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Extramedullary Plasmacytoma of Left Buccal Mucosa – A Rare Case Presentation

[1]C.R. Fathima Shirin, MDS, Department of Oral Medicine and Radiology, Government Dental College, Alappuzha

[2]Gopinadh Soumya, MDS, Department of Oral Medicine and Radiology, Government Dental College, Alappuzha

[3]Thomas Sherin Ann, MDS, Department of Oral Medicine and Radiology, Government Dental College, Alappuzha

[4]K.L. Girija, MDS, Department of Oral Medicine and Radiology, Government Dental College, Alappuzha

Corresponding Author: C.R. Fathima Shirin, MDS, Department of Oral Medicine and Radiology, Government Dental College, Alappuzha

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Type of Publication: Case Report

Conflicts of Interest: Nil

Abstract

Extramedullary plasmacytomas are uncommon, solitary soft tissue tumours that can be identified by histopathological analysis. They are caused by malignantly altered monoclonal plasma cell proliferation. Multiple myelomas, which are intimately linked to these lesions, should be ruled out in each of these instances by appropriate laboratory and radiographic examinations. This article reports a case of a 79-year-old man presented with a quickly expanding, painless swelling in the left cheek area. A diagnosis of solitary plasmacytoma was reached based on histological, radiological, and clinical data. It is crucial to diagnose extramedullary plasmacytomas as soon as possible. The treatment of choice is radiotherapy with or without adjuvant chemotherapy. Close patient monitoring is necessary following the conclusion of the therapeutic procedures since there is a chance that the condition will progress to multiple myeloma.

Keywords: Extramedullary plasmacytoma, Multiple Myeloma, Solitary Plasmacytoma of Bone

Introduction

The plasma cell neoplasms are tumors derived from bone marrow stem cells of the B-lymphocyte lineage. It is characterized by the expansion of a clone of immunoglobulin-secreting cells. It can appear as a solitary plasmacytoma of bone (SPB), an extramedullary plasmacytoma

(EMP), or as a component of the multifocal disseminated disease multiple myeloma (MM) [1]. The EMP makes up just about 3% of all plasma cell neoplasms mainly affecting men, in their sixth to eighth decades of life. [2] Extramedullary lesions may occur in the absence of bone involvement, especially in the head

and neck. Approximately 90% of EMPs are found in the head and neck region commonly involving nasal cavity, paranasal sinuses, tonsillar fossa, and oral cavity.[3] Viruses, excessive radiation, chronic stimulation, and gene defects in the reticuloendothelial system have been considered as the probable etiological factors, yet the exact etiology is still uncertain.[4] Extramedullary plasmacytomas have the tendency to transform into plasmacytomas of the bone and myelomas, both of which have a worse prognosis[5].

Case Report

A 79 year old otherwise normal male patient came to the outpatient department of Oral Medicine and Radiology with a chief complaint of swelling on left side the face since two months. Patient was using denture since 10 years and was apparently normal till he noticed the swelling two months back. Initially the swelling was small peanut sized which rapidly increased to the present size. Swelling was not associated with pain or pus discharge. His major issue was related to discomfort while using denture. His family, social and personal history were unremarkable and his vital signs were within normal limits.

On extra oral examination, a diffuse swelling was noted on left side of face extending superoinferiorly from the left lower eyelid to lower border of left side of mandible.[Figure 1] Anteroposteriorly it extended from ala of nose to 4 cm posterior to tragus of ear. Surface appeared to be of normal skin with no erythema, bleeding, pus discharge or visible pulsations. Drooping of eyelid and epiphora were noted in relation to the left eye. On palpation, all inspection findings were confirmed with respect to site, size shape and extent. The swelling was non tender and soft to firm in consistency with no bleeding or pus discharge. It was also non compressible, non-reducible and non-pulsatile. Skin over the swelling could be pinched off and there was no local rise in temperature.

On intraoral examination, completely edentulous upper and lower arch were noted. Upper and lower labial mucosa, right buccal mucosa, tongue, upper and lower vestibules and hard palate appeared to be normal. A well defined swelling of size approximately 7x5 cm was noted on left buccal mucosa.[Figure 2] Overlying surface appeared to be smooth and the colour was of normal adjacent mucosa. No bleeding, erythema or pus discharge were present. On palpation, it was non tender and firm in consistency with no bleeding or pus discharge. A differential diagnosis including minor salivary gland tumour and connective tissue neoplasms were made. Panoramic radiograph was taken to rule out any bony changes and showed no visible changes.[Figure 3] Routine haematological investigation results were within normal range. Incisional biopsy of the swelling was done under local anaesthesia. Histopathological examination revealed closely packed tumour cells proliferating in sheets. Most of the cells were round to ovoid in shape with eosnophilic cytoplasm and eccentrically placed nuclei exhibiting chromatin condensation in a cart wheel pattern. Perinuclear halo was seen in some of the cells.[Figure 4] Kappa-Lambda(FLC) ratio test was also done which showed an elevated FLC ratio of 3.982 mg/L, with a high serum kappa and lamda free light chains(174.00 mg/L and 43.70 mg/L respectively). Following this, a diagnosis of Solitary plasmacytoma was made and the patient was referred to higher centers for further evaluation and management.

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Discussion

Plasma-cell tumors are characterized by the expansion of monoclonal plasma cell proliferation. Plasmacytoma is divided into medullary and extramedullary types, which can be either solitary or multiple in distribution.

EMP makes upto 3% of all plasma cell tumors. The solitary variant of EMP makes up between 5% and 10% of all plasma cell neoplasms. There is a reported higher incidence among elderly individuals, mostly in fifth and sixth decades of life with a male predilection.[6] In our case also the patient was an elderly male with 79 years.

