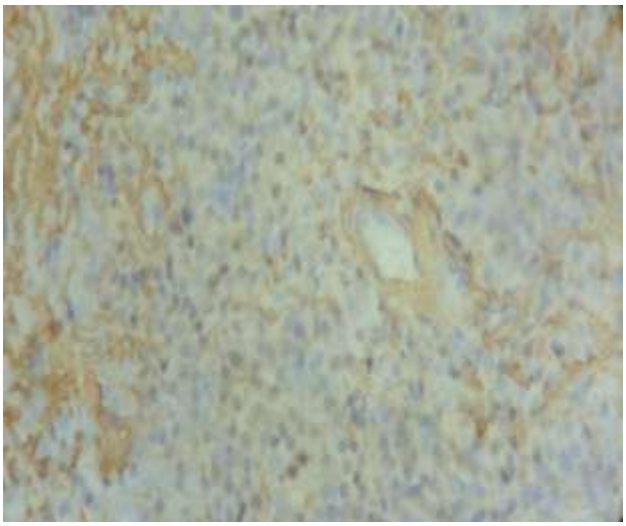

Fig8: Immunopositivity picture for Lamda light chain.

Biochemical investigations for Serum Calcium, phosphorous, urea, uric acid, etc were within normal reference range. Skeletal analysis was done to rule out multiple myeloma but lytic lesions were not found in any other sites. Serum electrophoresis revealed low levels of gammaglobulin. Based on clinical, histopathologic, radiographic, immunohistochemical and biochemical findings confirmatory diagnosis of solitary plasmacytoma was made. The patient underwent radiotherapy and was followed up on a monthly basis for 6 months and no further lesions were observed at the 1 year follow-up examination.

III. Discussion

Plasmacytoma is a rare solitary mass of neoplastic monoclonal plasma cells, first described by schridde in 1905[6]. The following variants of plasma cell myeloma have been described non-secretory myeloma, indolent myeloma , smoldering myeloma, plasma–cell leukaemia (PCL), in addition to extramedullary plasmacytoma, and solitary plasmacytoma of bone[1]. The exact distinction is based on clinical and radiographic features[1]. The median age of occurrence of solitary plasmacytoma is 50–70 years with a male: female ratio of 3:1[10]. Canger et al[7] noted slight male predilection with 2:1 male to female ratio. . The most common sites of SPB are long bones and vertebrae[9]. Korolkowa et al reported that 40% occur in the nasal cavity and paranasal sinus, 20% in the nasopharynx, and 18% in the oropharynx[8,9]. 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[7] Pisano et al[3] noted that SBP most frequently occurs at the posterior mandible, which is consistent with our case.

Clinical signs and symptoms of solitary plasmacytoma includes localized pain and paresthesia, but bone-impaired functions are also frequently reported[7]. The most frequent clinical symptoms of SBP are referred pain in the jaws and teeth. Swelling, soft tissue masses, and pathologic fractures are less commonly seen. No spontaneous jaw pain was observed in the present case. The tumors are found as unilocular or multilocular radiolucent destructive lesions on radiological analysis[21]. Plasma cells produce osteoclasts activating factor which promotes growth of osteoclasts leading to bone resorption[21]

Clinical and microscopic features may not be sufficient to distinguish plasmacytoma from other malignancies that commonly arise in the oral cavity, such as poorly differentiated carcinoma, and other types of Lymphoproliferative diseases[10]. The diagnosis of SBP requires a solitary bone lesion, with both confirmatory histopathological and immunohistochemical (IHC) findings with a definite support of haematological investigation[10].

Criteria for identifying solitary bone plasmacytoma vary among authors. However, the current criteria to perform a diagnosis of solitary plasmacytoma are the following:
Isolated area of bone destruction due to clonal plasma cells, bone marrow plasma cell infiltration not exceeding 5% of all nucleated cells, absence of further osteolytic bone lesions or other tissue involvement (ie, no evidence of systemic plasmacytoma), absence of anemia, hypercalcemia, or renal impairment attributable to myeloma, low concentrations of serum or urine monoclonal protein (ie, myeloma protein), or preserved levels of immunoglobulins[13.]

Differential diagnosis considered owing to the destruction and extensive tissue mass were Lymphomas, Peripheral Neuroectodermal tumours, Ewings sarcomas, Rhadomyosarcomas, Histocytomas and Neruoblastomas. Phenotypic studies positive for CD138, 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 tumors[10]. Plasmacytic infiltrates are also common in various 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 histopathologic evaluation of the present case. Moreover, plasma cells in inflammatory conditions are polyclonal and hence will express both kappa or lambda immunoglobulin light chain using immunohistochemistry, in contrast to SBP which is monoclonal which will express either kappa or lambda light chain. In the present case the monoclonal IHC cytoplasmic reactivity was positive only for the

lamda light chain thus distinguishing this lesion from an inflammatory condition (Fig7,8). This monoclonal reactivity along with CD138 positivity along with the criteria helped to arrive at a confirmatory diagnosis of solitary plasmacytoma.

