ORIGINAL RESEARCH PAPER
Volume - 12 | Issue - 02 | February - 2023 | PRINT ISSN No. 2277 - 8179 | DOI : 10.36106/ijsr
INTERNATIONAL JOURNAL OF SCIENTIFIC RESEARCH

EXTRAMEDULLARY PLASMACYTOMA OF THE ORAL CAVITY: THREE CASE REPORTS OF A RARE ENTITY

Oral Pathology

Sabu Paul Associate Professor, Department of Oral Pathology and Microbiology, Government Dental College Kottayam

Rasla P C* Junior resident, Department of Oral Pathology and Microbiology, Government Dental College Kottayam*Corresponding Author

Latha Mary Cherian Professor and HOD, Department of Oral Pathology and Microbiology, Government Dental College Kottayam

Indu Mohan Assistant Professor, Department of Oral Pathology and Microbiology, Government Dental College Kottayam

Manjusha Mohanan Junior resident, Department of Oral Pathology and Microbiology, Government Dental College Kottayam

Bhagya J Junior resident, Department of Oral Pathology and Microbiology, Government Dental College Kottayam

Sudharani Junior resident, Department of Oral Pathology and Microbiology, Government Dental College Kottayam

ABSTRACT

The plasma cell neoplasms are tumors derived from bone marrow stem cells of the B-lymphocyte lineage. Extramedullary plasmacytoma (EMP) is a monoclonal, immuno-proliferative B-cell disorder that develops outside of the bone marrow and lacks any outward signs of multiple myeloma. It makes up just about 3% of all plasma cell neoplasms. The majority of EMP (80%) occurs in the head and neck region especially in the nasopharynx, paranasal sinuses, and tonsils. Diagnosis of EMP is based on histologic confirmation of monoclonal plasma cell infiltration of a single disease site and by exclusion of systemic myeloma. Continuous patient monitoring is required since these individuals have a significant risk of developing multiple myeloma[1]. Three cases of EMP of oral cavity clinically diagnosed as benign soft tissue lesions were reported. After histopathological evaluation using Hematoxylin and Eosin and immunohistochemical examination, final diagnosis of Extramedullary Plasmacytoma was given for all the three cases.

KEYWORDS

Extramedullary plasmacytoma, Multiple myeloma, Plasma cell neoplasms, Solitary plasmacytoma.

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, making it uncommon. The majority (80%) occurs in the head and neck area, particularly in the tonsils (75%), nasopharynx, and paranasal sinuses. Men in their sixth to eighth decades of life are commonly affected. An EMP can develop either spontaneously (primary EMP) or develop over the course of multiple myeloma disease (secondary EMP). Viruses, excessive radiation, chronic stimulation, and gene defects in the reticuloendothelial system have been proposed as the most likely causative factors, yet the exact cause of this disease is still unknown. Extramedullary plasmacytomas have the potential to develop into plasmacytomas of the bone and myelomas, both of which have a worse prognosis[2]. We described three incidences of EMP in this paper in otherwise healthy people.

Case 1

A 69-year-old lady presented with pain and swelling in the left mandibular alveolar ridge for one month. The pain was more intense at night and was of throbbing nature. The swelling gradually increased in size and the patient felt difficulty while chewing. Upon inspection, no apparent facial asymmetry was noted. Intraoral examination revealed a 2 x 1 cm diffuse, slightly tender, firm to hard swelling on the left body of the mandible between 34 and 36 regions. The swelling extended buccolingually, involving the lingual sulcus and floor of the mouth. There was no discharge, and the swelling was not fixed to the underlying bone. The surface mucosa appeared to be normal. A slight obliteration of the buccal vestibule was present. An uneven lytic zone with respect to the edentulous area of 36 and 37 was visible on the panoramic radiograph. Blood investigations were within the normal range. Microscopic examination of the biopsied specimen revealed sheets of pleomorphic plasma cells, which were positive for CD138 and Kappa light chain and negative for Lambda light chain antibodies (Figure 1). Based on this, the diagnosis of extramedullary plasmacytoma with lambda light chain restriction was made.

