CASE REPORT

Oral Bullous Lichen Planus; An Unusual Variant: Case Report and Review

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Abstract:

Lichen planus is a chronic inflammatory mucocutaneous disorder that affects both skin and oral mucosa. The exact cause of oral lichen planus is unknown. It may or may not be associated with skin lesions. Oral mucosa exhibits a variety of clinical patterns, including reticular, plaque, erosive, bullous, and atrophic, with bullous lichen planus being a rare entity. We present a unique case of bullous lichen planus in a 69-year-old female patient with no skin manifestations, as well as a review of literature on bullous lichen planus.

Keywords: Bullous lichen planus, Burning sensation, Mucocutaneous.

Introduction

Oral Lichen Planus (OLP) is a chronic dermatologic disease that commonly affects the oral mucosa. Erasmus Wilson, a British physician, coined the unusual name for the condition in 1869. Lichen Planus (LP) is thought to be a T cell-mediated autoimmune disease in which cytotoxic CD8+ T cells are recruited into the skin, causing interface dermatitis. Viruses, drugs, and contact allergens have all been implicated in the development of LP. It is seen all over the world, mostly in the fifth and sixth decades of life, and is twice as common in women as in men. Purple, pruritis, polygonal, and papular lesions are common in LP skin lesions. Variations in morphology and location have also been observed, including linear, annular, circular and ulcerative[1].

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Bullous lichen planus (BLP) is a rare form of lichen planus. It is distinguished by the presence of vesicles or bullae, which typically develop in the context of pre-existing LP lesions. The formation of bullae in bullous lichen planus is consistent with extensive vacuolar change in the basal cell layer. Familial forms are inherited in an autosomal dominant pattern with variable penetrance. BLP usually appears as tense bullae on top of violaceous, polygonal LP lesions. These bullae have the potential to rupture, resulting in ulcerative areas. It commonly affects the dorsal aspects of the hands, feet, and trunk. The bullous variant of OLP is less common than the dermal variant[2]. We are reporting a case of BLP that was present on the oral cavity extensively.

Case report:

A 69 year old female patient reported to the Department of Oral Medicine and Radiology, Malabar Dental College and Research Centre Edappal with a chief complaint of recurrent oral ulcers and burning sensation of oral cavity while taking spicy food for the past 7years. The burning sensation was minimal in beginning and it made worse by spicy food. It was preceded by the eruption of fluid-filled vesicles that used to rupture in a matter of time. She had visited many doctors for the same and had taken medications by which she had only symptomatic relief. There were no dermal lesions and the family history was non-contributory.

Intraoral examination revealed, an eroded erythematous area with multiple ulcers on right lateral aspect of hard palate in relation to 16, 17, 18 teeth region (Figure 1). White radiating striations were seen over the left alveolar ridge in relation to 26, 27, 28 region which was seen extending to buccal vestibule. (Figure 2). This non scrapable lesion was tender on palpation and the surface was smooth and flat.

Based on clinical history and presentation, the lesion was provisionally diagnosed as BLP and a number of lesions with similar features like pemphigus, pemphigoid, allergic stomatitis, erythro leukoplakia and erythematous candidiasis were considered for differential diagnosis.

Incisional biopsy of the lesion was carried out and the tissue was sent for histopathological examination which revealed stratified squamous surface epithelium supported by connective tissue stroma. Epithelium was parakeratotic with absence of rete-ridges. It showed basal cell degeneration and large cleft like spaces focally. Subepithelial band of inflammatory cells predominantly of lymphocytes were distinct in most areas and diffuse inflammation in few areas.(Fig 4) Stroma was moderately fibro-vascular. Supra basilar split was also seen in few areas. (Figure 5).On the basis of above histopathological features, the lesion was diagnosed as BLP.

