CASE REPORT

Solitary Medullary Plasmacytoma of Mandible-A Case Report

Amit K. Roy[1], Jayanta Chattopadhyay[2], Soumi Ghanta[3], Suman Agrawal[4]

ABSTRACT

Introduction: Plasmacytoma is monoclonal neoplastic proliferation of plasma cells. It can be either solitary or multiple and skeletal or extraskeletal. Solitary plasmacytoma is most frequently seen in vertebrae and secondarily in long bones. Its presence in jaws is extremely rare.
Case Report: Here we report a case of plasmacytoma in the right side of mandible, a chronology for diagnosis of the lesion is also reviewed along with clinical, radiographic and histopathological evidence. Orthopantomograph showed ill-defined, radiolucent lesion in the body, angle and ramus region without sclerotic border. 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.

Key words: Dutcher bodies, Multiple myeloma, Mandible, Solitary bone plasmacytoma,

How to cite this article: Amit K. Roy, Jayanta Chattopadhyay, Soumi Ghanta, Suman Agrawal. Solitary Medullary Plasmacytoma of Mandible-A Case Report. International Journal of Contemporary Medical Research 2015;2(3): 518-521

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Source of Support: Nil

Conflict of Interest: None

INTRODUCTION

Plasmacytoma is a lymphoid neoplastic proliferation of B cells. It may occur alone in bone as solitary bone plasmacytoma (SBP) or in soft tissue as extra medullary plasmacytoma (EMP). The multifocal disseminated form of plasmacytoma is known as multiple myeloma (MM).[1,2]

Solitary Bone Plasmacytoma (SBP) arises from B-cells which have undergone terminal differentiation into plasma cells with a single bone involvement due to infiltration of those malignant plasma cells.[2] SBP will transform into multiple myeloma upto 50% cases. Oral manifestations may be the first sign of plasmacytoma. For this reason, early diagnosis of the disease by the dentists is very important.[1]

The incidence in males is three times that of females. The most affected age group is 50–70 years.[2] Here we report a case of SBP of a female patient with destructive radiographic and anaplastic cytological findings.

CASE REPORT

A 48 year female reported with a complaint of swelling in the right mandibular posterior region since 1 year. The swelling was static in size but a sudden increase in the size of swelling was noticed since two months. The patient was undergone extraction of teeth in that region 4 months back which were mobile at that time. The patient's medical history was non-contributory and physical examination revealed no other abnormality.

Extra oral examination revealed presence of swelling measuring 2.5cmx2.0cmx2.0cm at the right angle of the mandible (figure-1). The swelling was bony hard in consistency, non - tender on palpation and was not attached to the overlying skin. The right submandibular lymphnodes were palpable measuring approximately 1 to 1.5 cm in diameter,firm in consistency, non tender and mobile. Intraoral examination revealed partial edentulousness in right mandibular posterior region. Intraoral examination also revealed presence of firm swelling measuring 3cmx1.5cm extending from right mandibular premolar region to retromolar region (figure-2). Overlying mucosa was normal in colour. No signs of discharge were present. On palpation, vestibular obliteration was present in that region. The swelling was tender and both buccal and lingual cortical expansion was present.

Orthopantomograph revealed extensive solitary, ill-defined, radiolucent lesion in the body, angle and ramus region without sclerotic border (figure-3). Very thin radiopaque sptae are present in the radiolucent area. Root resorption was not present adjacent to radiolucent area.The skull and pelvis radiographic

survey failed to identify additional osseous lesions (figure-4). Haematological investigation revealed markedly increased leukocytic count and subnormal haemoglobin levels. The serum calcium levels were slightly deranged but alkaline phosphatase levels were within normal limits. The patient was tested for Hepatitis B and HIV I and II which were negative. Laboratory analysis of the patient's blood and urine revealed no monoclonal gammopathy or Bence Jones protein.

Biopsy was done and histopathological section showed dense cellular infiltrate, with sheets of malignant plasma cells, which are seen as diffuse,monotonous and variably differentiated (figur e-5 and figure-6).

The differential diagnosis was plasma cell tumorand rhabdomyosarcoma. The malignant cells were immunopositive for kappa light chain and CD38.

Based on the clinical presentation, radiological findings and histopathology the diagnosis of solitary plasmacytosiswas suggested. Bone marrow aspiration and biopsy revealed normocellular marrow with no evidence of involvement. Surgical excision of the lesion was done. After that, the patient was referred to an oncologist and underwent local radiotherapy.

