DOI: 10.7860/JCDR/2022/58839.17045

Case Report

Dentistry Section

Medullary Solitary Plasmacytoma of Maxilla- A Rare Case Report

JIGNA S SHAH[1], JAYA DUBEY[2]

ABSTRACT

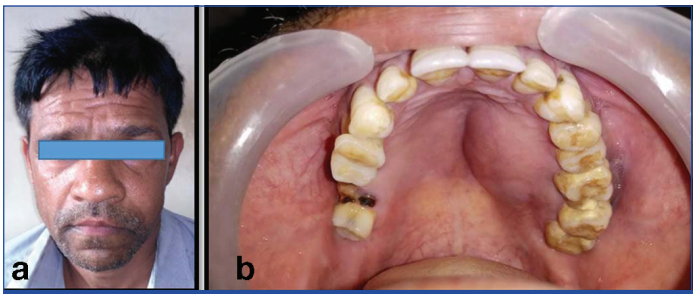
Solitary Plasmacytoma (SP) is a rare disease characterised by a localised proliferation of neoplastic monoclonal plasma cells, without evidence of systemic disease. The aetiology of SP remains unknown, but chronic stimulation, overdose irradiation, viruses, and gene interactions in the reticuloendothelial system have been suggested as the aetiologic factor. While pain is the most common symptom of osseous mandible SP, it is usually painless in the maxilla and may manifest orally with paresthesia, swelling, soft tissue masses, mobility and migration of teeth, haemorrhage, or pathologic fracture due to cortical destruction of bone. Because the amount of haemopoitic bone marrow in the maxilla is decreased, oral involvement of the maxilla is less common than that of the mandible. Here, authors reported a case of 46-year-old male patient presented with the chief complaint of mobility of his upper left back teeth and swelling over his left-side palatal area. Clinical examination was suggestive of benign odontogenic tumors and salivary gland tumors, while two-Dimensional (2D) and three-Dimensional (3D) imaging were suggestive of malignant tumors arising from either alveolar bone or maxillary sinus. The laboratory investigations and biopsy revealed solitary plasmacytoma of maxilla. There were clinical symptoms of paresthesia and mobility, but no other signs of malignancy were observed. As the case report concludes, all necessary investigations need to be conducted to exclude all other pathologies, so that treatment can begin as soon as possible.

Keywords: Extramedullary plasmacytomas, Hard palate, Multiple myeloma, Swelling

CASE REPORT

A 46-years-old male patient presented to the Department of Oral Medicine and Radiology with chief complaint of mobility of upper left-side of teeth and painless swelling extraorally over left-side of the face and intraorally over left palatal region for the past 3 months. Past history showed that patient first noticed mobility of teeth in upper left back tooth region. Then after one and half month he noticed paresthesia over left mid-face region and gradually increasing swelling in the same region. No history of any adverse oral habit, dental treatment or any systemic illness was present.

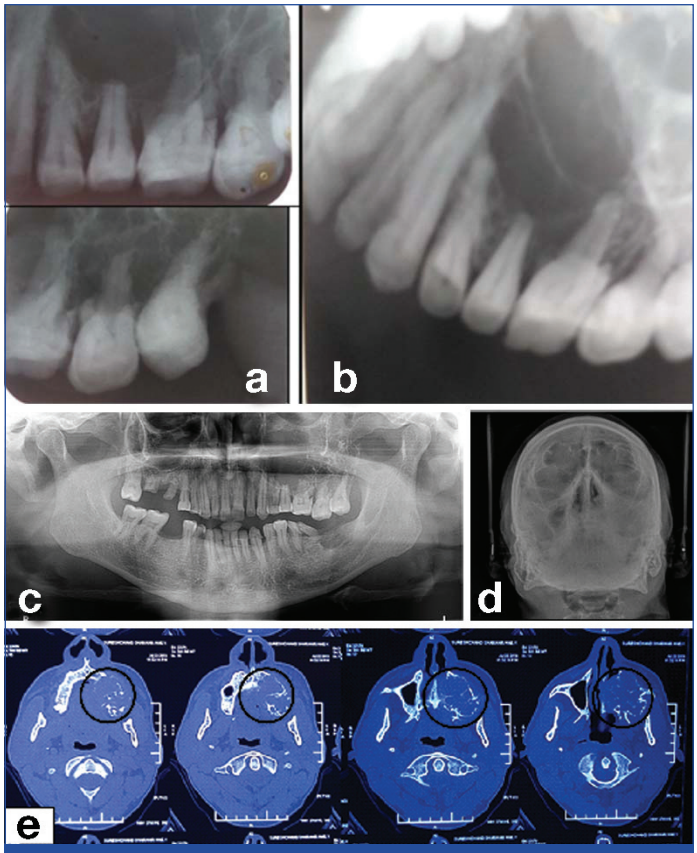
Extraoral examination, [Table/Fig-1a] showed facial asymmetry with diffuse swelling over left side of face with overlying skin was normal in colour, texture and afebrile temperature. Intraorally [Table/Fig-1b], a well-defined swelling of approximately 3×4 cm extending mesiodistally 1 cm away from midpalatal raphe to alveolar ridge and anterio-posteriorly from mesial surface of 24 to distal surface of 28 over left-side of hard palate was present. Overlying mucosa was normal with obliteration of depth of upper left buccal vestibule. Swelling was firm in consistency, non pulsatile, non tender, non haemorrhagic, non fluctuant and not fixed to overlying mucosa. Grade II mobility were present in 26,27 while Grade I mobility in 24,25,28. On the basis of history and clinical findings, benign odontogenic tumors and salivary gland tumors were suspected, then salivary gland tumors were excluded by the fact of presence of facial asymmetry. Single segment tooth mobility (in absence of periodontitis) and paresthesia raise the suspect of malignant lesions of alveolar bone or maxillary sinus and was further investigated by radiological, histopathological and haematological tests.