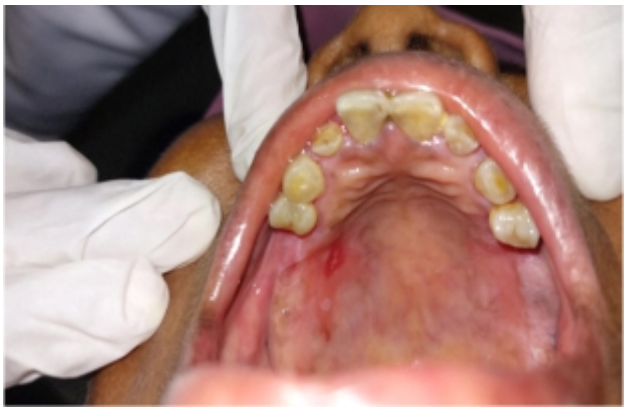

Figure 1: Lesion in right lateral aspect of hard palate

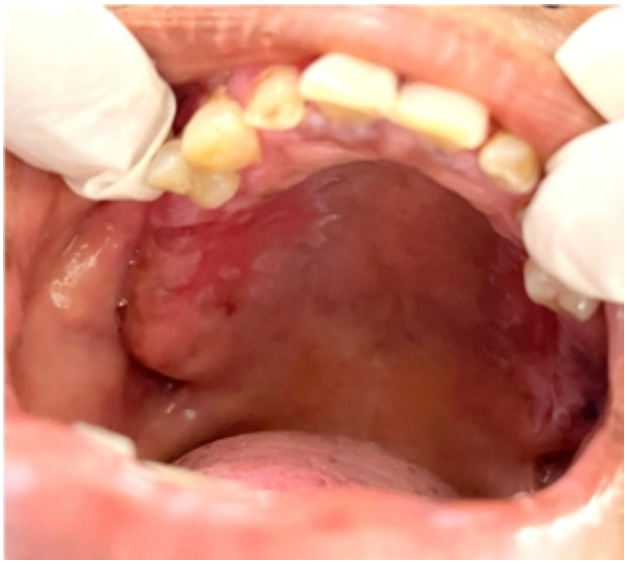

Figure 2: Lesion in alveolar ridge

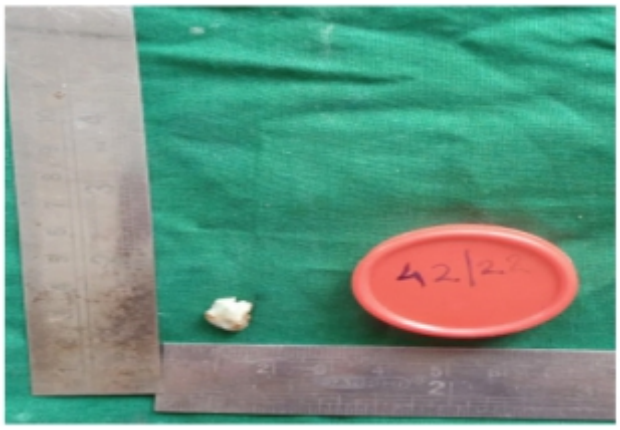

Figure 3: Collected Specimen

Discussion

LP is a common immune mediated mucocutaneous disease with characteristic clinical and histopathological

