Lichen Planus with Extensive Cutaneous and oral Manifestations - A Case Report And Review of Literature

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ABSTRACT:
Oral Lichen Planus is a common chronic inflammatory disorder affecting stratified squamous epithelium of skin, oral mucosa and genitalia. It is a T-cell mediated auto-immune disorder in which auto-cytotoxic CD8 T-cells triggers the apoptosis of oral epithelial cells and leads to chronic inflammation. Skin lesions are usually self limiting and cause pruritis, whereas oral lesions are chronic, rarely undergo remission, are potentially premalignant and may even cause death. Usually clinical presentation is sufficient for diagnosis, especially if characteristic skin and oral features are present. However, Biopsy and immunofluorescent studies are done for a confirmatory diagnosis. The mainstay of treatment remains the use of topical steroids but newer therapies are also been in current use. This paper presents a case of Lichen Planus with extensive oral and skin lesions. The patients were treated with topical corticosteroids for oral lesions and was referred to dermatologist for the skin lesions.

Key words: Lichen Planus, Desquamative gingivitis, Auto immune disorders, Topical steroids

INTRODUCTION
Lichen planus is a common muco-cutaneous disorder, more frequent within the oral cavity, where it appears as either white reticular, or erosive lesions. In the majority of patients with oral lichen planus (OLP) there is no associated cutaneous lichen planus or lichen planus at other mucosal sites. This may be called “isolated” OLP. Although the exact etiology is unknown, it is generally considered to be an immunological hypersensitivity reaction, characterized by an intensive T-cell infiltrate localized in the epithelium-connective tissue interface. However, there are various factors that may predispose to its etiopathogenesis. Most patients with lichen planus are middle-aged or older adults. Women predominate in most series of cases, usually by a 3:2 ratio over men. The oral lesions are reticular, papular, bullous, plaque-like, atrophic, erosive and ulcerated. A rarely encountered form of lichen planus is the bullous variant. Lesions are usually seen on the buccal mucosa and less common on the tongue, inner aspect of the lips and gingiva. Gingival manifestations of lichen planus are relatively rare. Lichen planus is often diagnosed...
based on clinical information only, but erosive and bullous variants of lichen planus always require laboratory evaluation. Histological examination, immunohistology, particularly immunofluorescence, is increasingly being used to more accurately diagnose such diseases. Direct immunofluorescence analysis is not only proving very useful for differential diagnosis, but also adds insight into possible pathogenic mechanisms of desquamative gingivitis and it is essential for diagnosis of lichen planus. Early recognition of lichen planus or the vesiculobullous disorders may prevent delayed diagnosis and inappropriate treatment of potentially serious dermatological diseases. Treatment includes correction of predisposing factors and the use of topical steroids. However when the mainstay of treatment fails, systemic steroids, various immunomodulatory agents, Retinoids and Thalidomide may also be used.

A 60 year-old patient reported to department of periodontics with the complaint of burning sensations in the gums for the past 09 months. History reveals that patient was asymptomatic 09 months back when she developed burning sensations in the maxillary gingival region. The burning sensations were particularly increased on intake of hot and spicy foods. History also reveals that the patient was under depression for some personal family reasons. There was no associated history of vesicle formation in any area of oral cavity. Extra oral examination reveals multiple papular lesions on the skin of lower back region with no relevant drug history or family history. (FIG. 1) On Intra-oral examination, there was area of desquamation involving marginal and attached gingiva in relation to maxillary and mandibular anterior teeth. The desquamated area was surrounded by minute radiating whitish striations. The desquamated area showed bleeding on probing. Gingiva was soft and edematous in consistency. Non scrappable, whitish interlacing, radiating striae (wickham’s striae) were seen on lower labial mucosa, right & left buccal mucosa. (FIG 3, 4 & 5). Melanin pigmentation was also seen on left buccal mucosal lesions. Whitish plaque like lesion was observed on the dorsum of tongue. (FIG.6). Considering the history of 9 month duration, areas of desquamation with radiating white striations without formation of vesicle, along with characterestic skin lesions, provisional diagnosis of Lichen Planus can be given. After the patient was informed about the disease and getting her approval, incisional biopsy was performed. Histopathological features were consistent with the diagnosis of Lichen Planus. (FIG 7). The patient was subjected for thorough oral prophylaxis and oral hygiene instructions. Thereafter, the patient was prescribed topical application of high potency steroids (Clobetasole propionate) 3 times daily for one month. Tablet Cetzine (antihistaminic) once daily for 15 days and once daily tablet of chelating agent (Supracal) was also given to the patient. The patient was reviewed every 2 weeks for the first one month. The patient was referred to dermatologist for the cutaneous lesions. The lesions had subsided with topical steroids within 4 weeks of starting the treatment. (FIG 8, 9,10 &11). The patient was asked to stop the topical application and reinforcement of oral hygiene instructions was given. Since the lesions can recur, the patient was under observation for 6 months and the lesions showed no signs of recurrence.