Majority of the EMPs are reported to originate from the upper respiratory tract (nose and paranasal sinuses being the common sites), followed by nasopharynx, tonsils and oropharynx.[7] Similar cases has been reported by Rawat et al. and Singh et al.[7,8] The involvement of buccal mucosa in EMP has been reported by Kerem Ozturk et al.[9] The reported cases of solitary plasmacytoma on the buccal mucosa are very few. Our case has been unique in this aspect.

In our case the chief complaint of the patient was a painless swelling of two months duration on the left side of the face. This goes in accordance with the findings of Ozdemir et al.[10] where pain is not a feature of EMP except when there is a secondary infection or bone destruction.

Plasmacytoma is clinically similar to chronic inflammatory diseases, plasma cell gingivitis, lymphomas etc. Histopathology is the only definite diagnostic tool for differentiating plasmacytoma from other lesions. Monoclonal proliferation is suggestive of neoplasm superimposed to inflammatory lesions. Radiographically, EMP can erode the bone and make it difficult to be distinguished from a bony lesion.[11] The absence of CRAB characteristics (increased calcium, renal failure, anemia, or numerous bone lesions) helps to distinguish solitary plasmacytoma from multiple myeloma. Tissue biopsy confirming monoclonal plasma cell histology, bone marrow plasma cell infiltration of less than 5% of total nucleated cells, lack of osteolytic bone lesions or other tissue involvement without evidence of myeloma, hypercalcemia, renal failure, and low serum M protein concentration are the diagnostic criteria for extramedullary plasmacytoma (EMP). Immunohistochemistry staining should be performed to ascertain the presence of a monoclonal plasma cell population. Positive plasma cells that express CD38 and simultaneously display kappa or lambda light chains in their cytoplasm can serve as a diagnostic marker[12]. The rate of progression of EMP to MM is 15-20%. Harwood et al. reported the high rate of conversion to MM if EMP involved the underlying bone. [13] According to Dingli et al., an abnormal FLC ratio at the time of diagnosis of solitary plasmacytoma would be an adverse prognostic factor for progression to myeloma and in our case FLC ratio showed an elevated value of 3.982 mg/L.[14]

Extramedullary plasmacytoma is highly radiosensitive[15]. So treatment of choice is radiotherapy. 40 to 60 Gy is the radiation dose advised. According to Weber et al., a radiation dose of 40 Gy in 20 fractions is enough for EMP less than 5 cm but a higher dose of up to 50 Gy in 25 fractions is recommended for EMP greater than 5 cm. In contrast to single-modality radiation therapy, Chao et al. found that surgical resection, either alone or in combination with adjuvant irradiation, demonstrated a better pattern of response to EMPs originating in the head and neck [15].

EMP in the head and neck has a good prognosis. The patient's prognosis depends on various factors i.e. type of plasmacytoma, tumour burden, synchronous tumours,

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specifc immunoglobulin expression, presence of signs of dissemination, myeloma conversion and pathological grade of tumours. Mock et al. observed that EMP expressing lambda light chains were more likely to evolve into MM, whereas none of the cases expressing kappa light chains progressed to multiple myeloma. Cases with lambda light chain restriction were more immature and, therefore, more likely to progress to MM[17]. In contrast, Knobel et al. found no correlation between light chain restriction and survival[18].

Conclusion

The EMP can evolve under unfavorable conditions. 35 to 50% of cases to regional localizations or develop into multiple myeloma. Localized plasma cell tumors may be the first sign of MM. Therefore proper evaluation should be done in every patient with oral plasmacytomas to rule out MM symptoms. Proper examinations including bone scan and, if necessary, and MRI and/or FDG-PET, should be adviced to rule out extramedullary localizations.

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Legend Figures

Figure 1: Extraoral swelling

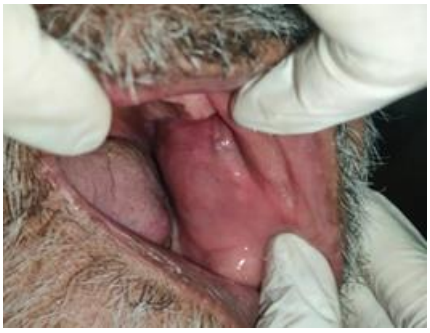

Figure 2: Intraoral swelling

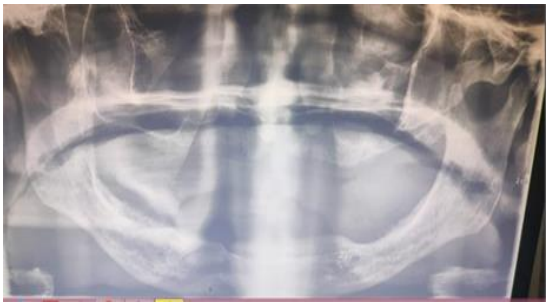

Figure 3: Panoramic radiograph showing no visible changes

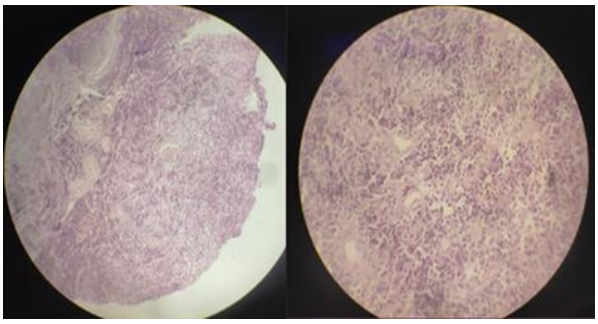

Figure 4: Histopathology revealing closely packed tumour cells

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