CASE REPORT

Plasmablastic extramedullary plasmacytoma

Irulandy Ponniah, Suhasini Rajan

Department of Oral and Maxillofacial Pathology, Tamil Nadu Government Dental College and Hospital, Chennai, Tamil Nadu, India

Address for correspondence:
Dr. Irulandy Ponniah, Department of Oral and Maxillofacial Pathology, Tamil Nadu Government Dental College and Hospital, Chennai - 600 003, Tamil Nadu, India. E-mail: salivaryduct@yahoo.co.uk

ABSTRACT

Extramedullary plasmactyoma is the solitary, soft tissue form of plasma cell neoplasm but lack the defining features of medullary or multiple myeloma. The diagnosis is difficult to make in routine practice setting due to the morphological and immunohistochemical overlap with plasmablastic lymphoma. We report a case of plasmablastic extramedullary plasmacytoma in a 52-year-old in the mandibular lingual gingiva and discuss its differential from plasmablastic lymphoma. The gingival mass regressed with primary radiotherapy.

KEY WORDS: Extramedullary, gingiva, lymphoma, plasmablastic, plasmacytoma

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INTRODUCTION

Plasmactyoma exists in three clinical forms, *viz.*, multiple myeloma (MM), medullary plasmacytoma (MP), and extramedullary plasmacytoma (EMP).[1] The latter is a localized collection of tumor cells in the soft tissue with plasma cell differentiation, but without evidence of the former two or lymphoma.[1] The adjective "plasmablastic" is used when there are more than 30% of plasmablasts in routine sections.[2] We report a case of plasmablastic EMP in the gingiva of a 52-year-old male and discuss its primary differential diagnosis.

CASE REPORT

A 52-year-old apparently healthy male presented with a painless, gingival mass of 3 months duration. His past medical history was not significant. He did not give any history of fever, night sweats, chills, or weight loss. On examination, a palpable cervical lymph node was noted on the right side of the neck. Intra-orally, a reddish, smooth surfaced, soft tissue mass involving the gingival/alveolar complex in relation to the right second mandibular molar, edentulous third molar, and retromolar region was noted [Figure 1]. There was no tooth mobility or bleeding on provocation. Panoramic and lateral skull radiographs revealed no bone lesion.

An incisional biopsy was performed under the impression of squamous cell carcinoma, which microscopically showed sheets of medium to large-sized cells with variable cytoplasm, eccentric nuclei with dense to light chromatin, variable perinuclear hof, single prominent nucleoli, binucleated cells, and occasional mitotic figures. The description indicates varying levels of cellular maturity [Figure 2]. The microscopical differential diagnosis included plasmablastic lymphoma (PBL) and plasmacytoma. Fine needle aspiration cytology of the cervical lymph node suggested lymphoma.

Complete blood count and biochemical values were within normal limits. Serum and urine electrophoresis for gammoglobulins and Bence-Jones protein were negative. Enzyme-linked immunosorbent assay for HIV was negative. No abnormalities were detected on skeletal survey. Plain chest radiograph and abdominal computed tomography scan were normal. No abnormalities were detected on bone-marrow examination.

A panel of commercially available immunohistochemical markers [Table 1] was used to characterize the lesion as to whether it was PBL or EMP, but the immunoprofile of the current case is consistent with EMP [Figure 3] and is also not inconsistent with PBL. The slides were interpreted by three pathologists, with and without immunohistochemistry, but no specific diagnosis as to PBL or EMP was rendered by each of them. Therefore, the diagnosis of EMP was based on the clinical and laboratory parameters in conjunction with morphological and immunohistochemcial features. The patient was treated initially by radiotherapy (40 Gy in 20 fractions over 4 weeks) followed by conditioning chemotherapy with oral mephalan plus thalidomide (six 4-week cycle). Follow-up showed regression of the oral lesion [Figure 4]. No changes were noted on laboratory investigation.