DISCUSSION

Oral lichen planus is a relatively common mucocutaneous inflammatory disease affecting 1% to 2% of the population. The term “Lichen Planus” was coined by the British physician Erasmus Wilson in 1869. Since Lichens are primitive organisms of symbiotic algae and fungus, and planus in latin refers to “Flat”. Most patients who experience this disorder are middle-aged or elderly, and 60 % are female.

ETIOPATHOGENESIS:

The exact etiology is not known. However, OLP is a T-cell mediated auto-immune disorder in which auto-cytotoxic CD8 T cells triggers the apoptosis of oral epithelial cells, leading to chronic inflammation.

PREDISPOSING FACTORS:

1) HEPATITIS C VIRUS Infection: HCV infection is more common in Erosive Lichen Planus. HCV viral sequences have been found in the serum of patients with OLP; and HCV was shown to occasionally replicate in Oral Lichen Planus tissue, possibly contributing to the pathogenesis of mucosal damage.9,10

2) PSYCHOLOGICAL FACTORS: OLP patients exhibit higher levels of anxiety, depression, stress and psychological disorders. The levels of anxiety and salivaory cortisol of OLP patients are high, thus establishing the relationship of OLP and stress.
3) **ORAL LICHENOID REACTIONS:**
   
a) Dental restorative materials:
   Amalgams, composite resins, cobalt, gold and even flavouring agents may lead to oral lichenoid reactions.  

b) Drugs: NSAIDS, ACE inhibitors (captopril), beta blockers, Penicillamine may be implicated as a cause. However, lichenoid reactions are usually unilateral and resolves on discontinuation of the offending factors.

4) **MECHANICAL TRAUMA:** Dental procedures, friction from sharp cusps, rough dental restorations, poorly fitting prostheses and deleterious oral habits are exacerbating factors. **KOEBNER'S PHENOMENON,** where the lesions develop in response to trauma, explains why erosive lesions are common in areas subjected to trauma.

5) **PLAQUE AND CALCULUS:** May result in worsening gingival lesions LP and are associated with a higher incidence of erosive lesions.

6) **DIABETES AND HYPERTENSION:** There is no literature supporting the association of LP with Diabetes and Hypertension. However, **GRINSPAN'S SYNDROME** is the association of LP, Diabetes and Vascular Hypertension.

A) **EXTRA-ORAL MANIFESTATIONS:**
1) **CUTANEOUS LESIONS:** consists of purple, pruritic, polygonal papules, often overlain by radiating lines (**WICKHAM'S STRIAE**). Skin lesions develop several months after the appearance of oral lesions and are usually self-limiting. Genital mucosa is the most common extraoral site of involvement.

   **VULVOVAGINAL-GINGIVAL SYNDROME** - is the association of LP of vulva, vagina and gingiva in female patients. Patients usually complains of burning, pain, vaginal discharge and dyspareunia.

   **PENO-GINGIVAL SYNDROME** - is the male counterpart of vullovaginal-gingival syndrome of LP.

2) **SCALP AND HAIR FOLLICLES:** Lichen planopilaris / Graham's little syndrome represents LP involvement of scalp and hair follicles, resulting in scarring alopecia.

3) **NAILS:** Thinning and ridging of the nail plate and splitting of the distal free edge of the nail. Healing with scar produces **PTYERGIUM**.