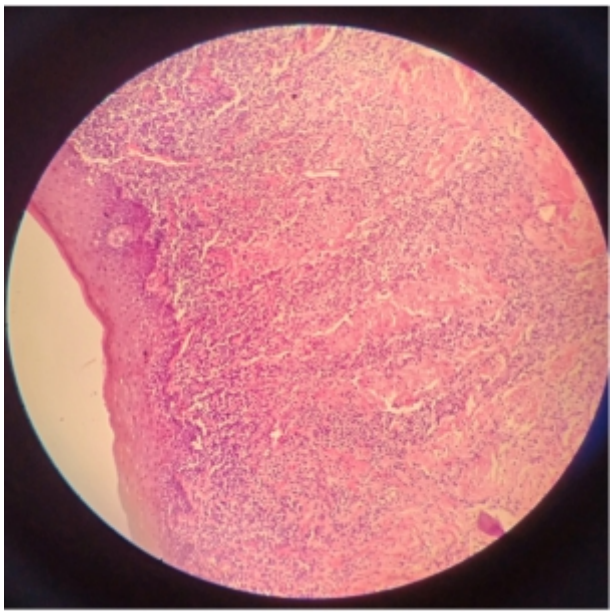

Figure 4

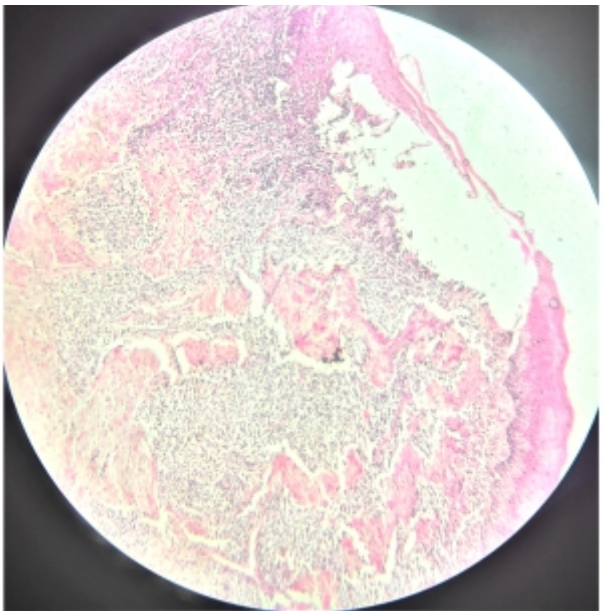

Figure 5

findings that affects between 0.5 and 1.2% of world population. OLP affects about 1.5-2% Indian population. Cutaneous LP is self-limiting and resolves within 6 to 14 months, whereas mucosal LP is chronic and may be refractory to treatment. Various etiological factors are considered for LP which include mainly genetic and environmental factors. T-cell mediated immunity is being projected as one of the reasons for its pathogenesis. Also HLA based susceptibility association studies had pointed towards a genetic pre-disposition for LP, since several HLA alleles are associated with LP. The Various environmental factors which can precipitate LP may include dental materials, drugs, infectious agent, autoimmunity, immune deficiency, food allergy, stress, habits, trauma, diabetes, hypertension, malignant neoplasm, and bowel diseases[2].

Clinical Features:

LP can affect individuals of all ages, but up to 95% of cases occur in adults, with most cases presenting between the third and sixth decades of life. Female sex predilection was observed than males.

Approximately 15% of patients develop cutaneous lesions. The classic appearance of skin lesions described by the six P's: planar, plaque, pruritic, purple, polygonal, and papular. Typically cutaneous lesions develop after the appearance of oral lesions.

The clinical picture of LP of skin is quite distinctive, nearly pathognomonic, and consists of flat-topped, polygonal, erythematous or violaceous papules with scaly surfaces. The papules frequently feature branny scaling that forms tiny, whitish streaks known as Wickham's striae on their dry, glossy surfaces. Although LP tends to favour the flexural surfaces of the forearms, wrists, and ankles; the dorsal surface of the hands; the shins; the trunk; and the sacral region, lesions are normally symmetric in distribution and can affect any part of the body[3]. BLP is a rare form of LP in which peri-lesional skin or pre-existing LP lesions develop vesicular and/or bullous lesions. Exact prevalence of BLP is unknown. Typically, the lesion is infrequent. A familial variant of BLP, which affects people of younger ages, causes widespread rashes that remain for a longer period of time. Legs and the mouth are the typical sites for familial blister development. Bullae are short-lived and burst, leaving a rough, ulcerated, or degraded surface[4].

Oral manifestations:

There are six distinct clinical forms of lichen planus: reticular, papular, plaque, atrophic, erosive, and bullous. The buccal mucosa, followed by the tongue, gingiva, and lower lip, is the most common location for the most prevalent kind, the reticular variety, which is characterised by the presence of Wickham's striae[5]. In contrast to the above findings, our case was seen on the hard palate.

BLP in the mouth has symptoms resembling those in the extremities. Most often, a pruritic, violaceous, papular look is present, along with bullae development that later become connected with pigmentations. Clear fluid may be present in the bullae. Most commonly affected intraoral site is buccal mucosa. Most often, bilateral incidence is recorded. It tends to have an unbroken bullae or erythematous area in the centre, with white striae around the edges[6].

