Case Report

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EXTRAMEDULLARY PLASMACYTOMA: AN UNUSUAL FINDING OF ORAL CAVITY, MIMICKING AN IRRITATIONAL FIBROMA

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ABSTRACT

Extramedullary plasmactyoma is a solitary soft tissue growth, resembling plasma cell neoplasm which truly lacks the features of multiple myeloma. It is difficult to diagnose it in a routine practice setting due its clinical presentation in the oral cavity. A 65-year-old female reported to our OPD with growth in the mandibular gingiva mimicking an irritational fibroma. Later the diagnosis was confirmed to be solitary extramedulary plasmacytoma after ruling out multiple myeloma as differential diagnosis.

KEYWORDS: Plasmacytoma; oral cavity; irritational fibroma

INTRODUCTION

Plasmacytoma or Multiple Myeloma is a neoplasm of bone that originates from cells of the bone marrow, bearing a remarkable resemblance to plasma cells. Plasma cells reside in the red pulp of the spleen, medulla of the lymph nodes, tonsils, lamina propria of the entiregastrointestinal tract, mucosa of the nose and upper airway, and sites of inflammation. They are termed as the terminally-differentiated B lymphocytes which produce immunoglobulins or antibodies.[1] Dalrymple and Henry Bence-Jones, in 1846, characterized it by marked proteinuria and bone pain and were the first ones to describe these neoplastic proliferation of plasma cells.[2] It is majorly described as a neoplastic proliferation of plasma cells in soft tissue.[3] 3% of all plasma cell tumors account to extramedullary plasmacytomas and 90% of them are found in the head and neck region commonly affecting the nasal cavity, paranasal sinuses, tonsillar fossa, and oral cavity.[4,5]

CASE REPORT

A 65 year old female patient presented with a complain of a growth in her lower jaw anterior teeth region since 4 months. She described noticing a small growth in the same region which gradually increased to the present size (Fig. 1) Earlier medical history was suggestive of a swelling on the left side of her neck a month back which was provisionally diagnosed as plasmacytoma on fine needle aspiration cytology report by a physician. Bone marrow aspiration showed 3% of plasma cells (Fig. 2). On intraoral inspection we found a solitary growth, firm in consistency, non-tender, not fixed to the underlying bone, approximately measuring about 2cm in diameter. It extended from the right mandibular central inscisor till the right mandibular 1st premolar approximately with the lateral missing on that side. The surface of the lesion toward the lingual facings of the teeth could be separated and teeth indentaions were present over it. Skin over the swelling appeared to be normal. Generalised attrition was observed along with periodontitis in the oral cavity. To rule out possibilities of multiple myeloma other investigations were carried out. None of the radiographs (skull, chest, sacral, lumbar and hip bone) showed any bony changes. Values of other investigations hematological & urine test were in normal range except for raised ESR. The provisional diagnosis was that of an irritational fibroma and differential diagnosis included pyogenic granuloma, peripheral giant cell granuloma, peripheral ossifying fibroma, soft tissue plasmacytoma and multiple myeloma. After biopsy (Fig. 3), the histopathological section showed sheets of plasma cells with eccentric nuclei and inflammatory cells indicating soft tissue plasmacytoma (Fig. 4 & Fig. 5). The final diagnosis was extramedullary or soft tissue

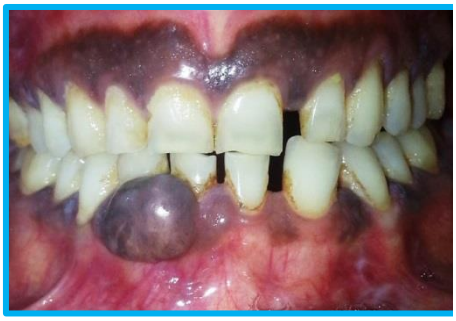

Fig. 1: Clinical photograph of the growing mass

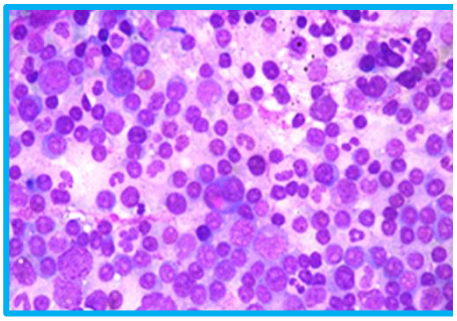

Fig. 2: Mature and immature plasma cells from bone marrow aspiration stained with leishman's

