Case Report

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Mandibular Plasmacytoma of Jaw – A Case Report

Dentistry Section

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ABSTRACT

The plasma cell neoplasm may present as Extramedullary Plasmacytoma (EMP) in soft tissues in bone as a Solitary Plasmacytoma of bone (SPB) or as a part of multi focal disseminated disease Multiple Myeloma (MM). The majority of 80% occurs in head and neck region. In our case, a 62-year-old male patient presented with a non tender swelling of short duration. The swelling was noted obliterating the vestibular depth in right lower mandibular region. The radiological features were non specific.

Keywords: Mandible, Plasmacytoma, Radiotherapy, Solitary

CASE REPORT

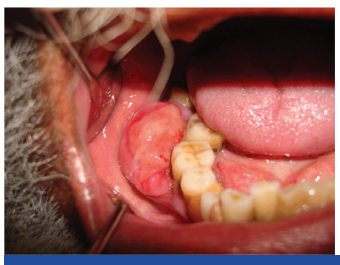
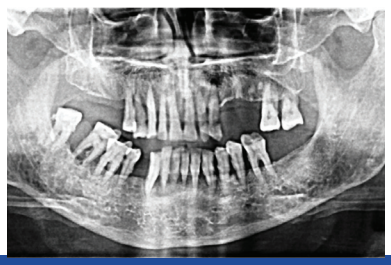
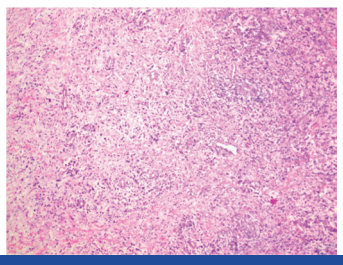
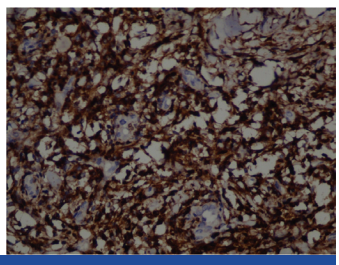
A 60-year-old male reported the OPD of Oral Medicine & Radiology of Gian Sagar Dental College & Hospital, Ram Nagar, Banur with a growth in the lower right back tooth region for the last 3 to 4 months which subsequently increased to the present size of 3x4 cms. Patient complained of dull ache and mobility of teeth in the affected region. On intraoral examination a well defined reddish pink pedunculated growth extending from 45 to 48 tooth region was seen [Table/Fig-1] which was obliterating the vestibular depth. The growth was firm in consistency and was attached to the underlying structures. The surface of the lesion was smooth with white ulcerated layer close to the occlusal region. Radiological examination revealed generalized bone loss more in relation to 45, 46, 47 and 48 [Table/Fig-2] .Routine hematological and biochemical investigations (S.Calcium, phosphorous, urea, uric acid, etc) were within normal reference range. Viral markers performed were negative. Bence Jones proteins were negative. Biopsy of the growth was performed and was sent for histopathological examination. Microscopic examination revealed dense cellular infiltrate with sheets of malignant plasma cells [Table/Fig-3]. These cells were monotonous and variably differentiated. These cells were large with round to oval with eccentrically placed vesicular nuclei. Russell bodies are also noted at places. Immunohistochemistry confirmed CD 138+ Bone plasmacytoma [Table/Fig-4].

DISCUSSION

Neoplastic proliferation of plasma cells was discussed by Dalrymple and Bence Jones in 1846 [1]. These are described as a group of clinical disorders characterized by uncontrolled clonal proliferation of plasma cells [2]. Plasma cell neoplasm's can be studied under three main groups : as Medullary Plasmacytomas ,as Extramedullary Plasmacytomas and as a component of Multiple Myeloma. Plasmacytomas are localized proliferations of plasma cells giving rise to a localized tumour mass which can be classified into two groups depending upon the site where they appear:

- Solitary plasmacytoma with single bone involvement (SPB)
- Extra medullary plasmacytoma (EMP) with soft tissue involvement [3].

These localized proliferations of monoclonal plasma cells can also occur as a component of multifocal disseminated disease Multiple Myeloma which represents the most important and common plasma cell dyscrasia [4]. In multiple myeloma neoplastic proliferation of highly specialized B-lymphocytes produces a single immunoglobulin. So, it is characterized by the disseminated monoclonal overproduction of immunoglobulin's in blood and/or by detecting Bence Jones proteins in urine [2]. Nearly 80-85% patients suffering from medullary plasmacytomas can have progression to multiple myeloma while extramedullary plasmcytomas usually remain localized to the site of occurance [1]. Common age of presentation of plasmacytomas is in 50-80 y [5]. Clinically these can be present with bone pain, localized swelling of the area involved or with pressure symptoms on the surrounding structures. Solitary bone plasmacytomas usually appear in the bones of vertebrae, ribs, pelvis and pectoral girdle while extramedullary plasmacytomas usually appear on the mucosal surfaces e.g. nasal sinuses, oropharynx, larynx etc. Histologically these tumours are comprised of well differentiated plasma cells with small eccentric nuclei and coarse chromatin which is condensed at the periphery. The tumours can also have immature plasma cells showing finely dispersed nuclear chromatin in eccentric nuclei having prominent nucleoli and abundant cytoplasm[3]. The incidence of plasma cell neoplasm's is about 2.6 to 3.3/lac population and higher incidence are seen in people of African- American origin [2]. Solitary bone plasmacytomas accounts for about 3-10% of plasma