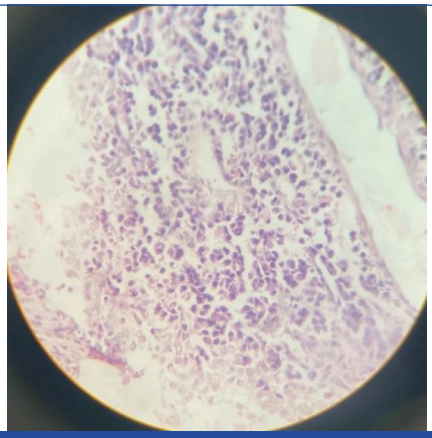
[Table/Fig-1]: a) Shows facial asymmetry with extraoral diffuse swelling present over left-side of face. Overlying skin was normal; b) shows intraoral well-defined swelling over left-side of hard palate and obliteration of left upper buccal vestibule.

Two-dimensional radiographs Intraoral Periapical (IOPA), lateral occlusal, Orthopantomogram (OPG) and Paranasal Sinus (PNS) [Table/Fig-2A,B,C,D], showed ill-defined multilocular radiolucency with irregular destruction of alveolar bone and floor of maxillary sinus. Internal structure showed very thin and incomplete septa. There was resorption of root apex 23,24,25,26 and 27 at right angle at apical third region. Contrast Enhanced Computed Tomography (CECT) of facial bone showed an ill-defined, [Table/Fig-2e] heterogeneous enhancing soft expansile lytic lesion of size 4.5×3.8×4.7 cm involving left maxillary sinus, hard palate and alveolar process with multiple internal septations area of cortical break. There was erosion of all walls of left maxillary sinus and floor of orbit with extension in nasal cavity. Present case also showed an air space in upper portion of the sinus. Radiological findings were suggestive of malignant tumors arising from either alveolar bone or maxillary sinus.

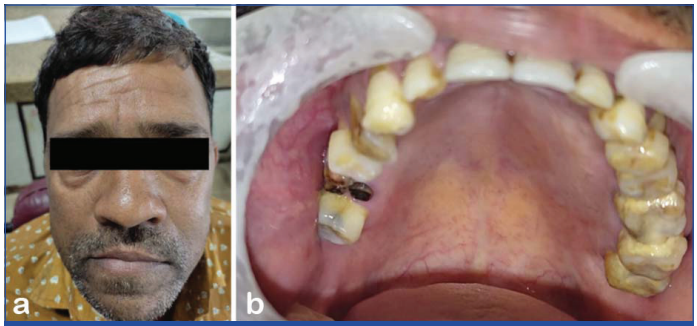
Routine blood investigations showed normal range of Haemoglobin (Hb), Red Blood Cell (RBC), Differential Leukocytes Count (DLC). There were no abnormalities identified in the liver and renal function tests, as well as no Bence Jone protein or any other abnormalities found in the urine examination. An incisional biopsy showed [Table/Fig-3] sheets of mature and immature closely packed cells resembling plasma cells with eccentrically placed nuclei exhibiting chromatin clumping in a 'cartwheel' or 'checkerboard' pattern. Immunohistochemistry results were reactive for Mum-1 and CD-138 and non reactive for CD-20. Final diagnosis was made as Solitary Plasmacytoma (SP). On diagnosis of plasmacytoma, the patient was referred to the Oncology Department for treatment, where he was given thalidomide 100 mg once a day until now. During the past two and a half years, the swelling has regressed in size, and the patient has been receiving regular follow-ups. [Table/Fig-4a,b].