DISCUSSION

Plasma-cell tumors represent a groupof related disorders each of which is associated with a proliferation and accumulation of Ig-secreting cells derived from B lymphocytes.[3,4]

Solitarybone plasmacytoma (SBP) is a solitary tumor resulting of an uncontrolled proliferation of monoclonal plasma cells.[5]

Multiple myeloma (MM) represents the disseminated form of this disorder and is by far the most common plasma-cell tumor. Plasmacytomas,on the other hand, less than 10% of patients present with a solitary plasmacytoma that may be either extramedullary plasmacytomas (EMPs) or osseous/medullary/bone plasmacytomas (SBPs), each withdifferent characteristics and clinical behaviour patterns.[1,3,5-7]

SBPs are most often found in the axial skeleton, usually the vertebrae and skull. The mandible is more frequently involved than the maxilla, most commonly in the bone marrow-rich areas of the body, angle and ramus of mandible. The oral manifestations of bone Plasmacytoma include jaw pain, tooth pain, parasthesia, swelling with or without local erosion, involvement of surrounding mucosa or tissue, pathologic fracture, mobility and migration of teeth because of localized monoclonal proliferation of plasma cells.[1,2,4-8] The clinical presentation in the present case was in harmony with literature right up to the site of occurrence.

Histologically confirmed single lesion or isolated area of bone destruction due to clonal plasma cells, bone marrow plasma cell infiltration not exceeding 5% of all nucleated cells , no evidence of systemic plasmacytoma or negative skeletal survey, absence of anemia, hypercalcemia or renal impairment attributable to myeloma, low concentrations of serum or urine monoclonal myeloma protein are taken into consideration for the diagnostic criteria for solitary plasmacytoma.[1,4,8,9]

Though the etiology remains uncertain, certain etiologic agents like radiation, exposure to chemicals, viruses and genetic factors have been implicated. Plasmacytoma has been reported in HIV positive patients. Cytogenetic studies reveal loss in chromosome13, 1p, 14q, gain in 19p, 9q, 1q and IL-6 is consideredas the principal growth factor in the pathogenesis.[1] Clonal plasma cells involvement frequently produces a monoclonal immunoglobulin as well as κ or λ light chains. A quantitative assay of plasma monoclonal immunoglubulin may be perfo-rmed and may also reflect tumor growth. In cases of light chain production, immunoelectrophoresis may reveal clonal activity in the patient's serum and urine.[8]

Radiologically, lesions are osteolytic with unilocular /multilocular radiolucency without periosteal reaction or as a protruding mass with cortical expansion. Osteoclastic activation bycytokines increases the osteoclast numbers in areas invaded by malignant plasma cells, resulting in an osteolytic lesion.[1,7] Plasma cells produce osteoclast-activating factors that promote the growth of osteoclasts and stimulate the bone resorption. Therefore, SBP usually appears on radiographic images as radiolucent areas without any reactive bone formation.[4] In our case, radiolucent lesion with thin radiopaque septae was present.

CT, MRI and more recently PET are useful to establish the characteristics of the lesion and tissues involved and to rule out other affected areas.[5,7]

Laboratory signs of solitary plasmacytoma are usually related to the immunoglobulin production which inclu des monoclonal gammopathy in serum electrophoresis, light chains production detectable in serum and/or urine, cryoglobulinaemia. Plasmacytoma is also related to blood calcium levels, kidney dysfunction and serum β-2-microglobulin levels.[4]

Microscopically, the plasma cells show varying degrees of differentiation with sparse stroma. These cells may present as mature plasma cells of normal spectrum or different degrees of maturity, from undifferentiated cells similar to lymphoid precursors (plasmacytoma) to intermediate forms between lymphocytes and plasma cells. The immature plasma cells

have larger or more irregular nuclei, less condensed chromatin and occasional nuclei. There are three different groups of differentiation, which are related to the rate of survival: 1) low dysplasia, with less than 10% of plasmoblastos 2) moderate dysplasia, with 10-50% of plasmoblastos, and 3) severe dysplasia with 50% of immature forms and worse prognosis than the others. The spherical nuclei are set eccentrically and show regular or irregular margination of chromatin often showing a cart-wheel pattern. Sometimes the chromatin may be coarsely clumped showing a clock-face pattern.[1,3,7-10] The cells show paranuclear globular, pale-staining, cytoplasmic space called hof. The plasma cells may contain intracytoplasmic acidophilic inclusions known as "Russell bodies" and nuclear inclusions called Dutcher bodies. Plasmacytoma has to be differentiated from reactive inflammatory lesions and lymphomas. Monoclonal proliferation is suggestive of a neoplasm over an inflammatory lesion and presence of nonplasmacytic neoplastic component and Ig M expression favours Lymphomas.[1,4,7,8]