4) **ESOPHAGEAL LESIONS:** Dysphagia is the commonest feature. Chronic pain and strictures may also be seen.

B) **ORAL MANIFESTATIONS:**
   
   The clinical evaluation of the oral lesions is based on the six clinical forms described by Andreason: reticular, papular, plaque, atrophic, erosive, and bullous. **Mucosal lesions**, which are multiple, generally have a symmetric distribution, particularly on the mucosa of the cheeks, adjacent to molars, and on the tongue mucosa, less frequent on the labial mucosa (lichenous cheilitis) and on the gums (desquamative gingivitis). The most common form is reticular type with characteristic slender white radiating interlacing striations. The lesions frequently occurs bilaterally and are mostly asymptomatic. Erosive LP most often appears as a mixture of intensely erythematous mucosa with large areas of irregularly shaped ulceration with a whitish-yellowish pseudomembrane. Erosive and atrophic LP results in burning sensations. Erythematous lesions that affect the gingival cause desquamative gingivitis, the most common type of gingival LP. The plaque like forms of LP may resemble leukoplakia, particularly proliferative verrucous leukoplakia and appears as slightly raised or flat area on oral mucous membrane. The most common site of plaque like LP is tongue. Bullous LP is extremely rare form in oral cavity. The bullae rupture almost immediately, leaving an ulceration on a bed of inflamed mucosa. Bullous LP most commonly affects the posterior buccal mucosa.

**MALIGNANT POTENTIAL:**

The most important complication of OLP is the development of Oral squamous cell carcinoma. The first case of carcinoma arising in LP of oral mucosa was described by HALLAPEAU in 1910. The risk of malignant transformation varies from 0.4% to over 5% a period of 5 – 20 years. Accumulation of inducible nitric oxide synthetase with 8-nitroguanine and 8-oxo-7,8-dihydro-2'-deoxyguanosine in oral epithelium in OLP may cause oxidative and nitrate damage to DNA and could be the basis of malignancy. The risk of malignant transformation is independent of the clinical type of OLP or the treatment used.
DIAGNOSIS:
1) CLINICAL DIAGNOSIS: is sufficient to establish a diagnosis of OLP, if characteristic oral and skin lesions are present.

2) HISTOPATHOLOGY:
   Essential features:
   - superficial band-like infiltrate of T lymphocytes
   - Basal cell liquefaction degeneration
   - Normal epithelial maturation pattern

   Additional features:
   - SAW TOOTH rete pegs
   - Civatte /colloid bodies
   - Separation of epithelium from lamina propria
   - Max joseph spaces

3) IMMUNOFLUORESCENCE OF PERILESIONAL MUCOSA:
   - Fibrin and shaggy fibrinogen in a linear pattern at basement membrane zone
   - Cytoids in the absence of deposition of fibrinogen

TREATMENT:
Generally, no medication is necessary for the benign form of this disease (reticular lichen planus). In the case of severe pain and a burning sensation, high-potency topical corticosteroids remain the most reliably effective treatment modality. Oral hygiene and corrective dentistry play a major role in the management of OLP and consultation with a dentist or oral medicine specialist is helpful.

DRUG TREATMENT:
Drug treatment with topical steroids is preferred due to fewer side effects. Systemic steroids may be used if the lesions are extensive, or there are recalcitrant disease.

TOPICAL CORTICOSTEROIDS:
Most effective topical steroids are the medium potent steroids (triamcinolone), high potent steroids (fluocinolone acetonide) and superpotent halogenated steroids (clobetasol propionate). Elixir forms are also used such as dexamethasone, triamcinolone and clobetasol. These are used for diffuse oral involvement, elderly patients or for patients having difficulty in applying the medications. The greatest difficulty in using topical steroids is the lack of mucosal adherence for a sufficient period of time. Therefore, topical steroids may be used with adhesive pastes (orabase). A regular follow up should be done for prolonged use of topical steroids for the following adverse effects:
   a) secondary candidal infection
   b) Tachyphylaxis (diminished biological effectiveness)
   c) Adrenal suppression
   d) Atrophy of the oral mucosa

INTRALESIONAL STEROIDS:
Used for intractable erosive OLP lesions. Triamcinolone acetonide (10-20 mg/ml) is used and repeated every 2-4 weeks. Frequent steroid injections are painful and may result in an unwanted systemic dose.

