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Case Report

The diagnostic surprise of solitary anaplastic myeloma of mandible: a rare case report

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ABSTRACT

Myeloma predominantly affects the elderly who develop a monoclonal protein which is found in their blood and/or urine manifesting as anemia, osteolytic lesions, hypercalcemia, bone pains and renal compromise. Osteolytic lesions are seen in more than 30% of patients with multiple myeloma. However, primary affection of mandible manifest as solitary plasmacytoma of bone occurs in only 4.4% such cases making it a highly unaccustomed entity. The aim of this case report is to present the clinico-pathological profile and the immunohistochemical features of anaplastic myeloma that will be of immense utility in diagnosing its affection of oral cavity and mandible precisely.

Keywords: Plasma cell neoplasm, Multiple myeloma, Plasmacytoma, Mandible, Jaw swelling

INTRODUCTION

Myeloma, a B-cell lymphoid neoplasia, accounting for 1% of all malignancies, has a reported annual incidence of around 0.7/100000 fresh patients in the India which translates 6800 new cases annually.[1] Its global prevalence varies between 2.6-3.3/100,000 population. Its initial mention is credited to Una et al and its first description was given by Schridde et al.[2]

Anaplastic myeloma is an extremely unaccustomed entity. In such scenario of its rarity, available literature tending to be in the form of isolated case reports, few case series and a high potential of this tumor getting misdiagnosed, every case needs to be reported. This will not only create awareness about this rare entity amongst otorhinolaryngologists, dental surgeons, pathologists but also facilitate a valid statistical analysis of this rare disease.

Authors report a rare case of a 72 years old male who presented to us with a non-healing ulcer associated with an ipsilateral mandibular swelling. Our case report highlights paramount noteworthiness of the rare entity getting diagnosed by the clinician early thereby ensuring timely institution of correct management protocol and a possible/ hypothetical enhancement of its prognosis.

CASE REPORT

72 years old male presented with complaints of a non-healing ulcer in the right half oral cavity of 2 months duration. It was associated with localized pain and dysphagia. Patient denied any constitutional symptoms. No history of past radiation exposure. Patient was a reformed smoker since last five years after he developed bronchial asthma. Prior to reformation patient had a history of smoking approximately 20 beedis per day for a decade.

After quitting smoking, he shifted to chewable tobacco which he continued till he developed the ulcer in the oral cavity. Patient also gave history of a tooth extraction from (R) lower jaw (46,47) a month back. Upon enquiry, patient mentioned that he had developed a right mandibular swelling which was evaluated by a dentist

and with the presumption of an infected tooth to be its cause, extraction was performed. However, when the mandibular swelling did not resolve after extraction and antibiotics, patient was referred to a tertiary care center. Local examination revealed poor oral hygiene with halitosis, nicotine-stained teeth. An ulcero-proliferative, non-friable, tender lesion 5×5 cm was present at right lower alveolus extending anteriorly till right lower canine, posteriorly till retromolar trigone (RMT) (Figure 1a).

The lesion bled on touch. Base of tongue was soft and supple. Neck palpation revealed a solitary approximately 4×3 cm swelling in right submandibular region which was hard and fixed to underlying tissues (Figure 1b). The overlying skin was pinchable. No other neck swellings/ cervical lymphadenopathy. Rest otorhinolaryngological (ORL) examination including an office fibreoptic videolaryngoscopy was normal. With the clinical impression of a neoplastic growth of oral cavity (subsite-right alveolus) patient underwent a punch biopsy under local anesthesia from the growth which was reported as granulation tissue.

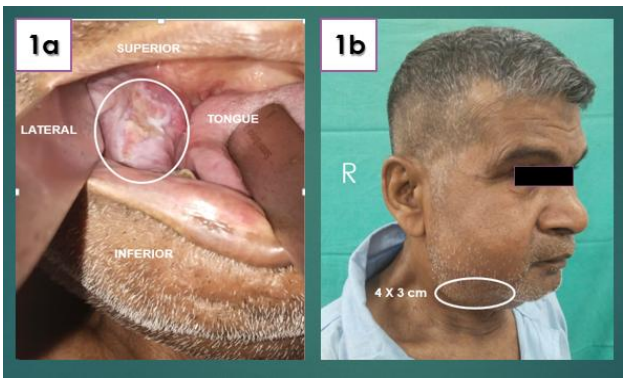

Figure 1 (a): Oral cavity examination showing an ulcero-proliferative, non-friable, tender lesion approximately 5×5 cm at right lower alveolus extending anteriorly till right lower canine; posteriorly till right RMT (white circle; (b): gross photograph showing a 4×3 cm swelling (hard consistency, fixed, tender) present in right submental region extending medially till midline and inferiorly till 5 cm above medial end of right clavicle, posteriorly till 1 cm short of angle of mandible right (white oval).