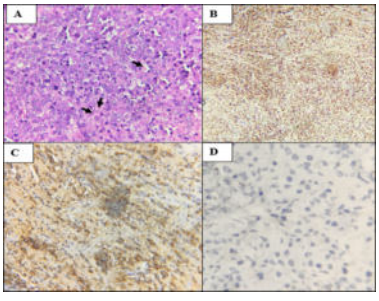

Figure 1: Photomicrographs showing (A) pleomorphic plasma cells and Russell bodies (black arrow), H&E 40x, (B) CD138 positive, 10x (C) Anti-kappa light chain positive, and (D) Anti-lambda light chain negative, 40x

Case 2

A three-week-old swelling in the mandibular anterior teeth region was noted in a 63-year-old man. There was occasional bleeding from the swelling while chewing and brushing. Upon inspection, a solitary, sessile, 0.8 x 0.7 cm ovoid swelling was found buccally over the interdental papilla of teeth 32 and 33. The swelling was firm and non-tender. The overlying mucosa appeared erythematous. A preliminary diagnosis of pyogenic granuloma was made based on the clinical presentation. On microscopic examination, the biopsied specimen revealed an ulcerated epithelium and a moderately collagenous connective tissue stroma with dense collection of plasma cells and

lymphocytes. Most of the plasma cells exhibited atypical characteristics and were positive with CD138 and kappa light chain and negative with lambda light chain antibodies

For more advanced care, the patient was transferred to a higher center. The patient developed back pain after a year, and subsequently, multiple myeloma was diagnosed. The patient was posted for radiotherapy and chemotherapy, but after three months of treatment, patient expired.

Case 3

A 47-year-old male presented with a proliferative growth in the left alveolar region for three weeks. He had undergone extraction of a grossly decayed mandibular third molar one month back. A growth was noted in the extraction site after one week of extraction and gradually increased in size for three weeks. Patient felt difficulty chewing. There was no associated pain. On examination, a firm, non-tender, mobile mass extending from 37 to the ramus region and the lingual vestibule was noted with bleeding on probing. Histopathology revealed stroma infiltrated with immature plasma cells with an eccentrically placed nucleus. Few cells showed vesicular nuclei and prominent nucleoli. Numerous mitotic figures were present. Tumor cells were positive for CD138, and Lambda light chain and negative for Kappa light chain (Figure 2). The patient was sent for radiotherapy and after 15 months, the lesion subsided. Patient is still on follow-up.

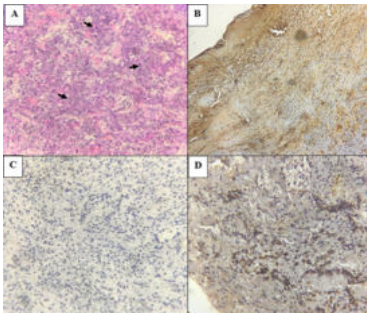

Figure 2: Photomicrograph showing (A) binucleated cells and mitotic figures (black arrow) (H&E, 40x), (B) Anti CD138 Positive (10x), (C) Anti-kappa Light chain Negative and (D) Anti-lambda Light chain Positive plasma cells (40x)

DISCUSSION

Plasma-cell tumors, which are characterized by the expansion of monoclonal plasma cell proliferation, comprise a wide range of neoplasms. According to the literature, solitary plasmacytoma (SP) is an uncommon form of plasma cell tumor that makes up between 5% and 10% of all plasma cell neoplasms. Based on where it is, it can be classified as either an extramedullary plasmacytoma (EMP) or a solitary plasmacytoma of bone (SBP)[1].

Extra-medullary plasmacytoma is a monoclonal, immuno-proliferative B-cell disorder that develops outside of the bone marrow and lacks any outward signs of multiple myeloma. It begins as a cluster of plasma cells that have undergone malignant transformation. They frequently move and come back to the bone marrow. In extremely rare cases, they localize in soft tissue or extracellular connective tissue. This is the source of extra-medullary monoclonal plasma cell foci, which are seen outside of the bone marrow. This might happen as a result of decreased expression of adhesion molecules, chemokine receptors, and tetraspanins, increased expression of the heparinase enzyme, or upregulation of activation pathways[2].

SBP is about 40% more common than EMP among plasmacytomas and has different presenting characteristics. The most common kind of plasma cell neoplasm is multiple myeloma (MM), which denotes the wide range of these tumors. Since SP frequently precedes multiple myeloma, a diagnosis of SP should only be made if no transformation takes place within a year. The average age of the patients with Solitary Plasmacytoma is 55 years, with male to female ratio of 2:1. The incidence rate increases exponentially with age, however, compared to multiple myeloma, it is less noticeable. Around 30% of people of African descent have SP, greater than the white race[3].