SYSTEMIC STEROIDS:
Should be reserved for recalcitrant cases of erosive or erythematous OLP or for widespread OLP with skin, genital, scalp or esophageal involvement. Daily doses of 40-80 mg is usually sufficient to achieve a response.

OTHER TOPICAL AGENTS:
   a) TOPICAL CYCLOSPORINE (100 mg/ml as a mouth rinse) - may be beneficial in recalcitrant OLP cases. Systemic absorption is generally low, but it is expensive and less effective than topical steroids in inducing clinical improvement in OLP.
   b) TOPICAL TACROLIMUS: is a steroid free topical immunosuppressive agent. It was used primarily for atopic dermatitis. The exact mode of action is not known they inhibit T cell activation and proliferation. Burning is the most common side effect with tacrolimus. Other side effects include carcinogenicity, mutagenesis and infertility.
   c) TOPICAL RETINOIDS: Tretinoin and isotretinoin are used for erosive-atrophic forms. Side effects include teratogenicity and liver dysfunction.

OTHER THERAPIES:
   - PUVA THERAPY: Phototherapy with 8-methoxy psoralen and long-wave ultraviolet light (PUVA) has been used successfully in the treatment of skin lesions and cutaneous lichen planus. It was first used in the treatment of recalcitrant OLP. Eighty-seven percent of patients treated with ultraviolet-A, without a systemic or topical photosensitizer, improved significantly. Some
studies have indicated that PUVA therapy might also have therapeutic effects. To avoid PUVA side effects, photosensitization with topical 0.01% trioxsalen can be used for the treatment, although oral mucosa seems more resistant to phototoxic damage in comparison to skin. PUVA with 8-methoxypsoralen has various side effects, such as nausea, dizziness, eye symptoms, paraesthesia, and headache. Photochemotherapy may be use-ful for severe forms of erosive OLP that do not re-spond to conventional treatment. Moreover, one matter of concern is that PUVA therapy has been shown to have oncogenic potential. CRYOSURGERY has also been used, particularly in erosive drug resistant OLP, but the lesions may develop in healing wounds and result in scars.

-LASER THERAPY- CO2 lasers are used to treat multicentric lesions or lesions in difficult areas.

ADDITIONAL DRUG THERAPY:
Griesofulvin, Thalidomide, Levamisole and Dapsone may also be used.

SURGERY:
Resection may be done for isolated plaques or non-healing erosions. Free soft tissue grafts have been used for localized areas of erosive OLP. However, periodontal surgery have been reported to provoke OLP.

CONCLUSION: OLP are a relatively common oral mucosal disease process encountered in clinical practice. Given the apparent risk of oral SCC, regular follow up of these patients is mandatory.

REFERENCES
16. Scott S; De Rossi; Katherine N Ciarrocca : Dental clinics of north America 2005; 49: 77-89
19. Chiayarit P; J ontell M; Hiraku Y et al; Nitratative and Oxidative damage of DNA in OLP; Cancer Sciences; 2005; 96: 553-559
20. Nazzaro G; Cestari R; Topical Tacrolimus ointment in ulcerative lichen planus; An alternative therapeutic approach; European journal Dermatology july 2002; 12:4:321
Figure 1: Papular lesions on the lower back skin.

Figure 2: Erythematous marginal and attached gingiva of maxillary anterior teeth.

Figure 3: Desquamative gingivitis affecting the mandibular labial gingiva with radiating whitish striaations of lower labial mucosa.

Figure 4 & 5: Whitish interlacing striae affecting the right & left buccal mucosa.

Figure 6: Plaque like lesions on the dorsal aspect of tongue.

Figure 7: Histopathology showing basal cell degeneration & chronic inflammatory cells.

Figure 8: Resolution of skin lesions.

Figure 9: Marked reduction in erythema of marginal and attached gingiva.

Figure 10&11: Resolution of lesions on lower labial mucosa and tongue.