A Rare Variant of Oral Lichen Planus-Bullous Form

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Lichen planus (LP) is an immune-mediated chronic inflammatory disease that is seen mainly affecting the skin, nails, hair, and mucous membranes. Oral LP (OLP) has about 0.4-5.3% potential to be malignant. If the disease is sufficiently characterized, the diagnosis can be made from the clinical features alone; but a biopsy is recommended to confirm the diagnosis and to exclude the presence of dysplasia and malignancy. OLP seems to be mediated by an antigen-specific mechanism, thereby activating the cytotoxic T-cells and non-specific mechanisms such as degranulation of mast cells and activation of matrix metalloproteinase. Pathogenesis of OLP may involve both antigen-specific and nonspecific mechanisms. Here, we report a case of bullous LP in a 44-year-old female patient, discussing clinical features and etiopathogenesis.

Key words: Bullous, Lichen planus, Mast cells, Oral, Pathogenesis

INTRODUCTION

Oral lichen planus (OLP) is an autoimmune chronic inflammatory disease of the mucous membrane. It is mostly a CD8+ T-cell mediated autoimmune response with unknown etiology and pathogenesis.¹ About half of the patients with skin lesions have oral lesions, whereas about 25% present with oral lesions alone.² OLP has several clinical subtypes including reticular, erosive, atrophic, papular, plaque-like, and bullous subtypes. OLP constitutes 9% of all white lesions. Its prevalence in the general population is around 1-2%, and there are a large number of cases seen affecting the females.³

CASE REPORT

A female patient aged 44 years, presented with recurrent history of occurrence of boils and ulcers in the cheek area came to our institution. The patient gave a history of water filled boils on the inner aspect of the cheek since past 2 years. These boils would subsequently rupture leaving red raw areas associated with pain and would give a salty sense of taste. There was the occasional occurrence of such boils on the lips, which were quite painful. Occurrences of vesicles and bullae were seen on the hands. The patient had sought medical advice, and vitamins were prescribed and injections were given. However, there was no resolution of the lesion.

Extraoral examination revealed the presence of a red erythematous lesion on the left lower lip with crust formation (Figure 1). The lesion was tender and was bleeding on touch.

Red erythematous area measuring 4 cm × 2 cm on the left buccal mucosa was evident intraorally. The periphery of the lesion was surrounded by a white lacy pattern and brownish pigmentation. The central part of the lesion appeared ulcerated and necrotic (Figure 2). Right buccal mucosa

Figure 1: Red erythematous lesion on the left lower lip with crust formation

Figure 2: Ulcerated and necrotic central part of the lesion.
showed the presence of red erythematous area measuring 3 cm × 1 cm with white lacy pattern and interspersed red area. Similar lesions were seen on the left and right lateral borders of the tongue, each measuring 2 cm × 2 cm. The lesions were tender on palpation. A provisional diagnosis of LP was given considering the association of cutaneous lesions.

In the present case, only oral involvement was appreciated at the time of initial presentation. Thus, an incisional biopsy of the lesion was done for histopathological examination to confirm the diagnosis of bullous LP and to differentiate it from other vesiculobullous diseases. Complete hemogram was also done which showed all parameters within the normal range.

Histopathology revealed stratified squamous epithelium, which was atrophic in nature. Basilar degeneration of the epithelium and cleft formation was evident. The cleft was observed with dense inflammatory infiltrate and extravasated red blood cell’s (Figures 3 and 4). Based on the above histopathological findings and clinical correlation to the presence of vesicles a final diagnosis of bullous LP was given.

**DISCUSSION**

LP in Greek means *tree moss* and Planus means *flat*. The term LP was first introduced in 1869 by Erasmus Wilson, thereby renaming a condition that had been described by Hebra as “Leichen Ruber.” Mostly seen in the fifth to sixth decades of life, and is twice as common in women than in men. The prevalence of OLP is 1-2% in the general population while its prevalence in Indian population is 2.6%. OLP usually occurs in a bilateral symmetrical pattern.

Well recognized clinical subtypes of OLP are reticular, papular, erosive, atrophic, hypertrophic and bullous OLP. The papular, reticular and plaque-like forms are often asymptomatic, whereas erosive, atrophic or bullous type lesions cause burning sensation and pain. Among these forms reticular is common and bullous forms are rare (Table 1).

Bullous OLP manifests as small vesicles or bullae that rupture easily and can range in size from few millimeters to several centimeters in diameter. It is commonly seen on the buccal mucosa, particularly in the postero-inferior areas adjacent to the second and third molars. The next most common sites of manifestation are the lateral margins of

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<th>Table 1: Distribution according to clinical types⁶</th>
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<td>Clinical type</td>
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<tr>
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<td>Ulcerous</td>
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*<0.001
the tongue, whereas lesions are rarely seen on the gingiva or lips.4

Vesiculobullous lesions such as pemphigus vulgaris, bullous pemphigoid, mucus membrane pemphigoid, and erythema multiforme are considered in the clinical differential diagnosis of the present case.

Several factors including stress, genetics, systemic diseases like diabetes and hypertension, dental restorative materials, drugs, infectious agent, autoimmunity, immunodeficiency, food allergy, trauma, malignant neoplasm, chronic liver disease and hepatitis C virus, tobacco chewing, graft versus host disease and bowel diseases have been implicated as causative agents.5,6 The pathogenesis of LP is explained in four mechanisms- antigen-specific cell-mediated immune response (heat shock proteins, CD4+ T-helper cells, CD8+ cytotoxic T-cells) non-specific mechanism (epithelial basement membrane, mast cells, chemokines, matrix Metallo Proteinases) autoimmune response, humoral mediated mechanisms (circulating autoantibodies to desmoglin 1 and 3).5,10

This further leads to the damage of the basal keratinocytes leading to apoptosis caused by the antigen inciting the cytotoxic T-cells, which could be any of the above-mentioned factors.

Histopathological criteria for erosive or bullous LP includes - Overlying keratinization, degeneration of the basal keratinocytes, bandlike T-lymphocyte infiltration, absence of epithelial dysplasia and sometimes presence of colloid (civatte, hyaline, cytoid) bodies that appear as homogenous eosinophilic globules within the epithelium. The ultrastructure of colloid bodies suggests that they are apoptotic keratinocytes, with DNA fragments in these cells. Colloid bodies may be positive for fibrin, immunoglobulin M, C3, C4, and keratin. Rarely, histological cleft formation (Max-Joseph space) with clinical blistering of the oral mucosa (bullous LP) may be seen due to weaknesses at the epithelial-connective tissue interface.4,11

An oral biopsy with histopathologic study is recommended to confirm clinical diagnosis and also to exclude dysplasia and malignancy.10

The range of malignant transformation of OLP is reported to be between 0.4% and 5.3%. Therefore, World Health Organization has now classified OLP as a potentially malignant disorder.3,5

CONCLUSION

Identifying and eliminating multifactorial agents associated with the disease is essential. Interaction of various factors is probably responsible for the initiation, aggravation and persistence of OLP. A proper understanding of the pathogenesis of the disease becomes important for providing the right treatment. This case provides further evidence that omp therapy improves the clinical outcome in patients with moderate to severe OLP and though the treatment was equally effective as topical triamcinolone acetonide, the response is earlier, especially in erosive and bullous forms of the disease. Long-term follow-up of the patient should also be kept due to malignant tendency of LP. All treatments are nonspecific and directed at eliminating inflammation and, therefore, are partially successful.

REFERENCES