Our case also had similar bilateral lesions with an erythematous core surrounded by white striae and light pigmentation. The palatal lesion was elevated with multiple minute round erythematous projections of varying sizes interspersed with white striae.

Investigations:

Hemograms, liver and renal function tests, anti-hepatitis B and C tests, and urine analysis are routinely carried out studies in BLP. If classic lesions are present, the distinctive clinical features are sufficient to determine the proper diagnosis. It is advised to do an oral biopsy with histopathologic analysis in order to confirm the diagnosis and rule out dysplasia and cancer. It is widely recognised that direct immunofluorescence is useful for diagnosing diseases, particularly those with non-diagnostic histopathologic characteristics and desquamative gingivitis[4]. In the case presented, biopsy was performed to confirm the diagnosis of BLP.

The characteristic histopathological features of LP consist of hyperkeratosis, increased granular cell layer, acanthosis with a so-called "saw tooth" appearance, liquefaction of basal cell layer and presence of band-like inflammatory cell infiltrate at the dermo-epidermal junction. Apoptotic or dyskeratotic keratinocytes, known as Colloid or Civatte bodies can be found at the lower dermis and superficial epidermis. Vacuolar degeneration of the basal layer is typical and results in separated spaces between the epidermis and the dermis, known as "Max-Joseph spaces"[1].

The dermo-epidermal junction is altered in BLP, and large Max-Joseph spaces are formed, causing liquefactive eruption of the basal layer cells, particularly in long-lasting lesions but also in skin that seems normal perilesionally. Due to the basal layer cells' detachment from the underlying basement membrane, tense bullae or vesiculobullous lesions that are intrabasal arise as a result. In these circumstances, the remaining histology results are LP-typical.[7]

Management:

There is currently no recommended standard treatment for BLP. Topical powerful corticosteroids can be used empirically because they are active against hyper-reactive type of LP. For moderate to severe BLP, mini pulse therapy with oral betamethasone has shown to be helpful. For severe refractory cases of lichen planus, systemic corticosteroids are the second line of treatment. Dapsone has proven to be useful for treating paediatric BLP. Resistant hypertrophic BLP was seen responding to mycophenolatemofetil. Bullous lichen-planus can be effectively treated with antimalarial medication as an alternative[7].

Conclusion

The term "OLP" refers to a mucosal lesion which affects a large group of people. It is crucial to locate and get rid of the disease's multiple causative agents. Topical steroids alone or in conjunction with other topical immuno modulatory medications can provide relief for the majority of individuals. Rarely do individuals need to take systemic drugs for an extended period of time. Due to the LP's propensity for malignancy, the patient should continue to receive long-term follow-up. Since all treatments are non-specific and focused on reducing inflammation, they are only partially effective.

Conflict of interest: Nil

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References:

1. Neville BW, Damm DD, Allen CM, Chi A. In Dermatology Diseases: Oral and maxillofacial pathology. First south Asia ed. Philadelphia: Elsevier Publication; 2016. p. 729-34.
2. Weston G., Payette M. Update on lichen planus and its clinical variants. Int. J. Womens Dermatol. 2015;1:140–149.
3. Bilgili ME, Yildiz H, Sarici G. Prevalence of skin diseases in a dermatology outpatient clinic in Turkey. A cross-sectional, retrospective study. J Dermatol Case Rep 2013; 7: 108.
4. Liakopoulou A, Rallis E. Bullous lichen planus–A review. J Dermatol Case Rep. 2017;11:1-4.
5. Andreasen JO. Oral lichen planus. 1. A clinical evaluation of 115 cases. Oral Surg Oral Med Oral Pathol. 1968;25:31-42.
6. Babu A, Chellaswamy S, Muthukumar S, Pandey B, Jayaraj M, Francis S. Bullous lichen planus: Case report and review. J Pharm BioallSci 2019;11:S499-506.
7. Patil A, Prasad S, Ashok L, Sujatha GP. Oral bullous lichen planus: Case report and review of management. ContempClin Dent. 2012;3:344-8.

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