[Table/Fig-2]: a,b) Shows Intraoral Periapical (IOPA) of lateral maxillary occlusal of maxillary posterior left back tooth region; c) OPG Showed ill-defined multilocular lytic lesion present over left maxillary alveolar region involving alveolar bone and left maxillary sinus; d) PNS view showed radiolucent areas in left maxillary sinus cavity with destruction of all walls of sinus; e) In the axial view of the CECT, a multilocular expansile lytic soft tissue lesion with internal septation and areas of cortical break was visible within the left maxillary alveolus and maxillary sinus. Erosion of all walls of maxillary sinus and floor of left orbit.



[Table/Fig-3]: Shows Haematoxylin and Eosin (H&E) (20x) sheets of mature and immature closely packed cells resembling plasma cells.



[Table/Fig-4]: a) Shows extraoral sign of regression of swelling; b) Shows intraoral regression of swelling by chemotherapy after two and half year follow-up.

DISCUSSION

Plasme Cell Neoplasms (PCN) are lymphoid neoplastic proliferations that have been grouped among B-cell peripheral lymphomas [1,2]. PCN are rare and uncommon type of haematological disorders which are classically categorised into four groups: Multiple Myeloma (MM), Plasma cell leukaemia, Solitary Plasmacytoma of Bone (SPB) and Extramedullary Plasmacytomas (EMPs) [3,4]. The aetiology of SP remains unknown, but chronic stimulation, overdose irradiation, viruses, and gene interactions in the reticuloendothelial system have been suggested as the aetiologic factor [2]. The male to female ratio of SP is approximately 2:1, with an average age of 55 years. Incidence rates varies from 8-15%. The most common site of occurrence is upper respiratory tract, especially the nasal cavity, oropharynx, nasopharynx, and sinuses [4]. The oral manifestations may vary from tooth pain, swelling or soft tissue mass, migration of teeth, root resorption, haemorrhage, or pathologic fracture [5]. The present case was reported in maxillary posterior region in a 46-years-old male patient having non ulcerated, painless firm palatal swelling and mobility of maxillary molars with paresthesia. Clinical differential diagnosis of palatal swelling includes odontogenic and non odontogenic, benign and malignant tumors of oral cavity.

SP exhibited a wide spectrum of radiographic findings, from ill-defined destructive radiolucency with ragged borders and no periosteal reaction to well-defined, unilocular radiolucency. The mandible is more likely to develop osteolytic lesions than the maxilla (more haematopoietic activity) [4]. The radiographic features in the present case were an ill-defined, multilocular radiolucency with straight septa that resembled an odontogenic myxoma. The 3D scan showed ill-defined lytic lesion with irregular destruction of the left upper alveolar bone, periodontium and the maxillary sinus suggestive of malignancy [6]. In a middle-aged adult with an expansile multilocular radiolucent lesion, a number of differential diagnosis are possible. The most common lesions based on the clinical manifestation and radiographic feature included odontogenic myxoma, ameloblastoma, and odontogenic keratocyst [2].

The differentiation of the origin of tumor may be radiographically difficult when bone destruction is associated with soft tissue involvement. Radiographically, on basis of two point, it can be differentiated whether the mass was of bone origin rather than soft tissue. The first point was that the mass originated in the sinus mucosa would have had a central area of necrosis on Computed Tomography (CT) and Magnetic Resonance Imaging (MRI) and second point was that in maxillary sinus squamous cell carcinoma, the sinus would first be filled by the mass, then begin to destroy the sinus wall radiographically. Present case showed an air space in upper portion of the sinus, and at the same time bony destruction was also present which suggests that the mass had originated in bone, not sinus mucosa [5,7].

The dense plasma cell infiltrate, commonly associated with inflammatory lesions, makes it difficult to diagnose SP. Therefore, Immunohistochemistry (IHC) is required to differentiate it with other lesions which shows plasma cell infiltration in bone [3]. SP shows similar histological features to MM. Only IHC is useful for identifying lymphomas and MM as well as other tumors [4]. In present case, histopathology showed classical features of SP but in some regions malignant spindle cells were present. IHC was used for confirmation of diagnosis. In IHC painting, CD-138, vimentin, Ki-67 and Epithelial Membrane Antigen (EMA) were positive. The results of the present case are in accordance with other studies [3-6,8,9]. SP, whether osseous or non osseous, is distinguished from MM by absence of hypercalcaemia, renal insufficiency and anaemia, normal skeletal survey. There was no evidence of these findings in the present case [9].

