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**Photo vignette**

**Unusual manifestation of mucosal plasmacytosis mimicking erythema multiforme**

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**Abstract**

Mucosal plasmacytoses constitute a group of idiopathic inflammatory disorders, characterized by a dense infiltrate of plasma cells at the mucocutaneous junction without any recognizable dermatosis or neoplasm. We report an unusual clinical presentation of mucosal plasmacytosis presenting with hemorrhagic crusting of the lips, mimicking erythema multiforme.

**Key words: erythema multiforme, mucosal plasmacytosis, plasma cell cheilitis, plasma cell gingivitis.**

**Introduction**

Mucosal plasmacytoses constitute a group of idiopathic inflammatory disorders, characterized by a dense infiltrate of plasma cells at the mucocutaneous junction without any recognizable dermatosis or neoplasm. The disease predominantly affects the lower lip with or without concurrent involvement of the gums, buccal mucosa, and tongue. Herein we report an unusual clinical presentation of mucosal plasmacytosis presenting with hemorrhagic crusting of the lips, mimicking erythema multiforme (EM).

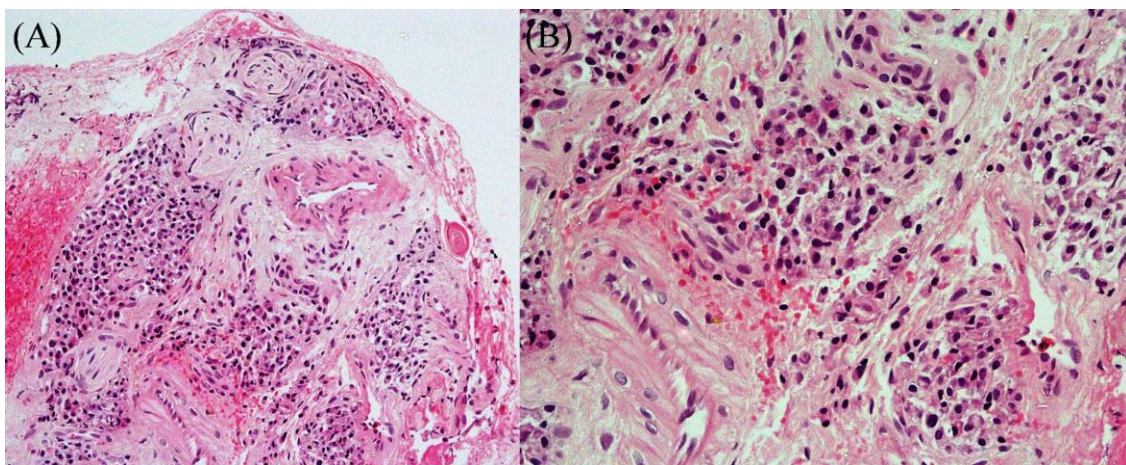
**Case synopsis**

A 38-year-old woman presented with painful, persistent erosions over lower lip and gingivae with intermittent bleeding for 3 months. She denied any preceding bullous lesions, drug intake in the recent past, excessive sun exposure, or use of lip care products. She was a tobacco and betel nut chewer since the age of 14 years. On examination, hemorrhagic crusts diffusely covered the lower lip, which on removal, revealed superficial glistening erosions. Both upper and lower gingivae were swollen and showed erosions over the gingival margin (Figure 1).

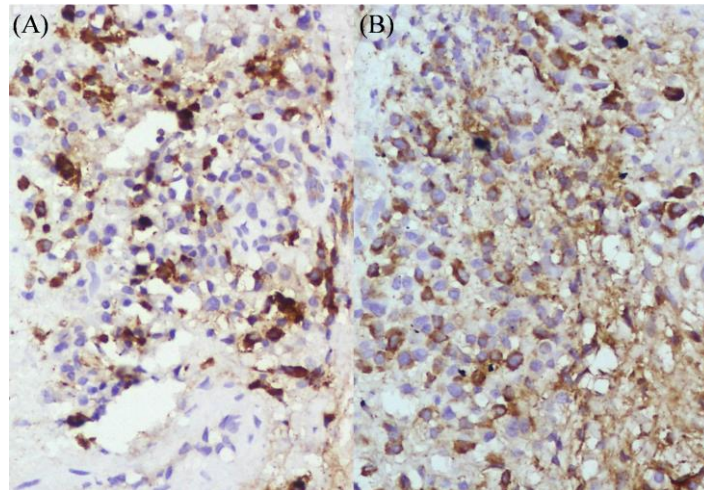


**Figure 1.** Erythematous, glistening erosion involving the upper gingiva and hemorrhagic crusting of the lips.

The rest of the mucocutaneous examination was normal. Local cervical lymph nodes were not enlarged. Routine biochemical and hematological investigations were within normal range. Venereal disease research laboratory (VDRL) test for syphilis was negative. A punch biopsy obtained from the lower lip showed a denuded mucosal lining epithelium with fibrin rich exudate covering the ulcer base. The subepithelium showed dense inflammatory infiltrate composed predominantly of plasma cells with a few scattered lymphocytes (Figure 2a). The plasma cells were mature-looking without any nuclear atypia (Figure 2b) and were found to be polyclonal with no light chain restriction (Figure 3a and 3b).