Contrast-enhanced computed tomogram (CECT) from skull base till diaphragm revealed a heterogeneously enhancing soft tissue density lesion (5.9×4.5×5.9 cm) with internal non-enhancing areas suggestive of necrosis seen on either side of mandibular ramus on right side with cortical expansion and permeative type of bone destruction (Figure 2a). It caused downward displacement of right submandibular gland with areas of infiltration. There was loss of fat planes with lateral pterygoid muscle superiorly. Multiple discrete sub-centimetric cervical lymph nodes (LNs) were seen in bilateral Ia, Tb, II and III stations; largest being 0.6×1.2 cm in right level IIa.

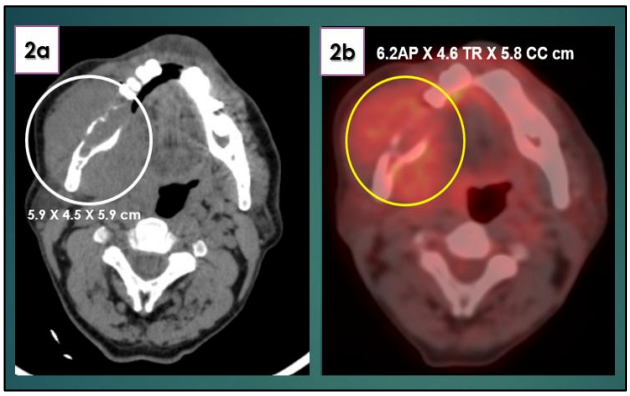

Figure 2 (a): CECT from skull base till diaphragm - heterogeneously enhancing soft tissue density lesion (5.9×4.5 ×5.9 cm) with internal non-enhancing areas suggestive of necrosis on either side of mandibular ramus on right side with cortical expansion and permeative type of bone destruction (white circle); (b): PET scan shows a metabolically active, lobulated soft tissue density mass lesion (6.2×4.6×5.8 cm, SUV max-8.2) in oral cavity on either side of right hemi-mandible causing cortical erosion. Anteriorly, the lesion infiltrated right submandibular gland; superiorly reached till hard palate without its obvious cortical erosion or involvement (yellow circle).

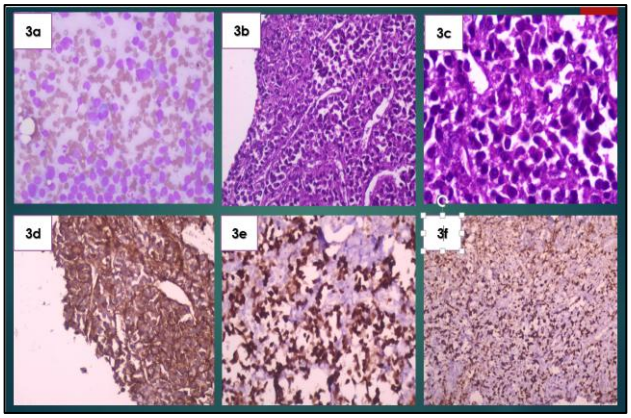

Figure 3 (a): Microphotograph (Leishman and Giemsa, 200 X) fine needle aspirate showing scattered atypical plasma cells and binucleate plasma in a background of RBCs; (b): microphotograph (Hematoxylin and Eosin, 200 X) showing sheets of atypical plasma along with scattered blood vessels; (c): Microphotograph (Hematoxylin and Eosin, 400 X) showing atypical plasma cells with increased N:C ratio perinuclear hof and amphophilic cytoplasm. Some of the atypical plasma cells appear binucleate; (d): microphotograph (200X) showing IHC positivity for CD 138. (e): Microphotograph (200X) showing IHC positivity for MUM1 and (f): microphotograph (200X) showing IHC demonstrating a Ki-67 proliferative index of 40-50%.