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[4].

Radiotherapy with radiation doses of 40 to 60 Gy is advised. According to Weber et al., a radiation dose of 40 Gy in 20 fractions is sufficient for EMP less than 5 cm; however, a higher dose of up to 50 Gy in 25 fractions is recommended for bulky EMP greater than 5 cm. For about 67% of patients, the right therapy leads to long-term stability and a chance of recovery. 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[5].

EMP in the head and neck has a good prognosis. Variations in the biological behavior of EMP and the limited number of documented cases make it difficult to determine prognostic factors. Age, tumor size, site of origin (extramedullary versus bone), grade, M-protein, light chains, and radiotherapy dose are all considered as prognostic factors. According to the study of Gerry D et al., head and neck EMP displays superior five-year overall and disease-specific survival rates compared with non-head and neck EMP[6]. Patients with lesions less than 5 cm had a local control rate of 100%, while patients with larger tumors had a local control rate of about 40%. 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 MM**.** Cases with lambda light chain restriction were more immature and, therefore, more likely to progress to MM[7]. In contrast, Knobel et al. found no correlation between light chain restriction and survival[8].

CONCLUSION

The EMP's evolution is unpredictable. After being identified and treated, the patient may go months or even years without exhibiting any symptoms. However, under unfavorable conditions, the tumor may grow, leading 35 to 50% of cases to regional localizations or developing into multiple myeloma. As seen in Case 2, localized plasma cell tumors can be the first sign of MM. Therefore, everyone involved in dental care should assess every patient with oral plasmacytomas for MM symptoms, including a bone scan and, if clinically necessary, an MRI and/or FDG-PET, among other tests. With the help of MRI and FDG-PET, clinicians may be able to rule out more extramedullary localizations[9].

REFERENCES

1. Dimopoulos MA, Moulopoulos LA, Maniatis A, Alexanian R. Solitary plasmacytoma of bone and asymptomatic multiple myeloma. Blood, The Journal of the American Society of Hematology. 2000 Sep 15;96(6):2037-44.
2. Kilciksiz S, Karakoyun-Celik O, Agaoglu FY, Haydaroglu A. A review for solitary plasmacytoma of bone and extramedullary plasmacytoma. The Scientific World Journal. 2012 Oct;2012.
3. Dores GM, Landgren O, McGlynn KA, Curtis RE, Linet MS, Devesa SS. Plasmacytoma of bone, extramedullary plasmacytoma, and multiple myeloma: incidence and survival in the United States, 1992–2004. British journal of haematology. 2009 Jan;144(1):86-94.
4. Bharathi MB, Malik E, Babu AR. Plasmacytoma of Head and Neck: Case Series. Journal of Case Reports. 2018 Jan 14;7(4):371-3.
5. Chao MW, Gibbs P, Wirth A, Quong G, Guiney MJ, Liew KH. Radiotherapy in the management of solitary extramedullary plasmacytoma. Internal medicine journal. 2005 Apr;35(4):211-5.
6. Gerry D, Lentsch EJ. Epidemiologic evidence of superior outcomes for extramedullary plasmacytoma of the head and neck. Otolaryngology–Head and Neck Surgery. 2013 Jun;148(6):974-81.
7. Mock PM, Neal GD, Aufdemorte TB. Immunoperoxidase characterization of extramedullary plasmacytoma of the head and neck. Head & neck surgery. 1987 Jul;9(6):356-61.
8. Knobel D, Zouhair A, Tsang RW, Poortmans P, Belkacémi Y, Bolla M, Oner FD, Landmann C, Castelain B, Ozsahin M. Prognostic factors in solitary plasmacytoma of the bone: a multicenter Rare Cancer Network study. BMC cancer. 2006 Dec;6(1):1-9.
9. Romano A, Marescalco MS, Liardo C, Villari L, Vetro C, Conticello C, Di Raimondo F, Ferlito S. Oral lesion as unusual first manifestation of multiple myeloma: case reports and review of the literature. Case Reports in Hematology. 2014 Nov 25;2014.]