[Table/Fig-1]: Pedunculated growth extending from 45 to 48 tooth region **[Table/Fig-2]:** OPG reveals generalised bone loss **[Table/Fig-3]:** Dense cellular infiltrate with sheets of malignant plasma cells **[Table/Fig-4]:** IHC immunopositivity to CD 138

cell neoplasms [2]. Mandible /Jaw is a rare site in head and neck region for plasmacytomas to occur [5].

Plasmacytomas occur due to clonal proliferation of plasma cells which are cytologically and immunophenotypically similar to myeloma cells. Presentation of these diseases show different groups of patients in relation to location, tumour progression and overall survival rate; while these patients have many similar biologic features of other plasma cell disorders [6]. Plasma cells produce osteoclasts activating factor which promotes growth of osteoclasts leading to bone resorption. So on radiographic study these pasmacytoma's appear as multilocular radiolucency without any reactive bone formation [5].

Skeletal plasmacytomas are characterized clinically by radiolytic lesion involving any part of skeleton, a clonal plasma cell infiltrate in the absence of disseminated bone marrow involvement [6]. Criteria for identifying solitary bone plasmacytomas vary among authors .While current criteria for diagnosis of solitary plasmacytoma is:

- 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 lesion or other tissue involvement (i.e. no evidence of systemic disease)
- No anaemia
- No hypercalcaemia or No renal involvement [7].

Solitary plasmacytoma of bone is a separate entity of plasma cell neoplasm. It constitutes approximately 3-10% of all the plasma cell tumours [2]. It may be located in bone or the extramedullary soft tissue in bone. Extramedullary plasmacytomas accounts for less than 3% of all plasma cell malignancies. About 80% to 90% of extramedullary plasmacytoma's occur in head and neck region. Out of these nearly 20% progress to multiple myeloma. Only 4.4% of solitary plasmacytoma's localize to the mandible/jaw particularly in the bone marrow rich areas of body, angle and ramus of mandible [4,5].

Plasma cell tumours usually occur in the age group of 50-80 y [5]. Patients diagnosed at age less than 60 y had significantly better 5 y survival (90% for patients aged 0-29 yrs & 80% for patients 30-59 yrs.) when compared to patients diagnosed at age more than 60 y (5 yr survival 45%). Patients with skeletal plasmacytoma had an overall survival of 57% at 5yrs.and 37% at 10y [6].

The Provisional and Differential diagnosis considered then owing to the destruction and extensive tissue mass were Lymphomas, Peripheral Neuroectodermal tumours, Ewings sarcomas, Rhadomyosarcomas, Histocytomas and Neruoblastomas. However Histocytomas and Sarcomas arise typically in younger patients and there was no known primary lesion suggestive of the malignant lesion. But the possibility of the aggressive appearance could not be excluded because of the aggressive appearance of the lesion.

Plasmacytomas are clinical entities where it is very difficult to confirm the diagnosis without radiological, histopathological, immunohistochemical and other supportive investigative modalities. In our case along with radiological, histopathological findings, support of Immunohistochemistry (IHC) positivity for CD138 was also done for establishing the final diagnosis.

Solitary plasmacytomas are highly radiosensitive lesions. Radiation therapy, radical extensive surgery or a combination of both is recommended as primary treatment. Radical radiotherapy comprising of 40-50Gy has shown 80% of local disease control. Surgical treatment is recommended to those cases where the whole tumour is to be removed to minimise cosmetic or functional deficit or in cases where pathological fracture is anticipated (to prevent that fracture and stabilize the fracture mandible). Research is being going on for the role of angiogenesis inhibitors, thalidomide, protease inhibitors or inhibitors of vascular endothelial growth factors in plasma cell neoplasm's as alternate mode of treatment [2].

CONCLUSION

Solitary bone plasmacytomas rarely occur in maxillofacial areas affecting mandible. In this case the patient was referred to cancer institute for complete treatment which includes radiation therapy, radical extensive surgery or a combination of both. Chemotherapy should be reserved for those cases progressing to multiple myeloma.

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