DISCUSSION

EMP is rare in the head and neck region, with an incidence of EMP to MM as 1:80

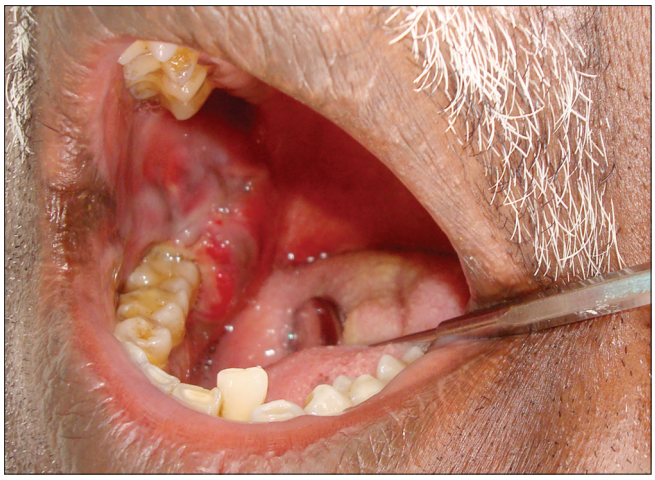

Figure 1: The clinical appearance of the lesion at presentation

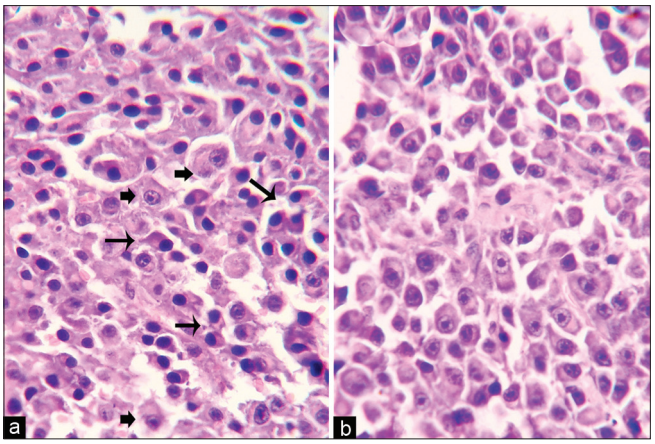

Figure 2: Neoplastic plasma cells with varying degrees of maturation with plasmacytes (arrow), intermediate form and plasmablasts (arrow head) in A, and plasmablasts in B (H and E; ×240)

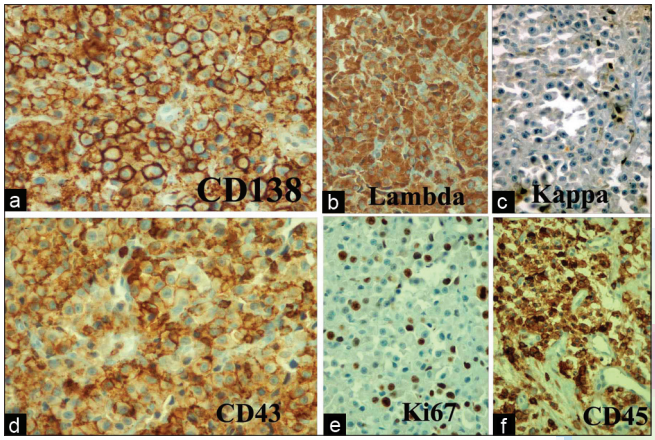

Figure 3: (a) Positive reaction with CD 138. (b) Positive reaction with Lambda. (c) Negative reaction with Kappa. (d)–(f) Positive reaction with CD 43, Ki-67, and CD 45 (IHC, ×400)