**Figure 2.** Photomicrograph showing ulcerated mucosal lining with sub-epithelium showing dense plasma cell rich inflammatory infiltrate (a) (H and E x200). Higher magnification showing mature looking plasma cells (b) (H and E x400).



**Figure 3.** Immunohistochemistry for light chain (a) kappa and (b) lambda did not show light chain restriction.

Direct immunofluorescence of peri-lesional skin showed no significant immune deposits. Based on the clinical and histological features, a diagnosis of plasma cell cheilitis was made. She was treated with topical mometasone furoate 0.1% and tacrolimus 0.1% once a day for 12 weeks with minimal improvement. She was further lost to follow-up.

## Discussion

Plasma cell cheilitis commonly involves the lower lip of the elderly, presenting as a flat to raised, erythematous plaque with a dry, atrophic, shiny surface and varying degrees of fissuring. Hemorrhagic crusting of lips is rare with only a single previous report [1]. Senol et al have reclassified plasma cell cheilitis under a unified term called mucous plasmacytosis, which also includes plasma cell gingivitis, Zoon balanitis, and plasma cell vulvitis [2]. It has also been occasionally described to involve the buccal mucosa, palate, nasal aperture, lips, tongue, epiglottis, larynx, and other orificial surfaces [3]. Mucous plasmacytosis in turn is classified under idiopathic plasma cell infiltrates, which also includes muco-cutaneous plasmacytosis, cutaneous plasmacytosis, and plasma-acanthoma.[2]

Mucosal plasmacytosis is considered to be a nonspecific inflammatory response to a wide range of exogenous agents including infections, trauma, and poor hygiene [4]. A few authors have implicated a hypersensitivity response to chewing gum and minor trauma related to dentures in the pathogenesis of mucosal plasmacytosis. When strongly suggested by history, discontinuation of these extraneous factors has induced remission in isolated patients [5, 6]. Betel nut and tobacco use may have been a factor in our patient.

Other clinical possibilities considered in our case were EM, erosive lichen planus, and paraneoplastic pemphigus (PNP). All these dermatoses share in common the presence of cheilitis and hemorrhagic crusting and need to be differentiated based on clinical and histological findings. A relatively long and persistent disease course, absence of skin lesions, and a dense inflammatory infiltrate with absent necrotic keratinocytes were against a diagnosis of EM. A rare paraneoplastic blistering dermatosis, PNP, is clinically characterised by recalcitrant mucositis, hemorrhagic crusting of lips, and polymorphic skin lesions. Absence of blistering and disease limited to lips and gingiva with no histological or immunofluorescence evidence of PNP helped in its exclusion. Erosive lichen planus can have similar clinical presentation, but a prominent plasmacytic infiltrate with minimal lymphocytes and absence of colloid bodies helped in its exclusion.

Other rare causes of predominant plasmacytic infiltrate of the lips include extramedullary mucocutaneous plasmacytoma and plasmocanthoma. Extramedullary mucocutaneous plasmacytoma histologically differs from our case by the monoclonal nature of the infiltrate and presence of atypical nuclei and mitotic figures. Plasmocanthoma presents as a circumscribed tumoral swelling of the lips, histologically showing acanthosis, papillomatosis, and dense infiltrate of plasma cells in dermis akin to plasma cell cheilitis. There is a report of both these entities occurring together speculating the possibility that they are diseases of the same spectrum [7].

Literature on therapeutic modalities of mucosal plasmacytosis is limited. Commonly used treatment options are summarised in Table 1. Corticosteroid administered topically or orally is the most common treatment modality. Other reported treatment options include topical tacrolimus, excimer light and surgical excision, but none is uniformly effective.

**Table 1:** Treatment modalities of mucosal plasmacytosis

<b>Treatment modality</b>	<b>Mode of administration</b>	<b>Formulation</b>	<b>Strength/ Concentration</b>	<b>Response</b>	<b>References</b>
Corticosteroids	Topical	Cream/ointment		Failure, partial improvement and complete response	1, 3, 4, 7-9
	Local	Injection	Triamcinolone 2.5mg/ml – 10mg/ml	Failure, Partial improvement and complete response	3, 4, 9
	Oral	Tablet	≤40mg/day	Partial to complete response	5, 10
Tacrolimus	Topical	Ointment	0.03%/1%	Failure, partial to complete response	9, 11-13
Pimecrolimus	Topical	Cream	1%	Partial response	8, 12
Cyclosporine	Topical	Solution	Not mentioned	Complete clearance	14
Monochromatic excimer light	Local		100-140mJ/cm <sup>2</sup> twice a week	Improvement followed by relapse	13
Surgical debulking	Local			Complete response	15, 16

## Conclusion

In conclusion, a diagnosis of mucosal plasmacytosis should be suspected in any patient presenting with erythematous, edematous, glistening plaque with or without erosions over the mucosal surfaces. The diagnosis is confirmed by clinico-pathological correlation, after excluding other dermatoses with similar presentation.

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