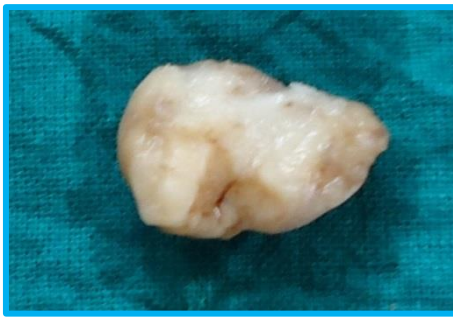

Fig. 3: Excised biopsy specimen

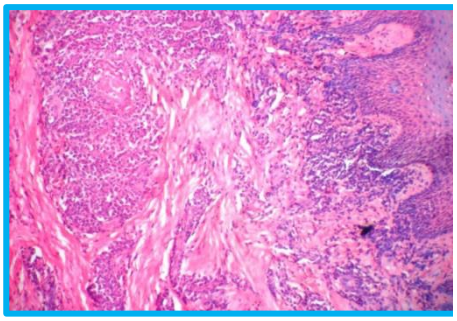

Fig. 4: Sheets of plasma cells with eccentric nuclei and inflammatory cells along with oral epithelium under low power view

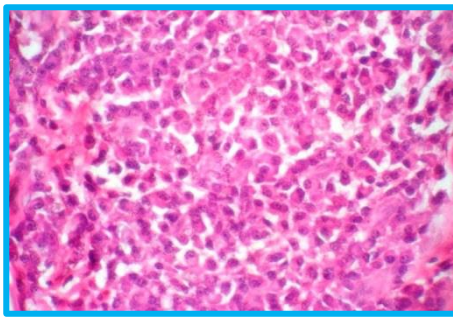

Fig. 5: Plasma cells with eccentric nuclei under high power view

plasmacytoma.

DISCUSSION

Plasma cells which are derived from B-cells, produce immunoglobulins (Igs) which contain heavy and light chains. Plasma cell neoplasms are divided into four groups multiple myeloma (MM), plasma cell leukemias, solitary plasmacytomas of the bone (SPB), extramedullary plasmacytomas (EMP).[6] Multiple myeloma (MM) is described by a clonal proliferation of neoplastic plasma cells(myeloma cells) in the bone marrow. It is associated with multifocal lytic lesions throughout the skeletal system.[1,7] Proliferation of myeloma cells is supported by IL-6, produced by fibroblasts and macrophages in the bone marrow stroma. Many myelomas have chromosomal translocations involving the IgG locus on chromosome 14. One fusion partner is the fibroblast growth factor 3 receptor gene on chromosome 4, which appears to be truncated so as to produce a constitutively active receptor. 60% of patients, M component is IgG; in 20-25% IgA; rarely, it is IgM, IgD, or IgE. Plasma cells produce only *k* or lamda chains, in about 15-20% of cases which, because of low molecular weight, are readily excreated in urine, where they are termed Bence Jones Proteins.[8] Extramedullary plasmacytomas are rare malignancies typically found in males of about 60 yrs age group.[9,10] These growth primary appears in the nasal, pharyngeal, or oral mucous membranes as sessile, grayish-red to dark-red swellings, that tend to be somewhat lobulated as they increase in size and they very seldom ulcerate. Till date the etiology of the extramedullary plasmacytoma is still unknown. Some of the commonly proposed risk factors include chronic antigenic stimulation such as osteomyelitis, cholecystitis, rheumatoid arthritis and bacterial flora. Inmunoperoxidase stains show mono or biclonality for light chains. Usually these lesions are solitary, and the routine investigations like the blood count, urine findings, and roentgen-ray examination of the bones negative.

Wiltshaw (1976) classified soft tissue plasmacytoma into 3 clinical stages:[6]

- Stage I - Limited to an extramedullary site
- Stage II - Involvement of regional lymph nodes
- Stage III - Multiple metastasis (although it is no longer a solitary plasmacytoma)

Criteria for diagnosis of EMP include:

- Biopsy proven plasma cell tumor,
- Absence of osteolytic bone lesions or other tissue involvement,
- Bone marrow plasma cell infiltration not exceeding 5% of all nucleated cells,
- Low serum electrophoresis M-protein concentration if present, and no hypercalcemia or renal involvement due to plasma cell dyscrasias

TREATMENT & PROGNOSIS

The accepted treatment for EMPs because of high radiosensitivity, has been radiotherapy. However, complete surgical resection have equal results to radiation alone. There is no published evidence that adjuvant chemotherapy is beneficial, although there may be a role in selected cases such as with large or high-grade tumors. Of all of the plasma cell tumors, EMPs have the best prognosis. Conversion to MM is less likely than in patients with SPB. While there are reports of EMP patients developing SPB, local recurrence or new solitary tumors at distant sites are seldom observed.[9,10]

CONCLUSION

Long term follow up in plasmacytomas is necessary to check disease progression. This includes bone marrow aspirations, skeletal survey with CT or MRI modalities, and serum and urine protein electrophoresis to detect the presence of M proteins. If MM is diagnosed, systemic treatment with chemotherapy is recommended using various alkylating agents in combination with glucocorticoids (2-3 years of survival rate). Other treatment protocols that are presently being considered for MM include, bone marrow transplantation, interferon, monoclonal antibodies, thalidomide and its analogs, and protease inhibitors.

CONFLICT OF INTEREST & SOURCE OF FUNDING

The author declares that there is no source of funding and there is no conflict of interest among all authors.

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