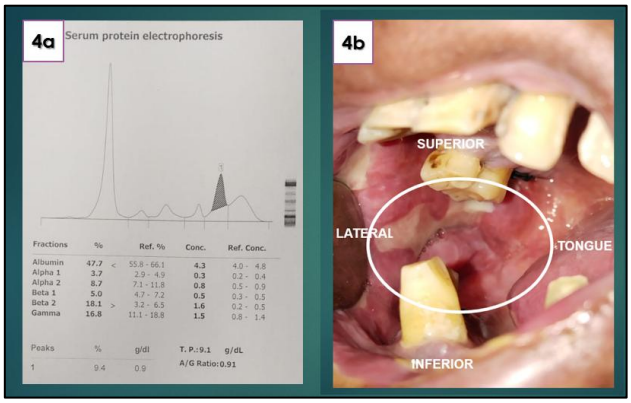

Figure 4 (a): 'M' spike seen in region of β microglobulin suggestive of monoclonal gammopathy (shaded region) and (b): post radiotherapy status.

Fine needle aspiration cytology (FNAC) from the lesion, submandibular swelling and sub-mandibular LN was done next which were consistent with hemato-lymphoid malignancy (non-Hodgkin's lymphoma). Positron emission tomography (PET) scan showed a metabolically active, lobulated soft tissue density mass lesion (6.2×4.6×5.8 cm, SUV max-8.2) in oral cavity on either side of right hemi-mandible causing cortical erosion.

Anteriorly, the lesion infiltrated right submandibular gland; superiorly reached till hard palate without its obvious cortical erosion or involvement (Figure 2b). USG guided trucut biopsy was taken from right submandibular LN that showed fragmented cores of plasmacytoid tumor cells arranged in sheets (Figure 3a- c). Immunohist-ochemistry was positive for cluster differentiation CD 138, vimentin and multiple myeloma oncogene 1 (MUM1) with Ki-67 proliferative index of 40-50% which was reported as plasma cell neoplasm of anaplastic morphology (Anaplastic myeloma) (Figure 3d-f). Haematologist's review was also sought to rule out disseminated form of the disease. Serum protein electrophoresis showed 'M' spike in region of β microglobulin suggestive of monoclonal gammopathy (Figure 4a). Markers for multiple myeloma (immunofixation and serum free light chain assay) were negative. Bone marrow biopsy that demonstrated 2-3% plasma cells. Complete blood counts, serum calcium and renal parameters were normal but hypoalbuminemia (Serum albumin- 3.2 gm%) was noticed. Urine protein to creatinine ratio was also normal.

The diagnosis was now revised to solitary plasmacytoma of mandible and radiation-oncologist's referral was taken. Patient received definitive radiotherapy (50Gy/25 fractions) over 5 weeks without significant side effects. Patient responded well to treatment and showed complete response. The mucosal component of the lesion completely resolved but bone destruction that had manifested continued be there without any major symptoms (Figure 4b). Presently patient has been on follow up for a year without any symptoms or evident recurrence.

DISCUSSION

Plasma cell myelomas (PCMs) are characterized by monoclonal, neoplastic plasma cell proliferation resulting in increased production of monoclonal antibodies. Clinically, the lesion may be single (solitary plasmacytoma) or multiple in its disseminated form affecting several bones (multiple myeloma, MM).[3] Solitary plasmacytoma is the localized form which is found to affect either bone or soft tissue (extramedullary plasmacytoma), have less than 10% plasma cells in the bone marrow and don't manifest systemic symptoms of MM.[4] Ordinarily, the bones afflicted by MM in the decreasing fashion are vertebrae, ribs, skull, mandible, clavicles, scapula and pelvis. Involvement of the mandible is infrequent and primary affection is even rarer. Solitary bone plasmacytomas account for 3% of PCMs with as high as 50% conversion rates into multiple myeloma within months or years after initial diagnosis.[2]