The ideal therapeutic approach is still controversial, however, radiotherapy seems to be the treatment that offers better clinical results since the SBP reveals itself as a radiosensitive lesion. The rates of local control of SBP with radiotherapy presented in the literature exceed the range of 80%. Surgical intervention should be carried out in situations where there is no prediction of functional or aesthetic damage. Chemotherapy is advocated only on the basis

of reports in the literature that showed improvement of local control and delayed development of MM. Radiation therapy in conjunction with chemotherapy is accepted treatment [10,11].

CONCLUSION(S)

The present study reports a rare case of medullary SP of the maxilla presenting with tooth mobility and palatal swelling as the first signs and suggesting that dentists should thoroughly examine patients and perform all investigations, particularly when the maxilla is involved in order to rule out any other possibilities. Early detection of malignant tumors is very important as survival is highly dependent on the stage of the tumor.

REFERENCES

[1] Grammatico S, Scalzulli E, Petrucci MT. Solitary plasmacytoma. Mediterr J Hematol Infect Dis. 2017;9(1):e2017052.
[2] Ozdemir R, Kayiran O, Oruc M, Karaaslan O, Koçer U, Ogun D. Plasmacytoma of the hard palate. J. Craniofac Surg. 2005;16(1):164-69.
[3] Oda T, Sue M, Sasaki Y, Kameta A, Okada Y, Tsuchimochi M, et al. Extraosseous plasmacytoma of the Maxilla: CT and MRI findings. Oral Sci. Int. 2017;14(1):18-21.
[4] Rezaei F, Nazari H, Izadi B. Solitary plasmacytoma in the mandible resembling an odontogenic cyst/tumor. Case Rep Dent. 2016;2016;3629047.
[5] Radhika MB, Thambiah LJ, Paremala K, Sudhakara M. Multiple myeloma: Periapical location can challenge diagnostic skills. JICDRO. 2010;2(1):49-54.
[6] Kasamatsu A, Kimura Y, Tsujimura H, Kanazawa H, Koide N, Miyamoto I, et al. Maxillary swelling as the first evidence of multiple myeloma. Case Rep Dent. 2015;2015. Doi: 10.1155/2015/439536.
[7] Matsumura S, Kishino M, Ishida T, Furukawa S. Radiographic findings for solitary plasmacytoma of the bone in the anterior wall of the maxillary sinus: A case report. Oral Surg Oral Med Oral Pathol Oral Radiol. 2000;89(5):651-57.
[8] Zhao XJ, Sun J, Wang YD, Wang L. Maxillary pain is the first indication of the presence of multiple myeloma: A case report. Mol Clin Oncol. 2014;2(1):59-64.
[9] Popovski V, Dvojakovska S, Benedetti A, Panchevski G, Stamatoski A, Janevska V. Mandibular involvement of plasmacytoma–Uncommon case report of rare entity. Ann Med Surg. 2019;45:95-97. Doi: 10.1016/j.amsu.2019.07.021.
[10] Shirani AM. The Diagnosis and treatment of a rare maxillary plasmacytoma: A case report. J Dent. 2020;21(3):239-43.
[11] Lombardo EM, Maito FL, Heitz C. Solitary plasmacytoma of the jaws: Therapeutical considerations and prognosis based on a case reports systematic survey. Braz J Otorhinolaryngol. 2018;84(6):790-98.

PARTICULARS OF CONTRIBUTORS:
1. Professor and Head, Department of Oral Medicine and Radiology, Government Dental College and Hospital, Ahmedabad, Gujarat, India.
2. Postgraduate Student, Department of Oral Medicine and Radiology, Government Dental College and Hospital, Ahmedabad, Gujarat, India.

NAME, ADDRESS, E-MAIL ID OF THE CORRESPONDING AUTHOR:
Dr. Jaya Dubey,
PG Girls Hostel, Civil Hospital Campus, Ahemdabad, Gujarat, India.
E-mail: jayadubey.dubey8.jd@gmail.com

PLAGIARISM CHECKING METHODS: [Jain H et al.]
- Plagiarism X-checker: Jul 04, 2022
- Manual Googling: Sep 09, 2022
- iThenticate Software: Sep 10, 2022 (20%)

ETYMOLOGY: Author Origin

AUTHOR DECLARATION:
- Financial or Other Competing Interests: None
- Was informed consent obtained from the subjects involved in the study? Yes
- For any images presented appropriate consent has been obtained from the subjects. Yes

Date of Submission: **Jul 02, 2022**
Date of Peer Review: **Jul 29, 2022**
Date of Acceptance: **Sep 12, 2022**
Date of Publishing: **Oct 01, 2022**