Plasmacytomasmay be confused with benign reactive plasmacytosis, undifferentiated carcinoma, non Hodgkin's lymphoma, malignant melanoma, or esthesioneuroblastoma and as such may present a diagnostic and therapeutic challenge to the head and neck surgeon.[3]

Other round cells tumor can be excluded by phenotypic studies positive for CD138, CD117, EMA and monoclonal cytoplasmic light chain expression of malignant plasma cells obtained by biopsy or fine needle aspiration from the lesion. 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. 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.[2,3]

The primary treatment of SBP is radiation therapy, radical extensive surgery or both. The optimal treatment is moderate-doseradiotherapy (RT), approximately 40 to 50 Gyadministered once daily at 1.8 to 2.0 Gy perfraction in a continuous course. It is not necessary to irradiate the entire medullary cavity of the bone in patients with SBP. Rather, the lesion may be treated with a more limited field with a 2- to 5-cm margin on the gross disease.[5,7]Aggressive subtotal resection of an incompletely resectable tumor followed by postoperative RT offers no improvement in the likelihood of long-term local control and should be avoided.[7] 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

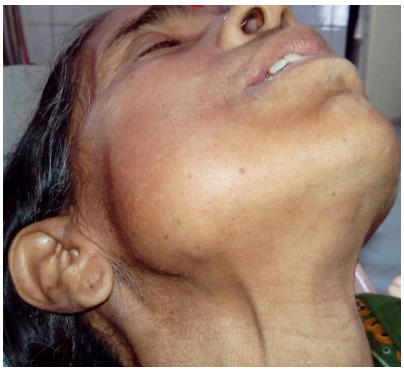

Figure-1: Extraoral view

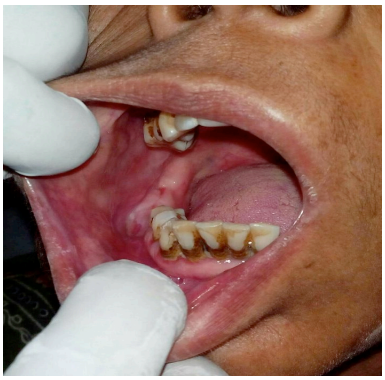

Figure-2: Intraoral view

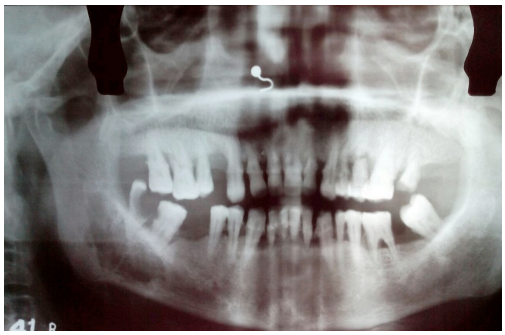

Figure-3: Orthopantomograph

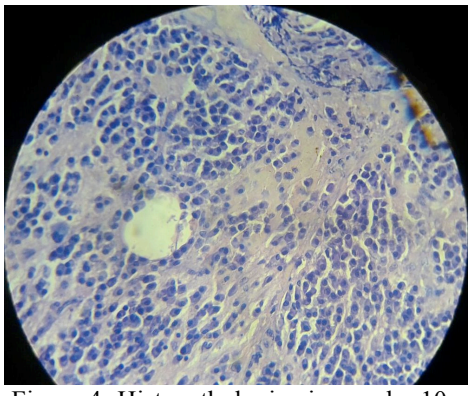

Figure:4- Histopathologic view under 10x

with radiotherapy alone, therefore, should be reserved for those cases progressing to multiple myeloma. 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 be an alternative treatment in future.[7]

The solitary plasmacytoma may develop a local recurrence, or recur in the regional nodes, or progress to multiple myeloma in case of treatment failure. The most common pattern of relapse is progression to MM.It is not possible to predict which case may transform although there are some risk factors: age (> 60 years), M component levels > 20 g / L up to one year following radiotherapy, large tumour neovascularization and tumor size> 5 cm. Therefore regular clinical follow up with monitoring of immunoglobulins and monoclonal proteins in serum and Bence-Jones proteins in urine is advised.[7]

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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