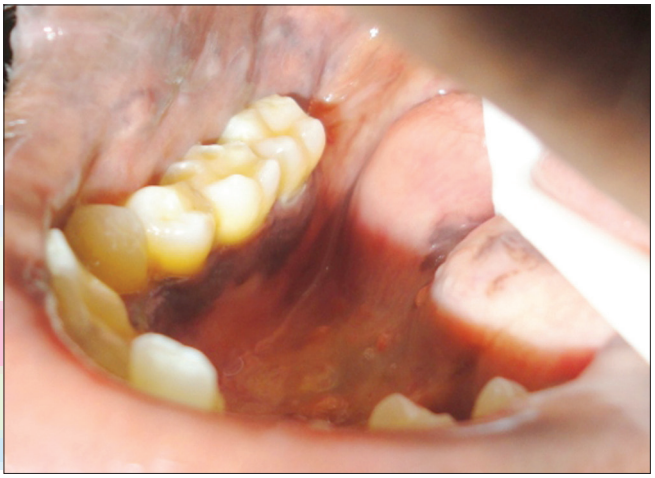

Figure 4: Resolution of oral lesion after radiotherapy

Table 1: Shows the panel of immunohistochemical markers employed

Marker	*Dilution*	*Clone*	*Result*
CD3	1 in 100	Polyclonal; DAKO	Negative
CD43	-do-	DF-T1; DAKO	Positive
LCA (CD45)	-do-	2B11+PD7/26; DAKO	Variable
CD56	-do-	1B6; Novocastra	Negative
CD20	1 in 200	L26; CELL MARQUE	Negative
CD79a	1 in 100	JCB117; DAKO	Negative
CD138	Ready to use	B-A38; CELL MARQUE	Positive
Kappa	Prediluted	Polyclonal; DAKO	Negative
Lambda	-do-	-do-	Positive
LMP-1	1:100	CS1-4; Dakocytomation	Negative
Ki-67	1 in 100	MIB-1; DAKO	20–25%
Cyclin D1	Ready to use	NCL-CYCLIN D1-GM; Novocastra	Negative

per year.[3] Within the head and neck region, EMP very rarely occurs on the gingiva[4] and about 75–85% are found in the upper aerodigestive tract with sites of predilection in the nasal cavity, paranasal sinuses, and nasopharynx.[3] Men are affected more frequently than women with peaks in the sixth to eighth decade of life. The disease may present as a localized mass, with lymph node involvement being not uncommon at the time of diagnosis.[3]

Histologically, the presence of neoplastic proliferation with features of mature plasma cells can establish a morphological diagnosis of EMP, but the diagnosis will be complicated when cytological variants coexist within the tumor infiltrates. This is especially true with PBL, which share similar features of monomorphic population of large cells (plasmablastic or immunoblastic) characterized by more or less eccentrically placed nucleus, little chromatin and single prominent centrally located nucleoli, or multiple peripherally located small nucleoli together with the presence of mature plasma cells.[5,6] However, the presence of cells with obvious plasmacytic differentiation is not typical of PBL,[7,8] but is the defining feature of EMP [Figure 2].[5,8]

The immunohistochemical panel used in the present case [Table 1 and Figure 3] may well exclude other tumors characterized by plasma cell cytology, but is unlikely to establish a diagnosis when the differential is PBL and EMP as both share identical immunophenotypes.[5,8] The low Ki-67 proliferation index (20–25%) is consistent with EMP compared

to higher expression in PBL (more than 80%).[7,9] Though in situ hybridization for EBER was not done in the present case, the reliance on EBER status to resolve the diagnosis of a case in routine practice is conflicting,[2,7,8] especially in non-HIV patients.[9]

The foregoing indicates that distinction between the two competing diagnoses may be best resolved when morphology is supplemented with clinical parameters such as aggressiveness, gingival presentation, and HIV status.[5-7,9,10] Interestingly, Kane *et al*.[7] is of the view that "plasmablastic EMP presenting as a gingival mass is more of a theoretical possibility rather than a practical problem." Although this is likely to be true in PBL in HIV positive patients, oral involvement is considered uncommon in non-HIV patients.[9] Moreover, PBL with light chain restrictions are considered as plasmablastic EMP.[6]

The lymph node involvement as noted in the present case has been reported to develop in 8% of EMP but has no prognostic implications.[3] Radiotherapy is the mainstay of treatment as it is a radiosensitive tumor. The optimal radiation dose adopted is in the range of 40–60 Gy given over a period of 4–6 weeks.[1] Good remission of the disease was achieved in the present case with primary radiotherapy. It has been reported that the type of light chain restriction has prognostic significance with better outcome when it is lambda light chain restriction,[3] a finding also found in the present case.[3]

In conclusion, we report a rare case of plasmablastic EMP in the gingiva with light chain restrictions that can be distinguished from PBL if subtle attention is paid to the morphological and clinical detail.

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