Solitary plasmacytoma of bone (SPB) occurs in slightly younger age group than multiple myeloma with average age at diagnosis of 55 years (6th decade of life) and is twice commoner in males.[5] SPB is uncommon in head and neck region with only 4.4% cases occurring in bone marrow-rich parts of mandible-retromolar area, angle and ramus.[6] Our reported case was at extreme of age (8th decade of life) which is contrasting to the available literature. SPBs show a very high (70%) rates of conversion to MM (average time span being 20.7 months from initial diagnosis). The 5 years survival rate of SPB is 60% falling drastically to 5.7% upon progression to MM.[7]

The etiopathogenesis of this rare entity remains an enigma with several proposed hypotheses like radiation, chemical exposure, viruses and genetic factors. Cytogenetic studies brought out loss in chromosome 13, 1p, 14q and gain in 19p, 9q, 1q. The principal growth factor implicated in this malignancy is interleukin-6. Malignant plasma cells secrete cytokines as well as osteoclast activating factor resulting in bone resorption.[8] Anaplastic variants likely develop in immunocompromised state and Epstein-Barr virus infection. In HIV patients a confounding factor is present. Routinely, HIV patients have reactive plasmacytosis in bone marrow. But, a highly aggressive myeloma, though rare, is still possibly be present.[9] Symptomatic patients of SPB mandible present with jaw pain, soft tissue /hard mandibular swelling, expansion of jaw, paresthesia, epulis formation, teeth mobility and pressure sensation.[10] Few case series also report symptoms of bleeding and pathological fractures.[8] Our case was unusual in presenting with a non-healing ulcer, an ipsilateral mandibular swelling with associated dysphagia which may make a clinician biased towards a primary oral malignancy. Diagnosis is essentially supported by radio-

imaging and clinched by histopathological examination (HPE). CECT shows well-defined osteolytic lesions having unilocular or multilocular radiolucency. These are multiple, rounded radiolucencies in staggered sizes and may be surrounded by a otosclerotic bone reaction.[8] Lae et al, described three types of radiographic patterns in SPB: multilocular soap-bubble lesions, unilocular radiolucency with cystic appearance and ill-defined destructive bone resorption.[1] In our case, it was ill-defined with bone destruction.

HPE revealed extensive laminae of plasma cells possessing architectural width extending between well-differentiated to anaplastic (blastic) end. Plasmoblastic morphology means high nucleocytoplasmic ratio round nuclei, fine chromatin and prominent nucleoli. These anaplastic cells may enlarge or become signet ring shaped mimicking a metastatic carcinoma, a poorly differentiated tumor from any cell lineage (neural, histiocytoma, fibrosarcoma, rhabdomyosarcoma) and melanoma thereby making the diagnosis tricky and difficult. In such situations, IHC facilitates correct diagnosis with melanoma being positive for S100, Melan A or HMB45 and pan-cytokeratin AE1/AE3 differentiating anaplastic myeloma from undifferentiated carcinoma.[3] In our reported case, IHC was positive for CD138, vimentin and MUM1 with the Ki-67 proliferative index between 40-50%. This index is a surrogate marker of likely growth and spread of a cancer. Index value above 30% is typically considered high.

Upon diagnosis, optimal treatment strategy must be instituted comprising of bisphosphonates (inhibit osteoclastic bone resorption) to prevent skeletal fractures. Definitive radiotherapy is offered for SPB and chemotherapy is reserved for advanced/ symptomatic myeloma.[7]

CONCLUSION

Plasmacytoma affecting head and neck region in absence of myelomatosis is extremely sporadic. Anaplastic variant of solitary mandibular plasmacytoma is a rare and aggressive variant that may mimic poorly differentiated carcinoma. Therefore, knowledge about oro-gnathic manifestations of such entities amongst otolaryngologists, dental surgeons and pathologists is undoubtedly mandatory. This surely should translate into early and correct diagnosis with institution of optimal treatment protocol that is essential to overall patient survival. Pathologists must have higher degree of diagnostic suspicion and knowledge to perform the arduous task of extensive search for plasmocytic cells with features of anaplastic variant. Upon confirmation, an elaborate diagnostic, multidisciplinary workup is the recommended approach towards its management involving an Otorhinolaryngologist, Onco hematologist, Onco pathologist and radiation oncologist. Radiotherapy by far has been the treatment of choice with an elevated 10 years survival value. Owing to a substantial risk of conversion into disseminated disease, authors propose a lifetime follow-up.

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