The characteristic numeric abnormalities seen in plasma cell neoplasms are losses in chromosome 13, 13q14, 8, 14, and X, and gains of chromosome 3, 5, 7, 9, 11, 15, 19, and 21[23].Translocations involving the immunoglobulin heavy chain (IgH) locus occur at high frequency (50%-70% of cases). In 40% of these cases, the translocation partner is one of five loci: 11q13 (bcl-1/cyclin D1 locus), 6p21 (cyclin D3), 4p16 (fibroblast growth factor receptor 3 and multiple myeloma SET domain), 16q23 (c-MAF), and 20q11 (MAFB)[23].

Systemic alterations such as hypercalcemia, impaired renal function, anemia, bone lesions, leukopenia, thrombocytopenia, and proliferation of monoclonal plasma cells in the bone marrow are frequently observed in MM patients[11]. Daghighi et al noted that bone damage may also be responsible for alteration of blood calcium levels, especially in MM cases compared with SBP. Therefore, complete blood cell count (CBC), calcium, phosphorous, C reactive protein (CRP), beta 2-microglobulin analysis, and skeletal bone surveys are highly recommended . On the other hand, SBP is a rare disease that constitutes approximately 3% - 10% of all plasma cell neoplasms and clinical signs and symptoms of SBP may be less common that differs from MM. Therefore an early diagnosis of SBP may be challenging to perform.[12]

The primary treatment of SBP is radiation therapy, radical extensive surgery, or both combined[15,16] Radical radiotherapy is the treatment of choice at doses of 40-50 Gy, showing an index of local disease control of 80%. There is no evidence of relationship for a 40-45 Gy dose response and tumor size[14]. Surgical treatment is elective in those selected cases where all tumour is removed with minimal cosmetic or functional deficit or those to prevent or stabilize a pathologic mandibular fracture[20]. The partial resection, with or without adjuvant radiotherapy, has not been shown to decrease survival or local control compared to radiotherapy treatment alone[20]. The role of adjuvant chemotherapy is at present not clearly defined. The addition of chemotherapy to radiotherapy in the treatment of SBP has not been shown to decrease local recurrence or increase survival rates compared to local treatment with radiotherapy alone, therefore, should be reserved for those cases progressing to multiple myeloma[17,18].

Currently, there is considerable interest in the role of angiogenesis inhibitors, thalidomide, protease inhibitors or inhibitors of vascular endothelium growth factor in plasma cell neoplasms, which could, in a future, be an alternative treatment[19]. Between 35 and 85% of SBP progress to MM arising in a period of few months to years after diagnosis, in fact, some authors consider the SBP an early stage of the disseminated disease. It is not possible to predict which case may transform although there are some risk factors like age (> 60 years), M component levels > 20 g / L up to one year following radiotherapy, large tumour neovascularization and tumor size> 5 cm. So after treatment, SPB cases must be closely followed up [20].

The median survival rate of a solitary plasmacytoma is longer than in multiple myeloma because of the absence of diffused bone marrow alterations, kidney damage, hypercalcemia, and related bone fractures[5]. However, full clinical remission is unusual. The prognosis of solitary plasmacytoma could be worse if recurrence is present as in cases of evolution toward systemic disease (multiple myeloma)[5]. Similarly, the evolution of solitary plasmacytoma of the jaw is usually better if there are no recurrences after treatment (local surgery, local irradiation, systemic chemotherapy, or a combination) or an evolution toward systemic plasmacytoma[5]. According to a long-term outcome analysis reported in the literature, radiation, chemotherapy, surgery, or a combined radiation and surgical approach gave good outcomes with a low rate of local recurrence[5]. Nearly 80% of patients did not show local recurrence after five years while 20% developed multiple myeloma[5].

IV. Conclusion

Solitary bone plasmacytoma is a localized form of plasma cell tumours but rare involvement in maxillofacial area. This entity requires a meticulous overview of the patient by the specialist and overall 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. Treatment includes radiation therapy, radical extensive surgery, or both combined. Chemotherapy should be reserved for those cases progressing to multiple myeloma. The course of SPB is relatively benign and 10 year survival rate is 50-80%. The prognosis is worse if recurrence is present as in cases of evolution toward systemic diseases.

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